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PEDIATRICS

Focus on Content Specs

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Board content specifications identified and explained in detail

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Growth and Development Milestones

Growth
Normal growth
Measurement

Meaning, uses, and limitations of bone age: Bone age correlates with pubertal development stage and is helpful in predicting adult height in early- or late-maturing adolescents. In familial short stature, the bone age is normal (comparable to chronological age). In constitutional delay, endocrinologic short stature, and under nutrition, the bone age is low and comparable to the height age. Skeletal maturation is linked more closely to sexual maturity rating than chronological age. It is more rapid and less variable in girls than in boys.

Types of measurements used in assessing nutritional status include weight, length/height, head circumference, body mass index [BMI], and skin folds. Data are plotted on growth charts according to age and compared with a reference population.

How to obtain accurate growth measurements:

-Weight: use properly functioning, well maintained scales which are calibrated regularly. Children should be in their diaper or underwear. If a child refuses to stay still, an acceptable estimate can be made by weighing the child in the parent's arms and then subtracting the parents' weight.

-Length: should be used for infants and toddlers under 2 years, taken in a supine position with fully extended legs and head up against an inflexible board.

-Height should be measured for children 2 years and older and should be taken standing, with a stadiometer mounted to the wall.

-If patient cannot stand, arm span or height estimation by adding base of heel to knee, knee to hip, hip to the top of the head measurements.

Role of body mass index in monitoring growth: BMI-for-age charts are recommended to assess weight in relation to stature for children ages 2 to 20 years. The weight-for-stature charts are available as an alternative to accommodate children ages 2 to 5 years who are not evaluated beyond the preschool years.

Linear growth and weight gain

Length at birth doubles by 3 to 4 years of age.

Normal and abnormal growth can be evaluated by plots on a growth chart: Height (or length), weight, and weight-for-height should be plotted on a growth chart for every patient. Abnormal growth can present as:

Stunting (length or height for age <5th percentile)

Underweight (Children <3 years with Weight-for-length <5th percentile Children ≥2 years with BMI-for-age <5th percentile)

Overweight (Children >2 years with BMI-for-age between 85th to <95th percentile)

Obese (Children <3 years with Weight-for-length >95th percentile or Children ≥2 years with BMI-for-age ≥95th percentile)

Most full-term infants will regain their birth weight within two weeks. Full-term infants will triple their birth weight by 1 year of age.

Head growth

Normal head circumference of a full-term infant at birth is 33 to 36 cm. Growth pattern of acquired microcephaly:

Acquired (or progressive) microcephaly is a condition in which the head circumference is within normal range at birth, and for a period thereafter, but then does not increase as fast as normal and crosses percentiles to below the second percentile.

An environmental insult to the brain that occurs later in life, particularly beyond the age of 2 years, is less likely to produce severe microcephaly. Serial head circumference measurements are more meaningful than a single determination, particularly when the abnormality is minimal.

Distinguish between hydrocephaly and macrocephaly:

Hydrocephaly = congenital or acquired enlargement of the ventricular system.

Macrocephaly = Head circumference > 2 SD above the mean based on age and sex; causes include hydrocephaly, megalencephaly, thickened skull, space occupying lesions.

Normal and abnormal variations in head shape:

Cranial growth is triggered by brain growth, two thirds of which occurs by two years of age. The metopic suture between the frontal bones closes at two years of

age; the other sutures remain open until brain growth stops in the second decade of life.

Fontanel size is influenced by brain growth, dural attachments, suture development, and osteogenesis. Size of the fontanel is calculated by the average of the antero-posterior and transverse dimensions.

Cranial index (CI), an objective measure used to quantify head shape, is the width of the head divided by the length multiplied by 100%. Normal CI ranges from 76% to 81% in prone sleeping infants, and 86% to 88% in supine sleeping infants. CI is heavily influenced by infant sleep position due to the malleability of the skull.

A CI of > 81% indicates a shortened anterior-posterior dimension and widening between the biparietal eminences, or brachycephaly.

Dolichocephaly, a narrow elongated head, is defined as a CI < 76%.

During infancy, increased supine positioning is a frequent cause of occipital flattening, also called deformational brachycephaly.

Plagiocephaly is a nonspecific term used to denote an asymmetric head, and has become more common since the 1990s "Back-to-Sleep" initiative to prevent sudden infant death syndrome (SIDS). It can be seen in normal infants, but is more prominent in hypotonic or weak infants.

The shape of the head should be documented carefully. A variety of abnormal head shapes can be seen when cranial sutures fuse prematurely = craniosynostosis.

An infant presenting with an abnormal head shape requires careful investigation. The etiology of cranial dysmorphology can be divided into 3 categories that include:

Malformations - occur in the first 8 weeks of pregnancy during embryogenesis. Malformations may be caused by either a genetic or environmental etiology, or with a combination of both.

Deformations - occur after embryogenesis, and result from nondisruptive mechanical forces, such as intrauterine or postnatal positioning, that cause abnormal formations or distortions of normal body parts.

Disruptions - result from a destructive process that occurs after normal organ formation.

Correct measurement of the head circumference should be performed at every visit for patients younger than 3 years and should be recorded on a suitable head growth chart.

The average rate of head growth in a healthy premature infant is 0.5 cm in the first 2 weeks, 0.75 cm in the 3rd week, and 1.0 cm in the 4th week and every week thereafter until the 40th wk of development. The head circumference of an average term infant measures 33-36 cm at birth, 44 cm at 6 months, and 47 cm at 1 year old.

Growth pattern of familial macrocephaly: These children are born with large heads and normal body size. During infancy, head circumference (HC) increases to greater than 90th percentile, increasing by 0.6 to 1 cm per week (compared with the normal 0.4 cm/week). Head growth velocity slows to a normal rate by approximately six months of age. Familial megalencephaly/macrocephaly can be confirmed by measuring the patient's parents' HCs and by using Weaver curves. If the child's HC falls within normal ranges using the Weaver curves, radiologic evaluation is not necessary.

Atypical growth

Failure to thrive (FTT) is a physical sign of under-nutrition in the majority of patients.

Extensive laboratory evaluation of FTT can be deferred until outpatient dietary management is tried.

Observational clues to FTT include oromotor dysfunction and mother-infant interaction.

Poor feeding technique, inappropriate formula preparation, and inadequate intake are common causes. FTT may be an expression of parental neglect/ inadequacy.

Long-term consequences of FTT in infancy include decreased postnatal brain development leading to short stature, and behavioral and academic difficulties.

Differential diagnosis for an infant with failure to thrive:

Surveillance and screening

Regular, periodic standardized developmental and behavioral screening for the well child population leads to increased early intervention, which leads to better developmental outcomes.

Commonly used developmental screening tests and their limitations:

Ages and Stages Questionnaires (ASQ): screens development in children from 4 to 60 months. Limitations include only 75-85% of children are identified with developmental problems, and the ASQ is written at a 3rd-12th grade reading level, which would be a problem for illiterate parents.

Brigance Screens-II: screens development in children from 0 to 90 months. The test identifies 70 to 82 percent of children with giftedness or developmental and academic problems, and correctly identifies 70 to 82 percent of children without such conditions.

Infant-Toddler Checklist for Language and Communications: screens development in children 6 to 24 months of age. Limitations include literacy issues as it is written at a sixth-grade reading level.

Parents' Evaluation of Developmental Status (PEDS): screen development in children from 0 to 8 years. The PEDS provides longitudinal surveillance and triage. Similar limitations as the above screening tools

Methods of developmental and behavioral screening: Screening tests draw on information reported by the parents and can be self-administered in waiting or examination rooms, attached to an appointment letter, administered online before an appointment, or delivered by interview in person or over the telephone.

Developmental delay is considered significant when performance is 2 standard deviations or more below the mean on appropriate standardized norm-referenced testing. Red flags include:

Motor: failure to have steady head control while sitting (assisted) at 4 months, inability to sit at 9 months, inability to walk independently at 18 months

Cognitive: lack of fixation at 2 months, lack of visual tracking at 4 months, not turning to sound or voice at 6 months, not using consonant sounds in babble at 9 months, not using single words at 24 months, and not speaking in three word sentences by 26 months

Social emotional: lacking smile or other expressions of joy at 6 months, lacking reciprocal vocalizations at 9 months, failure to respond to name when called or babble or perform reciprocal gestures (showing, reaching, waving) at 12 months, lack of proto- declarative pointing or showing gestures or single words at 15 months, not engaging in simple pretend play or using spoken language/gesture combinations at 18 months, lack

of two-word meaningful phrases at 24 months or at any point in development, the loss of any of the babbling, speech or social skills that were previously acquired

Distinguish among isolated, global, and atypical developmental delay:

Isolated developmental delay = restricted in language, learning or motor skills; result in lower academic performance than expected for age, IQ, and educational level.

Global developmental delay = significant delay in two or more domains including gross/fine motor, speech/language, cognition, social/personal, activities of daily living.

Atypical development = some aspect of a child's development is abnormal even if not meeting full criteria (i.e. two standard deviations below the mean or age-appropriate average) for diagnosing an isolated or global developmental delay.

Variations in development associated with prematurity: increased risk of cognitive and motor impairments. Interventions include teaching, home visits, child care at a child development center, and parental group meetings.

Variations in development associated with congenital visual impairment: Prevalence of congenital visual impairment is 1 per 1000 children. Impacts on attachment formation, social development

and later cognitive development; Vision screening may occur at birth, especially in a premature infant, but wellness visits as early as six months should also include basic vision screening.

Variations in development associated with congenital hearing impairment: Prevalence of congenital hearing impairment is 1 per 1000 children. The National Institute of Health has recommended hearing screens in the first 3 months of life for all infants, as well as screening at school-entry to detect those who have an acquired hearing loss. This is because hearing loss can lead to delays in language acquisition and cognitive development, and early intervention can aid in minimizing these delays.

Developmental milestones

Milestones for infants and young children
Neonatal period (first four weeks after birth)

Normal developmental milestones for the neonatal period include alerting to sound (bell or voice); demonstrating visual preference for human face; starting to smile by 1mo.

Two months

Normal motor developmental milestones include regarding objects, following 180 degrees, lifting head and shoulders off bed in the prone position, moving symmetrically, diminishing primitive reflexes.

Normal cognitive/behavioral milestones: smiles socially; coos; makes reciprocal

vocalizations; looks at parent; comforts self.

Lack of visual fixation by 2 months of age is abnormal.

Four months

Normal motor milestones: steady head control while sitting; holds head up; bears weight on forearms in the prone position; reaches for objects; moves symmetrically; begins to roll.

Normal cognitive/behavioral milestones: laughs out loud; initiates social interaction; responds to affection; indicates pleasure/displeasure.

Lack of visual tracking by 4 months of age is abnormal.

Lack of steady head control while sitting by 4 months of age is abnormal.

Six months

Normal motor milestones: transfers object between hands; rolls in both directions; sits with support.

Normal cognitive/behavioral milestones: turns directly to sound/voice; uses visual exploration; uses oral exploration; babbles consonant sounds; imitates speech sounds; shows pleasure interacting with parents/others.

Failure to turn to sound or voice by 6 months of age is abnormal.

Nine months

Normal motor milestones: feeds self with fingers; plays pat-a-cake; bangs two objects together; grasps pellet-like object with immature pincer; sits without support; crawls; pulls to feet with support.

Normal cognitive/behavioral milestones: says nonspecific "mama" and "dada" and repeats consonants; understands his/her own name; recognizes common objects (e.g., bottle) or people (e.g., daddy); plays peekaboo; looks at books; seeks parent for comfort; stranger anxiety; object permanence.

Inability to sit by 9 months of age is abnormal.

Lack of babbling consonant sounds by 9 months of age is abnormal.

Twelve months

Normal motor milestones: pulls to a stand and cruises; takes a few independent steps; neat pincer grasp of raisin or pellet; bangs toys together; drinks from a cup.

Normal cognitive/behavioral milestones: says "mama" and "dada" with specific meaning; babbles; copies sounds and gestures; follows simple commands; waves bye-bye; cries when parent leaves.

Fifteen months

Normal motor milestones: gives and takes a toy; drinks from a cup; makes a line with a crayon; makes two-cube tower; walks independently; stoops to floor/recovers to standing position.

Normal cognitive/behavioral milestones: says three to six specific words; follows simple commands; uses jargon; indicates some desires or needs by pointing; brings toys to caregiver; helps in the house; listens to a story.

Eighteen months

Normal motor milestones: feeds self with spoon; stacks tower of three cubes; runs; walks up steps with hand held.

Normal cognitive/behavioral milestones: imitates household tasks; says 7 to 10 words; uses words for wants or needs; identifies one or more body parts; laughs in response to others.

Inability to walk independently by 18 months of age is abnormal.

Twenty-four months

Normal motor milestones: washes and dries hands; removes clothing; stacks tower of four to six cubes; feeds self with spoon and fork; runs well; kicks ball; jumps with two feet off floor; turns book pages.

Normal cognitive/behavioral milestones: combines words into two- or three-word phrases; points to pictures named; uses vocabulary of 50+ words; uses "I," "me," and "mine" in speech; follows 2-step commands; plays pretend; plays alongside other children.

Failure to use single words by 24 months of age is abnormal.

Thirty-six months

Normal motor milestones: helps in dressing (unbuttons clothing, puts on shoes); stacks tower of 10 cubes; copies a circle; stands momentarily on one foot; walks up stairs with alternating feet; draws a person (usually 2 body parts); pedals tricycle; toilet- trained during daytime.

Normal cognitive/behavioral milestones: speech is 75% intelligible; speaks in sentences of five to eight words; knows meaning of simple adjectives (e.g., tired, hungry, thirsty); knows age and gender; names objects; imaginative play; self-care skills.

Failure to speak in three-word sentences by 36 months of age is abnormal.

Four years

Normal motor milestones: copies cross; draws simple figure of a person (head plus one other body part); hops on one foot; balances on one foot for 3 seconds; brushes own teeth; dresses self.

Normal cognitive/behavioral milestones: pretend play; speech fully intelligible; asks questions (e.g., when? why? how?); tells a story; counts four objects accurately; draws person (usually 3 body parts); interacts with peers; fantasy play.

Five years

Normal motor milestones: dresses and undresses; draws a person with 6 body parts; skips, alternating feet; balances on one foot for 2 seconds; able to tie knots.

Normal cognitive/behavioral milestones: plays board or card games; good articulation/language skills; draws person (6+ body parts); counts to 10; prints some letters/numbers; copies triangle/squares; follows simple directions; listens and attends.

Six years

Normal motor milestones: rides a bicycle without training wheels.

Normal cognitive/behavioral milestones: writes name; knows right from left across the midline ("Touch your right ear with your left hand."); knows color names; identifies letters and numbers.

Milestones for kindergarten readiness

Indications of social readiness to attend school: separate from parents for several hours at a time; plays well with other children; follows directions in a group activity; tells stories.

I. Nutrition and Nutritional Disorders

A. Normal nutritional requirements

1. General

One problem with introducing solid foods before 6 months of age is increased risk of gastrointestinal infection

Before 4 months of age, pancreatic function is not completely mature, so there is difficulty with completely hydrolyzing starches before reaching the colon, where bacteria are able to ferment them to produce gas.

The typical adolescent's diets often provide insufficient amounts of: Calcium (especially if vegan or lactose intolerant) Vitamin D

Whole grains

Fruits and vegetables Iron

Nutritional needs in adolescence

3.5-6.5 cups of fruits and vegetables

Half of grain servings should be whole grains (at least 3 oz) At least 3 servings of fat-free/low fat milk products

1300 mg calcium per day

600 IU vitamin D (New Institute of Medicine Recommendation)

Nutritional needs in sports participation

2 hours of athletic activity requires 800 to 1700 calories

Extra calories: 60% complex carbohydrates, 25% fat, and 15% protein

Some diets increase risk of infants having particular nutrient deficiencies.

Goats milk diet - deficient in folic acid, infants at risk for myeloblastic anemia Vegetarian diet - vitamin B12 deficiency

Infants breastfed by vegan mothers - vitamin B12 deficient (2-12 months)

Single nutritional source-exclusive breastfeeding or iron-deficient formula after 6 months of age: iron deficiency.

Family meals affect the diet of children and adolescents. Cultural dietary restrictions affect diet. Also, the traditional methods of cooking and ingredients used in meals typical to a given culture affect the diet of children.

2. Minerals

Full term neonates have acquired sufficient iron stores during in utero growth.

Iron deficiency anemia is common in young people in the U.S. Behavior changes:

fatigue, irritability and paleness (conjunctiva, gums, nail beds or palmar creases)

Iron is less bioavailable in cow's milk than in human milk, so infant formulas have a higher concentration of iron.

In preterm neonates (<36 weeks' gestation) who are fed breast milk, unless otherwise contraindicated, supplementation with elemental iron at 2 mg/kg per day by 1 month of age until 12 months is recommended. In neonates with very low birth weight (weight less than 1,500 g),

higher iron intakes of 2 to 3 mg/kg per day may be needed. Premature newborns who are formula fed may also require iron supplementation, depending on formula intake.

Preterm infants require extra phosphorous to allow for catch-up bone mineralization, but if excess phosphorus is consumed along with excess calcium, especially when using loop diuretics or glucocorticoids, calciuria or renal stones can form.

Low iron formulas do not reduce straining and constipation but do put the infant at risk for iron deficiency.

Most mineral accumulation occurs during the third trimester, therefore premature newborns are at risk for developing deficiencies of calcium, phosphorus, iron, copper, and zinc; other mineral deficits (eg, iodine) are possible, but there have been few if any clinical reports of these deficiencies.

The current recommendations are that premature newborns consume 150 to 200 mg/kg of calcium and 60 mg/kg to 75 mg/kg of phosphorus each day.

Fluoride is recommended beginning at 6 months of age. It is important to ask parents how they are preparing their infant's formula to help determine fluoride exposure. Fluoride has been added to approximately two-thirds of community tap water in the United States.

It is now recommended that fluoride toothpaste be used as soon as the first tooth erupts. Under 3 years of age, a smear of fluoride toothpaste or a grain of rice–sized amount should be applied, while over 3 years of age, a pea-sized amount should be used twice a day to brush the teeth.

Preadolescents and adolescents need 1300mg of each, calcium and phosphorous, daily.

3. Vitamins

Breast milk does not have significant amounts of vitamin D, so all breastfed infants should receive 400 IU Vitamin D.

Vitamin A

Absorption: carotenoids are absorbed in the terminal ileum.

Metabolism: converted into retinyl esters in the intestinal cells, which are transported to the liver, undergo hydrolysis to make retinol, and are made into a complex with retinal-binding protein.

Storage: transported to epithelial cells in the eyes, respiratory tract, urinary tract and digestive tract.

Vitamin D

Absorption: ergocalciferol (D2) and cholecalciferol (D3) are absorbed in the small intestine. Also, UV radiation converts 7-dehydrocholesterol to vitamin D3 in certain layers of the epidermis.

Metabolism: The liver converts D3 to 25-hydroxyl-D3 and the kidney converts this to 1, 25 dihydroxy-D3 which releases calcium from bones and causes kidneys to reabsorb more calcium.

Storage: fat tissues

Vitamin D intake of 600 IU per day is necessary in children and adolescents. This is a recent change made by the institute of medicine from previous recommendation of 400 IU per day. Darker skinned infants may require as much as 800 IU per day.

Babies should continue taking vitamin D unless weaned to 32 oz of vitamin D–fortified formula (approximately 1 L)

If exclusively formula-fed babies are consuming less than 32 oz of formula per day, or if older children are drinking less than 1 L of whole milk per day, they may also need a vitamin D supplement of 400 IU per day, depending on their intake of other dietary sources of vitamin D.

It is believed that some components of breast milk, including neutrophils, stem cells, and immunoglobulins, may be reduced by the pasteurization process, donor milk must be pasteurized. It is not recommended, however, that donor milk be obtained from the Internet or directly from an individual. Milk obtained from these sources may not have been safely handled or tested properly for bacteria, viruses (such as HIV, cytomegalovirus, hepatitis), or other sources of contamination.

Vitamin E

Absorption: tocopherols are absorbed in ileum by scavenger receptor SR-B1, after which it is transported in chylomicrons or ATP binding cassette transporters to liver.

Metabolism: vitamin E gets incorporated into high density lipoproteins in the liver.

Storage: liver

Vitamin K

Absorption: jejunum

Metabolism: it is recycled by vitamin K-epoxide reductase after being used as a cofactor for gamma glutamyl carboxylase

Storage: liver

Vitamin B1 (Thiamine)

Absorption: water soluble, absorbed in the jejunum by active transport when intestinal level is low and by a passive mucosal process when the concentration in the intestines is high.

Metabolism: it serves as a coenzyme which helps decarboxylate and transketolate alpha-ketoacids, and also plays a role in acetylcholine synthesis.

Storage: Little is stored in the body and it is excreted by the kidneys.

Vitamin B2 (Riboflavin)

Absorption: takes place in the proximal small intestine

Metabolism: involved in oxidative and reduction reactions in the mitochondria, as well as hydrogen transfer reactions. Specifically, it is important for fatty acid synthesis, amino acid synthesis, function of enzymes, producing energy, and reproducing glutathione.

Vitamin B3 (niacin)

Absorption: stomach and small intestine

Metabolism: involved in electron transport and glycolysis, synthesis of fatty acids and steroids, cell differentiation, DNA processing.

Vitamin B6 (pyridoxine)

Absorption: in large intestine, vitamin B6 from both diet and bacteria is absorbed

Metabolism: plays a role in amino acid metabolism and transport, glycogen metabolism, neurotransmitter synthesis, and activity of steroids; serves as coenzyme in these processes

Vitamin B 12 (cobalamin)

Absorption: in the small intestine, it binds to intrinsic factor (IF) (which is produced by parietal cells in the stomach); the IF bound vitamin B12 is absorbed by enterocytes in the terminal ileum, after which Vitamin B12 is released from IF

Metabolism: enterohepatic cycling (excreted in bile, reabsorption in terminal ileum) Storage: large stores in the liver

4. Fat

Preterm formulas: 40-50% fat content as medium chain triglyceride oil (directly absorbed into the portal vascular system), whereas term formulas often have 100% long chain triglycerides, which depend on bile acids and micellar solubilization.

Preterm infants have a decreased ability to absorb fats and fat soluble vitamins, and this has implications for the content of preterm infant formulas.

5. Protein

Preterm infants require 4 grams per kilogram per day of protein, and 3.4g/kg to 4.2g/kg/d if they need to achieve catch up growth. Full term infants require 1.5g/kg/d to 2g/kg/d protein.

Cow's milk should not be introduced into the diet before one year of age, as it does not contain the proper nutrient balance and places infant at risk of iron deficiency anemia.

6. Calories

Infants require 100 kcal/kg/day for the first few months, but by 3 months of age only require about 95 kcal/kg/day. From 4-36 months they require about 80 kcal/kg/d. As children age, their calorie requirement is related to energy expenditure, and active children require about 20 kcal/kg/d more than sedentary children. In early childhood, children require 50-60 kcal/kg/d. By adolescence, children only require 30-50 kcal/kg/d.

Preterm infants generally require 120 kcal/kg/d, but require more if they have short bowel syndrome, intrauterine growth restriction, chronic lung disease, congestive heart failure or infection.

B. Infant feeding

1. Breast-feeding

Human milk has a lower protein content than cow's milk but has a higher whey:casein ratio than cow's milk.

Secretory IgA and antibodies against bacteria and viruses are found in human milk.

These antibodies from human colostrum and milk can help defend the body against pathogenic organisms and prevent their entrance into the rest of the body.

Human milk has low vitamin K, and if the infant does not receive vitamin K from another source, he or she will be susceptible to hemorrhagic disease of the newborn.

Oligosaccharides in human milk promote growth of the gastrointestinal microflora, which differs between human milk-fed and formula-fed infants.

Storing breast milk

Contraindications in Breastfeeding

Infants with a diagnosis of galactosemia cannot breastfeed, they require lactose-free milk, and they are most often started on a soy-based formula.

Maternal infection with human T-cell lymphotropic virus type I or II, untreated brucellosis.

In the United States, HIV, are also contraindications to breastfeeding; in these cases, mothers should neither breastfeed nor provide expressed breast milk to their infants.

In mothers with untreated active tuberculosis, expressed breast milk may be provided, and breastfeeding may later begin once a mother receives treatment with antituberculosis drugs for approximately 2 weeks and is proven to no longer be infectious.

Mothers with the onset of varicella 5 days before a newborn's birth through 2 days after the birth will be separated from their newborn, but they may provide expressed breast milk; likewise, mothers with H1N1 influenza should be separated from their neonates until they are afebrile, but they can provide expressed milk.

Mothers with active herpes simplex lesions on the breast may also provide expressed breast milk.

Some drugs are contraindicated in breastfeeding. These include Amiodarone

Anticancer medications (Doxepin) Radioisotopes

Tetracyclines for more than 3-4 weeks Ergotamine

Cabergoline Bromocriptine

Sodium iodide or potassium iodide Methotrexate

Lithium

amphetamines, chemotherapy agents, ergotamines, and statins, for which risks associated with breastfeeding have been identified.

Drugs of abuse: cocaine, amphetamine, phencyclidine, heroin

NOTE: mothers on a stable methadone program are encouraged to breastfeed, and this has been shown to reduce symptoms of neonatal abstinence syndrome.

Mothers who wish to drink alcohol should drink no more than 0.5 g of alcohol per kilogram of body weight; alcohol consumption should occur soon after nursing the infant to ensure that there is a maximal time until the next feeding—ideally more than 2 hours—to reduce the alcohol concentration in the breast milk.

If breastfeeding mothers ingest medications that have sedating effects, the infant may experience sedation.

Group A streptococcus or Staphylococcus aureus mastitis until after 24 hours of maternal antibiotics and mother is comfortable.

Since breast milk is lower in iron, iron-fortified infant cereals or pureed meats are important first foods for breastfed infants. In addition, foods rich in vitamin C, such as cantaloupe, mango, strawberries, and broccoli, should be included in the diet, along with nonheme iron-rich foods (for example, prunes, lentils, and kidney or white beans) to improve iron absorption. Likewise, breast milk provides an adequate amount of zinc until 6 months of age, after which babies should be provided with additional zinc through complementary foods, such as meat and fortified infant cereal.

Human milk and infant formula are different in the following ways:

Human milk has 70:30 whey to casein ratio, cow's milk and many infant formulas

have lower ratio,formulas consist of approximately 80% casein and 20% whey protein.

Total protein content is higher in infant formula to compensate for the differences between protein composition of cow's milk and human milk.

Infant formulas are fortified with iron which has somewhat poor bioavailability, whereas human milk has lower amount of iron that is more bioavailable.

Infant formulas are supplemented with vitamin D, but human milk does not contain significant amounts of this nutrient.

Infants who consume human milk have fewer gastrointestinal infections.

Breast feeding can be temporarily suspended when there are breast lesions from: Herpes simplex, varicella zoster, smallpox virus or smallpox vaccine virus until treatment and/or prophylactic medication provided for mother and infant.

2. Formula-feeding

The following formulas contain full lactose:

Enfamil premium, Similac Advance, Similac PM 60/40 low iron, Similac Go and Grow

The following formulas contained reduced amount of lactose:

Enfamil AR, Enfamil Gentlease, Enfamil RestFull, Enfamil Premium Next Step, Enfamil Premature, Enfamil EnfaCare Enfagrow Gentlease Next Step,Good Start Gentle Plus, Good start Protect Plus, Good Start 2 Protect Plus, Good Start 2 Gentle Plus, Good Start Premature, Parent's Choice Gentle, Parents Choice Added Rice Starch, Parent's Choice Advantage, Parent's Choice Older Infants, Similac Sensitive (trace), Similac Sensitive for Spit-Up (trace), Similac Special Care, Similac Expert Care

Infants with an exclusive goat milk diet tend to develop folate deficiency and consequential megaloblastic anemia.

Protein hydrolysate formulas can be used in short bowel syndrome, hepatobiliary disease, pancreatic insufficiency, autoimmune diseases and immunodeficiency syndromes, patients with high risk of atopic disease who are not being fed with human milk and if it extensively hydrolyzed, in cow milk protein allergy.

IgE mediated cow milk protein allergy presents as flushing, urticaria, rash or angioedema.

Non-IgE mediated cow milk protein allergy presents with hematochezia, abdominal pain and failure to thrive, while lactose intolerance presents with abdominal pain, diarrhea, nausea, flatulence or bloating after eating products with lactose.

Although persons with IgE mediated milk protein only have cross-reactivity to soy 8- 14% of the time, those with non-IgE cow's milk protein allergy have 30-64% chance of allergic enterocolitis when exposed to soy protein.

Ways to increase caloric density:

Add human milk fortifier (HMF) to human milk (increases mineral content). Add standard formula powder to human milk

Mix standard formulas with less water content.

Higher calorie feeds necessitate assessment for infant dehydration, irregular output, urine specific gravity, renal solute load, serum electrolytes or intolerance to or toxicity of the nutrients delivered.

Soy formula is indicated in galactosemia, congenital lactase deficiency, for infants older than 6 months of age with signs of IgE mediated allergy, and when a vegetarian diet is preferred.

The AAP does not recommend the use of soy formulas in preterm neonates for several reasons. First, osteopenia may be seen in preterm neonates weighing less than 1,800 g who consume soy formula, despite supplementation with both calcium and vitamin D. Also, there is a concern of aluminum toxicity in premature newborns fed soy-based protein formula because of the high aluminum content in these formulas. Increased aluminum deposition can occur in bone and in the central nervous system, especially if the preterm neonate has reduced renal function. Aluminum competes with calcium for absorption, which may contribute to osteopenia. Soy-based protein formulas also contain about 1.5% phytates, which can bind zinc, phosphorus, and iron and prevent adequate absorption of these nutrients. Soy-based formulas are therefore now fortified with higher amounts of these nutrients to make up for these deficits.

There are some amino acid–based formulas are mixed at a 1:1 ratio. It is important to review mixing instructions with families. With the help of a dietitian, formula can be concentrated to as much as 30 kcal/oz to help in the management of failure to thrive; also, to decrease the volume of feedings for infants with poor growth due to GER disease (GERD). Inappropriately concentrated formula may lead to symptoms of vomiting, constipation, and excessive weight gain, and the high protein content may cause renal dysfunction. Formula that is diluted may lead to diarrhea, slow weight gain, and electrolyte imbalances, including hyponatremia that can lead to seizures.

3. Introduction of solid food

Infants should not initiate solid feeds before 4 to 6 months due to need for the ability to coordinate chewing and swallowing

Gastrointestinal tract maturity to digest and absorb the contents of solid foods

Single ingredient foods should be started first:

Only one food per week to identify possible allergies or intolerances; these foods include dry cereal in formula, pureed fruits, vegetables and meats.

There is no evidence that suggests food allergy can be prevented by delaying solid food introduction beyond 4 to 6 months, the American Academy of Allergy, Asthma, and Immunology recommends 4 to 6 months of age for the introduction of complementary foods.

Chopped foods may be given to infants at 10 to 12 months of age.

Premature solid food introduction can lead to aspiration events or increased risk of gastrointestinal infections, may interfere with an infant's ability to acquire the appropriate nutrients needed in breast milk or formula, and in babies fed formula, feeding complementary foods prior to 4 months is associated with increased odds of obesity at 3 years of age.

Starting solids later than 6 months may lead to inadequate iron and zinc intake, particularly in children who are exclusively breastfed, and may lead to rejection of foods by an infant accustomed solely to sucking from a breast or bottle.

Signs of readiness include good head control, ability to sit with support, becoming unsatisfied after a formula or breast milk feeding, shortened intervals between feedings, opening the mouth when presented with food, diminished extrusion reflex (where the tongue raises and pushes against any

object that is placed between the lips, resulting in food being pushed out), showing interest in what a caregiver is eating, and demonstrating cues that they are finished feeding, such as turning the head away from the breast or bottle when they are done.

C. Deficiency states and hypovitaminosis (including rickets)

1. Vitamin deficiency states

Premature infants with low calcium or phosphorous intake may develop rickets.

Vitamin D deficiency

Breast fed infants or formula fed infants.

Rickets hypocalcemia, convulsions, tetany, apnea, stridor, wheezing, hypotonia, weakness of muscles or brisk reflexes

Children

Rickets (irritable, delayed gross motor skills, bone pain, wide wrists and ankles, knocked knee or bow legged, rachitic rosary, delayed fontanelle closure, craniotabes, frontal bossing, delayed tooth eruption with poor quality tooth enamel, poor growth, increased infections.)

Vitamin D deficient rickets in childhood presents with:

Bowlegs

Kyphosis

Deformities of the pelvis Delay in tooth eruption Widened wrists

Enlarged costochondral junctions (rachitic rosary)

Harrison groove (groove at the lower border of the chest caused by the diaphragm pulling against the weakened bone)

Vitamin D deficiency rickets findings

Labs: Decreased serum calcium, decreased phosphate, increased alkaline phosphatase, increased parathyroid hormone, normal or decreased 1,25 OH-D3 Radiographs: knees, wrists and shoulders: widening of the distal ends with cupping and fraying, osteopenia and metaphyses without calcification.

In children taking anticonvulsants, the 25 hydroxyvitamin D level should be checked annually and maintained in the normal range. Vitamin D deficiency can occur regardless of the serum level of the anticonvulsant.

Adolescents

Hypocalcemic seizures Tetany

Vitamin K deficiency

Early Vitamin K deficiency bleeding (first 24 hours of life): usually due to intrapartum maternal medications, such as warfarin, certain anticonvulsants, and isoniazid, affecting vitamin K storage.

Cephalohematoma, intra- abdominal bleeding or intracranial bleeding

Classic Vitamin K deficiency bleeding (2 to 7 days of life): bleeding from umbilicus, GI tract or puncture sites, gastrointestinal (GI),circumcision-site bleeding.

Late Vitamin K deficiency bleeding (2 to 12 weeks of life): intracranial bleeding

Formula-fed infants are relatively protected from VKDB, and they achieve vitamin K levels approximately 10 times greater than exclusively breastfed infants.

Vitamin K deficiency can be diagnosed by a prolonged prothrombin time and an elevated INR, with PTT being prolonged in severe deficiency.

Subcutaneous, IM, or IV supplementation of vitamin K can reverse the coagulopathy quickly, within 4 to 6 hours. In a patient who is clinically stable, without signs of serious bleeding, such as intracranial hemorrhage, vitamin K supplementation is sufficient.

Fresh frozen plasma is indicated for severe bleeding or if the patient requires an invasive procedure. Close monitoring in an intensive care setting is required if the neonate has intracranial bleeding.

The prognosis for infants without intracranial hemorrhage is excellent if treated appropriately and immediately with vitamin K. Long-term sequelae of children with intracranial hemorrhage depend on the extent and location of the hemorrhage. These neonates are at risk for motor and cognitive deficits.

Vitamin E deficiency

Signs/symptoms: ataxia, vision loss, inability to sense vibration and position, reduced reflexes, weakness, hemolytic anemia (Vitamin E is an important antioxidant present in cell membranes that reduces free radical damage to unsaturated fatty acids) and myopathy

Causes: fat malabsorption, short bowel syndrome, cholestatic liver disease, Friedrich ataxia, abetalipoproteinemia, extreme prematurity with very low birth weight, CYSTIC FIBROSIS

Vitamin B12 Deficiency

A child with either inflammation of the ileum or resection of the ileum will require supplementation of B12; because B12 bound intrinsic factor is absorbed exclusively in this location. Therefore, supplementation with 0.2 micrograms/kg subcutaneous injection for two days of cyanocobalamin (to minimize risk of hypokalemia as side effect) followed by 1000 micrograms each day 2 through 7, then 100 micrograms per week for one month. Then, monthly injections may be required if process is ongoing.

Folate deficiency may occur in children with malabsorption from Crohn disease, celiac disease or small bowel resection.

Strict vegan diet can lead to vitamin B12 deficiency and inadequate omega-3 fatty acid consumption. Most vegans and vegetarians consume significant amounts of folate, which can mask the hematologic effects of vitamin B12 deficiency.

Vitamin B12 deficiency causes

A diet without animal meat or supplementation.
Decreased absorption (autoimmune pernicious anemia, chronic acid suppression with proton pump inhibitors, bacterial overgrowth, parasitic infection, terminal ileitis, celiac disease, or after ileal resection).
Imerslund-Grasbeck disease (inherited disorder with abnormal vitamin B12 receptors in the ileum).

Signs and symptoms of B12 deficiency

Hematologic: macrocytic anemia, hypersegmented neutrophils, low white blood cell count, low platelets, pancytopenia

Neurologic: developmental delay, deficits in sense of vibration or proprioception, hypotonia, seizures, ataxia, abnormal movements, paresthesias, loss of memory, dementia, weakness

Psychiatric: change in personality, depression, irritability, declining school performance

Constitutional: failure to thrive, poor appetite, fatigue Gastrointestinal: glossitis, vomiting, diarrhea Dermtological: hyperpigmentation of skin, icterus

It is recommended to check vit B12, MMA, and homocysteine levels when B12 deficiency is suspected.

Vitamin B3 (niacin) deficiency results in pellagra and is associated with diarrhea, dermatitis, and dementia.

Vitamin B6 (pyridoxine) deficiency results in refractory seizures, dermatitis, peripheral neuropathy, and microcytic anemia.

Vitamin C deficiency

Signs and symptoms: bleeding (hemarthrosis, ecchymosis, myalgias), wounds heal poorly, gum disease, tooth loss, follicular keratosis, osteopenia, irritability, developmental delay

Causes: absence of sufficient vitamin C in diet (overheating foods or boiling milk decreases vitamin C content), alcoholism, diabetes mellitus, smoking, AIDS, bowel resection, chronic diarrhea

Vitamin A deficiency

Signs and Symptoms: Xerophthamia, night blindness, keratomalacia, ,defective tooth enamel, follicular hyperkeratosis on the extensor surfaces of the body, failure to thrive, and more severe clinical presentation of certain illnesses due to decreased immune response (diarrhea, respiratory illness, and measles)

Causes: malnutrition, Crohn disease, celiac disease, liver failure

Folate deficiency

Signs and symptoms: macrocytic anemia with hypersegmented neutrophils, irritability, chronic diarrhea, failure to thrive

Causes: malabsorphion (Crohn disease, celiac disease, short bowel syndrome), hemolytic anemia, exfoliative skin diseases, medications (methotrexate, pyrimethamine, phenytoin) or exclusive goat milk diet

2. Mineral deficiency states

Zinc deficiency: diarrhea, skin disorders (red inflammatory pustules, vesicles and bullae around the mouth, around the anus and limbs), short stature, hypogonadism, developmental and cognitive impairment, peripheral neuropathy, poor appetite, poor wound healing, susceptibility to bacterial infections, in severe cases hypothyroidism.

Copper deficiency: microcytic hypochromic anemia.

Magnesium deficiency: it may occur in conjunction with protein-energy malnutrition, malabsorption or severe diarrhea, leading to convulsions in severe cases.

Chromium deficiency: poor growth, impairment in the metabolism of lipids, protein and glucose (leading to glucose intolerance and hyperglycemia).

3. Protein, calorie deficiency states

Protein deficiency may present with features such as slowed growth,FTT scaling skin, thinning hair with little pigmentation, edema from low albumin, distended abdomen with hepatomegaly, irritability, developmental delay.

Insufficient total calorie intake causes marasmus, which features restricted growth parameters, little subcutaneous fat, constipation and xerosis with no edema.

Protein-losing enteropathy can be due to:

Intestinal inflammation (celiac disease, IBD, allergic enteropathy) Infection
Obstruction of venous or lymphatic outflow (CHF, constrictive pericarditis, intestinal lymphangiectasia)
Abetalipoproteinemia
Malignant infiltrate in bowel or lymphatics Mucosal injury

To screen for protein losing enteropathy, collect random stool to measure fecal alpha-1-antitrypsin. Collection of 24 hour specimen for alpha-1-antitrypsin and serum alpha-1- antitrypsin can accurately show GI protein loss. Also labs will show low serum proteins i.e albumin and immunoglobulins.

4. Hypervitaminosis

Hypervitaminosis D causes loss of appetite, increased thirst; dehydration, hypertension, hypotonia, and clouding of the cornea.Symptoms of acute vitamin D intoxication are the result of hypercalcemia, which may lead to emesis, anorexia, pancreatitis, hypertension, arrhythmias, nephrolithiasis, renal failure, and central nervous system effects.

Hypervitaminosis A can cause drowsiness, hair loss, arthralgias, elevated intracranial pressure and carotenemia (leading to yellow-orange skin pigmentation).

A single dose of more than 200 mg (> 660,000 units) will cause symptoms of acute toxicity. Most cases of vitamin A toxicity are caused by long-term ingestion of more than 10 times the recommended daily dietary allowance.

Acute or long-term vitamin A excess may cause hepatotoxicity and increased intracranial pressure (pseudotumor cerebri). Symptoms are Headache, Fatigue, Malaise, Irritability, Anorexia,Vomiting, Bulging Fontanelle ,Diplopia, Papilledema,Pruritic desquamating skin, Angular cheilitis,Hepatomegaly, Painful bone abnormalities caused by hyperostosis (especially in the midshaft of the long bones),Alopecia or coarsening of the hair and ataxia.

Hypervitaminosis C can be followed by a “rebound” deficiency.

Acute vitamin E toxicity is rare. Long-term intake of excess vitamin E supplementation has been associated with an increased risk for sepsis in premature infants and increased risk for hemorrhage and mortality in others.

D. Principles of nutritional support

1. General

2. Tube feeding, enteral nutrition

INDICATIONS OF TUBE FEEDING:

Insufficient oral intake
As a primary therapy for e.g IBD,metabolic disorders.
Oral motor dysfunction
Abnormal GI tract
Injury, critical illness, surgery,sepsis etc.
Contraindications of TF
Nonfunctioning GI tract, GI obstruction, intestinal ischemia.
(Severe pancreatitis and intestinal fistulas are not contraindications anymore)

Types of enteral feeding methods.

Orogastric (OG)

Preterm (less then 34 wks before they develop gag reflex)

Basilar skull fractures

Obstructed nares eg cystic fibrosis

Nasogastric (NG)

Mainly indicated for short term feeding

For long term nocturnal nutrition support

Gastrostomy tube (G tube) if feeding required for more than 3 months; can be placed surgically, endoscopically or radiologically. Surgical placement is better option as it's a formal attachment between gastric and abdominal wall but cannot be used for many days.

Placement of a G tube should be confirmed radiologically.

G tube does not prohibit oral feedings and easy to be used by caregivers once they are trained however needs to be changed frequently, kept clean and monitored for obstruction,dislodge,breaks, leaks and granulation tissue formation.

Prepyloric feeding is more physiological and better tolerated, allows bolus feeds,better gastric contractility, protective against hyperosmolar formulas thus decreasing the risk of dumping syndrome (results from high volume or high osmolar formula entering the duodenum causing distension, pain and diarrhea from inappropriate GI hormone release.

Associated with risk of aspiration so not indicated in gastroparesis and history of aspiration

Postpyloric feeding is feeding in duodenum or jejunum past ligament of treitz via nasoduodenal or nasojejunal tubes, useful for patients with intolerance to gastric feeding, severe reflux , neurologically impaired, gastroparesis, emesis due to chemotherapy. Disadvantages are difficulty in placement, frequent dislodges and dumping syndrome.

Bolus feedings are better for ambulatory patients and continuous feedings are better for critically ill, severely malnourished and those who cannot tolerate bolus feeds.

Complications of tube feeding:

GI: nausea, vomiting, diarrhea, constipation

Mechanical: aspiration, occluded or dislodged tube, skin and mucosal breakdown Metabolic:

Dehydration (less free water, hyperosmolar high protein formula

Hyperglycemia (too much sugar, inadequate insulin control)

Hyperkalemia (acidosis, renal disease,high K formula)

Hyperphosphatemia (renal insufficiency)

Hyponatremia (overhydration)

Hypokalemia (refeeding syndrome, diarrhea, malnutrition, insulin)

Hypophosphatemia (refeeding syndrome,insulin)

Weight gain (too much water or calories)

Refeeding syndrome (hypokalemia, hypomagnesemia, hypophosphatemia)

Advantages of enteral feeding.

Enteral feeds have the advantage of promoting GI motility, decreasing mucosal atrophy, decreasing bacterial overgrowth, and prevent entry of bacteria into circulation through gastrointestinal tract. They do not require central venous access and do not have associated risks of parenteral nutrition (cholestasis, line infections).

(This section can be removed)Enteral nutritional support may be indicated for preterm infants, prolonged anorexia, malnourishment (severe protein energy malnutrition), neurologic impairment, short gut syndrome, critically ill children, cardiopulmonary disease, hypermetabolic states, renal disease, hepatic failure and inflammatory bowel disease.

Bolus feeding is more convenient for home care, but is not usually appropriate when there is a high risk of aspiration, need for intestinal feeding, or intolerance of bolus feeds.

3. Parenteral nutrition

Total and peripheral alimentation are indicated when nutritional needs cannot be met by the enteral route. Common reasons include:

Preterm infant with severe respiratory disease, congenital GI tract anomalies or inflammatory disease of intestinal mucosa (e.g. necrotizing enterocolitis)

Infants with pancreatitis, graft-versus-host disease or post-surgical in cases where there is risk of starvation without parenteral nutrition

Patients with problems with full absorption due to short bowel syndrome or chemotherapy

Cardiac or renal failure

Large body surface area burns

Complications of parenteral nutrition:

Parenteral nutrition associated liver disease (PNALD)-cholestasis (infants), steatosis/steatohepatitis (older children from excess energy infusion), and necrosis of hepatocytes or cirrhosis: Monitor for hepatomegaly, increased conjugated bilirubin, and then liver enzymes and alkaline phosphatase increase later.

Enteral side effects-intestinal mucosal atrophy, decrease in pancreatic secretion (tend to resolve with time after enteral feedings are being given).

Metabolic bone disease: osteopenia, hypophosphatemia, rickets due to inadequate calcium and phosphorous nutrition: Monitor for alkaline phosphatase increase.

Vit B 12 often deficient in TPN can cause macrocytic anemia in patients on long term TPN e.g. with short bowel syndrome.

The goal is to maintain triglycerides at less than 150 mg/dL (1.7 mmol/L).

E. Nutritional problems associated with specific diseases, conditions

1. Gastrointestinal disorders

Acute gastroenteritis can produce a temporary postviral lactose intolerance which resolves on its own, with diarrhea symptoms ending in about one week.

Reintroduction of food early in the course of acute gastroenteritis improves nutrition and reduces symptoms of diarrhea. Excess sugar should be avoided, but otherwise dietary restrictions are unnecessary and provide suboptimal nutrition.

Crohn disease is associated with low vitamin D levels and consequential hypocalcemia. Fat malabsorption leads to poor absorption of fat soluble vitamins (A, D, E, and K). Also, when taking sulfasalazine (which competes for uptake with folate), folate deficiency can occur.

Malabsorption of bile, disaccharides, and fats can result in worsening diarrhea in CD.

Disaccharides (lactose, fructose, sucrose, etc) are digested and absorbed in the duodenum and jejunum. Small bowel inflammation may result in injury to villi, causing a secondary disaccharidase deficiency resulting in diarrhea. Malabsorption of carbohydrates results in increased stool reducing substances, not seen in the child in the vignette. Fat is digested and absorbed in the proximal intestine, and can be malabsorbed in severe chronic inflammation, however, this is fairly uncommon. Elevated fecal fat levels would identify this as a possible etiology.

2. Renal disease

Restriction of protein intake is not recommended in children, in view of their unique needs for growth and neurocognitive development. Also, protein restriction has not been linked with improved outcomes in CKD. Protein intake between 100% and 140% of the dietary reference intake (DRI), based on age and sex, is recommended for children with CKD and a glomerular filtration rate (GFR) of 30 to 60 mL/min per 1.73 m2. In children with a GFR less than 30 mL/min per 1.73 m2, the recommended protein intake is between 100% and 120% of the DRI for age and sex. In children with a GFR less than 15 mL/min per 1.73 m2, water-soluble vitamin supplementation is indicated. However, vitamin A supplementation is not routinely recommended because of an increased risk for hypervitaminosis A, secondary to accumulation of vitamin A metabolites.

Children with renal insufficiency are often in a catabolic state which means they need more nutrition for continued growth. Protein can be limited to 0.8 to 1.1g/kg per day without negative impact on linear growth.

Inadequate nutrition and fluid and electrolyte abnormalities, including metabolic acidosis, osteodystrophy, and disturbances of the growth hormone/insulin-like growth factor I axis, contribute to growth impairment in children with CKD,Interventions for renal osteodystrophy include dietary phosphorus restriction, vitamin D supplementation, and oral phosphate binders. Renal disease may lead to the deficiency of protein and calcium.

3. Hepatic disease

In chronic cholestasis, anorexia and fat malabsorption lead to growth failure. Protein energy malnutrition is also common from increased catabolism with decreased hepatic protein synthesis.

Liver disease diet should include:

Low fat diet with formulas containing medium chain triglycerides.; supplementation with fat soluble vitamins (A, D, E, and K); sometimes IV/IM vitamin A or oral vitamin A with a water soluble form of vitamin E are used Calcium supplementation.

Fat malabsorption can happen with cirrhosis or chronic cholestasis due to decreased bile acid excretion, therefore less lipolysis, solubilization and absorption of the long-chain triglycerides. Fat soluble vitamins are therefore also poorly absorbed.

Hepatic disease leads to decreased bile excretion, poor fat absorption and therefore poor vitamin D absorption. Also, 25 hydroxylation of vitamin D occurs in the liver, which is necessary for the production of active 1, 25-OH vitamin D. Hypocalcemia, hypophosphatemia, tetany, osteomalacia and rickets result.

4. Cardiac disease

Caloric intake needs to be increased in hypermetabolic conditions, such as cardiac disease, and when fluid restriction is necessary, food must therefore be more calorically dense.

Children with cardiac conditions that cause hypoxemia, congestive heart failure, or pulmonary hypertension are at particular risk for growth failure, and these children usually require at least 140 kcal/kg per day to meet their energy requirements.

Glucose polymers or fats (eg, microlipid) increase the caloric density of formula without increasing the osmolar load.

In 1992, National Cholesterol Education Program for Children made dietary recommendations which include diet with 25-30% fat, <10% calories from saturated fat, and daily cholesterol intake less than 300mg per day.

5. Cystic fibrosis

Children with cystic fibrosis may have malabsorption of protein, fat and fat soluble vitamins due to impaired secretion of pancreatic hormones, as well as protein energy malnutrition due to inadequate intake.

Thirty-eight percent of infants with CF have evidence of vitamin E deficiency.

6. Hematologic-oncologic disease

Malnutrition is common at the time of cancer diagnosis, especially with solid tumors, and may worsen with treatment due to increased energy requirements and decreased nutritional intake. Adequate nutrition improves survival, growth and immune function.

Children with cancer experience malnutrition as a result of hypophagia and anorexia, increased caloric needs, and malabsorption.

Children with cancer have higher caloric needs than children without cancer.

7. Neurologically handicapped children

Neurologically impaired children generally have lower calorie needs, but are at risk for micronutrient deficiency (osteopenia, iron deficiency anemia) due to decreased intake. Children with dyskinesias, hypertonicity or high seizure frequency may actually need more calories and fluid.

8. Burns

Children with significant burns have increased calorie needs and may need increased calcium, magnesium vitamin D, zinc, and other micronutrients.

Burn injuries cause a hypermetabolic state that can last up to 12 months after injury due to chronic inflammation, stress hormones, and elevated circulating catecholamines.

Carbohydrates should account for 60% of energy requirement, 20% should be from fats, and 2.5 to 4 g/kg per day of protein should provide the remaining 20%. Overfeeding and excessive carbohydrate administration can also be harmful, leading to hyperglycemia, impaired wound healing, increased fat stores, and increased CO2 production leading to delayed ventilator weaning. The preferred route of nutritional support is enteral because of its role in maintaining intestinal integrity. If enteral feeds are not tolerated, parenteral nutrition can be provided.

Severe burn injury can lead to additional requirements of vitamins, minerals, and trace elements. Hypocalcemia and hypomagnesemia can occur even when calcium and magnesium supplementation is administered, possibly from an upregulation of the parathyroid gland calcium-sensing receptor.

Hyperkalemia can occur in burn injury from cell lysis and can cause arrhythmia. Hyperlipidemia can occur because of lipolysis in a catabolic state. Hypophosphatemia can be seen as a nutritional deficiency in burn injury.

9. Allergies

Restrictions in diet in response to multiple food allergies can lead to poor nutrition status and deficiencies in calcium and vitamin D. Height for age percentile is lower in children with multiple food allergies.

10. Athletes

Sports drinks help with hydration, electrolyte balance and glucose levels for athletes exercising more than one hour without side effect. However, the caffeine content of energy drinks has the potential for hypertension and other cardiovascular or behavioral side effects. Protein supplements have not been shown to be effective and have not been studied for safety.

Pediatric athletes are at risk or malnutrition, especially with restricting consumption to maintain a certain body figure for sports such as gymnastics, cheerleading, diving, distance running or weight lifting.

11. Vegetarians

Vegetarian and vegan diets always require the supplementation of vitamin B12 (since it is primarily in meats).

Folate can be obtained through enriched starch products, green leafy vegetables and orange juice.

Omega-3 fatty acids can be obtained through eating walnuts, flax seed, dark greens and tofu.

F. Malnutrition

Caloric deficiency: Bradycardia and acidosis (Marasmus)

Protein-calorie deficiency: Alopecia, abdominal distention, acidosis (Kwashiorkor)

Fat malabsorption: Pale and malodorous stools, Elevated PTT (CF; chronic liver disease)

Protein losing enteropathy: Diarrhea, and edema (Bowel infections, Celiac, IBD, CHF)

Increase caloric need: FTT (Chronic lung disease)

G. Obesity

Moderate exogenous obesity is due to a greater caloric intake than needed for metabolic needs and a sedentary lifestyle; generally tall for age (height ≥50th percentile due to excessive caloric intake).

Endocrine causes of obesity include hypothyroidism, Cushing syndrome, GH deficiency, craniopharyngioma, insulin resistance, Turner syndrome, pseudohypoparathyroidism, polycystic ovarian syndrome (PCOS); usually small for age (height < 50th percentile or poor height velocity).

Genetic risk factors

Defect of leptin signaling pathway

Strong correlation between BMI of parents and biological offspring vs. adoptees and adoptive parents; twin studies show similarity of BMI is about twice as great among monozygotic than dizygotic twins.

Complications: cardiovascular (e.g. hypertension), respiratory, sleep, endocrine (e.g. type 2 diabetes, metabolic syndrome, PCOS, dyslipidemia), orthopedics, neurological, psychological, gastrointestinal, & skin problems.

Differential diagnosis: Cushing syndrome, Diabetes mellitus type 2, fatty liver disease, GH deficiency, hypothyroidism, PCOS, medication side-effects like hormonal contraceptives, psychotropic drugs, anticonvulsants or antihypertensive drugs, or genetic diseases like Prader-Willi syndrome, Bardet-Biedl Syndrome, Cohen Syndrome or Fragile X Syndrome.

Children in the overweight category (BMI 85th to 95th percentile) should be seen quarterly with dietary counseling. Children in the obese category (BMI >95th percentile) should be seen monthly for evaluation of lifestyle modifications and dietary counseling, focusing on portion control and

decreased sedentary time. Motivational interviewing can be used as a tool to encourage these lifestyle modifications. Early intervention leads to long term decrease in morbidity.

Predictors of adulthood obesity include parental obesity and obesity in adolescents. Consequences of obesity include hypertension, diabetes, coronary artery disease, hypercholesterolemias and other dyslipidemias, left ventricular hypertrophy, obstructive sleep apnea, increased asthma severity, mechanical stress on the joints, slipped capital femoral epiphysis, Blount disease, pseudotumor cerebri, hepatitc steatosis, cholelithiasis, gastroesophageal reflux, insulin resistance with acanthosis nigricans, depression or low self-esteem.

Effective interventions for adolescent obesity include portion control diets and reduction in sedentary activities in a highly motivated adolescent who expresses readiness for change, as well as bariatric surgery in extreme cases. Ineffective interventions include fad diets or using body dissatisfaction as a motivator for weight loss.

Lifestyle choices that may contribute to obesity include inadequate physical activity and excessive "screen" time, (TV, computer).

BMI levels are important because they correlate with body fat and with coexisting health risks in the pediatric population.

Definitions: BMI between 85th and 94th percentile for age and sex is considered at risk of overweight, and BMI at or above 95th percentile is obese.

Environmental risk factors of obesity:

-Risk of persistence of obesity into adulthood higher among obese adolescents; adolescents engaging in high-risk behaviors (smoking, alcohol use, and early sexual experimentation) have greater risk for poor dietary and exercise habits.

-Extent and duration of breastfeeding are inversely associated with risk of obesity due to physiologic factors in human milk and nursing patterns.

-Timing of introducing solid foods or high protein intake may have effects on childhood obesity.

-Early puberty and menarche is associated with degree of overweight, possibly due to insulin resistance (twofold increase in rate of early menarche associated with BMI greater than 85th percentile).

Counseling a family with regard to obesity prevention and treatment includes: Routine assessments of eating and activity patterns.

Advocacy for obesity control by increasing awareness of food choices, activity levels, smoking cessation, and breastfeeding.

Promotion of fitness programs in schools, in after-school care centers/daycares.

Other interventions: Medications like sibutramine (SSRI) [withdrawn from the market in 2010 due to associated side effects] for adolescents >16yo, orlistat (lipase inhibitor) for patients >12 years, very-low calorie diet, and bariatric surgery (gastric bypass or banding).

AAP recommends children older than 2 years watch < 1 – 2 hours of quality TV per day.

Lifestyle choices which may contribute to obesity include insufficient physical activity, too much "screen time" at the TV or computer, the expense involved in many recreational activities, choosing high calorie-low nutrient foods, and excessive portion sizes early in life.

Calculation of BMI: ratio of weight in kilograms to the square of height in meters.

Adolescents with BMI > 95th percentile or between 85th and 95th percentile with additional risk factors should receive in-depth medical assessments for the sequelae of obesity.

BMI = (weight in kilograms)/height in meters2

Weight loss

Types of "fad" weight loss diets:

Lacto-ovo-vegetarian – omit meat, fish, poultry; milk and egg allowed Lacto-vegetarian – omit meat, fish, poultry, egg; milk allowed

Strict vegetarian – no animal products

Zen macrobiotic – vegetarian diet with progressive restriction to finally only cereal grains and limited water

Fruitarian – fruits, honey, nuts, & olive oil

Feingold diet – omit foods containing additives, salicylates and artificial colors Protein-sparing modified fast – mainly proteins, very low calorie

Megavitamin regimen – 10-100 times required doses of Vitamin B & C

Adverse effects of "fad" diets:

Deficient in nutritional ingredients required for growth & development of the child/adolescent.

The more restrictive the diets, example Zen macrobiotic or fruitarian, the more deficiencies exist in essential nutrients like calcium, iron, ascorbic acid, Vit B12 and Vit D.

Cereal and vegetarian proteins vary in essential amino acid proportions and are not suitable substitutes for milk and egg. In contrast, increased animal protein consumption is strongly associated with adult coronary disease, just as fat consumption.

Preventive Pediatrics

Immunizations
Indications and schedules

Immunization schedules: at birth, 2-, 4-, 6-, 12-, 15-, 18-months, and 4-6 years & 11-12 years.

Preterm infants should be immunized at the same postnatal age (i.e. the chronologic age) as full-term infants. Vaccine doses should not be reduced for preterm infants.

Indications and the schedule for hepatitis B vaccine:

Hepatitis B spread through body fluid contact with infected person or a contaminated object where the virus can survive up to 7 days.

Vaccine schedule: 3 doses of the vaccine at birth, 1 to 2 months, and 6 to 18 months. Anyone who is not vaccinated can receive the vaccine in 3 doses (1st dose, then 2 months later, then 6 to 12 months from the first dose).

Chronic hepatitis is more common among infants and children, who were affected via transplacental exchange or body fluid contact and have not received Hep B vaccine and Hep B immune globulin at birth.

The risk of being a "chronic carrier" is inversely proportional to age, i.e. the younger the age at the time of initial infection, the greater the chance of developing chronic hepatitis.

Indications and the schedule for the inactivated influenza (killed) vaccines:

Rates of infection are highest among children, and severity is worse among children, pregnant women, immunocompromised individuals, therefore these individuals have the highest priority for influenza immunization.

Due to mutation of the influenza viruses, annual vaccination is recommended. Can take up to 2 weeks for the vaccine to take effect; protection will last for approximately 1 year. Influenza season typically runs from October to May; optimal vaccination during this time is recommended.

May contain thimerosal in multiple dose vials; thimerosal-free formulations are available as well.

Children with history of anaphylactic reaction to eggs should not receive the vaccine.

Both TIV and LAIV are produced in eggs. TIV is well tolerated in recipients who have egg allergy if a single, age-appropriate dose is well tolerated.

Data are not available to support use of LVAIV in people with egg allergy.

Children 6 months and older should get the vaccine. Children < 9 years require 2 doses of the vaccine during their initial season undergoing vaccination to be protected, then one dose per year. Children > 9 years require 1 dose each year.

Indications & schedule for live attenuated/weakened (intranasal) influenza vaccines (LAIV):

Recommended for healthy patients from age 2 through 49 years, not for pregnant women or immunocompromised people; Children from 6 through 23 months of age should not get LAIV.

LAIV does not contain thimerosal or other preservatives.

Children <5 years with asthma or reactive airways (with wheezing) should not get LAIV. Children/adolescents on long-term aspirin treatment should not get the vaccine.

Indications and the schedule for pneumococcal vaccines: Pneumococcal Conjugate Vaccine 13 (PCV13):

New conjugate vaccine protecting against 13 of the 90 types of Streptococcus pneumoniae instead of the PCV7.

To protect against sepsis and meningitis in infants, toddlers, & immunocompromised

Given as a series of 4 doses at 2, 4, 6 and 12 - 15 months.

Children who began immunization series with PCV7 or who have completed the 4-dose series with PCV7 should receive an additional dose of PCV13.

Immunized children with sickle cell disease, asplenia or partial splenectomy, diabetes, HIV/AIDS, chronic heart or lung disease or on immunosuppressive medications should get an extra dose of PCV13 between the 2nd and 6th birthdays. On February 20, 2013, ACIP recommended children 6 to 18 years old with immunocompromising conditions, functional or anatomic asplenia, CSF leaks or cochlear implants who have not received one dose of PCV 13 should receive one dose now that PCV13 has been FDA approved in individuals 6-17 years old.

Pneumococcal Polysaccharide Vaccine 23 (PPSV23):

Offers protection against 23 types of Streptococcus pneumoniae bacteria.

Should be given to anyone over the age of 2 through 64 years if they are immunocompromised, on immunosuppressive medications, or with long-term health conditions.

Usually only one dose is required; however, in patients with asplenia, sickle cell disease, HIV/AIDS, bone marrow or organ transplant, cancers, or on

immunosuppressive medications, a second dose can be given 5 years after the first dose.

Indications and schedule for tetanus vaccine (e.g., routine booster and at the time of injury):

3 types of tetanus immunizations are available: Children < 6 years receive a 5-dose series of DTaP vaccinations starting at 2months of age, children >7 years, Tdap is recommended as a single booster dose usually around ages 11 or 12, and a Td booster dose is recommended every 10 years for adults >19 years of age.

Patients with puncture-type wounds or burns require either Td or Tdap as prevention.

Pregnant women who are not immunized should receive a dose of Tdap after the 20th week of gestation or during the third trimester or soon after delivery.

Indications and schedules for diphtheria, tetanus, and pertussis vaccines:

Diphtheria, caused by Corynebacterium diphtheriae, forms a thick, gray-black coating at the back of the throat causing respiratory problems. Incubation period is 2 to 5 days. Treatment is diphtheria antitoxin, IV hydration and antibiotics.

Pertussis, caused by Bordetella pertussis, is a highly contagious upper respiratory infection with uncontrollable, violent coughing. In infants, pertussis can cause permanent respiratory damage, and even death, if left untreated. Incubation period is approximately 7 days, with severe coughing episodes occurring within 10 - 12 days of exposure. Treatment with macrolides is usually recommended to reduce the spread of disease, and is important to treat family members.

Children < 6 years receive a 5 dose series at 2, 4, 6, 15 - 18 months, and 4 - 6 years.

Indications and schedule for mumps vaccine:

Incubation period is 12 - 24days. Some complications include deafness, meningitis, orchitis in males, painful ovarian swelling in females, and rarely sterility.

MMR vaccine given to children 12 – 15 months and a second dose at 4 – 6 years. Contraindicated in patients with allergies to Neomycin.

Indications and the schedule for the rubella vaccine:

MMR vaccine is given to children 12-15 months and a second dose at 4-6 years. Children younger than 12 months can receive the vaccine if they are traveling out of the country.

Contraindications of the (live virus) rubella vaccine:

Anaphylactic reaction or hypersensitivity to neomycin or another component of

MMR vaccine.

Pregnant women must wait till after delivery to receive the vaccination. Women should avoid getting pregnant for 4 weeks after an MMR vaccination.

Patients in immunocompromised states with leukemias, lymphomas, blood dyscrasias, or bone marrow neoplasms, acquired or primary immunodeficiency syndromes like HIV/AIDS, or post-transplant patients, or patients on immunosuppressive therapies.

Indications and the schedule for the measles vaccine:

The rates of measles infection are on the rise as parents choose not to vaccinate from unfounded fears that they cause autism.

Complications include subacute sclerosing panencephalitis (SSPE), which usually manifests by age 2 with progressive personality changes, seizures, myoclonus, ataxia, and coma.

MMR vaccine should be given to children 12-15 months and a second dose at 4-6 years.

Indications and the schedule for the poliovirus vaccine:

Polio, caused by poliovirus, is spread by fecal-oral route. Incubation period is 7-14 days.

Immunization for children < 6 years is a 5 dose series at 2-, 4-, 6-, 18 months, and 4-

6 years.

Two vaccination forms exist: Inactivated polio vaccine (IPV) and live oral polio vaccine (OPV). IPV, used in the US since 2000, is usually combined with DTaP and Hib and given at 2-, 4-, 6-, and 18-months with a booster at 4-6years. Children may receive one extra dose of IPV with the "combination" vaccines; this does not cause any problems. OPV has not been used in the US since 2000 to avoid the risk of developing vaccine-associated paralytic poliomyelitis (VAPP); however, it is still used in polio-endemic regions of the world due to its benefits of intestinal immunity and secondary spread of the vaccine to unprotected contacts. OPV is contraindicated in pregnant women, immunocompromised patients, or those with persistent diarrhea or vomiting.

If patients are allergic to neomycin, streptomycin, or polymyxin B, they should not receive the IPV vaccine.

Indications and schedule for H. influenzae type b vaccine:

Haemophilus influenzae is a Gram-negative coccobacillus that possesses a polysaccharide capsule serotyped into 6 types (a-f); Hib as the most virulent strain

causing sepsis, cellulitis, pneumonia, meningitis, and epiglottitis.

Incubation period is unknown. Complications from Hib meningitis include partial or total sensorineural hearing loss, developmental delay, language delay, behavioral abnormalities, ataxia, seizures, and hydrocephalus.

Vaccine schedule: at 2-, 4-, 6-, and 12-15 months. It can be given as a "combination" vaccine with DTaP and IPV. Children less than 6 weeks of age should not get Hib vaccine.

Vaccines for families traveling to foreign countries may require > 1 month inducing a sufficient immune response, depending on the vaccine and the number of doses in the series.

CDC website has all necessary information for traveler's vaccinations based on The Red Book recommendations.

Indications and schedule for varicella vaccine:

Chickenpox, caused by Varicella-Zoster virus, where the patient is contagious 1-2 days before rash appears. Incubation period is approximately 10-21 days from exposure. Complications include myocarditis, Reye's syndrome, pneumonia, and shingles in adulthood.

Vaccine schedule: 1st dose by 12-15 months of age and 2nd dose by 4-6 years. Children > 13 years who have not been vaccinated should get 2 doses at least 28 days apart.

Contraindications include pregnancy, patients with anaphylactic reaction to previous varicella vaccine or to gelatin or neomycin, immunodeficiency syndromes, or receiving transfusions.

Indications and schedule for hepatitis A vaccine:

Persons who should be vaccinated include children between 12 to 23 months, travelers to disease endemic areas (Central or South America, Mexico, Asia, Africa, & Eastern Europe), children/adolescents who live in states or communities with high disease incidence, drug abusers, and patients with chronic liver disease or with clotting factor deficiencies.

Vaccine schedule: For children, 1st dose is between 12 to 23 months of age and 2nd dose at least 6 months apart. Unvaccinated children >2 years can still be vaccinated at later visits. Vaccine is not available for children <1 yr.

Contraindications include anaphylaxis to previous dose of Hep A vaccine or to a component of the vaccine esp. latex allergy (Hep A vaccines contain alum and sometimes 2-phenoxyethanol). Caution if pregnant.

Indications and the schedule for the meningococcal vaccines:

Two vaccines available: Meningococcal conjugate vaccine (MCV4) for persons between 9 months-55 years and Meningococcal polysaccharide vaccine (MPSV4) for persons >55 years.

Prevent against only 4 types of meningococcal disease.

Vaccine schedule: 2 doses of MCV4 for adolescents from 11-18 years with 1st dose at 11-12 years and booster dose at 16 years. Adolescents with HIV/AIDS should receive 3 doses, 1st and 2nd doses should be 2 months apart given at 11-12 years and a booster dose at 16 years.

Contraindications include allergic reactions to previous vaccine doses or components, severely ill patients, or with other vaccines in children with sickle cell disease or splenectomy. No contraindication for pregnant women to receive the vaccine.

Recommendations and schedule for rotavirus vaccine Rotavirus vaccine is an oral vaccine.

Vaccine schedule: 2-, 4- and 6-months (if needed); may be given with other vaccines. The 1st dose can be given as early as 6weeks of age, and the last dose should be given by 8months.

Contraindications include patients with severe reaction to previous dose of rotavirus vaccine, anaphylaxis to a component of the vaccine, history of intussusception, severe combined immunodefiency (SCID), HIV/AIDS, cancer, or on immunosuppressive therapies.

Recommendations and schedule for the human papillomavirus vaccine:

HPV vaccines can protect both males and females against 4 strains of HPV, 6, 11, 16 & 18. Vaccine schedule: given as series of 3 injections at 0, 2 and 6 month intervals.

Girls and women: Two vaccines are available, Gardasil and Cervarix, which protect against 2 HPV strains, types 16 & 18, causing 70% of cervical cancers. Gardasil also protects against HPV types 6 & 11. Available for girls from age 9-26years, but the recommended age is 11-12 years. All three doses of the vaccine are required to be most effective. For sexually active girls and women, yearly Pap smears must be done to check for changes in cervical mucosa.

Boys and men: Gardasil is available for males to protect against genital warts and cancers. It should be given at age 11-12years, but can be given up to 26years of age.

Contraindications include pregnant women, patients with allergic reactions to previous dose or components of the vaccine. Women breast-feeding can receive the vaccine.

General contraindications

Immune deficiency

Those that should not receive a live-virus vaccine include patients with leukemias, lymphomas, blood dyscrasias, or bone marrow neoplasms, acquired or primary immunodeficiency syndromes like HIV/AIDS, or post-transplant patients, or patients on immunosuppressive therapies.

Egg allergy

Not all egg-based vaccines (which include influenza, MMR, rabies, and yellow fever vaccines) are contraindicated for children after an anaphylactic reaction to eggs:

Influenza and MMR vaccines are cause for concern for allergic reactions due to their recommended administration to children younger than age 2 years when egg allergy is most common.

A child who has a known history of egg allergy should receive vaccine based on severity of allergy and clinical indication.

Measles component of MMR is grown in chick fibroblast cultures, which do not contain egg antigens. Most egg-allergic children can be vaccinated routinely with MMR as a whole dose.

Yellow fever vaccine has the most potential risk for allergic reactions as it is grown in chick embryos that are homogenized, thereby containing about 10-fold the amount of egg protein as does the influenza vaccine. Children requiring the yellow fever vaccination should be referred to an allergist.

Rabies vaccine presents very little risk to egg-allergic individuals. Of the three available rabies vaccines, only one is grown in chick fibroblasts and contains picogram amounts of egg protein.

Patients with a history of anaphylactic reaction to eggs should generally not receive inactivated influenza or yellow fever vaccine.

Prevention by active immunization

Influenza vaccine

Vaccine contains antigens for influenza A and B and should be administered yearly due to changes in antigens, especially to children with CHD, CF, BPD, and post- transplant patients.

Safety of the inactivated influenza vaccine includes lack of significant neurologic complication and non-communicability.

Meningococcal vaccine

Serotypes contained in the meningococcal vaccine include strains A, C, W-135, and

It does not cover serotype B, which accounts for 50% of infant cases and <20% of teen cases. Reasons for not including serotype B in vaccine:
B has poor immunogenicity in vaccine
Risk of cross-reactivity with neural tissue

Pneumococcal vaccine

Either conjugated or non-conjugated and multivalent, i.e. acts against several types of

Streptococcus pneumoniae bacteria.

The polysaccharide pneumococcal vaccine is less immunogenic in children younger than 24 months.

Benefits of pneumococcal conjugate vaccine include prevention of pneumococcal pneumonia and pneumococcal meningitis; however, it is less effective in prevention of otitis media due to other organisms that may cause OM.

The pneumococcal conjugate offers protection against the serotypes included in the vaccine but not those which are not included in the vaccine.

Hepatitis vaccines

Newborn infants exposed to hepatitis B infection via maternal placental transfer are at greatest risk for infection and becoming chronic carriers of the disease with a greater chance of developing chronic hepatitis infection.

Hepatitis B vaccine is made of a recombinant DNA-produced HBsAg. Premature infants weighing <2000g depend on maternal HBsAg status:

If maternal HBsAg is positive, CDC recommendation is to administer Hep B

immunoglobulin and a single-antigen Hep B vaccine within 12hrs of birth. The infant should still receive 3 additional doses of the vaccine at 2, 4, and 6 months of age, and then be tested for HBsAg and HBsAb 1-2 months after the completion of the hep B vaccine series.

If maternal HBsAg status is unknown, follow above directions.

If maternal HBsAg is negative, the 1st dose of Hep B vaccination can be given at 1month of age or upon discharge from hospital, and patient will complete series with 2, 4, and 6 month vaccinations.

Tetanus vaccine

Tetanus toxoid is available alone or in various combinations with diphtheria toxoid, acellular pertussis, and IPV, Hep B and Hib vaccines.

Permanent immunity does not result from C. tetani infections treated with antitoxin. Hence, updated immunization is necessary for prevention of tetanus infections.

Adverse effects of excessive tetanus immunization include serum sickness, brachial plexus neuropathy, encephalomyelitis and transverse myelitis, which have rarely been reported in association with tetanus vaccination. More frequent boosters may

lead to severe local and systemic reactions.

Diphtheria-tetanus combination

The difference between DT and Td: DT does not contain pertussis, and is used as a substitute for DTaP in children <6years who cannot tolerate pertussis vaccine. Td is a tetanus-diphtheria vaccine given to adolescents and adults as a booster shot every 10 years, or after an exposure to tetanus under some circumstances.

Pertussis vaccines (cellular and acellular)

In 1991, safety concerns of the cellular pertussis vaccines led to the development of a more purified (acellular) pertussis vaccine that has fewer side effects and contains less endotoxin than whole-cell pertussis vaccines.

Composition: chemically or genetically detoxified pertussis toxin and one or more bacterial components such as filamentous haemagglutinin (FHA), 69kDa outer- membrane protein known as pertactin, and fimbrial-2 and fimbrial-3 antigens in different concentrations.

The pertussis vaccine is80-85% effective at the completion of the primary series, and many who did still become infected with pertussis have a milder course. Possible complications of the vaccine include fever and hypotonic-hyporesponsive episodes which were more common with the whole cell vaccine.

Contraindications: patients with unstable or active CNS disease, immediate anaphylaxis, or encephalopathy within seven days.

Use of the pertussis vaccine is not contraindicated in stable CNS disease, family history of seizures, SIDS in a sibling, or low-grade fever.

DTaP and Tdap vaccines

Administering decreased volumes of the DTaP vaccine because of prior reactions is inappropriate.

Measles vaccine

The appropriate use of the measles vaccine during an outbreak:

In outbreak situations, local health departments may recommend measles vaccination of infants between 6-12 months of age, even though they may fail to respond to the measles component of the vaccine.

Infants who were immunized for measles when younger than 12 months of age may not be properly immunized and may be susceptible to measles or its complications later in life. These infants should receive MMR vaccine at 12-15months followed by the 4-6yr revaccination.

The contraindications of the (live virus) measles vaccine include:

Pregnant women, patients with anaphylactic reactions to eggs or neomycin, patients in immunocompromised states with leukemias, lymphomas, blood dyscrasias, or bone marrow neoplasms, acquired or primary immunodeficiency syndromes like HIV/AIDS, post-transplant patients, or patients on immunosuppressive therapies.

Administration of a single dose of MMR vaccine induces ⩾95% immunity when administered at age 12–15 months. Since measles is highly contagious in school settings and in crowded urban areas, the 2-dose strategy is in effect with the 1st dose given between 12-15months and the 2nd dose at 4-6years prior to beginning school.

Rubella vaccine

Although the rubella vaccine is not recommended for use in pregnant women, there has never been a reported case of congenital rubella syndrome caused by the vaccine virus.

Poliovirus vaccine

The efficacy of the poliovirus vaccine:

IPV is highly effective in immunizing against the polio virus, and protecting from paralytic poliomyelitis. 90% or more of vaccine recipients develop protective antibody to all three poliovirus types after 2 doses of the vaccine, and at least 99% are immune following 3 doses. However, duration of immunity is unknown.

OPV, also highly effective against poliovirus, produces immunity in 50% of patients who received a single dose of the vaccine. Three doses produces immunity to >95% of recipients; Immunity is possibly life-long as it is a live virus vaccine.

The oral polio vaccine can cause poliomyelitis in a minority of persons receiving the vaccine. The inactivated polio vaccine does not cause vaccine-associated paralytic poliomyelitis, but can cause local reactions, and in individuals sensitized to streptomycin, polymyxin B or neomycin, an allergic reaction can occur.

Poliovirus is transmissible by fecal-oral route.

It is important to know whether everyone in the home has been immunized against polio, as unimmunized persons would put the family at a greater risk during the rare polio outbreaks.

Hemophilus influenzae type b vaccine

Composition and use of the H. influenzae type b vaccines:

Hib is composed of Hib polysaccharide (purified polyribosylinhobitol phosphate) bound with protein carriers.

Recognize the changes of epidemiology of H. influenzae infection secondary to widespread use of the vaccine.

Varicella vaccine

Indications for the use of varicella vaccine after exposure: those who have not been immunized should receive varicella vaccine after exposure to chickenpox. Vaccine should be given 3-5 days after exposure, thereby preventing chickenpox or making it less severe.

Catch-up immunizations

Plan an immunization schedule for a patient born three months prematurely. Vaccination for premature infants should proceed according to chronologic (not adjusted) age with the exception of hepatitis B in infants weighing less than 2,000 g, where immunization is dependent on maternal hepatitis B status.

The Advisory Committee on Immunization Practices (ACIP) supports rotavirus vaccination of premature infants if they are at least six weeks of age, discharged from the NICU, and clinically stable even though very low birth weight premature infants can have increased adverse reactions because of lower maternal antibody to rotavirus.

Respiratory syncytial virus immune globulin should be given to premature infants

<34weeks gestational age. Palivizumab (Synagis), licensed to prevent RSV infection, is administered once per month for five months beginning in early November and ending in March. The five doses should provide adequate coverage for the RSV season.

Plan an immunization schedule for children or adolescents who begin their immunizations late or whose immunizations are delayed:

CDC has a catch-up schedule for those children or adolescents who have delayed immunizations. There is no need to restart a vaccine series regardless of the time that has elapsed between doses. Please see catch-up schedule on CDC website:
http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6105a5.htm.

Live vs. killed vaccines

Vaccines that are live include MMR, yellow fever, live influenza virus, varicella, oral poliovirus, BCG and typhoid. Vaccines that are killed include inactivated influenza virus, inactivated poliovirus, inactivated pertussis, cholera, and rabies.

Screening

Principles of screening tests
Blood pressure

Blood pressure should be measured during every health care visit in children 3 years and older. Indications for the mandatory blood pressure check in children younger than 3 years are children with history of prematurity, congenital heart disease, renal disease, solid organ transplant, cancer, taking drugs known to cause high blood pressure, other illnesses associated with hypertension or evidence of raised intracranial pressure.

Average systolic blood pressure (SBP) and /or diastolic blood pressure (DBP) that is

≥95th percentile for age, sex and height on >3 occasions is diagnosed as hypertension. Average SBP or DBP that are ≥90th but <95th is considered as prehypertension. White coat hypertension is recognized when BP is ≥ 95th percentile only at a medical setting but normal outside.

Blood pressure has to be measured at repeat visits before diagnosing hypertension in children.

Blood pressure cuff size should be appropriate for patient size in order to prevent over diagnosis as inappropriate cuff may give falsely elevated BP reading.

Hematocrit

There is usually a difference between a centrally and a peripherally drawn hematocrit, with capillary samples being as much as 15% higher than venous samples.

Lead

Capillary blood obtained by finger stick is the initial screening test for lead level, but venipuncture is more accurate.

Screening examination for lead should be incorporated into early periodic screening at 1 and 2 years of age.

Children may have multiple non dietary sources of exposure to lead mostly from the environment.

Lead poisoning may cause serious cognitive impairment.

Effects of chronic mildly increased blood lead concentrations include but not limited to low IQ, poor concentration, impaired hearing and balance.

Lead concentration more than 45 microgram/dl requires treatment with succimer. Lead level higher than 70 microgram/dl merits treatment with EDTA and hospitalization.

Hearing

Conductive hearing loss results when the mechanical transmission of sound in the outer ear, tympanic membrane, or middle ear is impaired.

Common causes of conductive hearing loss in external ear are atresia or stenosis, cerumen or foreign body impaction. The causes in middle ear are perforated tympanic membrane, otitis media with effusion, cholesteatoma, osteosclerosis and discontinuity or fixation of the ossicular chain.

The natural history of conductive hearing loss depends upon the etiology, but if a source of the conductive hearing loss is left untreated, language delays will occur.

Brain stem evoked response audiometry involves electroencephalographic response to auditory stimulus.

Vision

Infants are capable of differentiating colors and patterns and also able to recognize facial expressions.

Cataracts, retrolental fibroplasia, retinoblastoma may be detected by periodic ophthalmoscopic examinations.

Ocular tropias and phorias using a cover test.

Cover tests (cover-uncover test and alternate cover test) are done to screen for strabismus in children. The cover-uncover test is used to differentiate tropias from phorias.

Hypercholesterolemia/hyperlipidemia

Indications for evaluation for hyperlipidemia are: children with positive family history of coronary artery disease before age 55, children whose parent has cholesterol>240mg/dl or children with high risk whose family history is unobtainable.

Clinical manifestations of hypercholesterolemia could be skin (hands, elbow,

knee, buttock) or tendinous xanthoma and corneal arcus.

Risk factors for hypercholesterolemia/hyperlipidemia are obesity, hypertension, type II DM, cigarette smoking, and concentration of low-density lipoprotein (LDL), a low concentration of high-density lipoprotein (HDL) and family history of premature cardiovascular disease (CVD) or high blood concentrations of cholesterol.

Disease prevention

Heart disease

Risk factors associated with coronary artery disease are increasing age and male gender, tobacco use, sedentary lifestyle, diet rich in saturated fat, obesity and a positive family history. Children born large for gestational age and low birth weight infants are at increased risk for developing congenital heart disease later in life.

Osteoporosis

To prevent osteoporosis children need to have diet rich in calcium (>1200mg/day in adolescent), vitamin D (400-800 IU daily) and encouraged to do weight bearing exercises. Examples of excellent source of calcium are dairy products, bony fish, green vegetables and calcium fortified drinks. Children with lactase deficiency can take yogurt and cheese to meet need for calcium. Tobacco and alcohol are risk factors for osteoporosis.

Respiratory disease (e.g., smoking)

Children exposed to passive cigarette smoke in the home have higher chances, frequency, and duration of lower respiratory tract illness.

Common indoor exposures like wood fires and stoves, cooking spray, hair spray, animal dander, cigarette smoke can produce respiratory symptoms.

Anticipatory guidance

Safety
General

Ways of preventing head injury include wearing appropriate size helmet during

activities like riding bicycle, skiing, and skating etc.

Parents and caregivers should be aware of potential hazards in a child's environment and take safety measures to prevent potential unintentional injuries.

Automobiles

Parents should be instructed to keep their children in rear-facing car seats until age 2 or until they reach the maximum height and weight for their seat. Children who have outgrown the rear-facing

weight or height limit for their car safety seats should use a forward-facing car safety seat with a harness for as long as possible. Children will need to ride in a belt-positioning booster seat until they have reached 4 feet 9 inches tall and are between 8 and 12 years of age. Children should not ride in the front seat of a vehicle until they are 13 years old. When children are old enough and large enough to use the vehicle seat belt alone they should always use lap-and-shoulder seat belts.

The qualities of an adequate car seat includes the capability to reduce risk of ejection during a crash, ability to better distribute the energy load of the crash through bones rather than soft tissues, limit the crash forces by increasing the time of deceleration, and limit the contact of the occupant with interior vehicle structures.

Drunk driving is a major cause of automotive accidents among young drivers.

Premature infants have the possibility of oxygen desaturation often associated with bradycardia while restrained in a car seat.

Non-crash automobile accidents for young children may occur from being ejected from the back of a pickup truck. Riding in the back of a pickup truck predisposes to head injury and carbon monoxide poisoning from faulty exhaust system.

Stairways

Dangers of using walkers with wheels is not limited to falls, walkers can delay normal motor and mental development. A gate mounted to the wall should be installed in front of stairs.

Bicycles

Bicycle safety may be ensured by following traffic rules and safety guidelines like using protective gears such as helmets, mouth and wrist guards, knee and elbow pads, eye protection and personal protective devices.

Rollerblades, skateboards

Using safety equipment such as helmets, mouth and wrist guards, and personal protective devices during skating or using rollerblades is not negotiable.

Poison prevention

A six-month well-child visit should include a discussion on the storage of poisonous substances at home in anticipation of the infant becoming more

mobile in the following months.

Child resistant caps can prevent accidental ingestion of drugs

Burns

Use of smoke detractors and regulating water heater temperature plays a major role in reducing burn injuries. Also parents should test smoke detectors monthly, replace batteries yearly, avoid drinking hot fluids near children, never leave the stove unattended, and cover electric outlets with protective devices, never smoke in bed and need to establish a fire escape plan. Matches, lighters, appliance cords, pot handles, grills and fireplaces must be out of reach of children.

Hot water heaters should be set at temperatures no higher than 120°F (48.9 ◦ C).

Bites and stings

Anaphylaxis from an insect sting should be managed with injection of intramuscular epinephrine if available and presentation to the emergency department for further evaluation and treatment.

The safe method of removing tick is by grasping the mouth parts with a pair of forceps at the site of attachment. Safe retraction may be achieved by applying gentle but steady pressure. After removal of tick the skin should be disinfected, hands should be washed to prevent accidental inoculations and tick should be either soaked in alcohol or flushed down the toilet. Twisting during removal should be avoided to prevent incomplete removal. Squeezing or crushing the tick must be avoided to prevent infection from fluid.

Parents should be informed about the risks of having potentially biting pets at home, about certain breeds that have higher inclination to inflict injuries and protective instincts of nursing animals. Parents should supervise children in presence of animals, and teach them to respect animals. Children should be cautioned that animals have the potential to inflict injury, and should not harbor exotic animals for pets.

Water safety

Drowning is one of the leading causes of unintentional injury and death in children in the U.S. More than 1100 children died from drowning in the year of 2005. Number of deaths from drowning peaks in the toddler age group and adolescents. The overall rate of drowning in African American children is 1.7 times higher than that for white children in the United States. Children younger than 1 year of age usually drown in bathtubs, buckets, and toilets. Unsupervised children 1 to 4 years of age often drown in swimming pools. In the adolescent and young adult age groups between ages 15 to 24 years, common site for drowning are in natural bodies of water.

Home pools should be enclosed on all four sides with fences at least 4 feet high and have self-closing gates.

Children should wear life jackets on boats and when playing near water and supervised closely (adult within one arm's reach of a child in or near water). Parents should inform children about water safety and provide swimming lessons.

Sun exposure

Children need to apply sunscreen cream/lotion when exposed to sun to prevent skin cancers. Babies under the age of 6 months need to be covered with clothes or kept under shades and may apply sunscreen only on the face and hands.

Firearms

Having firearms in the home increases the risk of adolescent suicide 3 to 10 fold and the risk of homicide up to 4 fold.

In US 35% of households own firearms. And 50% of homes have at least 1 firearm stored unsafely.

Firearms must be locked, unloaded with ammunition locked in a separate location.

Personal safety (e.g., strangers)

"Screen" time (e.g., TV, computer)

Children's "X" (e.g., TV, computer, cell phone, video games) time should be limited to 2 hours.

Sleep (e.g., SIDS, obstructive sleep apnea, normal patterns)

SIDS can be reduced when infants sleep on their backs and in the same room with

parents but not in the same bed.

A bed time routine helps to establish longer sleeping stretches. Children need to be put to bed at the same time consistently in a quiet and dark room. There should be a quiet hour before going to bed preferably reading a book together. No television should be allowed in the bedroom and bedroom should not be the site of punishment or timeout.

Parents often report snoring as a symptom of obstructive sleep apnea. Therefore, all children should be screened for snoring. Patients with cardiorespiratory failure must be evaluated. Polysomnography is used to differentiate between primary snoring and OSA. Primary treatment is adenotonsillectomy and continuous positive airway pressure when surgery is contraindicated. High risk patients need to be monitored for cardiorespiratory failure at an inpatient setting with adequate follow up after surgery.

Poisoning and Environmental Exposure to Hazardous Substances

General
Prevention and risks
Poison control centers, online resources

Poison control centers provide information on prevention of accidental ingestion and treatment of poisoning. If a child is unconscious, not breathing, or having convulsions or seizures due to poison contact or ingestion, 911 should be called. If a child has come in contact with poison or there is suspicion that a child may have swallowed a button- cell battery, and has mild or no symptoms, poison control center must be called.

Therapeutic options for poisoning

Management of poisoning includes reporting symptoms, careful physical exam, laboratory evaluation and diagnostic tests to identify toxin followed by decontamination, elimination, antidote and supportive care.

Recognition of ingestion of unknown substance(s)

In case of poisoning by an unknown agent history should include a list of all medications in the child's environment, location of poisoning, estimated time of ingestion and amount ingested.

Specific acute poisonings, ingestions, and exposures

Nonsteroidal analgesics

With acetaminophen toxicity there may be an asymptomatic period followed by nausea vomiting, anorexia, malaise, pallor and diaphoresis. The second stage includes right upper quadrant abdominal pain and tenderness. In the third stage fulminant hepatic failure, multisystem organ failure and death may occur.

Initial management of suspected or confirmed acetaminophen poisoning focuses on airway, breathing and circulation (ABC) and decontamination with activated charcoal if patient present within 1-2 hours of ingestion. The specific antidote for acetaminophen poisoning is N-acetylcysteine (NAC).

Opiates

Classic opioid toxidrome includes respiratory depression, sedation and miosis. Bradycardia, hypotension and hypothermia are signs of more severe toxicity. There is risk for prolongation of QTc interval and torsade de pointes.

Management of an opiate overdose can include respiratory support as well as cardiovascular support.

Anticholinergic substances

Signs and symptoms of anticholinergic substance exposure include diarrhea, urination, miosis, bronchospasm, bradycardia, emesis, lacrimation and salivation. Tremor, muscle weakness and fasciculation, hypoventilation, hypertension, tachycardia and dysrhythmia are the nicotinic manifestations of insecticide poisoning. Severe symptoms are coma, seizures, shock, arrhythmias and respiratory failure.

Salicylates

Early signs of salicylate poisoning are nausea, vomiting, diaphoresis and tinnitus. Moderate salicylate toxicity may present as tachypnea, hyperpnea, and tachycardia and altered mental status. Signs of severe poisoning include hyperthermia, coma and seizure.

There is no specific antidote for salicylate poisoning. Supportive care with antiemetic and acid blockers is main stay of therapy. Activated charcoal may be given if patient presents within 1-2 hours of ingestion.

Over-the-counter medications may have harmful additives (e.g. salicylate in antidiarrheal products.)

Antihypertensives

Signs and symptoms of ingestion of antihypertensive are bradycardia, hypotension, variety of dysrhythmias and heart block.

Tricyclic antidepressants

A danger of tricyclic antidepressant (TCA) treatment is accidental ingestion by siblings.

Cardiac dysrhythmias may be a later effect of ingestion of tricyclic antidepressants.

Signs and symptoms of TCA include delirium, mydriasis, dry mucous membranes, tachycardia, hyperthermia, mild hypertension, urinary retention

and slow GI motility. Cardiovascular toxicity may manifest as sinus tachycardia, wide QRS complex, premature ventricular contractions and ventricular arrhythmia. The CNS symptoms include lethargy, coma, myoclonic jerks and seizures.

Treatment of TCA toxicity is focused on maintaining airway and ventilator support as needed along with continuous cardiac monitoring and serial ECGs. Activated charcoal may be given for gastric

decontamination in appropriate situations. Specific antidote is sodium bicarbonate. Indications for using sodium bicarbonate are QRS duration >100 ms, ventricular dysrhythmias and hypotension. Norepinephrine is the vasopressor used in case of hypotension.

Ethanol

Ethanol intoxication may result in hypoglycemia.

Ethanol intoxication may mask signs of toxicity from other drugs.

Over-the-counter medications may have harmful additives (e.g. Alcohol in mouthwash, salicylate in antidiarrheal products).

Early signs of methanol ingestion are drowsiness, mild inebriation, nausea and vomiting.

The delayed effects are severe metabolic acidosis and visual disturbances which may result in irreversible blindness.

Specific antidote for methanol ingestion is Fomepizole. Hemodialysis is indicated if methanol level is >50mg/dl, metabolic acidosis, severe electrolyte imbalance and renal failure.

Hydrocarbons

Management of a child who has ingested a substance containing a hydrocarbon is largely supportive. Airway Breathing and Circulation (ABC), Charcoal is ineffective. Monitor signs of aspiration pneumonitis and arrhythmias.

Organophosphates

Signs and symptoms of organophosphate poisoning: Cholinergic Toxidromes – increase urination, defecation, salivation, lacrimation, bronchorrhea (bronchospasm), Miosis, bradycardia. (mnemonics: SLUDGEM/DUMBBELS).

Management of organophosphate poisoning include avoid further exposure, antidotes: Atropine and Pralidoxime.

Carbon monoxide

Signs and symptoms of carbon monoxide poisoning depend upon level of carboxyhemoglobin: shortness of breath, headache, nausea, vomiting, and skin bullae, cherry-red lips, altered mental status, seizure and coma. Management includes 100% O2 or Hyperbaric O2.

Acids, alkali, and alkaloids

Common household sources of acids are cleaning agents (e.g. toilet bowl cleaners), anti-rust compounds, etching compounds, automotive batteries, and pool sanitizers.

Common household sources of alkali are drain openers, automatic dishwasher detergents, alkaline batteries, toilet bowl cleaners, swimming pool products and radiator cleaning agents.

Gastric lavage is contraindicated in a caustic ingestion.

Corrosive material such as hydrochloric and sulfuric acids can be transported to the stomach with few or no esophageal burns, causing severe gastritis, perforation, or late stricture formation.

Endoscopy after a corrosive ingestion permits grading (on scale of o to 3) of the Injury and guide further management.

Button batteries

Management of a child who has ingested a button battery includes: Radiograph of neck, chest & abdomen. Button batteries lodged in esophagus must be removed endoscopically as soon as possible. Batteries detected in stomach usually traverse the gastrointestinal tract without incident, with >80% passing within 48 hours. In asymptomatic patient, at 48 hour battery location should be assessed radiographically and if object has not passed the pylorus, removal is recommended. In symptomatic patients, urgent endoscopic removal is indicated.

Coins

Management of child who has ingested a coin includes: Neck, chest & abdomen Radiograph. For an asymptomatic child with coin located in the stomach or small

bowel, no immediate management is required. Parents should be instructed to examine child's stool in 4-6 days. However, if nothing is recovered, a follow up radiograph at 2 weeks should confirm coin passage vs retention and guide further therapy. If coin is located in esophagus or child is symptomatic, indicating the need for urgent endoscopic removal.

Iron

Management of child with iron ingestion depends upon amount of elemental iron ingested. If patient is asymptomatic, ingestion < 40 mg/kg of elemental: observe at home.

In patient with significant ingestion (serum iron > 350 mcg/dl) Labs: CBC, CMP, serum iron; in a symptomatic patient, abdominal radiograph to look for retained tablets; If present, gastrointestinal decontamination using whole-bowel irrigation is indicated. Patients who have severe symptoms, anion gap acidosis, serum iron

> 500 mcg/, or a significant number of pills visible on abdominal radiograph should be treated with deferoxamine to chelate free circulating iron.

Ethylene glycol

Initial treatment is based on clinical suspicion (history, progressive lethargy, ataxia, seizures) before alcohol values are available. Indirect laboratory evidence of alcohol toxicity includes a high anion gap metabolic acidosis and a high osmolar gap greater than 10 mmol/L.

Initial treatment involves stabilization of vital functions, administration of sodium bicarbonate to correct acidosis, and administration of the antidote fomepizole (or ethanol, if fomepizole is unavailable).

Plants

Signs and symptoms following ingestion of some of the potentially toxic plants are:

Complementary and Alternative Medicine (CAM)

Complementary medications have often not been approved through the Federal Drug Administration's process to ensure certain standards for safety and efficacy, as well as establish accurate dosing. However, some of these medications may have side effects or affect the pharmacokinetics of other medications, so it is important to identify complementary medications being used by patients.

Over-the-counter medicines (e.g., cold remedies)

Over-the-counter cough and cold preparations have not been adequately studied in children younger than 6 years of age and that they are not recommended for use for the common cold.

The active ingredients and the common adverse effects and toxicities of over-the- counter cough and cold preparations are:

Pseudoephedrine, which may have sympathomimetic effects, such as tachycardia, arrhythmia, hypertension, central nervous system stimulation, and rarely seizures.

Antihistamines such as diphenhydramine, brompheniramine, chlorpheniramine, and carbinoxamine can lead to central nervous system agitation or depression, dysrrhythmias, hypertension, seizures, and in extreme cases, respiratory depression.

Guaifenesin, the most commonly used expectorant, is generally well tolerated but may cause mild gastrointestinal discomfort.

Dextromethorphan can cause euphoria, hallucinations, lethargy, coma, nausea, dizziness, drowsiness, ataxia, nystagmus, and urinary retention. It does have abuse potential because of the euphoric effects it may produce. In lower doses, codeine can lead to somnolence, ataxia, gastrointestinal distress, and pruritus; in higher doses, it leads to depressed mental status and respiratory depression.

Exposure to toxic substances in the environment

Age-related risk and impact of exposure

Infants are at greater risk than adults from toxic substances in the environment because infant and children eat more food, drink more water, and breathe more air than adults on a per-kilogram body weight basis. Consequently, if food supplies, water, or air contain a contaminant, exposed children receive a higher dose. Diet of young infants is limited to single foods (e.g., human milk or infant formula) if contaminated infant receive higher doses of contaminant.

Obtaining an exposure history

Obtain history of:

Habits and products consumption- tobacco, fish, occupational exposure to toxic substances, Hobbies involving toxic substances, organic versus conventional foods, Plastics: Bisphenol A (BPA) and phthalates.
Home: Water damage, leaks, floods, or mold growth; Active or recent renovation or remodeling in home built before 1978; Friable ceiling material or degraded insulation around pipes, boilers, and furnaces; Drinking water source; Carbon monoxide detector; Wood stove or fireplace use; Pesticides in home or on lawn/garden, storage.
Community: Near industrial site, hazardous waste site, landfill; near pesticide-treated agricultural fields; school and child care.

Contaminants in drinking water

Common contaminants potentially found in drinking water (e.g., E. coli, Cryptosporidium, trichloroethylene, perchloroethylene).

Contaminants in food

Common toxic substances that may contaminate food sources (e.g., mercury, E. coli).

Chemical hazards in the community

Common toxic substances that may contaminate the environment and affect the health of children (e.g., pesticides, industrial waste).

Chemical exposures in the home

Following are the potential occupational exposures that directly or indirectly affect the health of children:

Lead - Lead service lines or corrosion of plumbing systems leach lead into water, lead- containing paint may contaminate house dust or soil.

Arsenic- Natural sources in rock/sediment leach into water supplies, foods. Nitrite- Nitrate-containing fertilizers, sewage and septic tanks.

Pesticides- Conventional agriculture results in residues in foods and potential water contamination in agricultural areas.

Most common exposures that are associated with house renovation and repair are lead exposure. Low-level lead exposure is associated with a reduced intelligence quotient and behavioral problems, including attention deficit hyperactivity disorder (ADHD).

Exposures from terrorism (e.g., anthrax, smallpox)

Characteristic skin lesions of anthrax: boil-like skin lesion that eventually forms an ulcer with a black center (eschar). The black eschar often shows up as a large, painless necrotic ulcer.

Differentiate the skin lesions of varicella from those of smallpox.

Fetus and Newborn Infant A.Normal newborn infants

Delivery room management
Temperature control

Newborn infant is prone to heat loss because of a high surface area-to-body mass ratio.

Newborn infant who is cold stressed rapidly depletes essential stores of fat and glycogen.

Heat loss in the delivery room can be reduced by the use of a radiant warmer, drying, and swaddling.

Hazards and benefits associated with the use of radiant warmers for neonates. The hazards include hyperthermia, insensible water loss, and first degree burn. The benefit includes maintaining adequate body temperature.

Assessment

Normal newborn infant can fixate. Failure to do so by 2 month is pathological.

Components of the Apgar score are: A - Appearance – color, P – Pulse - heart rate, G

- Grimace - reflex irritability, A - activity – muscle tone, R – Respiration.

Significance of the one- and five-minute Apgar scores: <3 –critically low, 4 to 6- fairly low, 7 to 10- generally normal. A low score on the one-minute test may sugges6t that the neonate requires medical attention but is not necessarily an indication that there will be long-term problems, particularly if there is an improvement by the stage of the five-minute test. If the Apgar score remains below 3 at later times such as 10, 15, or 30 minutes, there is a risk that the child will suffer long-term neurological damage.

Fetal assessment

Non-stress test

Non-stress test monitors fetal heart rate reactivity in response to fetal activity.

Stress test

Stress test is used to evaluate uteroplacental insufficiency.

Biophysical profile

Factors used by obstetricians in evaluating fetal well-being are fetal activity, amount of amniotic fluid and heart rate.

BPP = non stress test + 4 more parameter monitored by ultrasound. (fetal breathing movement, gross body movement, fetal tone and presence of amniotic fluid pockets>2 cm).

Fetal arrhythmias: Although most fetal arrhythmias are benign, some types cause fetal hydrops and can lead to fetal death. Detailed analysis of the type of arrhythmia in utero is possible using M-mode and Doppler echocardiography.

Common causes of fetal tachycardia (ventricular heart rate >180 bpm), are paroxysmal SVT and atrial flutter. Most fetuses with tachycardia are successfully treated in utero by transplacental administration of antiarrhythmic drugs. Digoxin is widely accepted as a first-line antiarrhythmic drug. Sotalol, flecainide and amiodarone are used as second-line drugs when digoxin fails.

Fetal bradycardia is diagnosed when the fetal ventricular heart rate is slower than 100 bpm, mainly due to AV block. Common causes are associated congenital heart disease or maternal SS-A antibody. Beta stimulants and steroids have been reported as effective transplacental treatments for fetal AV block.

Maternal screening (see IX.I.30)

First trimester: ABO, Rh, hemoglobin, glucose, Iron, STD's (syphilis, hepatitis B, HIV, GC/Chlamydia), Rubella, Toxoplasmosis, Urinalysis, Ultrasound – crown rump length, first trimester screen (b-hcg, pregnancy-associated plasma protein – A [PAPP-A], nuchal translucency).

Second trimester: Quad screen (AFP, b-hcg, estriol, inhibin-A), ultrasound (to rule out or to evaluate for presence of cardiac anomalies).

Third trimester: non-stress test, biophysical profile, glucose tolerance test, Group- B strep screen.

Transition

Maturational assessment
AGA, LGA, SGA

Need to plot anthropometric measurements (height, weight, head circumference) against gestational age on a graph.

Preterm, term, postterm

Preterm infant is a baby born before 37 completed weeks of gestation. Different physical and neuromuscular characteristics that vary according to fetal age and maturity; Ballard score (physical and neuromuscular criteria for assessment of maturity and gestational age of infant).

Physical criteria mature with advancing fetal age, including increasing firmness of pinna of the ear; increasing size of the breast tissue; decreasing fine, immature lanugo hair over the back; and decreasing opacity of the skin.

Neuromuscular criteria mature with gestational age, including increase flexion of the legs, hips, and arms; increasing tone of the flexor muscle of neck; and decreasing laxity of the joints.

Distinguish between small-for-gestational age (SGA) and preterm gestation in low-birth-weight (LBW) infants:

SGA: Infants born who weighs <2500 g at birth and falls below the 10th percentile on the growth chart for gestational age are small-for-gestational age (SGA); they can be preterm or term.

LBW: defined as infant having birth weight <2500 grams. IUGR is the most common cause of LBW in developing countries, whereas prematurity (infant born at <37 completed weeks) is the cause in developed countries.

Physical and behavioral characteristics of a full-term infant: crackling, pale area, rare vein visible skin; creases anterior 2/3rd of sole; no lanugo; raised areola, small breast bud; ear formed and firm, instant recoil; testes down, good rugae in male scrotum and labia majora large and minora small in female.

Physical and behavioral characteristics of a post-term infant: cracked, dry, peeling skin; creases covering the entire sole of the foot; mature and long fingernail; absence of lanugo over the back; palpable breast bud; stiff cartilaginous ear with ready recoil, a well pigmented and rugated scrotum in male and labia majora that entirely cover labia minora and clitoris in female.

Routine care

General

Evaluation of an infant with physiologic breast hypertrophy: No evaluation required. Benign condition that resolves on its own. Neonatal breast hypertrophy occur due to constant exposure to maternal estrogen during pregnancy and after birth testosterone secretion in boys and estradiol secretion in girls surges due to neonatal hypothalamic-pituitary-gonadal axis activity.

Family members eager to resolve breast development by “extracting the milk” may prolong or exacerbate physiologic neonatal gynecomastia because breast stimulation promotes secretion of prolactin and oxytocin by the pituitary gland. Expressing milk from the neonatal breast also may result in mastitis or galactocele formation.

Vitamin K

Prophylactic administration of vitamin K will prevent classic hemorrhagic disease of the newborn.

Presenting signs and symptoms of classic hemorrhagic disease of the newborn: more common in breastfed infants, occurs in first few weeks of life. Rare in infants who receive prophylactic IM vitamin K on first day of life; It usually marked by generalized ecchymoses, gastrointestinal hemorrhage, or bleeding from a circumcision or umbilical stump; intracranial hemorrhage can occur, but is uncommon.

Maternal exposure to drugs that may affect coagulation can result in early hemorrhagic disease of the newborn.

Eye prophylaxis

Prophylaxis of ocular gonorrheal infection in a newborn infant should include silver nitrate solution in single-dose ampoules or single-use tubes of ophthalmic ointment containing erythromycin or tetracycline.

However, although silver nitrate is effective against N gonorrhoeae, it is ineffective against Chlamydia. In fact, silver nitrate may injure epithelial cells, thus rendering them more susceptible to other infectious agents. Therefore, erythromycin or tetracycline ointments are recommended for routine prophylaxis.

Silver nitrate and tetracycline ointment no longer available in the United States so erythromycin ointment is widely used for eye prophylaxis in newborn

Feeding requirements

Calories

Caloric requirement per kilogram for adequate growth is greater for preterm infants than for full-term infants.

Fluid

Preterm infants have a greater daily fluid requirement per kilogram than full term infants.

Insensible water loss is increased with prematurity and the use of radiant warmers.

Screening

General
Glucose

Rapid assessment of whole blood glucose concentrations for example, glucose oxidase test strips, may yield falsely high or low values.

Hematocrit

In the newborn, the normal hematocrit range is 54% – 68% on day 1, to 37% - 49% by week 4.

Preterm infants have lower hematocrit values than full-term infants.

In the full term infant, physiologic anemia occurs between 8-12 weeks of age when the hemoglobin is between 9-11g/dL.

In the preterm infant, physiologic anemia occurs by 3-6 weeks of age with hemoglobin levels decreasing to 7-9 g/dL.

Serologic test for syphilis

The presenting signs and symptoms of congenital syphilis are divided into early and late signs.

Early: During first 2 years of life: 30% - 40% are stillborn, 75% of liveborn infants are asymptomatic at birth.

General: Fever, weight loss, anorexia, fatigue.
CNS – elevated cell and protein in CSF.
HEENT – Chorioretinitis, glaucoma, syphilitic rhinitis or "snuffles."
Respiratory – pneumonia.
GI – hepatomegaly – most consistent feature, splenomegaly, jaundice, bile stasis, fibrosis, extramedullary hematopoesis, gastroenteritis, peritonitis, pancreatitis.
GU – testicular masses.
Heme – Coombs negative hemolytic anemia, thrombocytopenia.
MS – Osteochondritis: usually around 8 months of age, esp. wrists, elbows, ankles, knees; painful –which causes pseudoparalysis of Parrot. Periostitis
especially long bones, skull (rare).
Derm – mucocutaneous rash with erythematous maculopapular or bullous lesions, desquamation of hands and feet, mucous patches, condylomatous lesions, non-immune hydrops.
Renal – nephritis, nephrotic syndrome.

Late: Not infectious. During the first 2 decades – manifestation of chronic inflammation especially bone, teeth, CNS.

MS – frontal bossing, Olympian brow (bony prominence of the forehead), clavicular or Higoumenakis sign (unilateral of bilateral thickening of the sternoclavicular third of the clavicle. Saber shins (anterior bowing of the midportion of the tibia).Scaphoid scapula (convexity of the medial scapular boarder). Clutton joint (synovitis, usually knee, with painless joint swelling
spontaneously resolves.)
HEENT – Dental caries, Hutchinson teeth (peg/barrel-shaped central upper incisiors), notch along biting surface due to abnormal enamel. Mulberry molars (small biting surface with excessive cusps on first molars.) Saddle nose (depression of nasal root resulting from destruction of bone and cartilage secondary to rhinitis). Perforated nasal septum.
Eye – photophobia, lacrimation, corneal opacification, blindness, choroiditis, retinitis, vascular occlusion.
Ear – Vertigo, high tone hearing loss, eighth nerve deafness.
Derm – Rhagades (linear scars from mucocutaneus fissures in the mouth, anus, genitalia.
Soft tissue gummas.
CNS – Juvenile paresis (latent meningovascular infection presents in adolescents; seizures, behavior changes, loss of intellectual function.), hydrocephalus, seizures, Juvenile tabes.
Renal – Paroxysmal cold hemoglobinuria.

Expanded Metabolic Screening

Thyroid function

Possible causes of a decreased serum thyroxine concentration in a neonate include:

Congenital hypothyroidism
Central (hypopituitary) hypothyroidism
PIT-1 mutation, PROP-1 mutations, TRH deficiency, TRH unresponsiveness, TSH deficiency, multiple pituitary deficiencies, TSH unresponsiveness.
Primary Hypothyroidism
Thyroid dysgenesis, Thyroid hormone synthesis defects, Thyroid hormone transport defects, Iodine deficiency, maternal antibodies, maternal medications.

In preterm infants, the possible causes of a decreased serum thyroxine concentration are similar to those for term infants; however, there is an increased frequency of transient primary hypothyroidism. In preterm infants less than 28 wks gestation: immature hypothalamic-pituitary-thyroid axis plus loss of maternal thyroid hormone.

Phenylketonuria (PKU)

PKU screening allows for the mass screening of newborns for increased concentration of phenylalanine. The test is useful in that it is relatively inexpensive, and only requires a few drops of blood from the newborn. Although PKU levels may be elevated at 4 hours of age, the recommendation is to obtain the sample within the first 24 – 48 hours to reduce the rate of false negatives. The screening test must be followed by a confirmation with quantitative measurement of plasma phenylalanine.

Hearing

Otoacoustic emission (OAE) devices for neonatal hearing screening are inexpensive, easy to administer, and the test is performed quickly. They provide a sensitive indicator of hearing loss as there is no response if hearing is worse than 30-40 dB. Children who fail can then be referred for Auditory Brainstem Evoked Response (ABR) testing.

Cord care

The umbilical stump should be kept clean and dry and allow to fall off naturally in 10-14 days. The previous practice of using triple dye, alcohol, or antibiotic ointment is no longer indicated.

Physiologic events (1). Stool

Newborn infants should typically pass the first stool within 24 – 48 hours

following birth. The delayed or absent passage of meconium is associated with colonic obstruction (e.g., meconium plug syndrome, Hirschsprung disease, imperforate anus).

Breast-fed infants have frequent stool which typically occur with each feeding (approximately every 2 hours.). Bottle-fed infants have less frequent stools, and feedings generally occur every 3 – 4 hours.

Urination

A newborn infant who does not urinate by 24 hours of age warrants evaluation.

The evaluation of an anuric infant should include a careful reevaluation of the maternal history and any problems during pregnancy, especially oligohydraminos or any signs of renal obstruction seen on prenatal ultrasound. Feeding should be assessed, and records should be examined for the possibility that the infant voided at birth. A bag or cotton balls may be placed in the diaper to

observe urine. Finally, catheterization, renal and bladder ultrasound, urology consultation, and renal function tests may be obtained.

Vital signs

Blood pressure values vary directly with gestational age.

Spitting vs vomiting

Bilious vomiting is a common finding in infants with small bowel obstruction.

Jaundice

Bilirubin is produced as a result of the hemolysis of red blood cells. It circulates bound to albumin until it is transported into hepatocytes. In the liver, uncongugated bilirubin is conjugated with glucuronic acid by UDP- glucuronoyl transferase. The now soluble form is excreted into the biliary system and into the intestine. In the intestine, half of the bilirubin is converted to urobilinogen by bacteria. It is then oxidized to stercobilin in the feces. Some of the urobilinogen in the intestine is reabsorbed into the blood. A small amount is excreted by the kidneys in the urine in the form of urobilin.

Physiologic jaundice refers to the increase in uncongugated bilirubin believed to be a result of increased breakdown of fetal red blood cells coupled with limited conjugation of bilirubin due to immaturity of the liver in the newborn. Typically the uncongugated bilirubin level rises at a rate of < 5mg/dL per 24 hours and becomes clinically apparent on day 2-3. It usually peaks between 2-4 days with a level of 2-4mg/dL and falls below 2mg/dL between 5-7 days.

In preterm infants, the rise in uncongugated bilirubin is of longer duration than in term infants. Typically, the bilirubin level peaks on day 4-7 with levels between 8-12mg/dL.

Discharge plans

Qualifications for consideration of early discharge of a newborn infant:

> Early discharge is defined as discharge of the newborn before 48 hours. An infant is ready for discharge if: newborn care tasks have been completed, parental education has been completed, and there is a well-established feeding plan. There must be a plan in place for patient follow up within 72 hours, preferably within 24-48 hours.
> Benefits of early discharge: elimination of hospital practices which may interfere with parent-infant bonding and breast-feeding.
> Complications of early discharge: increased risk for rehospitalization, hyperbilirubinemia, sepsis, failure to thrive, dehydration, missed congenital anomalies. Follow-up after early discharge of a newborn infant is important within 48 hours.
> Home births can be facilitated by a home nurse and/or midwife. In hospital routine screening and prophylaxis are still considered the norm despite the different environment.

Abnormal newborn infants

> General

All infants with neonatal abstinence syndrome (NAS) should receive supportive care. Supportive care includes: providing a quiet environment, swaddling, pacifier use, gentle handling. The infant

should be monitored for regulatory problems including: sleep/wake states, responses to stimuli, ability to express needs. Medications may be used to treat the neurobehavioral symptoms.

Formulate a differential diagnosis of lethargy and coma in a neonate

Resuscitation

Ventilation

A normal newborn infant establishes regular respirations by 1 minute of age.

A newborn infant who has a slow heart rate and impaired ventilatory effort requires immediate positive-pressure ventilation.

Establish a patent airway before applying positive-pressure ventilation. Initial lung inflation may require increased pressure for the first breath.

Suctioning

A newborn infant's larynx needs to be visualized and the trachea suctioned if thick or particulate meconium is present in the amniotic fluid and the infant is not vigorous.

Perfusion

External cardiac massage of a newborn infant during resuscitation is indicated if the heart rate does not increase above 60 beats/min after effective ventilation with oxygen has been established.

The proper technique for external cardiac massage in a newborn infant is achieved by encircling the thorax with both hands, placing both thumbs on the midsternum and compressing the chest wall at least 1/3 of the AP diameter. An alternative is to do compressions using 2 fingers on the midsternum.

The metabolic consequences of continued poor perfusion in a newborn infant are acidosis and hypoglycemia.

Very-low-birth-weight infant

An Apgar score greater than 6 often cannot be achieved in very-low-birth-weight infants because they are neurologically immature (e.g., hypotonic, blunted response to noxious stimuli).

Initial care of very-low-birth-weight infants includes all the care required for term infants such as clearance of the airway, initiation of breathing, umbilical cord and eye care, and vitamin K administration. Specifically, these infants require administration of a parenteral glucose solution prior to the establishment of enteral feeds, and

maintenance of a thermoneutral environment.

Initial care of a very-low-birth-weight infant includes monitoring of blood glucose and arterial oxygen concentrations.

Initial care of the very-low-birth-weight infant includes evaluation for sepsis if appropriate.

Prognostic factors for very-low-birth-weight infants include: weight, prematurity, bronchopulmonary dysplasia, necrotizing enterocolitis, and nosocomial infection. There is also an increased incidence of failure to thrive, SIDS, child abuse and inadequate maternal-child bonding in premature infants.

Conditions, diseases

Hypoxia, ischemia

Hypoxic-ischemic encephalopathy is the most frequent cause of neonatal seizure in a full-term infant.

Neonatal seizures secondary to hypoxic-ischemic encephalopathy characteristically occur within 24 hours of birth.

The majority of full-term newborn infants who have neonatal seizures secondary to hypoxic-ischemic encephalopathy do not manifest long-term neurodevelopmental sequelae.

Intrapartum asphyxiation can cause injury to multiple organ systems such as: kidney, lung, intestine, liver, brain, and heart.

Polycythemia, hyperviscosity

Newborn infants with polycythemia are at risk for hypoglycemia and hyperbilirubinemia and should be managed with these considerations.

The treatment for symptomatic polycythemia is a partial exchange transfusion.

Nonphysiologic jaundice

The newborn patient with hyperbilirubinemia should be treated with phototherapy. For severe hyperbilirubinemia, exchange transfusion should be performed.

Clinical manifestations of acute bilirubin encephalopathy include: poor sucking, stupor, hypotonia, seizures on day 1-2. By the middle of the first week, hypertonia of extensor muscles, opisthotonos, retrocollis, and fever are observed. After the first week, hypertonia is the dominant feature.

The permanent clinical sequelae of bilirubin toxicity, kernicterus, include: hypotonia, hyperreflexia, obligatory tonic neck reflexes, delayed motor skills. After the first year, movement disorders, upward gaze and sensorineural hearing loss are noted.

Strategies for prevention of severe hyperbilirubinemia in newborn infants include the following: newborn infants should be examined for any signs of jaundice and screened for hemolytic disorders, ABO incompatibility. Breast-feeding infants should be fed 8-12 times daily in the first few days.

Intracranial hemorrhage

Plan the evaluation and management of a neonate with intracranial hemorrhage.

- Premature infants less than 34 weeks gestation should be screened with cranial ultrasound through the anterior fontanel on a routine basis. Infants less than 1000 grams are at highest risk. Serial ultrasounds are used to monitor progression, regression or posthemorrhagic hydrocephalus. Infants who develop hydrocephalus may require ventriculoperitoneal shunt placement.

Most infants with intraventricular hemorrhage are asymptomatic. Clinical and laboratory findings associated with intracranial hemorrhage in a neonate include: apnea, pallor, cyanosis, poor suck, abnormal eye movements, high pitched cry, muscle twitching, convulsions, decreased muscle tone, metabolic acidosis, shock, decreased hematocrit.

Small-for-gestational age

Small-for-gestational-age infants have a higher neonatal mortality rate than appropriate-for-gestational age infants.

Small-for-gestational-age infants are prone to fasting hypoglycemia, polycythemia, and temperature instability.

Perinatal asphyxia is a frequent complication of intrauterine growth restriction.

Respiratory distress (1). General

Normal arterial blood gas values for a healthy newborn infant: pO2 60 to 90 mm Hg, pCO2 32 to 45 mm Hg.

Respiratory distress syndrome

Surfactant administration in an infant with respiratory distress syndrome improves

survival and reduces pulmonary air leaks.

Characteristic clinical signs of respiratory distress syndrome include tachypnea, grunting, retractions, nasal flaring, and duskiness. Radiographic appearance of respiratory distress syndrome features a granular appearance of the lung fields, and air bronchograms.

Appropriate treatments for respiratory distress syndrome include respiratory support with warm humidified oxygen, mechanical ventilation, and surfactant administration.

Pneumothorax

Pulmonary air leaks are common in newborn infants who are treated with assisted ventilation.

On radiograph, a pneumothorax in a newborn infant is significant for visualization of the lung edge standing out in relief against the hyperluscent area.

Meconium aspiration syndrome

Clinical features: respiratory distress within the first hour with grunting, tachypnea, retractions, and cyanosis.

Radiographic appearance of meconium aspiration syndrome is a coarse granular pattern and irregular aeration.

Congenital pneumonia

Neonatal pneumonia can mimic respiratory distress syndrome.

Transient tachypnea of the newborn

Transient tachypnea of the newborn is characterized by tachypnea, occasional grunting or retractions, and may present with cyanosis. Lung sounds are typically clear and Radiograph chest radiographshows increased vascular markings, fluid in intralobar fissures, overaeration, and rarely pleural effusions. Symptoms are relieved by low supplemental oxygen (< 40% oxygen.) The condition typically resolves within 3 days.

Cyanosis (nonrespiratory)

Peripheral cyanosis is a common finding in healthy full-term newborn infants.

It may be difficult to distinguish between persistent pulmonary hypertension without meconium aspiration and cyanotic congenital heart disease.

Infants with persistent pulmonary hypertension following meconium aspiration typically present with cyanosis, grunting, nasal flaring, retractions, tachycardia, and shock.

Bronchopulmonary dysplasia (BPD) (see XIII.E.1)

Sepsis

Treatment with ampicillin and either gentamicin or cefotaxime is the appropriate antibiotic treatment for suspected sepsis in the immediate newborn period.

Prolonged premature rupture of the membrane (PROM)≥ 18 hours increases the risk of sepsis.

Use of intravascular catheters increases the risk of sepsis.

TORCH infection including HIV

Cytomegalovirus infection may be acquired in utero, during delivery, or in the neonatal period, such as from breast milk, blood products, saliva, cervical and vaginal secretions, urine ,semen, tears, or organ allographs.

Characteristic signs and symptoms include: IUGR [symmetric], prematurity, hepatosplenomegaly, jaundice, blueberry muffin like rash, thrombocytopenia, purpura, microcephaly, intracranial calcifications, chorioretinitis, sensorineural hearing loss, mildly elevated CSF protein, neurologic sequelae and psychomotor retardation. 90 % of the infants born with CMV have subclinical infection and are asymptomatic, 5 % are severe, 5 % are mildly symptomatic. Sensorineural hearing loss occurs in 7 to 10% of all infants with CMV whether they are symptomatic at birth or not.

Majority of newborn with congenital toxoplasmosis are asymptomatic in neonatal period.

Recognition and stabilization of surgical emergency

In the evaluation of a full-term infant who has severe respiratory failure at birth that does not respond to intubation and assisted ventilation, it is important to assess for mechanical problems, such as incorrect endotracheal tube placement, pneumothorax or pleural effusion, as well as to pursue cardiac etiology. A chest radiograph, arterial blood gas, preductal and postductal pulse oximetry and echocardiogram can be done. Sepsis evaluation, including blood culture, blood count, lactate, glucose and tracheal aspirate may be helpful in identifying an infectious cause.

Necrotizing Enterocolitis

Usual presentation of NEC and initial management:

Infants with NEC have a variety of signs and symptoms and may have an insidious or

sudden catastrophic onset.

Initial presentation:

GI: Abdominal tenderness, Abdominal distension, delayed gastric emptying, vomiting, occult blood in stools [25% of patients], Abdominal wall discoloration. Systemic: temperature instability, acting “not right”, lethargy, apnea/respiratory distress, acidosis, glucose instability, DIC, poor perfusion, shock.

Initial management:

Mainly supportive care including cessation of oral feedings, nasogastric decompression, intravenous fluids, surgical consultation, monitoring respiratory status, obtaining coagulation profiles, acid base balances, and blood cultures. Start broad spectrum antibiotics with coverage for gram positive, gram negative and anaerobic organisms. Indications for surgery include failure of medical management and perforation.

Pneumatosis intestinalis [air in the bowel wall] is the hallmark of diagnosis of NEC and confirms the diagnosis. Portal venous gas on KUB radiology is a sign of severe disease and pneumoperitoneum indicates perforation.

Complications

Early: post-operative wound infections, wound dehiscence, stomal problems

Late: intestinal stricture, Short bowel syndrome, risk for adverse neurodevelopmental outcome.

Intestinal obstruction

Clinical signs and symptoms of congenital bowel obstruction include: vomiting, abdominal distension, no meconium in first 24 hours, pneumoperitonium and air under the diaphragm on abdominal radiograph.

Treatment of abdominal distention caused by a congenital small bowel obstruction includes nasogastric decompression, cessation of feeds and intravenous fluids.

Meconium ileus:

Etiology: obstruction of the terminal ileum caused by hyperviscous secretions from the intestinal glands coupled with lack of pancreatic enzymes. 90 % of meconium ileus is associated with cystic fibrosis.

Diagnostic evaluation:

> Abdominal radiograph:
> Soap bubble appearance of the bowel caused by trapped air within the meconium.
> Large dilated loops of bowel with few air fluid levels.
> Barium enema shows a microcolon and pellets of inspissated meconium at the site of distal bowel obstruction.
> Tests for cystic fibrosis in a confirmed case of meconium ileus include sweat chloride test and genetic testing.

Treatment:

Initial management:

Nasogastric decompression, iv hydration, electrolyte replacement, half strength Gastrografin enema.

Gastrografin draws fluid into the bowel lumen, dilutes viscous meconium and facilitates passage.

Surgical treatment is indicated in complicated meconium ileus causing obstruction.

For a simple meconium ileus, enterotomy with extraction and irrigation of meconium from the bowel is performed.

Complicated ileus requires resection of the bowel segment and ostomy.

Tracheoesophageal fistula

Signs and symptoms of esophageal atresia with distal tracheoesophageal fistula include pregnancy complicated by polyhydramnios, excessive salivation and inability to swallow feeds after birth, with attempts to feed resulting in regurgitation, cyanosis, choking, and coughing.

Diagnosis of a tracheoesophageal fistula is established by attempting to pass an orogastic/nasogastric tube and meeting resistance at 10 to 12 cm from the nares or 8 to 10 cm from the mouth followed by chest radiograph which demonstrates recoiling of the tube. H type fistula is diagnosed by barium swallow. However, bronchoscopy with esophagoscopy is the best diagnostic test.

Abdominal-intestinal wall defect

Gastroschisis is centrally located full thickness abdominal wall defect with 2 types of anatomical features

> The extruded intestine never has a protective sac covering it.
> Umbilical cord is intact and the defect is towards right of the umbilicus opening in the abdominal wall. It can be 2 to 4 cm in diameter with solid organs residing within the peritoneal cavity.

Omphalocele: Herniation of the abdominal contents into the base of the umbilical cord usually enclosed by a membrane. Elements of the umbilical cord course individually over the sac and come together at its apex to form a normal appearing umbilical cord.

Extrophy of the bladder: is a congenital malformation of the lower anterior abdominal wall where the internal surface of the posterior wall of the urinary bladder extrudes through the abdominal wall defect. It occurs in varying degree of severity ranging from epispadias to extroversion of the bladder with exposure of urethral orifices.

Cloacal exstrophy: Rare but devastating complex of anomalies including imperforate anus, exstrophy of the bladder, omphaocele and vesicointestinal fistula, frequently with prolapse of the bowel through the fistula into the bladder mucosa.

Infants affected by maternal disorders (e.g., diabetes, SLE]

Infant of a diabetic mother is at risk for hypoglycemia, hypocalcemia, polycythemia, and neonatal small left colon syndrome.

In mothers with poorly controlled DM, the fetus in utero becomes hyperglycemic due to transplacental transportation of glucose, which stimulates the fetal pancreatic cells and increases insulin levels. After delivery glucose is abruptly removed, hyperinsulinemia persists causing hypoglycemia.

Congenital anomalies are more frequent among infants of diabetic mothers than among normal control infants.

Management of newborn born whose mother has Type 1 DM (absolute insulin deficiency)

Infants are prone to develop macrosomia, hypoglycemia, hypocalcemia, hypomagnesemia, perinatal asphyxia, RDS, TTN, and hypertropic cardiomyopathy.

Initial evaluation:

Upon delivery monitor blood glucose levels and hematocrit. Observe for jitteriness, tremors, convulsions, apnea, weak cry or poor sucking. A thorough physical exam with attention to heart, lungs, kidneys and extremities should be done.

Metabolic management:

Hypoglycemia: if symptomatic start 2ml/kg of 10% glucose at a rate of 1 ml/min followed by continuous infusion at a rate of

6 to 8 mg /kg/min to maintain normal glucose above 40 to 50 mg/dl - consider trial of steroids in persistent hypoglycemia.

Hypocalcemia: 10 % iv calcium gluconate. Monitor Calcium every 12 hrs. Hypocalcemia should respond in 3 to 4 days.

Hypomagnesemia: PO or IV magnesium sulfate 50 %, 0.2ml/kg/day. Management of cardiorespiratory problems:

Hyaline membrane disease: assess amniotic fluid for fetal lung maturity. Manage with surfactant and administer respiratory support as needed.

Cardiomyopathy: treatment of choice is propranolol; digoxin is contraindicated because of possible ventricular outflow tract obstruction.

Monitor and manage hyperbilirubinemia and polycythemia.

Renal venous thrombosis: fluid restriction, close monitoring of electrolytes and renal status as well as supportive therapy.

Management of morphologic and functional problems:

macrosomia and birth injury include fractures and Erb's palsy which are treated with immobilization and range of motion exercises

Infants affected by maternal medications

The use of some anticonvulsants during pregnancy is associated with an increased risk of fetal anomalies.

Isotretinoin is a potent teratogen which causes microtia, hydrocephalus, nonspecific facial aberrations and cardiac defects.

Effects of drugs given to the mother during labor on the fetus/neonate Opiates:

May cause respiratory depression, hypotension; Synthetic opiates like fentanyl may cause muscle rigidity when administered rapidly.

Beta-adrenergic agents:

Albuterol is considered safe during pregnancy.

Ritodrine and terbutaline sulfate used as tocolytics causes rebound hypoglycemia, increase fetal aortic blood flow, increased cardiac output which leads to increased systemic blood pressure which can increase incidence of intracranial hemorrhage in immature fetal brains.

Tocolytic agents:

Magnesium sulfate can cause decreased muscle tone, drowsiness, and decreased calcium levels.

Effect on a fetus of lithium use during pregnancy include cardiac anomalies [Ebstein's anomaly], skeletal and craniofacial anomalies, abnormalities of vascular development.

Effects on a fetus of warfarin use during pregnancy are dependent upon timing of exposure. First trimester administration cause nasal hypoplasia and epiphyseal stippling. In third trimester it is known to cause eye and central nervous system abnormalities.

Infants affected by maternal substance use

Fetal alcohol syndrome is a frequently documented cause of mental retardation. Moderate or high alcohol intake (≥7 drinks per week or ≥3 drinks on multiple occasions) is a risk for fetal alcohol syndrome.

Physical features of fetal alcohol syndrome are short palpebral fissures, epicanthal folds, flat nasal bridge, simple smooth philtrum, a thin upper lip, small hypoplastic nails, microcephaly, small for gestational age, increased association with congenital heart defects, and hirsuitism.

The association between the maternal use of alcohol is dose dependent, and there is no safe limit. Withdrawal symptoms may occur within 24 hours of life including tremors, irritability, opisthotonos, and seizures.

Marijuana may increase the risk for preterm labor and reduced birth weight in neonates. However there is no evidence of long term withdrawal. Marijuana in pregnancy is NOT associated with an increased risk for birth defects, dysmorphic features or developmental delay in exposed offspring.

Maternal use of tobacco causes increased risk for intrauterine growth retardation (IUGR), premature labor, abortion, abruptio placenta, and facial clefts.

Maternal use of opiates is associated with increased incidence of IUGR and perinatal distress, as well as signs and symptoms of withdrawal in 60-90% of exposed infants. Onset of withdrawal varies from 2 days to 2 weeks. Restlessness, agitation, tremors, wakefulness, feeding problems may persist for 3 to 6 months. They have increased risk for sudden infant death syndrome (SIDS) and strabismus.

Methamphetamines increase rates of fetal distress and growth restriction. Also, there have been isolated case reports of cardiac defects, cleft lip, and biliary atresia, as well as some neonates with decreased arousal, increased stress, poor movement quality and the occasional withdrawal syndrome.

Maternal use of barbiturates leads to withdrawal symptoms beginning at the end of 1st week and last up to 6 weeks of age. Symptoms are similar to the narcotic withdrawal.

Maternal use of cocaine may lead to preterm labor, spontaneous abortion, IUGR, abruptio placenta, fetal asphyxia. In infants it may lead to developmental delay, learning disabilities, impaired auditory information processing.

Teratogenic effects are controversial and presumed to be due to vasoconstictive effects of cocaine. These include central nervous system and cardiovascular abnormalities, limb reduction defects, intestinal atresias, genitourinary abnormalities. Withdrawal is unusual, but when it does occur, it is similar to narcotic withdrawal.

Cigarette smoking is associated with an increased risk for miscarriage, reduced fetal weight, and abnormal placentation. There may be an increased risk for facial clefting, but cigarette smoking is not otherwise associated with major congenital anomalies.

Maternal drugs of abuse include opiates, barbiturates, alchohol, amphetamine, cocaine, benzodiazipines and they are notorious to cause neonatal abstinence syndrome. Symptoms include CNS hyperexcitability (tremors, hyperactive moro's, hyperreflexia, irritability, yawning, seizures, sleep disturbances), fever, excessive sweating, mottling, tachypnea, nasal congestion, sneezing and gastrointestinal symptoms (poor feeding , excessive sucking, suck-swallow incordination, vomiting, diarrhea, poor weight gain, and dehydration.)

Multiple congenital anomalies (See VII)

Oligohydramnios

Oligohydramnios is associated with Potter's facies: Low set ears, wide set eyes, broad flat nasal bridge, receding chin, parrot beak nose. Infants with features of the oligohydramnios tetrad (Potter facies) warrants a detailed evaluation of the genitourinary system, pulmonary system as they are associated with renal agenesis, pulmonary aplasia, cryptorchidism, imperforate anus, meningocele, and club feet .

Deformations:

Mechanical forces that alter the structure of intrinsically normal tissue are called deformations.

Amniotic bands

Amniotic bands can become adherent to any part of the fetal body, thereby causing a variety of disruptions (e.g., ring-like constrictions of limbs, amputation of digits, disruptive clefts of the face).

Positional deformations (e.g., hip dysplasia, clubfeet, torticollis)

Newborn deformations are more common due to in utero fetal constraints in the context of multiple births and first pregnancies or in any situation where the fetus is crowded (e.g., uterine leiomyoma, abnormal uterine structure, large fetus).

Development of congenital anomalies in fetal deformation and/or malformation:

The four primary mechanisms of unusual embryonic and fetal formation are malformation, deformation, disruption, and dysplasia.

Malformations, such as cardiac defects, are due to intrinsic problems in the developing tissue.

Deformations, such as pugilistic facies, are caused by extrinsic forces acting upon an otherwise normally developing structure.

Disruptions, such as amniotic bands, are due to the destruction of previously normally developed tissue.

Dysplasias, such as hemangiomas, occur when organization of cells into tissue is abnormal.

Fluid and Electrolyte Metabolism

Composition of body fluids:

The body contains two major fluid compartments: the intracellular fluid (ICF) and the extracellular fluid (ECF). The ICF comprises of two-thirds of the total body water (TBW), while ECF accounts for the remaining third. The ECF is further divided into the interstatial fluid (75%) and plasma (25%). The TBW comprises approximately 70% of body weight in infants, 65% in children and 60% in adults.

Intracellular, extracellular

The ICF and the ECF are in osmotic equilibrium as the cell membrane is permeable to water. Decrease in protein concentration may lead to a reduction in plasma volume and an increase in interstitial volume.

The volume of plasma water can be altered by pathologic conditions, including dehydration, anemia, polycythemia, heart failure, abnormal plasma osmolality, and hypoalbuminemia.

Equilibrium between extracellular fluid and intracellular fluid is maintained by the movement of water in response to alteration of osmolality of either compartment. For example: when ECF osmolality decreases there is a shift of water into ICF. When ECF osmolality increases fluid shifts from ICF into ECF.

Normal plasma osmolality is 285-295 mosm/kg. Is estimated by the formula:

Plasma Osmolality = 2 [Na] + [Glucose]/18 + [BUN]/2.8

Glucose and urea normally contribute little to the plasma osmolality; multiplication of the sodium value by 2 provides an approximation of the osmolality. Uremia does not affect plasma osmolality significantly, however hyperglycemia increases plasma osmolality significantly. The calculated osmolality is usually slightly lower than the measured osmolality. The difference between measured and calculated osmolality constitutes osmolar gap which represents measured osmules.

Sodium forms the major component of ECF. Therefore chronic sodium depletion may result in intravascular volume depletion due to decreased osmolality of ECF.

Electrolytes (sodium, potassium, chloride)

Physiologic requirements for sodium are 3 to 4 Meq/100 ml/day and potassium requirement 2 Meq/100ml/day.

An electrolyte [Na,K] concentration does not reflect its total body content.

Sodium is predominantly intracellular while potassium is extracellular predominantly.

Protein

Hypoproteinemia causes generalized edema due to decrease in the intravascular oncotic pressure of intravascular fluid/plasma.

Acid-base physiology

Normal mechanisms and regulation

Normal pH ranges b/w 7.35 – 7.45 after BLANK hours of birth. Acid [HA] is a substance than donates hydrogen ion H+ and base is a substance that accepts hydrogen ion H +. HA ⍰ H + A.

Pulmonary mechanism plays an important role in regulating acid-base physiology:

Carbon dioxide is a weak acid and a physiologic buffer that regulates acid base system along with Bicarbonate [HCO3]. The lungs prevent an increase in the partial pressure of CO2 (Pco2) in the blood by excreting the CO2 via ventilation.

CO2 + H2O ⍰⍰ H + HCO3. An increase in ventilation decreases the Pco2leading to respiratory alkalosis and a decrease in ventilation increases the Pco2 leading to respiratory acidosis.

Anion gap [AG] is calculated by subtracting anions from Cations. Normal anion gap is between 8 to 16

AG = [Na] + [K] - [Cl] + [HCO3]

Acidosis, alkalosis

Metabolic acidosis [pH < 7.35]

Clinical presentation results from either a net reduction in bicarbonate (HCO3) or an increase in acids related to increased production, decreased excretion, or increased intake. Most common cause is diarrhea. Symptoms depend on the degree of severity

of acidosis, pulmonary and renal compensation. Presentation also includes impaired cardiac contractility, an increased risk of arrhythmias; decrease in the cardiovascular response to catecholamines which may lead to hypotension, volume depletion or shock. It can also lead to vasoconstriction of the pulmonary vasculature.

Laboratory presentation: pH <3.5; HCO3 < 22.

Serum findings in clinical disturbances of acid- base balance in the simple disorders, evaluating pH, PCO2, and bicarbonate.

Initial therapy for severe acidosis (metabolic) when rapid respose is required is intravenous bicarbonate [1meq/kg] administration and then treatment of the underlying cause.

Most effective therapeutic approach for patients with a metabolic acidosis is correction of the underlying disorder, if possible.

For example: insulin, restoration of potassium, intravenous fluid in diabetic ketoacidiosis; oral base therapy [sodium bicarbonate or citrate] in renal tubular acidosis, and hemodialysis in severe renal insufficiency.

Metabolic acidosis associated with a high anion gap includes ketoacidosis, renal failure, lactic acidosis, and certain toxins CUTE DIMPLES: -

Cyanide, Uremia, toluene, ethanol, diabetic ketoacidosis, isoniazid, methanol, propylene glycol, lactic acidosis, ethylene glycol, salicylates.

Chronic volume contraction can lead to alkalosis. Pulmonary compensatory changes seen:

In primary metabolic alkalosis includes respiratory acidosis with increase in pCO2 and decrease in pH.

In primary metabolic acidosis includes respiratory alkalosis with decrease in pCO2 and increase in pH.

Renal compensatory changes seen in primary respiratory acidosis includes HCO3 absorption followed by excretion of H+ ions in urine.

In primary respiratory alkalosis includes decrease in the proximal tubule bicarbonate resorption.

Parathyroid hormone decreases proximal tubule bicarbonate resorption; hyperparathyroidism may cause a mild metabolic acidosis.

Diuretics that produce metabolic alkalosis: loop diuretics [Furosemide], thiazide diuretics [metazolone, hydrochlorthiazide]

Diuretics produce metabolic acidosis: acetazolamide [Proximal RTA type 2] (similar mechanism as proximal renal tubular acidosis [p-RTA] [type 1]), spirinolactone [distal RTA type 4] (similar mechanism as distal renal tubular acidosis [d-RTA] [type 4].)

Differential diagnosis of acidosis with a normal anion gap: gastrointestinal loss of bicarbonate [diarrhea], renal loss of HCO3 [RTA 2], renal dysfunction [RTA type 1], ingestions [ammonium chloride, acetazolamide, ifosphamide], hypoaldosteronism, hyperalimentation fluids [TPN].

Useful mnemonic to remember this is FUSEDCARS (fistula (pancreatic), uretero- enterostomy, saline administration, endocrine (hyperparathyroidism), diarrhea, carbonic anhydrase inhibitors (acetazolamide), ammonium chloride, renal tubular acidosis, spironolactone).

Electrolyte abnormalities

Sodium
Hypernatremia

Extracellular fluid volume is relatively spared in hypernatremia as sodium is a major component of ECF , hypernatremia increases ECF osmalilty which draws fluid into ECF from ICF leading to relative sparing of extracellular fluid volume. For example: severe hypernatremia can draw fluid from brain cells leading to hemorrhage.

Symptoms of hypernatremia: irritability, lethargy, weakness, polydypsia, high pitched cry, hyperapnea, fever, hyperglycemia, mild hypocalcemia. Complications include seizure, stroke, brain hemorrhage, and extrapontine myelinosis.

Hyponatremia

Diseases associated with hyponatremia and increased sodium in the urine: autosomal recessive polycystic kidney disease, renal tubular acidosis type 2 , cerebral salt wasting, tubulointerstitial nephritis, obstructive uropathy, juvenile nephronophthisis, polyuric phase of acute tubular necrosis, postobstructive dieresis, osmotic diuresis, thiazide or loop diuretics, absence of

aldosterone (e.g., 21-hydroxylase deficiency), Pseudohypoaldosteronism type I, urinary tract obstruction with or without infection.

Causes of factitious hyponatremia: [normal plasma osmolality with low sodium]: hyperglycemia, administration of mannitol, sucrose, hypovolumic hyponatremia [excessive sweating, burns, vomiting, diarrhea].

Urinary sodium concentration and urinary osmolality is very useful in evaluation of hyponatremia:

Chronic diuretic therapy can produce hyponatremia .

It is important to estimate sodium intake and output in evaluating patients with hyponatremia.

Distinguish between dilutional hyponatremia and a total body deficit of sodium. Dilutional Hyponatremia: sodium loss can lead to a state of volume depletion serving as signal for the release of ADH (anti-diuretic hormone). As a result of ADH-stimulated water retention, blood sodium becomes diluted.

Sodium deficit is calculated using the formula: 0.6x weight [kg] x [140- plasma[Na]].

Potassium

Hyperkalemia

Emergency treatment of hyperkalemia:

Stabilizing the heart to prevent arrhythmia.
Removal of Potassium from the body.

#1: IV Calcium gluconate or calcium chloride over 2 to 5 min: Calcium stabilizes the cell membrane of heart cells, preventing arrhythmias.

#2: Shift potassium from extracellular to intracellular compartment, achieved by either

Insulin and glucose administration or
Sodium Bicarbonate or
Nebulized Albuterol
all of the above.

Three options exist for removing potassium from the body:

Removal from the gastrointestinal tract with a cation-exchange resin (sodium polystyrene sulfonate administered orally or rectally).

Removal into the urine with diuretics.

Bloodstream removal for patients who have limited urine output or renal function with dialysis.

Severe cardiac rhythm changes may begin abruptly in patients with hyperkalemia.

Peak T waves are the first sign of hyperkalemia followed by a prolonged PR interval and, when most severe, a prolonged QRS complex:

Signs of hyperkalemia:

The most important effects of hyperkalemia are due to the role of potassium in membrane polarization.

Cardiac changes are the main concern.

EKG changes begin with peaking of T waves followed by ST segment depression, an increased PR interval, flattening of the P wave, and widening of the QRS complex. This process can eventually progress to ventricular fibrillation. Asystole may also occur.

Paresthesias, fasciculations, weakness, ascending paralysis can occur but cardiac toxicity usually precedes these clinical symptoms.

Plan the treatment for a patient with hyperkalemia:

Long term therapy includes reducing intake via dietary changes and eliminating or reducing medications that cause hyperkalemia.

Medications to increase potassium excretion, such as sodium polystyrene sulfonate [kayexalate] and loop or thiazide diuretics.

Infants with chronic renal failure may require dialysis.

Fludrocortisone replacement therapy may help in aldosterone deficiency.

Hypokalemia [K+ less than 3.5 mEq/L]

Hypokalemia in a patient especially in children is most likely due to gastroenteritis.

The electrocardiographic rhythm abnormalities in patients with hypokalemia include flattened T wave, depressed ST segment, appearance of a U wave [located between the T wave (if still visible) and the P wave].

Emergency treatment of hypokalemia: Intravenous potassium [KCL] 0.5 -1 Meq/kg over 1 hr.

Symptoms of hypokalemia:

Muscle weakness, cardiac arrhythmias.

Ventricular fibrillation and torsades de pointes may occur if patient has an underlying heart disease.

Heart may become susceptible to digitalis-induced arrhythmias, such as supraventricular tachycardia, ventricular tachycardia, and heart block.

Chloride

Association of chloride and acidosis in the differential diagnosis of metabolic acidosis: there is increase in Chloride in normal anion gap acidosis. Lowered bicarbonate concentration, which is counterbalanced by an equivalent increase in plasma chloride concentration; Chloride levels remain unchanged in high anion gap acidosis.

Urinary screening examination for diuretics is needed in the evaluation of hypochloremia as diuretic use increases chloride excretion in urine. Urine chloride is typically >20 meq/L.

Disease states, specific therapy

Pyloric stenosis

Pyloric stenosis leads to hypochloremic, hypokalemic metabolic alkalosis due to loss of hydrochloric acid during emesis.

Hypokalemia occurs as kidneys retain H+ ions by losing K+. Management:

The initial goal of therapy is appropriate intravenous rehydration, with gradual correction of any electrolyte disturbances. Definitive management is surgery [Ramsted Pyloromyotomy].

In infants with normal electrolytes: maintenance intravenous fluid.

In infants with dehydration and electrolyte abnormality: Ensure the kidneys are functional and add KCL to IVF.

**Correction of electrolytes before surgery is very important as alkalosis can cause post operative apnea.

** It is important to note that the serum potassium concentration rises as the serum pH normalizes.

Gastroenteritis

Management of acute gastroenteritis:

Antidiarrheal agents are not indicated in the treatment of AGE.

Mild to moderate dehydration from diarrhea can be treated by Oral rehydration. As a guideline for oral rehydration, 50ml/kg of ORS should be given within 4hrs to patients with mild dehydration and 100ml/kg over 4hrs to those with moderate dehydration. An additional 10ml/kg of ORS is given for each stool. Breast feeding should be allowed after rehydration in infants who are breast fed; in formula fed patients, their usual formula, milk or feeding should be offered after rehydration. Early refeeding decreases the duration of the diarrhea. Once rehydration is complete, maintenance therapy should be started. Patients with mild diarrhea can be treated at home with 100ml/kg/24hrs of ORS until the diarrhea stops.

Intravenous hydration may still be required for severe dehydration.

Acute renal failure

Changing fluid requirements in patients with severe oliguria:

Place the patient with severe oliguria on one-third maintenance fluids to replace insensible losses. No potassium to be added.

Also replace urine output ml/ml with ½ NS.

Coexisting volume depletion should be corrected in patients with acute renal failure.

In patients with acute renal failure, output may slowly increase leading to dehydration and worsening ARF if patient is just replaced for insensible losses. Replacement solution of ½ NS is usually appropriate initially, although its composition may need to be adjusted if urine output increases significantly.

Shock

The clinical signs of shock due to fluid loss include dry mucous membranes, lack of tears, scant and concentrated urine, fluctuating mental status, sunken fontanel, cool, pale or mottled extremities and skin, cyanosis, poor cap refill, weak pulses, poor muscle tone, hyperpnea, tachycardia, hypotension without signs and symptoms of congestive heart failure.

The type of fluids to be administered in the treatment of shock:

0.9% NS 20ml/kg x 3 can be given for initial fluid replacement in shock.

If shock is because of acute blood loss, packed red blood cell (pRBC) if available should be given.

Central line to be placed and pressor support to be started if unable to maintain heart rate and blood pressure.

Try to treat the underlying cause of shock.

Frequent clinical assessment is required in the treatment of shock which includes monitoring HR, BP, I&O's, heart rate, blood pressure, fluid balance (input and output)..

Immediate fluid resuscitation of infants in shock may require more than 20 mL/kg of fluid to improve their clinical conditions.

Syndrome of Inappropriate Anti-Diuretic Hormone (SIADH)

Serum and urine abnormalities in SIADH include:

HYPONATREMIA, and decreased serum osmolality, Inappropriately concentrated urine with high specific gravity, inappropriately high urine osmolality (>200mOsm/kg) in face of hyponatremia and high urinary sodium secretion >30 mEq/l.

Clinical abnormalities, disease conditions and medications associated with SIADH are: Central nervous system disease - tumor, trauma, infection, cerebrovascular accidents (CVA), subarachnoid hemorrhage, Guillain-Barré syndrome, delirium tremens, multiple sclerosis.

pulmonary disease - pneumonia, chronic obstructive pulmonary disease, lung abscess, tuberculosis, cystis fibrosis, positive-pressure ventilation.

Carcinoma - Lung, pancreas, thymoma, ovary, lymphoma.

Drugs - Exogenous vasopressin, nonsteroidal anti-inflammatory drugs, nicotine, diuretics, chlorpropamide, carbamazepine, tricyclic antidepressants, Selective

Serotonin Receptor Inhibitor (SSRIs), vincristine, thioridazine, cyclophosphamide, clofibrate.

Surgery - Postoperative Idiopathic (most common)

The treatment of SIADH and importance of fluid restriction in the management of SIADH:

FLUID RESTRICTION!!! 50-75 % of management including oral intake (if doesn't work, consider wrong Dx Cerebral salt wasting).

In severe hyponatremia, use of furosemide with sodium supplementation may help. In adults demeclocycline and lithium are used but are problematic in children.

Plasma volume is increased in SIADH:

In SIADH there is secretion of ADH (vasopression) that is not inhibited by expanded intravascular volume or low serum osmolality.

Therefore the child is unable to excrete water. This results in hyponatremia. Also, expansion of extracellular volume due to retained water causes a mild increase in intravascular volume.

Differentiate SIADH from hyponatremic dehydration:

Hyponatremic dehydration occurs when the lost fluid contains more sodium than the blood (loss of hypertonic fluid).

Relatively more sodium than water is lost. Because the serum sodium is low, intravascular water shifts to the extravascular space, exaggerating intravascular volume depletion for a given amount of total body water loss. Head trauma can lead to diabetes insipidus or SIADH.

Cystic fibrosis

Hypochloremic and hyponatremic dehydration seen in cystic fibrosis (CF).

CF patients have abnormally high sodium in sweat. These can lead to hypochloremia and hyponatremia. Metabolic alkalosis is often seen because of loss of chloride in sweat. These infants are also prone to volume depletion. Sweat test- measures sweat chloride and the reference value is less than 40 mmol/L, and a value of more than 60 mmol/L of chloride in the sweat is consistent with a diagnosis of CF.

Dehydration

The clinical and laboratory abnormalities of hyponatremic dehydration:

Clinical: Initially presents with Nausea, vomiting, headache malaise. As the hyponatremia worsens, confusion, diminished reflexes, convulsions, stupor and coma may occur. Laboratory: decreased serum sodium and chloride. Increased BUN and urine specific gravity, and decreased urinary sodium.

The clinical and laboratory abnormalities of hypernatremic dehydration:

Clinical: Severe hypovolemic hypernatremic dehydration induces brain shrinkage, which can tear the cerebral blood vessels, leading to cerebral hemorrhage, seizures, paralysis, and encephalopathy. Infants who have severe hypernatremia (serum sodium, >160 mEq/L [160 mmol/L]) often present with muscle weakness, a high- pitched cry, insomnia, or lethargy. Younger children tend to have both a higher morbidity and mortality with severe hypernatremia. Clinical conditions include diabetes insipidus (by pure water loss), vomiting or diarrhea (hypotonic fluid loss), improperly prepared infant formula (hypertonic sodium gain). Laboratory: increased serum sodium, chloride, and BUN and urine specific gravity.

Laboratory abnormalities of isotonic dehydration:

Normal range serum sodium (135-150), high urine osmolality, low urine sodium, high BUN, serum bicarbonate is low.

Management of hypernatremic dehydration: Emergency phase:

Restoration of vascular volume with 10 to 20 mL/kg of isotonic intravenous fluid such as lactated Ringer solution with 130 mEq/L of sodium or normal saline with 154 mEq/L of sodium followed by a rehydration phase.

Rehydration phase:

The sum of the free water deficit and maintenance fluid requirements and administered evenly over 48 hours.

Free water deficit (L) = 0.7xwt (kg) [1–current sodium/desired sodium]. During the rehydration phase, 5% dextrose in 0.2% normal saline (31 mEq/L of sodium) is the usual IV fluid composition.

DO NOT CORRECT hypernatremia too rapidly as it can lead to brain edema, seizures, and death.

Goal is to decrease serum Na by less than 12mEq/l every 24hrs or 0.5mEq/l/hr. Because giving large amounts of NS may actually cause serum sodium to fall too rapidly. Therefore, various amounts of 3% normal saline (513 mEq/L sodium) should be added such that the IV fluid sodium concentration is approximately 10 to 15 mEq/L lower than the serum sodium level.

Management of hyponatremic dehydration:

Avoid overly rapid correction of hyponatremia because it may cause Central Pontine Myelinosis.

The cornerstone of therapy is to replace the sodium deficit and any water deficit that is present.

First step is to restore the intravascular volume with isotonic saline.

Hypertonic saline (3% sodium) is used to replace sodium if rapid correction is needed (if Na<120 or s/s). These are symptoms and signs of the electrolyte disorder.

Management of isotonic dehydration:

Mild-Moderate – Oral rehydration therapy is the best.

Severe: isotonic fluid bolus at 20ml/kg, then reevaluate after 30-60min.

Additional bolus if required then IVF to deliver maintenance fluids + additional volume for ongoing losses.

Effectiveness of oral rehydration solutions in treating acute diarrheal dehydration:

Oral rehydration therapy [ORT] is effective for the treatment of mild to moderate dehydration.

Patient's willingness to drink needs to be evaluated before choosing this option for rehydration.

Continued intermittent emesis is NOT a contraindication. Generally, if tolerating ORT, no need to check electrolytes.

Calculate fluid deficit and replace over about 2- 4 hrs. Mild=50ml/kg, moderate

=100ml/kg.

Should be administered in small volumes very frequently to minimize gastric distention and reflex vomiting.

Generally, 5 mL of oral rehydration solution every minute is well tolerated. Replace ongoing losses from stool and emesis.

Once rehydration is adequate, may return to a normal diet.

Differences between and rationale for the composition of oral rehydration solutions:

The basic rationale is to replace sodium, chloride, potassium, bicarbonate, glucose and water.

Sodium and glucose in the correct proportions can be passively cotransported with fluid from the gut lumen into the circulation.

If this proportion is off, e.g. apple juice, with a much higher carbohydrate concentration, then water absorption will be impeded and diarrhea will continue/worsen.

Hypotension is a very late sign of dehydration

The signs and symptoms of dehydration are related to changes in extracellular fluid volume:

5% □ no signs and symptoms,

10% (mod)□tachycardia, orthostatic hypotension, dry MM, weak pulses, oliguria, slight reduction in skin turgor, slightly depressed fontanel.

>15% (severe) □ hypotension/shock, decreased skin perfusion, sunken fontanel.

Intracranial hemorrhage may occur during the development of hypernatremic dehydration.

Seizures can occur in an infant with chronic hypernatremia who is being rapidly rehydrated:

As hypernatremia develops, the brain generates idiogenic osmoles to increase intracellular osmolality and prevent loss of brain water. If serum sodium concentration is lowered too quickly, there is movement of water from the serum into brain cells to equalize the osmolality in the two compartments. The resultant brain swelling is clinically similar to consequences of acute hyponatremia and most commonly manifests as seizures in infants.

Differentiate diabetes insipidus from hypernatremic dehydration (i.e., urine specific gravity, urine and serum osmolalities):

Diabetes Insipidus: Serum osmolality high, increased sodium, chloride, BUN,

urine specific gravity low

Hypernatremic Dehydration: Increased serum osmolarity, increased sodium, chloride, BUN with high Urine specific gravity

Hyperosmolar non-ketotic coma

Appropriate fluid therapy is essential for a patient with hyperosmolar non-ketotic coma to prevent the development of cerebral edema.

(Type 2 diabetics who are insulin resistant with high glucose may present with non- ketotic hyperosmolar diabetic coma).

Treatment is directed at rapid repletion of the vascular volume deficit and very slow correction of the hyperosmolar state.

Rapid volume expansion with 20ml/kg of NS boluses in the first hour. Fluid replacement should take place over 36-48hrs.

A higher initial volume may be necessary in patients with severe volume depletion. Slower initial rates may be appropriate in patients with significant cardiac or renal disease or in those who are not urinating.

Caution should be taken to not correct hypernatremia too quickly, as this could lead to cerebral edema.

After bolus one would likely switch to 0.2 NS and dextrose may be added once serum glucose <300.

Add 20meq/L KCl to prevent hypokalemia. Insulin can be given by continuous iv infusion starting with the second hour of fluid. Dose should be 0.05U/kg/hr of regular insulin.

Genetics and dysmorphology

General

Mendelian inheritance

The inheritance pattern associated with an autosomal dominant disorder with incomplete penetrance (example: Charcot Marie Tooth).

Incomplete penetrance is the complete absence of symptoms despite carrying an autosomal dominant condition or trait. However, there is still some potential for an affected person to develop symptoms later in life.

Autosomal recessive inheritance involves mutations in both copies of a gene.

Characteristics of autosomal recessive traits include horizontal transmission, the observation of multiple affected members of a kindred in the same generation, but no affected family members in other generations

Recurrence risk of 25% for parents with a previous affected child; Males and females being equally affected, although some traits exhibit different expression in males and females and increased incidence,

The inheritance patterns of X-linked recessive disorders (example Hemophilia)

The classic features of Rett syndrome include relatively normal neurodevelopmental progress for the first 6 to 18 months after birth, followed by a period of stagnation and subsequent developmental regression. Episodic apnea and hyperpnea are typical features, as are seizures, ataxia, and acquired microcephaly.

It is seen almost exclusively in girls and is associated with stereotypic hand movements (such as handwringing) and loss of purposeful hand movements. Now that the molecular defect has been characterized, clinicians are beginning to appreciate the much broader clinical spectrum of the disorder. Many affected girls also have unusual behavioral patterns, with episodes of unprovoked screaming or crying, bruxism, or panic attacks.

Mutations in the MECP2 gene are not only associated with classic Rett syndrome but are also found in girls who have atypical features and are often misdiagnosed with Angelman syndrome.It is important to consider further testing in a girl who has autism, microcephaly, concordant behavioral disturbances, or neurologic findings suggestive of atypical Rett syndrome.

Recognize the clinical features associated with an X- linked dominant disorder.

Diagnostic testing

Prenatal

Prenatal diagnosis of Hemoglobin disorders by mutation-specific DNA methods is available especially for sickle cell disease and thalassemia. It is accomplished by chorionic villus biopsy in the first trimester and by amniocentesis in the second trimester.

Important tests that can predict the absence of fetal lung immaturity :

The presence of phosphatidyl glycerol on the thin-layer chromatogram AND L/S ratio >2.0, indicates fetal pulmonary maturity with virtual certainty in any patient. A recently developed

method under investigation is the lamellar body count test. Thresholds representing fetal lung maturity are still under investigation.

Small-for-gestational-age fetuses are seen in women with chronic systemic illnesses
Maternal chronic hypertension, diabetes mellitus, congenital anemia, cyanotic heart disease, antiphospholipid antibody syndrome, and other autoimmune disorders are the most common chronic maternal diseases influencing fetal growth that can lead to small for gestational age fetuses.
Serial growth can be assessed by ultrasonography of the fetus. Second trimester ultrasonographic biometry measurements are employed every 2 to 4 weeks to assist dating of the pregnancy and estimating fetal weight. Factors such as head circumference, biparietal diameter, femur length, or abdominal circumference, can help calculate gestational age and fetal weight.
Prenatal ultrasonography can detect major fetal anomalies as early as 16 weeks gestation. These include hydrocephaly, anencephaly, myelomenigocele, congenital heart defects, gastrointestinal or genitourinary abnormalities.
Maternal serum alpha-fetoprotein (MSAFP) elevations can result from open neural tube defects such as spina bifida or anencephaly, when the protein leaks from the fetal tissue into the amniotic fluid through the amniochorion and into the maternal system. A lower-than-expected MSAFP suggests Down syndrome, Trisomy 18.
The measurement of maternal serum alpha- fetoprotein concentration is a useful screening test for the diagnosis of open neural tube defects in the etus.
Postnatal
Fluorescence in situ hybridization (FISH) has a higher resolution than chromosome

analysis and is excellent in detecting missing, extra, or relocated chromosomal regions. However, it does not detect single-gene mutations, and it requires that the physician request testing for a specific region, or regions, of interest.

FISH is useful for identifying contiguous gene deletions, as can be seen in Prader- Willi, Angelman, Williams, Smith-Magenis, and Miller-Dieker syndromes.
Microarray-based comparative genomic hybridization (CGH) is a revolutionary testing strategy for the detection of DNA copy number variations throughout the human genome that may be associated with disease.
Comparative genomic hybridization has replaced high-resolution chromosome analysis to screen patients suspected of having a chromosome abnormality in most cases except
Chromosomal abnormalities
Autosomal
Clinical features of Downs syndrome include:

Craniofacial: flat face, epicanthal folds, upward slanting palpebral fissures, Brushfield spots, flat nasal bridge, small dysplastic ears.

CNS: hypotonia, poor Moro reflex

CVS: Endocardial cushion defects, VSD, ASD, PDA, pulmonary hypertension

MSK: short fifth digit with clinodactyly, joint hyperflexibility, short neck, redundant skin, single tranverse palmar crease, wide gap between 1st and 2nd toes, short sternum

GI: Duodenal atresia, annular pancreas, tracheoesophageal fistula, hirschsprung disease, imperforate anus

Children with Trisomy 21 are more prone to congenital heart defects (50%) such as atrioventricular septal defects, ventricular septal defects, isolated secundum atrial septal defects, patent ductus arteriosus, and tetralogy of Fallot. Congenital and acquired gastrointestinal anomalies and hypothyroidism are common Other abnormalities include leukemia, immune dysfunction, diabetes mellitus, and problems with hearing and vision. Most males are sterile, but some females have been able to reproduce.
Prominent features of trisomy 13(Patau syndrome) include cleft lip,cleft palate, cutis aplasia, microcephaly, microphthalmia, capillary hemangiomas, deafness, congenital heart defects(VSD, PDA, ASD), hypoplastic nails, clinodactyly, polydactyly, renal abnormalities and severe developmental delays. Only 5% live >6

months.

DNA point mutations (substitutions, deletions, or insertions of a single base-pair) can cause a wide range of outcomes, some of which are dramatic. Point mutations can result in the production of nonfunctional proteins (nonsense mutations), different amino acids than expected (missense mutations), and alterations in the amino acid or protein that have no functional significance (silent mutations).
Some examples of a point mutation causing severe disease is that seen in sickle cell disease (SCD), achondroplasia.
The risk of having another child with Down syndrome is greater for a young woman who is a balanced translocation carrier than for a middle-age woman.
If a structural chromosomal abnormality is found in an infant (e.g., unbalanced translocation),this necessitates chromosomal analysis of both parents if future children are planned or possible.
There are associated risk factors with subsequent pregnancies when an infant is born with a chromosome abnormality
Contiguous gene syndromes include Prader Willi, Angelman, Beckwith Wiedemann, Di George, Smith-Magenis, and Miller-Dieker Syndromes.
Prader Willi syndrome: Major criteria include: neonatal hypotonia with poor suck, feeding problems, and FTT in infancy; onset of rapid weight gain between 12 months and 6 years of age, which causes obesity; hyperphagia, hypogonadism; characteristic facial features (dolichocephaly, almond-shaped eyes, down-turned mouth); and developmental delay with mental retardation. Molecular genetic testing confirms the diagnosis. Prader-Willi syndrome provides an example of the effects of genetic imprinting or the differential expression of genetic information, depending on the parent of origin. The condition results from the lack of a paternally derived gene normally present on chromosome 15.
Angelman syndrome: results when information on the maternally derived chromosome 15 is either missing or malfunctioning. Although genetically considered a “sister” syndrome to Prader-Willi syndrome, the two disorders have very different clinical manifestations. Referred to as “happy puppets,” children who have Angelman syndrome are characterized by severe mental retardation, midface hypoplasia, prognathism, seizures, uncontrollable bouts of laughter, poor or absent speech and hypotonia.

Di George: Deletion of 22q11. Affected infants often present with hypocalcemic tetany in the newborn period. Cardiac malformations include interrupted, right- sided, or double aortic arch, truncus arteriosus, pulmonary atresia, VSD, TOF. Facial dysmorphic features include hypertelorism; antimongoloid slant of the eyes, midline facial clefts, micrognathia, and shortened philtrum of the lip, cleft palate,

and choanal atresia, low-set posteriorly rotated ears, and notched ear pinnae. Can have T-cell deficiency. Moderate–to-severe developmental delays, renal anomalies, and dysphagia are common, and hearing loss may be present. Autoimmune disease, including autoantibody-mediated cytopenias, thyroid disease, and juvenile rheumatoid arthritis.

Sex chromosomes

A buccal smear is insufficient to diagnose Turner Syndrome (TS).

The definitive diagnosis of Turners requires a karyotype which should evaluate at least 30 cells (metaphases). Some girls who have TS may have mosaicism, with some cells having normal chromosomes and others having a TS karyotype. Therefore, if the initial chromosome analysis produces normal results, and there still is concern for the diagnosis, an extended FISH study should be performed to evaluate for low-level mosaicism.

Features in the newborn infant of Turner's include small for gestational age, webbing of neck, protruding ears and lymphedema of hands and feet, although many newborns can be phenotypically normal.

Congenital heart defects (coarctation of aorta and bicuspid aortic valve), structural renal abnormalities (horse shoe kidneys), MSK increased carrying angle of elbow, shield chest, Madelung deformity, scoliosis, short 4th metacarpal, widely spaced nipples are some other features in Turner syndrome.

All girls with Turner Syndrome will have gonadal dysgenesis; the gonads are usually streaks of fibrous tissue. There is primary amennorhea, lack of secondary sexual characteristics and infertility.

Older children and adults may manifest with short stature. Growth retardation may be the only clinical sign of Turner Syndrome.

Growth hormone may be an effective treatment for Turner syndrome.

A diagnosis of Klinefelter syndrome is often made at puberty. The testes and penis remain small, and secondary sexual changes fail to occur. They have tall stature. Testosterone levels are low, and follicle-stimulating hormone (FSH) and luteinizing hormone (LH) are elevated. Men with Klinefelter syndrome may have normal sexual function, especially when treated with testosterone supplementation, but are infertile. Developmentally, boys with Klinefelter syndrome may have some minor motor and verbal delays, and increased rates of learning disorders and ADHD The diagnosis of Klinefelter syndrome is made by karyotype examination of chromosomes.

Allergic and immunologic disorders

General

Epidemiology

Prevalence is 6%–8% in pediatric population

Most common allergens in children: Milk, eggs, peanuts, tree nuts, walnut, soy, and wheat

Prevention by dietary restriction

Breastfeeding for the first six months after birth with supplementation with a complete, nutritionally balanced hypoallergenic formula will decrease the severity and delay the onset of allergic disease.

Relation to environmental exposure

Early exposure to tobacco smoke, especially the mother's, increases the frequency of asthma in children.

Passive exposure to cigarette smoke exacerbates asthma and allergic rhinitis.

Influence of genetics on development of allergy

Several major groups of genes are associated with allergic diseases: genes that regulate systemic expression of atopy (increased IgE synthesis, eosinophilia, mast cell responses) that are commonly expressed among various allergic diseases, genes that control barrier function in specific target organs , and genes encoding pattern-recognition receptors of the innate immune system.

Atopic diseases have a strong familial predisposition, with approximately 60% heritability found in twin studies of asthma and atopic dermatitis.

Children with one component of atopy syndrome (allergic rhinitis, asthma, eczema) have a threefold greater risk of developing a second component.

Allergic rhinitis

There is an association between allergic rhinitis and sinusitis and/or otitis media.

Perennial allergic rhinitis is usually caused by indoor allergens such as dust mites and animal danders. Cockroach is probably the most important allergen for inner city children.

Treatment of allergic rhinitis includes allergen avoidance, antihistamines, and intranasal corticosteroids.

Environmental control measures to reduce exposure to dust mites include removal of carpeting, reduce indoor humidity, encase bedding in allergen–impermeable covers, washing bed linen every week in hot water (>130F). Avoid furred pets, Use HEPA filters in living areas to reduce exposure to mold. Control timing of outdoor exposure to reduce exposure to pollen.

Second generation antihistamines like Cetirizine, Levocetrizine, Fexofenadine, Loratidine and Desloratidine are preferable for treatment in children. It has much less sedative side effect.

Intranasal corticosteroids are the most effective therapy for allergic rhinitis. These include nasal sprays like fluticasone, Mometasone and Budesonide.

Asthma

Etiology, epidemiology, natural history

Characteristics of exercise-induced asthma: coughing and wheezing after 5 to 6 minutes of exercise, gradual improvement after 20 to 30 minutes of rest.

Smog increases the severity of exercise-induced asthma.

Asthmatic patients may have exaggerated obstructive responses to exercise, viral upper respiratory infection, allergen exposure, weather changes, smoke pollutants and other irritants, aspirin, and beta adrenergic blocking drugs.
Children with early-onset asthma (< 3 years of age) who have a parental history of asthma, a confirmed diagnosis of atopic dermatitis, or sensitization to aeroallergens are unlikely to outgrow asthma.
Asthmatic patients have bronchial hyperresponsiveness, with exaggerated bronchial obstructive response to environmental changes.
IgE-mediated allergen challenge induces immediate obstruction, which resolves and is followed in 4-12 hours by persistent late-phase obstruction.

Both viral infections and airborne allergens can precipitate a biphasic response .The IgE-mediated "early-phase" or "immediate" response to an allergen challenge causes mast cells and basophils to degranulate, precipitating bronchospasm as well as the release of proinflammatory cytokines and chemokines. This cascade of inflammatory responses results in the subsequent "late-phase" obstruction of air flow, which occurs 4 to 12 hours following exposure to the environmental insult.

There is an increased frequency of recurrent wheezing in infants with RSV bronchiolitis.
Asthma Education Recommendations for Patients Discharged from the Emergency

Department or Hospital:

Provide a written asthma action plan
Train for self-monitoring (e.g., peak flows)
Educate and reinforce correct inhaler technique
Refer to an asthma education program
Involve patients in decision-making processes
Schedule telephone and office follow-up
Consider the home environment (tobacco, allergens, etc)
Schedule telephone and office follow-up

Diagnosis

Atelectasis associated with an acute exacerbation of asthma does not usually require bronchoscopy, antibiotics, or chest physiotherapy.
The signs of severe obstruction in an acute exacerbation of asthma are severe retractions, inability to speak whole phrases, cyanosis(Pa02<60mm Hg), PC02

>42mm Hg ,quiet breath sounds in presence of other signs of obstruction, peak expiratory flow rates less than 30% of predicted, increased respiratory rate

>30breaths/min , pulse rate >120, pulsus paradoxus, mental status changes.

The majority of asthmatic school-age children have positive immediate type allergy skin tests.
The new guidelines for assessing Asthma severity separate children into three groups: birth through 4 years, 5 through 11 years, and 12 years and older.
Characteristics of intermittent asthma:
daytime symptoms <2 days/week, nighttime symptoms 0-4yr: 0,>5yr:<2 x/month, short acting B2 agonist use:<2days/week, no limitation of normal activity, FEV1 Normal.

Characteristics of persistent asthma based on asthma severity are as follows
Mild: >2day/wk but not daily, nighttime awakening 0-4 yr: 1-2/month; >5yr:

3-4x/month, short acting B2 agonist use : >2days/wk but not daily, not more than 1 time/day ,minor limitation with normal activity, FEV1 >80% predicted.

Moderate: Daily, nighttime awakening 0-4yr: 3-4x/month; >5yr:>1x/week but not daily, short acting B2 agonist use: Daily, some limitation with normal activity, FEV1 60-80% predicted.
Severe: Through out the day, nighttime awakenings: 0-4 yr: >1x/week; >5yr: often 7x/week, short acting B2 agonist use: several times/day, FEV1 <60% predicted.

Treatment

Exercised-induced asthma may be a sign of poorly controlled asthma.
Initial management of a patient presenting with an acute exacerbation of asthma should be aimed at decreasing airway obstruction, correcting hypoxemia, and preventing further progression of symptoms. Immediate goals include close patient monitoring, oxygen supplementation to maintain oxygen saturation at 90% or greater, and administration of bronchodilators. Inhaled beta2 agonist via nebulization or a metered-dose inhaler administered every 20 minutes for 1 hour should be the initial therapy. Oxygen should be continued at least for 20 minutes after SABA administration to compensate for V/Q abnormalities caused by SABA's.
Inhaled anticholinergics may provide additional bronchodilation and have been shown to improve lung function and reduce hospitalization rates.
Administration of intravenous or oral corticosteroids is recommended during an asthma exacerbation; both delivery methods result in similar outcomes in children hospitalized for asthma.
The appropriate treatment for a patient with an acute exacerbation of asthma that is severe and unresponsive to adrenergic agonist therapy include supplemental oxygen, intravenous corticosteroids. Increase frequency of SABA delivery: can be given continuously (5-15mg/hr) or every 20 minutes to every 1 hour. Watch hydration status, administration of fluids at maintenance is recommended. Parenteral epinephrine, B-agonists (Terbutaline sulfate), methylyxanthines, Magnessium sulphate, inhaled heliox may be required for severe asthma. Respiratory failure may require intubation and mechanical ventilation.
Chest physical therapy, incentive spirometry and mucolytics are NOT recommended during acute exacerbations- they trigger severe bronchoconstriction.
Corticosteroids in an acute exacerbation of asthma can increase adrenergic response, improve FEV1, and improve oxygenation.
The clinical manifestations of toxicity to adrenergic agonists: muscular tremor, tachycardia, hypokalemia, irritability.
The benefits of inhaled corticosteroids (ICS): It is the recommended therapy for all patients with persistent asthma. It improves lung function and reduces asthma symptoms. They help reduce ER visits, hospitalizations, prednisone use for asthma by 50%.May also lower risk of death due to asthma.
Risk of adverse effects from ICS therapy is related to the dose and frequency with which they are used. High doses (> 1,000 mcg/day) and frequent use (4 times/day) are more likely

to have local and systemic effects. Local effects include: oral candidiasis, dysphonia. There is a risk of growth suppression and osteopenia.
The characteristics of a child at increased risk of ICU hospitalization because of asthma include: one or more life-threatening episodes, severe asthma requiring chronic steroids, poor control of daily symptoms, abnormal FEV1, poor adherence, depression and stress.
The kinetics of short-acting beta agonists(SABA's) and long-acting beta-adrenergic agonists (LABA's).
LABA's like Salmeterol (prolonged onset of action, onset of action is within one hour).Formoterol (shorter onset of action within 5-10 minutes). Both have prolonged duration of action, at least 12 hours. Only use for patients who have suboptimal control with ICS.
SABA's like albuterol, levalbuterol (less tremors, less tachycardia), terbutaline, pirbuterol have short onset of action 30 minutes -2 hours, duration of action 3-4 hours.
Step-up, step down approach: Start with higher level controller therapy, once good

asthma control has been achieved then can decrease the number and dose of daily controller medication. Wait at least 3 months if patient is well controlled. If patient is poorly controlled, treatment goes up 2 steps, plus short course of oral corticosteroids, with reevaluation in 2 weeks. Other treatments that can be used in Step 2 include Cromolyn or Montelukast. Theophylline can be added on for patients

>12 years.

Excessive daily use of beta adrenergic agonists has been associated with increased mortality and with diminished symptom control in asthma.
Corticosteroids interfere with the late-phase but not the immediate response to allergen exposure.
Long-term treatment with inhaled corticosteroids decreases bronchial inflammation and bronchial hyper-responsiveness.
Leukotrienes cause mucous secretion, airway edema, bronchospasm. Leukotriene receptor antagonists (LTRA's) Montelukast and Zafirlukast help by causing bronchodilation, anti-inflammatory effects, reducing exercise, aspirin and allergen induced bronchospasm. Montelukast (children>1yr, once daily). Zafirlukast (>5yr, twice daily).Use as alternative treatment in mild persistent or add on for moderate persistent with ICS.
Medical and self-assessment in a patient with asthma: Two to four asthma checkups per year are recommended for reassessing and maintaining good control. Asthma control can be assessed by determining the: (1) frequency of asthma symptoms during the day, at night, and with physical exercise; (2) frequency of "rescue" SABA medication use and refills; (3) quality of life; (4) lung function measurements for older children and youths; (5) number and severity of asthma exacerbations; and

(6) presence of medication adverse effects since the last visit. Peak expiratory flow monitoring is feasible in children as young as 4 years who are able to master this skill. Use of a stoplight zone system: The green zone (80-100% of personal best) indicates good control; the yellow zone (50-80%) indicates less than optimal control and necessitates increased awareness and treatment; the red zone (<50%) indicates poor control and an exacerbation requiring immediate intervention.

Patient education is very important in asthma management. Outline goals of asthma management. Written asthma management plan with a daily "routine "for regular medication use and also an action plan to manage worsening asthma. Address concerns about potential adverse effects. Teach, demonstrate and have the patient show proper technique for: inhaled medications, use of spacer, peak flow measures.
Routine pulmonary function testing (spirometry) is recommended at least annually and more often if asthma is inadequately controlled.

Atopic dermatitis

Atopic dermatitis (AD) is a chronic, relapsing dermatosis that features dry skin (xerosis), pruritus, and a personal or family history of eczema, allergic rhinitis or allergies, or asthma. Children who have one component of the atopic triad (AD, asthma, allergic rhinitis) are three times as likely to develop a second component.
There is no sex predilection, and the onset frequently is in infancy. Although many affected children outgrow the condition by age 5 years, AD may persist into adolescence and adulthood.

Urticaria, angioedema, anaphylaxis:

Agents that cause urticarial, anaphylaxis and angioedema:

Egg, milk, wheat, peanuts, tree nuts, soy, shellfish, fish, strawberries (direct mast cell degranulation)
Medications
Hymenoptera (honeybee, yellow jacket, hornets, wasp, fire ants), biting insects (papular urticaria)
Infections: Bacterial (streptococcal pharyngitis, Mycoplasma, sinusitis); viral (hepatitis, mononucleosis [Epstein-Barr virus], coxsackievirus A and B); parasitic (Ascaris, Ancylostoma, Echinococcus, Fasciola, Filaria, Schistosoma, Strongyloides, Toxocara, Trichinella
Contact allergy
Latex, pollen, animal saliva, nettle plants, caterpillars
Chronic urticaria rarely responds favorably to dietary manipulation. Removal of recognized urticarial aggravators. The mainstay of therapy is the use of nonsedating or low-sedating H1 antihistamines. In those patients not showing response to standard doses use higher than recommended doses of H1 blockers. The combined use of H1- and H2 antihistamines is helpful to control chronic urticaria when H1-type alone at higher doses do not work .Doxepin is an antagonist of both H1 and H2 receptors and can be helpful, but its usefulness is limited by adverse effects. H2-type antihistamines alone may exacerbate urticaria. Antileukotriene agents in combination with antihistamines are occasionally helpful. If hives persist after maximal H1- or H2-receptor blockade has been achieved, a brief course of oral corticosteroids may be considered. Immunomodulatory agents that have been used but effectiveness unknown, include hydroxychloroquine, sulfasalazine, colchicine, dapsone, mycophenylate and omalizumab (anti-IgE).Intravenous immunoglobulin, and plasmapheresis have been used to treat autoimmune chronic urticaria refractory to other therapies.

Chronic urticaria does not warrant allergy testing.

Anaphylaxis is highly likely when any one of the following three criteria is fulfilled:

Acute onset of an illness (minutes to several hours) with involvement of the skin or mucosal tissue (e.g., generalized hives, pruritus or flushing, swollen lips, tongue, uvula)

AND AT LEAST ONE OF THE FOLLOWING:

Respiratory compromise (dyspnea, wheeze, stridor)

Reduced BP or associated symptoms of end-organ dysfunction

Two or more of the following that occur rapidly after exposure to a likely allergen for that patient (minutes to several hours):

Involvement of the skin/mucosal tissue (e.g., generalized hives, itch/flush, swollen lips/tongue/uvula).

Respiratory compromise

Reduced BP following exposure to known allergen for that patient (minutes to several hours):

Infants and children: low systolic BP (age-specific) or >30% drop in systolic BP Anaphylaxis is a medical emergency requiring aggressive management with:

Intramuscular or intravenous epinephrine (0.01 mg/kg up to 0.3 mg)
Intramuscular or intravenous H1 and H2 antihistamine antagonists
Oxygen, intravenous fluids
Inhaled β-agonists
Corticosteroids (methylprednisone, prednisone)

More than 90% of biphasic responses occur within 4 hours, so patients should be observed for at least 4 hours before being discharged from the emergency department.

Adverse reactions to substances

Food
Common foods causing allergic reactions include milk, soy, eggs, peanuts, seafood, wheat, and tree nuts.
It is important to distinguish between anaphylaxis and food poisoning.
Some patients with moderate or severe eczema have positive skin tests to food, may or may not have acute symptoms on ingesting these foods, and experience improvement in their eczema after eliminating these foods.

Food allergies can be present in up to 40% of patients who have Atopic Dermatitis (AD). Symptoms include pruritus, urticaria, contact dermatitis, and exacerbation of AD as well as wheezing, asthma, and anaphylaxis. The symptoms can be immediate or delayed. Allergy skin prick testing usually is more reliable after age 2 years; specific radioallergosorbent testing can be performed in infancy.

More than 90% of food-allergic individuals demonstrate clinical responses to only 1 or 2 foods. The most common food allergens in the pediatric population include cow milk, eggs, peanuts, tree nuts, soy,wheat, peanuts, tree nuts, fish, and shellfish are implicated most commonly in adults.
Most milk, egg, and soy allergies are outgrown by 5 years of age.
Most allergies to peanuts, tree nuts, and seafood are not outgrown.

Drugs

Contrast media
Reactions to contrast media (RCM) are not IgE mediated and can be prevented by pre-treatment with corticosteroids and antihistamines. This can reduce the reaction risk to less than 1%.
The osmolarity of the solution is the primary cause. Desensitization is a procedure used to allow safe administration of medications that cause IgE-mediated reactions. It currently does not have a role in the treatment of RCM reactions because most of these reactions are non-IgE-mediated.

Issues in diagnosis and treatment of allergic disease

Skin testing

Indications for immediate type skin testing include:

Identification of aeroallergen triggers in patients who have asthma
Allergic rhinitis that is not controlled with usual medications or if specific avoidance (e.g., pet dander) is desired
Food allergy
Insect sting allergy
Vaccine, drug, or latex allergy
Evaluation for moderate-to-severe atopic dermatitis
Other conditions, including allergic fungal sinusitis, allergic bronchopulmonary aspergillosis, and eosinophilic esophagitis
Current guidelines recommend that patients who experience less severe reactions (local or normal reactions, large local reactions, or cutaneous-only symptoms in children younger than than 16 years of age do not require testing because of their low risk for future anaphylaxis.
Antihistamines are the most commonly used medicines that alter the results of allergy skin testing.

In vitro testing

RAST (radioallergosorbent test) testing correlates closely with results of skin tests.
In vitro testing is indicated:
1) when antihistamines cannot be stopped for allergy skin tests
2) for patients who have severe atopic dermatitis or dermatographism
3) Those that have experienced life-threatening reactions to the allergen

Immunotherapy

Immunotherapy is most effective in treating allergic rhinitis.

Allergen immunotherapy (AIT) is currently used for the treatment of allergic asthma, allergic rhinitis, allergic conjunctivitis, and stinging insect hypersensitivity.
Anaphylaxis is a risk during Allergen Immunotherapy. A mandatory waiting time of

30 minutes after administration, prompt recognition of symptoms, and prompt administration of intramuscular epinephrine are key steps to preventing the rare outcome of fatal anaphylaxis. Other treatment should include supplemental oxygen, oral or intramuscular antihistamine, oral corticosteroids, and nebulization therapy with a short-acting beta-2 agonist.

Other drawbacks to AIT include requiring patients to receive therapy in a physician's office, discomfort from a subcutaneous injection, and local erythema and swelling at the injection site. Local reactions do not portend more severe reactions on subsequent injections, but they can reduce adherence with AIT if large or bothersome.

Immunodeficiency disorders

Presenting signs and symptoms of potential immunodeficiency
The clinical characteristics of antibody deficiency syndromes present after 4 to 6 months of age and include severe first infections and chronic and recurrent bacterial infections in more than one anatomic site. The history of recurrent sinusitis, pneumonia, and decreased immunoglobulin concentrations should prompt evaluation for a possible humoral immunodeficiency.
The clinical characteristics of cellular immunodeficiency present in the first few

months after birth with failure to thrive, chronic diarrhea, overwhelming infections with viral, bacterial, and opportunistic infections.

Leukocyte Adhesion Deficiency (LAD) 1, 2 AND 3 are rare autosomal recessive disorders chararcterized by recurrent bacterial and fungal infections associated with a lack of pus formation, in spite of blood neutrophilia. There is usually a history of delayed separation of umbilical cord with omphalitis of cord stump. Typical signs of inflammation may be absent.
Chediak-Higashi syndrome is a rare autosomal recessive disorder characterized by increased susceptibility to infection, mild bleeding diathesis, partial oculocutaneous albinism and progressive peripheral neuropathy. Patients have light skin, silvery hair. Frequent infections involving mucous membranes, skin and respiratory tract, most common being S. aureus infection. The most life threatening complication is hemophagocytic lymphohistiocytosis (HLH).

Screening tests

Laboratory evaluation of antibody function include
Quantitative immunoglobulin concentrations.
Antibody response to protein (diphtheria tetanus toxoid) and polysaccharide vaccines (23-valent pneumococcal vaccine). Measuring baseline antibody titers, vaccinate the patient, and repeat antibody measurements in 3 to 4 weeks. The recommended appropriate protein vaccine response in adolescents consists of a fourfold increase in titers.
Exclusion of other causes of hypogammaglobulinemia

Genetic analysis for mutations in the Bruton tyrosine kinase (Btk) gene. Patients who have low-to-absent B-cell concentrations or who are younger than 2 years of age should be evaluated for X-linked recessive (Bruton) agammaglobulinemia.
Many clinicians assess IgG subclasses (i.e., IgG1, IgG2, IgG3, and IgG4), but the clinical significance of a low IgG subclass value is unclear
Laboratory evaluation of cell-mediated immunity includes tests for lymphocyte counts and lymphocyte function.
Baseline CBC.
Flow cytometry uses technology that can detect specific cell surface markers of T cells (CD3, CD4, CD8), B cells (CD19), and natural killer (NK) cells (CD16, CD56).
Lymphocyte proliferation studies.
Anergy testing for delayed-type hypersensitivity is a good qualitative screening of the cellular immune system but is limited by higher false-negative rates in infants compared to older children.
Measurement of T-cell receptor excision circles (TRECs) has been evaluated. TRECs are formed during the normal maturation of T cells; low-to-absent TRECs are consistent with T-cell lymphopenia.

Infectious Diseases

Public health considerations; prevention of infectious diseases
In child-care centers

Children in childcare centers may reflect or even cause infectious disease in the community.

A child should be excluded from daycare if there is a chance to decrease secondary cases, and he/she has any condition suggesting the possibility of acute illness (fever, lethargy, or any other condition that causes difficulty in participating with daily activities).

A child should not be excluded from daycare if child has had a febrile respiratory

illness.

The most important way to prevent transmission of pathogens in childcare centers is with hand hygiene.

Hospital and office infection control

Standard precautions (e.g. hand hygiene) are warranted with all patient care.

Airborne precautions differ from standard precautions by requiring a single room that is negative pressure to provide airborne infection isolation and a N95 mask for protection (airborne precautions are required when dealing with mycobacterium tuberculosis, varicella, and measles).

Droplet precautions differ from standard precautions by requiring a single room and the use of a protective mask (droplet precautions are required when dealing with adenovirus, strep pyogenes, influenza, nisseria meningitides, mycoplasma pneumonia, parvoviurs B19, Bordetella pertusis, rhinovirus).

Contact precautions differ from standard precautions by requiring a single room, along with the use of a gown and gloves (contact precautions are required when dealing with clostridium difficile,

enteroviruses, hepatitis A, herpes simplex, herpes zoster, multidrug-resistant organisms, noncontained abscess, cellulites, or decubitus ulcer, parainfluenza virus, RSV, staph infections, salmonella, scabies, shigella).

Recognize that all office and hospital personal should have influenza immunizations annually.

Through breast-feeding

Bacterial infections that should interrupt breast-feeding include: tuberculosis, staph or group A strep (of note, once appropriate antibiotics have been initiated then breast- feeding may resume).

Chronic viral infections that should interrupt breast-feeding include: HTLV-1 or -2, HIV, CMV, West Nile Virus, and viruses that cause skin lesions which may come in contact with a breast-feeding infant (lesions on the mother's nipple or breast)

Recognize that human milk provides protection against many gastrointestinal and respiratory infections.

Medical evaluation of internationally adopted children

For internationally adopted children who have a history of questionable medical care antibody tests to some vaccines are available to assist in evaluating immunization status (e.g. tetanus and measles).

The validity of immunizations records for internationally adopted children may be deemed valid if the number of doses, interval between doses, and age at immunization are appropriate.

Prevention of vector-borne diseases

Measures to prevent tick-borne and mosquito born infections include the use of protective clothing (e.g. long-sleeved shirts, pants, and socks), mosquito netting during sleep, along with permethrin-impregnated clothing and fabrics.

When applying topical insect repellants precautions that should be used include applying low concentrations of DEET (<30%), along with avoiding any application to children's hands for fear of ingestion.

Prevention of infection associated with recreational water use

- Contaminated recreational water by be a source for pathogen transmission.

Pool-associated outbreaks of acute gastroenteritis may be caused by pathogens that are resistant to chlorination.

Precautions that should be implemented to prevent infections from recreational water use include:

Proper chlorination of the water, avoid ingesting water, proper hand hygiene, monitoring children that are not toilet-trained with frequent bathroom breaks and checks for soiling.

Antibiotics
Aminoglycosides

Major toxicity associated with aminoglycosides includes:

Ototoxicity (specifically high frequency hearing loss), vestibular toxicity, nephrotoxicity.

To modify aminoglycoside doses one should take into account the bactericidal properties of these drugs are dependent on peak serum concentration.

Aminoglycosides are used to treat serious community acquired infections and most nosocomial gram negative bacterial organisms (including neonatal meningitis, community acquired UTIs in older children).

Beta-lactam antibiotics

Mechanism of action involves inhibiting bacterial wall synthesis, by binding to penicillin-binding proteins.

Third generation Cephalosporins provide broad spectrum coverage, as well as good penetration into CSF.

Advantages of Cephalosporins over aminoglycosides include:

Greater activity in deep-tissue infections, less toxicity, avoidance of need to monitor renal function and drug concentrations.

Recognize that penicillinase-resistant pencillins are used to treat pencillinase- resistant strains of staphylococci.

Ampicillin and amoxicillin should be used when treating gram positive penicillins susceptible bacteria along with some gram negative organisms (e.g. Escherichia coli, H. flu, Proteus, Salmonella, and Shigella).

Second generation cephalosporins are used as second-line options to treat skin, soft-tissue, AOM, respiratory infections, pneumonia, and acute bacterial sinusitis .

First generation cephalosporins are not used to treat infections involving the CSF due to poor penetration into the cerebrospinal fluid.

Clindamycin

Adverse reactions to clindamycin include Clostridium difficile enterocolitis.

Uses for clindamycin include:

Anaerobic infections (e.g. Bacteroides), Staph and Strep infections (exceptions include enterococci).

Uses for macrolides include: Erythromycin can be used to treat:

Individuals with allergies to penicillin, along with more severe infections (e.g.

atypical pneumonia) due to slightly broader coverage and efficacy than penicillin.

Adverse reactions to macrolide antibiotics include:

Macrolides may cause altered cardiac conduction in rare cases, along with nausea in certain individuals. Also of note erythromycin is contraindicated in infants due to a relationship with pyloric stenosis.

Azithromycin can be used to treat:

Pertussis at any age, community acquired pneumonia, sinusitis, acute otitis media, strep pneumonia (if penicillin allergy), atypical pneumonia, and sexually transmitted disease.

Clarithromycin can be used to treat:

Pertussis after 1 month of age, disseminated mycobacterium avium-intracellulare complex (MAC), along with all the above mentioned bacteria mentioned for Azithromycin.

Rifampin

Can be used forChemoprophylaxis for close contacts of known invasive meningococcal infections (e.g. neisseria meningitis), as a part of standard tuberculosis treatment regimens, in combination with vancomycin to treat staph infections, in combination with beta lactam antibiotics to eliminate persistent group A strep pharyngitis.

Quinolones

Should be used when a pathogen is multidrug-resistant and there is no safe and effectivealternative (examples include urinary tract infections or osteomyelitis caused by pseudomonas aeruginosa); also can be used to treat or prevent anthrax.

Tetracyclines

Generally used for treatment of UTIs in children older than 8 years of age, tetracyclines most common use today is for the treatment of severe acne and rosacea.

Trimethorpim with sulfamethoxazole Can be used to treat:

Pneumocystis, along with other infections with adequate susceptibility (including acute UTI, IBD, burns, nongonocaccal urtheritis due to Chlamydia, toxoplasmosis, bacterial conjunctivitis, stenotrophomonas matophilia, Burkholderia cepacia, Burcella, and methicillin resistant staph aureus).

Recognize that the major adverse reactions associated with trimethoprim sulfamethoxazole include: rash, neutropenia, Stevens-Johnson syndrome.

Vancomycin

Originally used to treat penicillin resistant staphylococcus aureus, vancomycin is now used for broad spectrum coverage typically as a last line antibiotic in order to help limit growing resistance to the drug.

Most common adverse reaction associated with vancomycin is Red Man Syndrome (reddening of the skin due to release of histamine associated with vancomycin administration).

Note that inappropriate use of vancomycin may result in resistance especially with organisms such as enterococci and staph aureus.

Antivirals
Antivirals can be used to treat varicella zoster and herpes (HSV 1 and 2) infections.
Recognize the major adverse reactions associated with acyclovir include gastrointestinal symptoms (nausea, vomiting, and diarrhea) and malaise. Of note rare cases of acute renal failure due to acyclovir precipitation in the renal tubules has been reported.
Oseltamivir and Zanamivir are commonly used to decrease the duration of symptoms of the influenza virus. These two medications should be administered within 24 hours of symptom onset.

Antiparasitics

Metronidazole should be used in the treatment of anaerobic bacteria, microaerophilic bacteria, clostridium, trichomonas vaginalis, treponema pallidum, garderella vaginalis, oral spirochetes, H. Pylori, campylobacter fetus, and certain other parasitic infections.

Mebendazole should be used in the treatment of various types of amebic infections including ringworm.

Permethrin should be used in the treatment of scabies.

Malathion should be used in the treatment of lice, due to its ovacidal activity.

Chloroquine should be used in the treatment of malaria.

Adverse effects associated with choloroquine include gastrointestinal problems, itch, headache, and blurred vision.

Mefloquine is administered in the treatment of malaria.

Atovaquone/proguanil is used to help treat or prevent pneumocystis pneumonia, malaria, toxoplasmosis, or babeosis.

Antimicrobial resistance

One way antibiotic resistance has developed in the community includes the use of excessive antibiotics.

Excessive use of cephalosporins has contributed to broad-spectrum antibiotic resistance in nosocomial pathogens.

If treated by antibiotics children are at increased risk of becoming carriers to resistant strains of bacteria.

Antimicrobial treatment is not indicated in a variety of pathologies including: bronchitis, middle ear effusion of short duration, most cases of pharyngitis (exception includes group A strep), mucopurlent rhinitis of short duration.

Antifungals

Amphotericin B

Typically administered in one daily dose, amphotericin B is useful in treating a wide array of fungi (e.g. Candida, aspergillus, zygomycetes, histoplasomsis, coccoides immitis.) Adverse effects of amphotericin B include hypokalemia and multisystem toxicity (especially in the kidneys.)

Fluconazole

Maybe used to treat tinea capitis in any age group, used in the treatment of oral candidiasis refractory to nystatin drops.

Griseofulvin

Should be used in the treatment of tinea capitis, or ringworm of the head (most commonly caused by trichophyton tonsurans) in children over 2 years of age.

Of note when administered in the recommended 8 week course no assessment of liver enzymes is necessary in patients with no current history of liver disease.

Adverse effects with griseofulvin are typically mild and may include gastrointestinal upset or rash (may cause disulfiram reaction when used concomitantly with alcohol, and may have cross reactivity between penicillins).

Infections in immunocompromised hosts

Malnutrition is a significant risk factor for morbidity and mortality associated with infections.

- it is estimated that 35% of child deaths are caused by malnutrition which includes lack of macro and micro nutrients, lack of adequate caloric intake, and lack of breastfeeding. Also of note, infection in malnourished children is associated with long term cognitive impairment and poor school performance.

Central nervous system disease

Neonates and children may not manifest fever in central nervous system diseases because the thermoregulatory center of the hypothalamus may be immature/abnormal.

Asplenia

Children with asplenia are at increased risk to have complications, including possible death, from infection with encapsulated organisms.

Malignancy

In order to diagnosis pneumonia in immunocompromised host aggressive procedure (e.g. bronchoscopy) may be required.

Neutropenic patients that become febrile are at high risk for bacteremia. Causes of infections in neutropenic patients are caused by a wide variety of organisms including staphylococci and enteric gram negative organisms, therefore treatment should include broad spectrum antibiotics such as cefepime.

Specific viral pathogens
Cytomegalovirus

Overall CMV can be congenital (1-2% of newborns worldwide) or acquired (~100% population will have acquired CMV by 70 years of age).

Congenital CMV- gold standard for diagnosis is to detect the virus in a urine (or saliva) sample by cytopathic effect in tissue culture.

Acquired infection may be detected by seroconversion (presence of IgM antibodies). Of note, due to the presence of IgM in acute and reactivated infection the gold standard to detect acute infection is by the presence of anti-CMV lgM antibodies and anti-CMV low avidity IgG antibodies.

Recognize that once an individual is CMV-seropositive they will always be a carrier and may shed the virus through saliva, urine, and genital secretions.

Recognize that CMV infection may cause severe illness in an immuno-compromised host. Clinical manifestations of postnatally-acquired CMV infection may include “blueberry muffin” syndrome due to extramedullary hematopoiesis, biliary obstruction, and in 66% of infected children neurologic symptoms (e.g. seizures, chorioretinitis, hypotonia, poor suck). 80-90% of CMV infections, congenital and acquired, are asymptomatic.

CMV may be spread through a variety of different methods including: horizontally (virus-containing secretions via person-to-person transmission), vertically (mother to infant), transfusions andr transplantations.

Definitive diagnosis of congenital CMV infection must be established within the first 3 weeks of life because after this period it will be unclear whether the infection was congenitally acquired or due to exposure in the maternal cervical vaginal tract.

Epstein-Barr virus (EBV)

In immune-competent hosts EBV infections are typically self-limited. In immune- compromised hosts EBV may cause a variety of pathologies including: virus-associated hemophagocytic syndrome, posttransplant lymphoproliferative syndrome (PTLS), lymphoid interstitial pneumonitis, oral hairy leukoplakia.

Short course (< 2 weeks) of corticosteroids in the treatment of EBV are indicated for only certain complications including: Upper airway obstruction, thrombocytopenia complicated by bleeding, autoimmune hemolytic anemia, seizures, and meningitis. Of note corticosteroids should not be routinely used for uncomplicated cases due to potential oncologic complications associated with EBV.

Recognize that individuals with EBV inappropriately treated with Ampicillin, and less frequently with other beta-lactam antibiotics, will develop a pruritic maculopapular rash up to 80% of the time.

Rash is self-limited and thought to be due to an immune-mediated response that will resolve after discontinuation of the antibiotic.

EBV has the designation of a lifelong infection with over 95% of the world's population being seropositive and chronically infected. More specific epidemiology of EBV includes:

Transmission- EBV is shed in oral secretions and may also be found in male and female genital secretions.

Incubation period- 30-50 days in adolescents and adults and potentially shorter in children.

Communicability- EBV is shed in high concentrations in oral secretions for 6 months after acute infection and then intermittently at lower concentrations throughout life (reactivation and increased shedding of virus later in life is facilitated by immunosuppression).

Clinical manifestations in children vary by age group:

Young children (<4 years) infected with EBV are typically asymptomatic or produce symptoms indistinguishable from other febrile childhood illnesses.

Adolescents and adults have a characteristic course with infectious mononucleosis which includes illness onset over 1-2 weeks, with nonspecific systemic complaints (malaise, fatigue, and low grade fever), accompanied by sore throat, lymphadenopathy, and often with headache, nausea, abdominal pain, and myalgias.

Acute infection can be distinguished from past infection via serologic tests including:

Acute infection: characterized by rapid IgM and IgG antibody responses to viral capsid antigen. IgM may be detected for as long as 4 weeks to 3 months.

Chronic infection: The presence of EBV-determined nuclear antigen (EBNA) help distinguish chronic infection due to the fact they present gradually at 3-4 months after initial infection and stay

present at low titers for life (absent EBNA with other EBV- specific antibodies present implies recent infection).

Management of EBV:

Observation and symptomatic treatment of symptoms (e.g. sore throat), bed rest for debilitating fatigue; infected individuals may continue normal activities, however, most practitioners will advise stopping all contact sports or strenuous activity during the first 2-3 weeks of illness or as long as splenomegaly is present.

Potential complications of EBV in a "normal" host include:

Neurologic symptoms: headache (50% of cases), seizures, ataxia, meningitis, Guillan-barre syndrome, Reye syndrome, transverse myelitis, encephalitis, along

with a rare perceptual distortion of sizes, shapes, and spatial relationships ("Alice- in-Wonderland syndrome" or metamorphopsia).

Hematologic: hemolytic anemia, aplastic anemia, commonly mild thrombocytopenia and neutropenia.

Heterophile antibody test-

can detect antibody in 90% of EBV cases associated with infection mononucleosis in adolescents and adults .

Can detect antibody in 50% of children < 4 years due to low titers of disease (not recommended in this population).

Herpes simplex virus (HSV)

Newborns are more likely to be infected during birth if their mother has a primary herpes infection as opposed to a recurrent genital herpes simplex outbreak.

Diagnosis of HSV can be made by a variety of ways including: viral cultures, Herpes PCR (nucleic acid amplification), Tzanck smear, HSV direct fluorescent antibody assay (DFA)- commonly used as the initial test due to rapid immunologic identification of HSV. Treatment with acyclovir of neonatal herpes simplex infection should begin immediately when infection is suspected, even before test results are available, in order to mitigate sequelae associated with herpes.

HSV can be transmitted from a person with primary or recurrent infection regardless of presence of clinical symptoms.

Clinical presentation of the herpes virus in the neonatal period is varied, and my not include skin lesions. A high degree of suspicion for herpes simplex should be used in dealing with neonates presenting with rule out sepsis or other signs that may point to herpes virus.

Clinical presentation of the herpes virus in children and adolescents typically involves the presence of skin lesions along with neurological symptoms (e.g. seizure, abnormal mentation) if encephalitis is present.

Varicella-zoster virus (VZV)

Know that varicella zoster immune globulin (VZIG) is not recommended for normal infants over the age of 2 days.

In the same family as herpes, the varicella virus can cause two diseases. The first is varicella (chickenpox) which is characterized as a primary illness that causes general symptoms. The second infection is zoster (shingles) which is a secondary infection caused by reactivation of VZV from latency.

Recognize that if VZIG is to be given it should be administered within 96 hours of exposure to varicella in order to be effective.

Epidemiology: The primary mode of transmission for varicella zoster is via skin vesicles to the respiratory tract of susceptible individuals (children who have no humoral or cellular immunity to VZV). The incubation period is anywhere from 10-23 days, and may be spread from infected individuals until vesicles have crusted.

Symptoms of VZV include characteristic vesicles present over infected individual's entire body. Zoster may also present with characteristic vesicles, unilaterally located on an individual's dermatomes.

Individuals who develop VZV should be isolated which includes keeping children out of daycare and school until their vesicles have completely crusted over. If hospitalized, VZV patients should be placed into isolation within negative pressure ventilation rooms.

Congenital varicella infection occurs in approximately 2% of infants born to mother's who acquired the disease during the 2nd or 3rd trimester. Symptoms of congenital varicella infection are varied, but may include cicatricial skin scars, hypoplastic limbs, chorioretenitis, micropthalmos, Horner's syndrome, cataract, nystagmus, zoster, or even early death.

Recognize that individuals who are immunocompromised are at risk of developing severe disease if infected with VZV.

Varicella-zoster immunoglobulin is used to treat individuals who have never had VZV, have been exposed to disease, and are at high risk of developing severe infection.

One should use antivirals to help treat VZV in normal adolescents and young adults when the risk for developing moderate illness is present. Acyclovir should be started within 24 hours of symptom onset in order to be effective. Overall, IV therapy is suggested for individuals who have or are at risk of developing severe disease.

Measles virus

Immunocompromised patients and infants who have not been immunized against measles should receive intramuscular immune globulin if they are exposed to measles.

Complications of measles include possible death with most of these occurring in endemic areas.

Individuals infected with measles easily transmit the virus with the main method being by direct droplet contact or airborne spread.

Clinical manifestations of measles includes: cough, coryza, conjunctivitis, characteristic maculopapular rash starting in the hairline, and a pathognomonic enanthem (koplik spots).

In order to help control the spread of measles all individuals should be immunized; however, when dealing with an infected individual isolation (airborne precautions) along with care for those exposed to the infection should be standard.

Mumps virus

Member of the paramyxovirus typically characterized by unilateral or bilateral parotid gland swelling.

Uncommon presentations involve symptoms ranging from orchitis to CNS infections (meningitis or encephalitis) and even possible pancreatitis.

Rabies virus

Recognize that post-exposure rabies prophylaxis should include both passive and active immunizations.

Recognize that a wide variety of hosts can transmit active rabies infection to humans including wild and even domestic animals.

Recognize that the current post-exposure immunizations guidelines for bat exposure include immediate rabies immunizations and rabies immunoglobulin (RIG). This should be implemented in all bat exposures unless in a region known to be free of rabies or the animal has been proven to be rabies negative by laboratory analysis.

Rubella virus

Transmission: via droplet inhalation from the respiratory tract of uninfected hosts, along with mother to fetus with transmission during the first trimester being the most devastating.

Incubation period: postnatally acquired infection is 2-3 weeks. Communicability: several days prior up to 2 weeks after the onset of rash. Clinical manifestation of rubella include:

Congenital rubella: sensorineural hearing loss, heart disease, cataracts, and psychomotor retardation.

Postnatally acquired disease: postauricular, cervical, and suboccipital lymphadenopathy, with low grade fever and rash for 1-5 days.

Adolescents and adults: prodromal eye pain, headache, sore throat, anorexia, nausea, fever, and lethargy.

Of note, up to 50% of individuals with postnattaly acquired infection present with no obvious clinical manifestations.

In order to prevent spread of rubella susceptible individuals should be kept out of contact with anyone affected until the exanthem has cleared. Further, healthcare providers should take responsibility to report active cases of rubella to appropriate healthcare agencies.

Even with proper vaccinations some women of childbearing age will remain susceptible to rubella.

Parvovirus B19 (erthyema infectiosum, fifth disease) Clinical manifestations of fifth's disease include

Recognize that complications of parvovirus B19:

Fetal hydrops, aplastic crisis in children with hemolytic disease, severe anemia in children with HIV infection and AIDS

Human herpes virus type 6 (Roseola)

Clinical manifestations of roseola include febrile seizures and roseola.

Human Immunodeficiency Virus (HIV)

Infants born to HIV-positive mother's should have an HIV DNA or RNA PCR in their health maintenance care with testing occurring at: 14-21 days, 1-2 months, and finally at 4-6 months. HIV can be excluded for the most part after 2 negative tests, however all positive tests require repeat confirmatory testing. Testing at with the first 48 hours is optional due to low test sensitivity. If testing is done, cord blood should not be used due to possible maternal blood contamination.

AIDS is transmitted via sexual contact or via infected blood.

The best way to prevent sexual transmission of AIDS is abstinence or the use of condoms.

Epidemiology: approximately 33 million people worldwide live with AIDS, 2.5 million of which are less than 15 years old.

Transmission: sexual contact or via infected blood.

Incubation: After infection seroconversion (positive test) may occur as early as 10-14 days, with most occurring within 3-4 weeks.

Communicability: HIV is considered a lifelong disease due to memory T cells. A newly infected individual, prior to antibody response, is considered at highest risk for transmission due to high viral loads.

Neonates with HIV and AIDS may present in a variety of ways including persistent infections, or febrile illnesses of unknown origin (fever over 8 days).

HIV antibody titers is the most appropriate screening test to use on children older than 18 months of age.

In order to decrease the risk of vertical transmission HIV positive mothers or mothers with AIDS should deliver via caesarean section and be treated with antiretroviral drugs.

HIV may be transmitted from mother to newborn via vaginal delivery, breast feeding, transplacentally, and intrapartum.

Nucleic acid amplification test is the most preferred method for diagnosing HIV during infancy.

Management of an infant born to a mother with unknown HIV status includes giving zidovudine.

Enterovirus (echo-, coxsackie-, numbered entero-, poliomyelitis)

Enteroviruses are primarily transmitted via the fecal-oral route, with peak transmission during summer months.

Neonates infected with echo- and coxsackievirus commonly present with a wide range of symptoms including nonspecific febrile illness, encephalitis, or even a possible sepsis-like illness. This is in contrast to older children who typically present mild febrile illness, viral exanthems, herpangina, and aseptic meningitis (rarely myocarditis or encephalitis).

Diagnostic tests for enteroviruses include culture and nucleic acid amplification (polymerase chain reaction). Of note the PCR is more sensitive and takes less time at detecting enteroviruses.

Influenza virus

Epidemiology: Influenza is primarily spread via respiratory droplet transmission between individuals or by contact with contaminated surfaces with highest rates of infection occurring in school-age children (1% of these children are hospitalized). Peak seasonal epidemics typically occur during winter months in temperate climates. Finally, the incubation period of influenza is 1-4 days.

In order to diagnosis one should monitor for symptoms including fever, malaise, myalgia, headache, nonproductive cough, sore throat, and rhinitis. Of note younger children may have an atypical presentation that may include febrile seizures or sepsis- like symptoms.

Laboratory tests used to diagnosis influenza include rapid antigen-detection tests (50- 70% sensitivity, and 90-95% specificity), immunofluorescence, viral culture, and reverse transcriptase polymerase chain reaction (RT-PCR).

Use of antiviral medications are advocated in children when there is a high risk for complications, or within 48 hours of symptom onset in otherwise healthy children (may decrease duration of symptoms by 1 day). Finally, prophylaxis with antivirals maybe indicated in high-risk children for 2 weeks following immunization during flu season in the community, or for those high-risk patients that are unable to be immunized.

Complications that may arise with influenza infections include encephalitis, primary viral pneumonia, and secondary bacterial infections (e.g. pneumonia, sinusitis, and otitis media).

Factors that may contribute to increased rates of complications and hospitalizations for patients with influenza virus include: children younger than 2, asthma, BPD, cystic fibrosis, hemoglobinopathy, malignancy, diabetes, congenital heart disease, chronic kidney disease.

Parainfluenza virus

Clinical manifestations of parainfluenza virus include

Similar symptoms to influenza virus infection, e.g. malaise headache, sore throat, rhinitis: “barky” cough should also be monitored for due to the association with larynotracheobronchitis.

Adenovirus (respiratory)

Epidemiology: Adenovirus is transmitted from person to person via fecal-oral route, respiratory secretions, and fomites. Further, adenoviral infection can occur at any time of year; however, most outbreaks are seen between winter and early summer. The incubation period varies from 2-14 days for respiratory infections and 3-10 days for gastrointestinal disease.

In order to diagnose adenovirus, clinicians should monitor for a variety of symptoms including cervical lymphadenopathy, nonspecific febrile illness, otitis media, exudative tonsillitis, and even pneumonia. Of note adenovirus symptoms may be confused with Kawasaki disease and should be quickly distinguished due to treatment requirements associated with a diagnosis of Kawasaki disease.

Respiratory Syncytial Virus (RSV)

Epidemiology: RSV is transmitted via respiratory droplets infecting the epithelial cells lining the small airways of the lungs; the incubation period for RSV is within 4 to 6

days of infection. Children infected with RSV are usually contagious for 3 to 8 days. However, some infants and children with weakened immune system can be contagious for as long as 4 weeks.

The presentation of a child with RSV infection varies, but typically includes rhinitis, cough, tachypnea, accessory muscle use, hypoxia, and along with intermittent wheezes/crackles on examination

The two ways to diagnosis RSV by lab testing is: culture and antigen detection

Management for RSV is primarily supportive care (supplementary oxygen and nasal suctioning as needed). A trial of alpha or beta agonist may be given but has limited benefit in treatment of RSV.

To help control spread of infection patients should follow strict hand washing along with isolation protocol if hospitalized

Individuals at high risk for morbidity and mortality from RSV include those with congenital heart disease, bronchopulmonary dysplasia, and prematurity or low birth weight. Prophylaxis with Palivizumab has been approved for premature infants less than 35 weeks, chronic lung disease, and those with hemodynamically significant congenital heart disease.

Recognize that lower respiratory infections in infancy are most often caused by RSV infection.

Rotavirus

Epidemiology: worldwide rotavirus accounts for 25% of all deaths due to diarrheal causes. The main mode of transmission is fecal-oral, incubation period is 2-4 days after infection when symptoms manifest, with worst symptoms occurring in children aged 4- 24 months. Finally, peak season for rotavirus varies depending on climate, with most incidences occurring in winter and spring months.

Best way to diagnosis rotavirus is via antigen testing, however a specific diagnosis is not needed in order to start implementing management.

Symptoms may vary from infected persons being asymptomatic to the classic presentation which includes a history of gastroenteritis, vomiting for 1-3 days, watery diarrhea for 5-6 days, and fever for 1-2 days. Respiratory symptoms maybe seen in 20- 50% of cases; Rare associations with seizures, gastric rupture, and central pontine myelinolysis are reported.

Miscellaneous enteric viruses (adenovirus, norovirus)

Recognize that enteric adenovirus and norovirus may be the cause of diarrheal disease in children.

Arbovirus

Epidemiology: The mode of transmission for most arboviruses is via a secondary transmission from arthropods; however, dengue fever is one virus that can be

transmitted from human to human. The peak season for arboviruses varies in the United States with the northern states primarily having highest infection in summer and fall and more southern states occurring all year long. The main areas infected will vary year to year depending on climate and number of arthropods in the area.

Recognize that arboviruses may be a cause of encephalitis.

Recognize that the incidence of dengue fever is increasing in tropical countries, and should be on the differential for individuals who have traveled to these countries presenting with acute febrile illness.

Most individuals with west Nile virus will be asymptomatic with only 20% presenting with fever (West Nile fever). Along with fever individuals will have "flu-like" symptoms which include myalgias, headache, along with weakness often associated with abdominal pain, nausea, vomiting and diarrhea.

The epidemiology of west Nile virus includes: most common mode of transmission is via the Culex mosquito, with most cases occurring in warmer months.

Hepatitis A virus (HAV)

Indications for intramuscular immune globulin and HAV vaccine for postexposure prophylaxis involve stopping the spread of a food borne or daycare outbreak. Vaccine should be given to individuals younger than 40 and has been shown to be effective if given within 2 weeks of exposure. Only in certain circumstances, such as children younger than 1 month, should the immune globulin be used for postexposure prophylaxis.

Epidemiology: Spread via the fecal-oral route the incubation period of Hepatitis A is 2- 3 weeks before symptoms may or may not manifest (not enough infected hepatocytes to create a response). The virus may be shed in infected individuals feces for weeks making HAV easily transmitted. Finally, once infected patients will have life long immunity to infection.

Infected individuals will typically be asymptomatic, however, if symptoms do manifest they will typically be flu-like symptoms. These include: fever, malaise, anorexia, nausea, vomiting, and eventually jaundice, which is seen in 40-70% of infected patients above 15 years old, and in less than 10% of children younger than 4 years of age.

Recognize that the best test for diagnosing HAV is via serology: HAV IgM for acute infection and HAV IgG for immunity status.

Hepatitis B virus (HBV)

Treatment for infants born to a mother who is HBV carrier includes administering a combination of Hepatitis B vaccine plus Hepatitis B immunoglobulin. These infants should than be completely vaccinated with the Hepatitis vaccine series and then should have their HBsAb and HBsAg checked. If HBsAb is low then patient should be

reimmunized, and if HBsAg is elevated then steps should be taken to monitor for complications related to chronic HBV infection. Of note, 98% of vertical transmission can be prevented by postnatal prophylaxis.

Epidemiology: HBV infection is acquired via percutaneous or mucosal exposure to body fluids of an infected person, most commonly via sexual contact, infected needles/blood, or vertical transmission. The incubation period of the virus is 2 to 6 months. Communicability is life long as is the disease, with periods of high and low infectivity based on serology of the individual infected.

Infected individuals will typically present with hepatic complaints, and will have arthralgias and possibly an urticarial rash prior to onset of symptoms. Hepatic symptoms of the disease vary from being self-limited to fulminant hepatitis and hepatocellular cancer.

Screening for Hepatitis B is best done via serology, however, it may take up to 14 days before an infected individual will show positive serology markers.

One group at risk of acquiring hepatitis B includes children born in endemic areas. Over 70% of adults have positive serology in areas known to be endemic to HBV, with most of these individuals having acquired the virus during birth or childhood.

Perinatally-acquired hepatitis B are more likely to become chronic infections and experience fatal complications of HBV compared to individuals infected later in life.

Periodic screening for complications should be routine for patients infected with HBV as treatment regimens are now available for many of these symptoms.

Hepatitis C virus:

Risk factors for acquiring hepatitis C infection are blood transfusion, intravenous or intranasal drug abuse, multiple sexual partners, homosexual activity, infant whose mother has hepatitis C infection, patients receiving hemodialysis.
The long-term outcome of hepatitis C infection are chronic carriers, chronic hepatitis, cirrhosis, hepatocellular carcinoma.
The current clinically available assays for diagnosis of hepatitis C infection are based on the detection of antibodies to HCV and measurement of RNA by PCR, also known as nucleic acid amplification. Enzyme Immuno-Assays (EIA)s have increased sensitivity but can produce false negative results due to low antibody concentration for the first 1 to 3 months after infection.
Babies born to mother who have HCV infection should be tested for anti-C antibodies starting at 18months of age. But testing for HCV RNA can be undertaken as early as 1 month of age.
Chronic HCV infection is defined as persistently elevated tromsaminase (ALT) concentration in the presence of hepatic fibrosis found on liver biopsy and positive HCV RNA in the blood. Biopsy remains the only means of following disease progression.
The treatment regimen available is pegylated interferon alfa 2b dosed weekly and daily oral ribavirin for 48 consecutive weeks. Successful treatment is achieved when the HCV RNA remains undetectable 6 months after treatment.

Human papillomavirus (HPV):

Clinical manifestations of HPV is cutaneous warts, epidermodysplasia verruciformis, anogenital warts, recurrent respiratory pappilomatosis, condylomata acuminata, caervical carcinoma.
Prevalence of HPV infection are as follows: common warts constitute 71% of all cutaneous warts with prevalence rate of 4 to 20%, the prevalence rate for cervical HPV DNA is 27%, the prevalence rate of anogenital wart is approximately 1%.
Risk factors for HPV infection are multiple sex partners, passage through infected birth canal.
Mode of transmission of HPV infection is close personal contact, nosocomial transmission.

The strains 16 and 18 are responsible for two-thirds of all cervical cancers worldwide. The types 6 and 11 are responsible for anogenital warts.

Human metapneumovirus (HMPV):

A) Humans are the only source of HMPV infection. In temperate climates, infection usually occurs during RSV season. It usually causes pneumonia, croup, upper respiratory infection (URI) with concomitant acute otitis media (AOM). Diagnosis is by rapid diagnostic immunofluorescent assays (IFA).

Bacterial pathogens:

Anaerobes:

Clinical manifestations of anaerobic infections are sinusitis, cervical

adenitis, chronic otitis media, Ludwig angina, dental abscess, and gingivitis. They are also are associated with infections of the central nervous system (brain abscess, subdural empyema, epidural empyema), pleura and lungs (aspiration pneumonia, pleural empyema, lung abscess), gastrointestinal tract (peritonitis, appendicitis, intra-abdominal abscess, pelvic inflammatory disease, ascending cholangitis), skin and soft tissues (necrotizing fasciitis, infected bite wounds, cellulitis or abscesses), and bactermia. Serious infections are seen more often in immunocompromised hosts or neonates.

Arcanobacterium haemolyticum:

Arcanobacterium Haemolyticum is a gram positive bacillus responsible for pharyngitis in teenagers and young adults and accounts for 0.5% to 3% of all cases of acute pharyngitis. It causes fever, pharyngeal exudates, cervical lymphadenopathy with maculopapular rash similar to that caused by group A streptococcus. Severe infection such as septicemia, pneumonia, brain abscess, meningitis, and endocarditis occurs in immunocompromised person.

Brucella (brucellosis)

Brucellosis should always be considered in the differential diagnosis of fever of unknown origin. It is a gram negative coccobacilli that usually infect wild and domestic animals. Human contract brucellosis by consuming unpasteurized milk or cheese or direct contact with infected animals; usually manifest with fever, myalgias, arthralgias, fatigue, abdominal pain, headache, anorexia, weight loss. Mild hepatosplenomegaly and lymphadenpathy may be present. Diagnosis is established by antibody testing and blood culture. Main treatment modality is by using bactrim or tetracycline for 6 weeks.

Campylobacter species:

Treatment of a Campylobacter infection starts with rehydration; however certain medications can be used to shorten the duration of illness. Drug of choice for treatment of campylobacter is macrolides such as azithromycin and erythromycin, other drugs which can be used are fluroquinilones or extended spectrum cephalosporins.
Clinical manifestation of Campylobacter infection is fever; abdominal pain and bloody diarrhea and bactermia in immunocompromised patients. Guillian berre syndrome and reactive arthritis have also been associated with campylobacter infection.

Handling uncooked poultry, ingestion of contaminated poultry (chicken and turkey) and raw milk are the most common sources of C jejuni infection.

Bartonella henselae (cat-scratch disease)

The most frequent clinical manifestation of cat scratch disease is fever with lymphadenitis. However it can also present with encephalitis, pneumonia, granulomatous hepatitis, neuroretinitis, and osteomyelitis.

Differential diagnosis in a patient with suspected cat-scratch disease is nontuberculous mycobacterial infection, tuberculosis, sarcoidosis, lymphoma, leukemia, EBV infection, Staph or strep infection.

Diagnosis is usually established by measuring antibody titer (Ig M and Ig G titers against B henselae). PCR can also used to diagnose cat scratch disease and it is used for detection of B Henselae DNA in clinical specimens.

Cats are reservoir of B henselae with up to 75% of pet cats being seropositive. The cat flea is most common vector. The disease is usually spread by cat bites.

Chlamydia and Chlamydophila (chlamydial infections)

Neonatal chamydial conjunctivitis develops 5 to 14 days after delivery and may be unilateral or bilateral. Initially presents with watery discharge and then progresses to mucopurulent discharge; other signs include eyelid edema, pseudomembrane formatiom and bloody discharge. Untreated infections may result in corneal and conjunctival scarring.

Mode of transmission of Chlamydia trachomatis are vertically from infected mother to infant, as sexually transmitted infection.

Diagnostic tests for different sites of Chlamydia trachomatis infection:

Culture: Approved for use from all collection sites, gold standard.

Nucleic acid amplification (PCR): tested using urine specimen.

DNA probe, direct fluorescent antibody titer: are some other tests available for detecting C trachomatis infection.

Trachomatis-specific IgM for diagnosis of pneumonia: a positive nasopharyngeal culture is considered diagnostic of infection.

Treatments of a chlamydial infection are,

Conjunctivitis: Erythromycin PO 50mg/kg per day in 4 divided doses

for 14 days.

Pneumonia: Erythromycin for 7 days or Azithromycin 20mg/kg once daily for 5 days.

Genital tract infections: Adolescents: Doxycycline 100mg po bid for

7 days or Azithromycin 1g single oral dose. For children: erythromycin or azithromycin can be used.

Clinical manifestations of Chlamydia trachomatis pneumonia in young infants presents as a subacute infection 2 to 19 weeks after birth. Signs and symptoms include tachypnea, staccato cough, apnea, wheezing.

Chlamydia pneumonia causes an atypical pneumonia with insidious or sudden onset of symptoms which includes cough for 2 to 6 weeks, sore throat and laryngitis. It can also cause nonexudative pharyngitis, bronchitis, acute otitis media and sinusitis. It is seen in

children between 5 to 15 years of age. Diagnosis is by serologic testing. Treatment is by azithromycin or erythromycin.
Clinical manifestations of genital tract infections caused by Chlamydia trachomatis are vaginitis, urethritis, cervicitis, epididymitis, endometritis, perihepatitis, and chronic pelvic inflammatory disease leading to infertility.

Clostridium botulinum (botulism)

Epidemiology of botulism:
Food borne: from ingestion of improperly packaged or incorrectly stored food.
Wound: from systemic spread of the organism from an infected wound.
Infantile: From intestinal colonization in infants after consumption of foods like honey.
Clinical manifestations of botulism is constipation, poor feeding, progressive descending paralysis, hypotonia, decreased gag reflex, ocular palsies, weak cry.
Infant botulism is usually caused by toxins A or B. The diagnosis is usually established by toxin assay and stool culture.

Clostridium difficile

Asymptomatic carriage of toxin producing strains of C difficile can occur

at any age but it is most common in newborns and infants younger than 1 year of age. So if the infants test positive for C. diff they don't necessarily need to be treated.

Clinical manifestations of Clostridium difficile infection are fever, abdominal pain, watery diarrhea, pseudomembranous enterocolitis leading to toxic megacolon, intestinal perforation.
Diagnosis of Clostridium difficile infection is usually established by testing stool for the presence of C. difficile toxins.
Treatment of a Clostridium difficile infection is with metronidazole, 30mg/kg in 4 divided doses, po or iv for 10 days, other alternative is oral vancomycin, 40mg/kg in 4 divided doses for 10 days.
Infection control measures for Clostridium difficile infection are proper hand washing using soap and water, limiting use of antibiotics, properly disposing of contaminated materials, adequately cleaning contaminated surfaces using bleach solutions. Children who have C. Diff should be prevented from participating in group activities including day care.

Corynebacterium diphtheriae (diphtheria) (See also III.A)

Clinical manifestations of diphtheria are low grade fever, malaise, sore throat, difficulty swallowing, and formation of thick membrane consisting of fibrin, bacteria and dead cells adhering firmly to underlying tissue. Removal of membrane leads to bleeding; it can be localized or extensive causing small airway edema. Severe disease may present with extensive neck swelling, upper airway obstruction, myocarditis, renal tubular necrosis, peripheral neuropathy.
Treatment is mainly by diphtheria antitoxin, erythromycin po or iv for 14 days or penicillin G intramuscularly for 14 days.

Enterococcus

Treatment of enterococcal infections:

Drug of choice: Ampicillin.
Alternative drugs: Imipenem, Meropenem, Zosyn, Moxifloxacin, extended spectrum penicillin/inhibitor combination antibiotic, vancomycin resistant enterococci (VRE) associated infections can be treated with quinupristin-dalfopristin and linezolid.
Ineffective drugs: Cephalosporins, Aminoglycosides, Bactrim, Clindamycin.
Clinical syndromes usually associated with enterococci are urinary tract

infections, bacteremia with and without endocarditis, meningitis, peritonitis, diverticulitis.

Escherichia coli

Hemolytic-uremic syndrome is characterized by triad of hemolytic anemia, thrombocytopenia and acute renal failure. Most of the cases are preceded by diarrheal illness and are caused by Shiga toxin producing E.coli.(O157:H7)
Patients whose diarrhea is due to enterotoxigenic strains of E. coli (ETEC) often have a moderately severe, self-limited illness characterized by abdominal cramping and watery diarrhea. Infection caused by this pathotype of E coli accounts for 30% to 40% of cases of traveler's diarrhea. ETEC usually is acquired by ingestion of contaminated food and water and person-to-person transmission is uncommon.
Enteropathogenic strains of E coli (EPEC) produce severe watery

diarrhea, predominantly in children younger than 2 years of age in developing countries. It is not a cause of traveler's diarrhea. The illness can result in significant dehydration and may lead to death. Chronic, persistent infection can lead to severe wasting.

Enteroinvasive E coli (EIEC) strains can cause mild-to-severe dysentery. Common clinical manifestations include fatigue, fever, abdominal cramping, and tenesmus. Watery, nonbloody diarrhea usually progresses to blood-streaked, mucoid diarrhea but rarely becomes grossly bloody. Fecal leukocytes may be present. The illness usually lasts 1 to 2 weeks.
Illness caused by enteroaggregative E coli (EAEC) strains usually presents as watery diarrhea without fever or fecal leukocytes. The diarrhea can occur in children of all ages in developed and developing countries and can result in a prolonged illness of more than 14 days' duration.

F. Nondiarrheagenic strains of E coli are associated with infections of the gastrointestinal tract (peritonitis, intra-abdominal abscess, ascending cholangitis), skin (neonatal breast abscess, necrotizing fasciitis), genitourinary tract (urinary tract infection, pelvic abscess), lungs (neonatal pneumonia), central nervous system (neonatal meningitis), bones and joints (neonatal osteomyelitis and septic arthritis), and bloodstream (neonate, immunocompromised host).

Neisseria gonorrhoeae (gonococcal infections)

Clinical manifestations of Neisseria gonorrhoeae infections:
Neonates: conjunctivitis, scalp abscess, vaginitis, bacteremia,

arthritis, meningitis.

Males: Pharyngitis, urethritis, epididymitis, proctitis, disseminated gonococcal infection.

Females: Pharyngitis, cervicitis, salpingitis, pelvic inflammatory disease, proctitis, perihepatitis, menometrorrhagia, disseminated gonococcal infection.
N.gonorrhae can be isolated in culture media or identified by using nucleic acid techniques (NAATs). The gold standard for diagnosing a gonococcal infection is growth in chocolate agar or Thayer-martin agar. Nucleic acid amplification technique (NAATs) can be used to detect N.gonorrhoeae from endocervical or urtheral sites. The other method to consider is gram stain which may be performed directly on exudates from several sites. The presence of gram negative intracellular diplococcic on microscopy suggests the diagnosis of a gonococcal infection.
Treatment of N.gonorrhoeae is based on the site and type of infection. The third generation cephalosporin like ceftraxione can be used for most of the infection caused by N.gonorrhoeae.
Neonatal infection is also treated using ceftriaxione but patients should always be monitored for disseminated infection. Pelvic inflammatory disease (PID) is treated using single dose of ceftraxione or cefoxitin and doxycycline. Always consider coinfection with C trachomatis in patients who have N.gonorrhoeae infection.
Control measures for gonococcal infections are prevention of neonatal ophthalmia by prophylaxis using erythromycin eye ointment. Mothers who are diagnosed with gonococcal infection should be treated with third generation cephalosporin, infants who are born to mothers with infected gonococcal infection should be monitored closely for any signs and symptoms of infection and also apply the eye prophylaxis. Sexual partner(s) of patients who are diagnosed with N.gonorrhoeae infection should be treated and also both the partners should be educated regarding various STDs.

Hemophilus influenzae

Antibiotic prophylaxis with rifampin is indicated for all household contacts in cases of invasive Hib infection if there is at least one contact younger than 4 years of age who is incompletely immunized or unimmunized. Prophylaxis is also indicated if there is a child in the household who is younger than 12 months of age and has not completed

the primary Hib vaccine series or if there is an immunocompromised child in the household. Antibiotic prophylaxis is also indicated in nursery school and child care center contacts when two or more cases of invasive HiB disease have occurred within 60 days.

Clinical manifestations of Hemophilus influenzae type b infection are upper respiratory infection (URI) symptoms followed by meningitis, seizures, altered mental status, cellulitis, arthritis, pneumonia, epiglotitis, pericarditis, urinary tract infection, cervical adenitis, and glossitis. Nontypable Hemophilus influenzae infection usually presents with conjunctivitis, neonatal sepsis, acute otitis media, pneumonia, URI symptoms, sinusitis, and bronchitis.
Treatment of a nontypable Hemophilus influenzae infection is by using oral antibiotics such as amoxicillin, augmentin, cefaclor, bactrim, azithromycin.

Helicobacter pylori

First line treatment regimen for a Helicobacter pylori infection in children includes clarithromycin with either amoxicillin or metronidazole and a protein pump inhibitor (PPI).

For children older than 8 years of age tetracycline, metronidazole, bismuth subsalicylate and histamine-2 (H-2) blocker can be used.
Risk factors for Helicobacter pylori infection includes residence in a developing country, lower socioeconomic group, family overcrowding, child care attendance, poor hygiene, living with infected family member, ethnic groups including Asian American, African Americans, Hispanic living in North America.
Helicobacter pylori infection may be asymptomatic in children or may cause chronic active gastritis increasing the risk of duodenal and gastric ulcers. The development of gastric adenocarcinoma, gastric lymphoma also has been associated with chronic H.pylori infection.

Kingella kingae

The most common infections associated with Kingella kingae are pyogenic arthritis, osteomyelitis, diskitis, bactermia, endocarditis.

Listeria monocytogenes

Listeria monocytogenes causes neonatal sepsis within 1 to 2 days after delivery. Severe disease is associated with granulomatous infantiseptica.

Neonatal late onset disease is associated with sepsis, meningoencephalitis, rhombencephalitis, and endocarditis.

Mode of transmission of Listeria monocytogenes is by crossing placenta. Most maternal infection occurs during third trimester of pregnancy, when T cell immunity is most impaired. Infected women develop nonspecific flu like symptoms and remain asymptomatic.

Borrelia burgdorferi (Lyme disease)

When a patient presents with arthritis there is a board differential diagnosis including septic arthritis, PRSA, Lyme disease, JIA. The diagnosis of JIA requires symptoms present for more than 6 weeks and ruling out other causes of arthritis.

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Mode of transmission of Lyme disease is by tick bite (ixodes scapularis) and it is caused by Borrelia burgdoferi.
Lyme disease consists of 3 stages: early localized, early disseminated and late chronic. About 7 to 10 days (up to 36 days) after the bite, 60% to 80% of affected individuals develop the early localized form, which is the erythema migrans rash and flulike symptoms (fever, arthralgias, and myalgias). Erythema migrans is described as a solitary, enlarging, annular plaque with a bull's-eye zone of central clearing that is painless and nonpruritic. Approximately 20% of persons who have erythema migrans have more than one lesion. The next stage, early disseminated Lyme disease, consists of arthritis, which occurs in 50% to 60% of untreated cases, facial nerve palsy, aseptic meningitis, and carditis. If still untreated, Lyme disease can progress to the late chronic stage, characterized by prolonged arthritis, encephalopathy, and acrodermatitis chronica atrophicans.
The diagnosis usually is made by the appearance of the typical rash. Without a clear rash, the diagnosis must be made by using serologic testing. Antibodies usually are not present in

the early stage because of a slow host antibody response to B burgdorferi. IgM appears 2 to 4 weeks after the rash and IgG after about 4 to 6 weeks. The IgM concentration declines after about 4 months, but IgG remains present even after treatment. Laboratory testing involves two steps. Serology is determined first by using a screening test such as a polyvalent ELISA. If the ELISA findings are positive or equivocal, an immunoblot (western blot) for both IgM and IgG is performed as a confirmatory, more specific serologic test. If the laboratory test is performed more than 4 weeks after the symptoms began, the IgG immunoblot must be positive for a laboratory-confirmed diagnosis.

E.Treatment of Lyme disease depends on the stage. Early stages can be treated with amoxicillin, cefuroxime, or doxycycline for a 14-day course (avoid doxycycline in children younger than 8 years). If there are neurologic sequelae, ceftriaxone is given for a 14-day course. For late stages, 28 days of amoxicillin, cefuroxime, or doxycycline should be given. In the late stage, if there is neurologic involvement, intravenous antibiotics for 2 to 4 weeks are needed. Prophylaxis is usually by doxycycline 200mg dose for adults, 4mg/kg of doxycycline for children older than 8 years within 72 hours of the time the tick is removed.

Neisseria meningitidis (meningococcal infections)

Vaccine against N meningitides four capsular groups (A, C, Y, and W-

135) is available in preventing meningococcal disease in the United States. MCV4 is routinely recommended at 11 to 12 years of age. But there are no vaccines available against N meningitides Serogroup B.

Household contacts of patients who have meningococcal disease should be treated to eradicate nasopharyngeal carriage of the organism. Those who have child care contact and those who have direct exposure to an index case's oral secretions (such as personnel providing mouth-to-mouth resuscitation) during the 7 days before the onset of illness also require chemoprophylaxis. Rifampin, ciprofloxacin, azithromycin, or ceftriaxone should be used for chemoprophylaxis.

Clinical syndromes of Neisseria meningitides are

Meningococcemia: characterized by sudden onset, rapid progression and systemic manifestation of the disease without any localizing symptoms. Patients usually presents with high fever, severe myalgias, shaking chills. At this stage patients may be misdiagnosed has having viral illness. Within 6 hours, patients develop rash gradually progressing into hemorrhagic rash and also associated with sudden drop in blood pressure due to acute adrenal hemorrhage.

Meningitis: usually associated with more localized symptoms of meningitis such as headache, fever, meningeal signs, lethargy, nausea, vomiting, petechial rash, muscle aches and sometimes may be associated with signs of increased ICP such as bradycardia, hypertension and respiratory depression.

Diagnostic tests for invasive meningococcal disease are Gram stain and antigen detection assays of CSF, polymerase chain reaction, antigen detection, culture from blood and CSF.

With introduction of vaccines against H, influenza and Strep Pneumonia, eisseria meningitidis is the leading cause of bacterial meningitis in young children in the USA.

Immunocompromised patients, patients with terminal complement component deficiency, patients with asplenia, people living in military recruits, college freshmen living in dormitories are at increased risk of developing invasive meningococcal disease.
Neisseria meningitidis infection is spread by droplet aerosolization or direct contact with secretions. Crowded living conditions such as military recruits, college dormitories, and residential camps predispose to transmission of organism.
Treatment of a neisseria meningitidis infection includes fluid and electrolyte management, airway management, antibiotics such as cefotaxime or ceftriaxone, penicillin G, corticosteroids and vasoactive agents if needed for invasive meningococcemia.

Mycobacterium tuberculosis

The intradermal Mantoux test is the most reliable tuberculin skin test (TST) diagnostic for tuberculosis (TB). It should be placed and evaluated by trained personnel to avoid errors based on incorrect technique. The test consists of 0.1 mL of 5 tuberculin units of purified protein derivative (PPD) injected intradermally on the volar aspect of the forearm, forming a 6- to 10-mm wheal. The area is inspected at 48 to 72 hours; indurations, not erythema, is measured transversely to the long axis of the forearm and the results recorded in millimeters. Intrepreation of test results is as follows:
5mm of reaction: Recent exposure to TB, patients with HIV, immunosuppression, or patients with clinical features of tuberculosis and chest radiograph consistent with TB.
10mm of reaction: High risk of disseminated TB (<4 years of age, lymphoma, diabetes, renal failure, malnutrition, or other predisposing condition), patients with frequent visit to endemic areas.
15mm of reaction: More than 4 years of age with no identifiable risk factor
When TST is positive, chest radiograph has to be performed to confirm the diagnosis of TB. The abnormal finding in chest radiograph would be hilar or mediastinal lymphadenpathy, infliterates, atelectasis, pleural effusion, military tuberculosis, cavities, calcifications.
BCG vaccination can cause reaction to tuberculin skin test. So if the TST is positive chest radiograph should be done to rule out TB.
Health-care worker who have positive TST results should have chest radiograph. If the chest radiograph is negative they can be offered therapy with 9 month of isoniazid or 6 months of rifampin. If the chest x ray is positive than further investigation has to be performed to determine the source of infection and healthcare worker is treated with isoniazid, rifampin, pyranzamide, ethambutol.
False-positive TST results can occur in children recently exposed to nontuberculous mycobacterium and also in children who have received BCG vaccination. False negative TST results can be seen in association with recent measles infection, high dose corticosteroid treatment, irradiation, other immunosuppressive therapy, and patients with military or disseminated tuberculosis due to anergy to skin test.

F. TB is transmitted via airborne spread. Inhalation of mycobacteria into the terminal airway results in formation of Gohn complex which includes the initial focus of infection, the draining lymphatic vessels, and enlarged regional lymph nodes. Following this stage, the infection can be contained, spread rapidly, or reactivate at a later point in life.

Clinical manifestations of mycobacterium tuberculosis are pneumonia, lymphadenitis, arthritis, meningitis, osteomyelitis, gastrointestinal, renal, pleural and miliary disease.
TB exposure is a category used to describe asymptomatic children who have had contact with a person suspected of having TB disease and in whom the TST result and chest radiograph are normal. Children younger than 4 years of age and immunocompromised should be started on INH, until second TST is negative. Once repeat TST is negative medication can be discontinued. In children older than 3 years and immunocompetent can be observed off of medication pending repeat TST results.
Children hospitalized because of cavitary or extensive pulmonary involvement, AFB smear-positive TB, or laryngeal TB should be isolated in negative-pressure isolation rooms and healthcare workers should use N95 respirators while taking care of such patients. Such children should remain isolated until effective therapy has been initiated, cough has diminished, and sputum AFB smears convert to negative.

Latent tuberculosis infection in an individual is when TST is positive but no symptoms, or physical findings or radiographic anomalies are noted which is consistent with active disease. It is recommended that most children who have LTBI receive a course of therapy to prevent the development of TB disease in the future. Tuberculosis disease is seen in patients with clinical or radiographic manifestation of the disease. These patients need treatment with multiple drugs.
Foreign-born individuals in the United States have TB rates 9.5 times higher than those in United States-born persons, with most cases occurring in immigrants from Mexico, the Philippines, Vietnam, China, and India. The risk factor for development of LTBI or TB in USA are untreated HIV infection, immunocompromised patients, recent LTBI, intravenous drug abuse, patients with medical conditions such as renal failure, diabetes mellitus, etc.
LTBI is treated with Isoniazid for 9 months or rifampin for 6 months. Tuberculosis is treated by using medications such as isoniazid, rifampin, ethambutol (for 6 months) and pyrazinamide (for first 2 months).The pulmonary and extra pulmonary TB s are treated for 6 months, but disseminated and CNS TB needs treatment for 9 to 12 months. Multiple Drug Resistant (MDR) TB is defined as s resistance to at least two of the first-line TB medications, isoniazid (INH) and rifampin. In this case, medication has to be individualized based on the exact drug resistance pattern. The other medications which are available for treatment are amikacin, streptomycin, kanamycin, ciprofloxacin, levofloxacin.

Mycoplasma pneumoniae

Clinical manifestations of mycoplasma infections are acute upper or lower respiratory tract infection presenting with symptoms of fever, cough, sore throat, cold symptoms and rash.
Laboratory tests for mycoplasma pneumonia are isolation, serology for detection of Ig M and Ig G antibody against Mycoplasma, polymerase chain reaction
Treatment of Mycoplasma pneumoniae infection is by Azithromycin, erythromycin and Fluoroquinolones such as ciprofloxacin.
Mycoplasma pneumoniae is a leading cause of pneumonia in school-age children and young adults and infection is especially prevalent in persons living in group settings such as dormitories, military institutions, and other residential facilities.

E.Extrapulmonary manifestations of mycoplasma are pharyngitis, rash, Stevens-Johnson syndrome, hemolytic anemia, arthritis, carditis, pleural effusion, and CNS disease (encephalitis, neuropathy, and transverse myelitis).

Nontuberculous mycobacteria

Cervical adenitis secondary to nontuberculous mycobacteria is usually diagnosed clinically. The main modality of treatment is surgical excision which helps in definitive diagnosis. Needle aspiration is not indicated as it leads to chronic draining fistula. Antimicrobial therapy with clarithromycin or azithromycin in combination with ethambutol or rifampin or rifabutin may be beneficial when complete resection is not possible or in cases of recurrent disease. Isoniazid is not beneficial.

Major clinical manifestations of nontuberculous mycobacteria in immunocompetent children are cutaneous infection, lymphadenitis, otologic infection; catheter associated blood stream infection, pulmonary infection, and skeletal infection. It is usually seen in children age less than 5 years and mycobacterium avium complex is most common cause of lymphadenitis in USA.

Pasteurella multocida

Mode of transmission of Pasteurella multocida is through animal bite, lick or scratch such as dogs and cats.

Most common clinical manifestation of a Pasteurella multocida infection is cellulitis at the site of an animal bite that develops within 24 hours of injury; complications include tenosynovitis, osteomyelitis, abscess, septic arthritis.

Treatment of Pasteurella multocida infection are penicillins, other alternative agents are trimethoprim-sulfamethoxazole (TMP/SMX), extended-spectrum cephalosporins, doxycycline, and fluoroquinolones.

Bordetella pertussis (pertussis)

Adolescents and adults are important sources of pertussis in infants and children. In the United States, the highest incidence occurs in infants younger than 6 months of age and in 10 to 1-year old children. Outbreaks also have been described in childcare facilities and secondary schools.

There is no lifelong immunity against pertussis despite immunization or natural infection. Chemoprophylaxis should be indicated for close contacts of patients who have pertussis even if the exposed individual is fully immunized. Azithromycin is the drug of choice for chemoprophylaxis.

Pertussis is spread by aerosol droplets expelled while coughing or sneezing in proximity to others.

Clinical manifestations of pertussis are:

Catarrhal phase: This phase lasts for 1 to 2 weeks and consists of non specific symptoms such as fever, cough, and nasal symptoms.

Paraxosymal phase: This phase lasts for 2 to 6 weeks and is characterized by paroxysms of cough often described as “rapid fire” or “staccato.” Classically, as many as 5 to 10 uninterrupted coughs occur in succession, followed by a “whoop” as the patient rapidly draws in a breath. It is associated with salivation, lacrimation, post tussive emesis. In infants less than 6 months of age may present with apnea, gagging or choking episodes.

Convalescent phase: lasts for weeks to months, cough gradually resolves during this phase of illness.

Diagnostic tests available for pertussis are isolation from nasopharyngeal swab or from aspirated specimen, polymerase chain reaction, serology, direct fluorescent antibody testing, culture, leukocytosis, together with an absolute lymphocytosis on a peripheral complete blood count.

Treatment of pertussis is by using azithromycin, erythromycin, and bactrim

If left untreated, most individuals will clear of B pertussis spontaneously from the nasopharynx within 2 to 4 weeks of infection. However, nasopharyngeal carriage can persist for 6 weeks or more. During this period, individuals remain contagious and can spread the illness to others. When started early in the course of the illness, during the catarrhal

stage, antibiotics can shorten the course and attenuate the severity of pertussis. Once the paroxysmal phase has started, however, antibiotics are not effective in altering the course of the disease.

It is very essential that family members of newborn infants be vaccinated against pertussis as pertussis can be very fatal in newborns with mortality rate of upto1%. Complications of pertussis include apnea, pneumonia, seizures, encephalopathy, and death.

Pseudomonas species

Risk factors for development of severe pseudomonas infections are cystic fibrosis, cancer patients with neutropenia, hospitalized patients receiving broad-spectrum antibiotic therapy, hospital-acquired infections in debilitated patients such as ventilator-associated pneumonia or infections in burn patients.

Antibiotics which are generally effective against Pseudomonas infections are penicillins (piperacillin, ticarcillin) fourth generation cephalosporins such as cefepime, ceftazedime, Quinilones such as ciprofloxacin, levofloxacin, Carbapenems (imipenem and meropenem), aminoglycosides (gentamicin, tobramycin, amikacin).

Rickettsial diseases (Rocky Mountain Spotted Fever [RMSF], Ehrlichiosis)

Clinical manifestations of Rocky Mountain Spotted Fever are prodrome of fever, headache, myalgias, abdominal pains, malaise. Approximately 3 to 5 days later the classic rash develops. The rash begins as a macular or maculopapular rash on the wrists and ankles and spreads centrally as well as to the palms and soles. As it spreads it gradually becomes more petechial and purpuric.

RMSF infection is diagnosed by serologic assays that detect immunoglobulin G and M antibodies to the organism using an indirect fluorescent antibody or enzyme-linked immunosorbent assay. However, antibodies may not be detectable until 7 to 10 days after the onset of illness. Polymerase chain reaction (PCR) amplification tests for specific Rickettsiae rickettsii DNA are available through the CDC. The highest yield from PCR is on a tissue sample (eg, skin biopsy or autopsy specimen).

Mode of transmission of Rocky Mountain spotted fever is by tick bite.

Treatment for RMSF should be initiated immediately when it is suspected. The most commonly used drug of choice is doxycycline. In children less than 8 years of age Chloramphenicol can be used.
Human Ehrlichiosis caused by Rickettsiae species is similar clinically to Rocky Mountain spotted fever but rash is rarely petechial and has a variable distribution.
In the United States, RMSF is most prevalent in the southeastern and south central states, with peaks occurring in spring to fall, when the tick vector is active. In these areas, the dog tick (Dermacentor variabilis) is the primary vector. In the mountain states west of the Mississippi, the Rocky Mountain wood tick (Dermacentor andersoni) is the primary vector.

Salmonella species

The main mode of transmission of nontyphoidal Salmonella species is food of animal origin (poultry, egg, beef and dairy products) or other foods (fruits, vegetables) contaminated by infected animal product or humans. Other modes of transmission include ingestion of contaminated water; contact with infected reptiles or amphibians and possibly rodents; and exposure to contaminated medications, dyes, and medical instruments. In contrast to nontyphoidal Salmonella serotypes, S typhi is found only in humans, and infection occurs through direct contact with an infected person or with an item contaminated by a carrier.
Clinical manifestations of typhoid fever are high fever, general ill feeling, abdominal pain, diarrhea, rose spots, bloody stools, chills, confusion, and delirium, difficulty paying attention, nose bleeds, and severe fatigue.
Treatment includes hydration, correction of fluid electrolyte imbalance, antipyretics and antibiotics. The most commonly used antibiotics are ampiclin, bactrim, cefotaxime, ceftriaxone, and are used for 7 to 14 days based on the severity of disease.
Healthy patients with uncomplicated Salmonella gastroenteritis don't need to be treated with antimicrobial therapy.
Infection with non-typhoidal Salmonella gastroenteritis can be asymptomatic or can present with fever, acute gastroenteritis, bactermia, focal infections (osteomyelitis). Stool examination reveals the presence of polymorphonuclear leukocytes, blood, and mucus.
Uncomplicated Salmonella gastroenteritis should not receive antimicrobial therapy. The main modality of treatment is supportive therapy with hydration and antipyertic.
Invasive Salmonella infections are seen in young infants (< 6 months of age), elderly, immunocompromised patients, and patients with hemoglobinopathy.

Shigella species (shigellosis)

Mode of transmission of Shigella is typically by ingestion (feco-oral contamination).
Clinical manifestations of Shigella are fever, bloody diarrhea, nausea, vomiting, abdominal pain, flatulence, seizures.
Severe cases of shigella dysentery are treated by using antibiotics such as ampicillin, bactrim, and fluroquinodones such as ciprofloxacin. A mild case is managed supportively and doesn't need antibiotic treatment because of high rate of resistance among sheigella species.

Treponema pallidum (syphilis)

Congenital syphilis may be due to transplacental transmission of Treponema pallidum at any time during pregnancy. Only one third of infants who have congenital

syphilis are symptomatic at birth, with most identified by maternal screening studies during pregnancy or at the time of delivery. Clinical findings of congenital syphilis include jaundice, petechiae, hepatosplenomegaly, desquamation of the hands and feet, mucous patches, rhinitis, and osteochondritis. Radiologic evaluation may reveal periostitis. Affected infants also may present with severe luetic pneumonia. If not treated, the symptoms develop further and evolve. Late findings of congenital syphilis include bony changes (Saber shin, saddle nose), dental anomalies (Hutchinson teeth) and neurologic abnormalities (juvenile paresis, tabes dorsalis).

Acquired syphilis almost always is contracted by direct exposure to ulcerative lesions of the skin or mucous membranes during sexual activity. Primary acquired syphilis presents as a chancre (a painless ulcer) located on the genitalia, mouth, breast, or rectum, with localized lymphadenopathy. If untreated, secondary syphilis develops 2 to 10 weeks after the chancre heals. A nonpruritic, maculopapular rash may erupt that involves the entire body, including the palms and soles. One to two months after the onset of the rash, the infection becomes latent, with intermittent relapses and some affected individuals developing tertiary syphilis. Sexual abuse must be considered when acquired syphilis is diagnosed in children.

10-day course of parenteral penicillin G continues to be the preferred treatment for congenital syphilis and covers potential central nervous

system involvement. Alternative treatment strategies, including penicillin G benzathine.

Yersinia enterocolitica:

Clinical manifestations of Yersinia enterocolitica infection are fever, bloody diarrhea. It can range from mild self limiting entero colitis to terminal ileitis. It can also cause pseudoappendicitis where it causes enlargement of mesenteric lymph nodes leading to abdominal pain similar to appendicitis. In immunocompromised patients it can cause abscess of spleen and liver.

Infection with yersinia is self limiting and usually doesn't require treatment. However, in more disseminated infection, especially in immunocompromised patients treatment is indicated with doxycycline in combination with aminoglycoside. Other drugs which can be used for treatment are Bactrim, fluroquinolones, ceftriaxone, etc.

Fungal pathogens

Candida species

Factors that predispose a patient to the development of candidiasis are repeated use of antibiotics, immunosuppression, burns, indwelling catheters, total parenteral nutrition, contaminated bottle nipples, pacifiers, or droppers can be sources of candidal infection in infants.

In an infant younger than 6 months of age, various conditions that predispose to persistent or recurrent candidiasis of the oral cavity are maternal breast colonization, contaminated vitamin dropper, long term antibiotic use, and pacifier use.

In children older than 6 months of age, various conditions that predispose to persistent or recurrent candidiasis of the oral cavity are systemic immune deficiency, AIDS, long term antibiotic use, inhaled steroid use without adequate rinsing afterwards, and nutritional deficiencies.

Treatment for a patient with a candida infection: Topical agents such as nystatin or clotrimazole can be used. Systemic antifungal agents which can be used are fluconazole, itraconazole, ketoconazole.

Coccidioides

Coccidiodes is a group of dimorphic ascomycete, causes coccidioidomycosis also known as San Joaquin valley fever. The disease is acquired via respiratory inhalation of spores. The most common clinical manifestation is acute respiratory illness characterized by fever, cough, pleuritic pain, erythema nodosum. It can cause a severe and difficult to treat meningitis in AIDS patients, and also can cause acute respiratory distress syndrome and fatal multilobar pneumonia. Treatment is usually by oral fluconazole, itraconazole, ketoconazole or IV Amphotericin B.

Aspergillus

Aspergillosis is caused by a fungus (aspergillus) which is commonly found growing on dead leaves, stored grain, and compost piles. The disease caused by aspergillus is

Pulmonary aspergillsois: allergic bronchopulmonary type is an allergic reaction to the fungus that usually develops in people who already have a lung problem.
Aspergilloma, which develops in area of past lung disease. Pulmonary aspergillosis: invasive type seen in immunocompromised patients. Diagnosis is usually established by aspergillus antibody testing, chest radiograph, CBC, CT scan, sputum stain and culture for aspergillus; Treatment is voriconazole, Amphotericin B, or itraconazole.

Parasitic pathogens (protozoa, metazoa)

Giardia lamblia (giardiasis):
Giardiasis typically affects travelers and hikers who drink untreated water that has been contaminated with infected animals such as beavers, muskrats, and sheep. Widespread outbreaks also occur through sewage contamination of water supplies. Unprotected anal sex is also a source of transmission. Among children important mode of transmission is outbreaks of diarrhea through feco-oral transmission.
Laboratory tests for Giardia lamblia infection (giardiasis) are microscopic stool examination; duodenal aspirate or biopsy if repeated stool examinations are negative.
Treatment is usually indicated for symptomatic patients. The drugs of choice are 5 days course of metronidazole, single dose of tinidazole, or 3 day course of nitazoxanide. Other alternative drugs which can be used are albendazole, paromomycin, furazolidone.
Prevention is mainly by strict hand hygiene in day care centers, boiling water for at least 1 min

before using, avoiding drinking water from untreated streams, lakes etc.

Clinical manifestations of Giardia lamblia infection: Most of the patients are asymptomatic. Symptoms of acute infection include watery diarrhea, abdominal pain, nausea, vomiting, and weight loss. With more chronic infection children may develop failure to thrive, impaired cognitive function.

Toxoplasma gondii (toxoplasmosis)

Clinical manifestations of congenital toxoplasmosis are hepatosplenomegaly, jaundice, generalized lymphadenopathy, thrombocytopenia, and meningoencephalitis leading to microcephaly, chorioretinitis, hydrocephalus, seizures, or deafness. Cerebral calcifications may be seen on ultrasonography or computed tomography scan. Early diagnosis and treatment with sulfadiazine and pyrimethamine plus supplemental leucovorin may improve long-term outcome in such patients.

Acute Toxoplasma infection in immunocompetent hosts acquired after birth is generally asymptomatic, but when symptomatic infection occurs, it may present as a mononucleosis-like illness, with generalized lymphadenopathy, fever, chills, malaise, headaches, myalgias, pharyngitis, or hepatosplenomegaly.

Epidemiology of toxoplasmosis: Hosts: Felines are the natural host for T gondii. Intermediate hosts: Cats excrete infectious oocysts in their feces, which can be ingested by other animals and disseminated to their tissues, including skeletal muscle and other organs. Modes of transmission: vertical transmission from mother to infant, ingestion of cysts from contaminated food or soil, ingestion of undercooked meat (especially ground beef or lamb) and drinking unpasteurized goat milk, gardening in soil contaminated with cat feces also can lead to the transmission of the pathogen.

Trichomonas vaginalis (trichomoniasis)

Epidemiology of Trichomonas vaginalis: The incidence of Trichomoniaisis is highest among women who have multiple sexual partners. Most men and 30% of women are asymptomatic.

Clinical manifestations of Trichomonas vaginalis infection are copious malodorous gray frothy vaginal discharge, vulvovaginal irritation, dysuria, dysparenuia, lower abdominal pain, postcoital bleeding. Infected males are usually asymptomatic.

Diagnosis of Trichomonas vaginalis infection is by examination of a wet-mount preparation of vaginal secretions, enzyme linked immunosorbent assay and direct fluorescent antigen testing of vaginal secretions, culture of the organism.

Treatment of Trichomonas vaginalis is by using metronidazole or tinidazole. Sexual partner should also be treated at the same time to prevent reinfection.

Pneumocystis jiroveci (carinii)

Clinical manifestations of Pneumocystis jiroveci (carinii) infection are subacute diffuse pneumonitis with dyspnea, tachypnea, oxygen desaturation, nonproductive cough, and fever. Chest radiographs often show bilateral diffuse interstitial or alveolar disease.

Pneumonia caused by Pneumocystis jiroveci (carinii) occurs almost exclusively in immunocompromised patients. Diagnosis is usually established by clinical history, chest radiograph, and demonstration of P jiroveci organisms in lung tissue or respiratory tract secretion, the organism can also be isolated from open-lung biopsy and transbronchial biopsy, bronchoalveolar lavage, deep endotracheal aspiration with intubation, and induced sputum.

Trimethoprim with sulfamethoxazole (Bactrim) is effective for the prophylaxis as well as treatment of a Pneumocystis jiroveci (carinii) infection.

F. Enterobius vermicularis (pinworms)

Enterobius vermicularis infection is usually seen in preschool and school age children. It is usually asymptomatic but may present with rectal itching, abdominal pain, rectal bleeding, and hemorrhagic colitis.

Diagnosis is usually established by cellophane tape test which is

applied to perianal skin, than the tape is placed onto a slide to visualize the bean shaped eggs under microscope. The test should be performed after the patient has been asleep for some time or in the early morning. The treatment is by using mebendazole, pyrantel pamoate, albendazole.

G. Plasmodium species (malaria)

Malaria parasites have developed resistance to many drugs. Prophylactic regimens are determined by the resistance patterns in the country of destination. When a person decides to travel to such areas, the primary doctor should go through CDC specific regimen for that particular area and then start the chemoprophylaxis before travel. Most commonly used drugs in children more than 8 years are chloroquine, mefloquine, and atovaquone-proguanil.

Malaria should be considered a possible diagnosis in a febrile patient who has traveled to an endemic region.

Pasmodium falciparum produces the most severe disease because it has the potential to affect the highest percentage of red blood cells. It can produce a variety of clinical syndromes that can be fatal unless recognized and treated promptly. Cerebral malaria, which can include a decreased level of consciousness, seizures, retinal hemorrhages, and hemiplegia, is the most common syndrome seen, with mortality estimated at 20% to 40% if untreated. Other potential life-threatening symptoms are hypoglycemia, low-output renal failure, noncardiogenic pulmonary edema, respiratory failure, and shock.

Ascaris (ascariasis)

Ascariasis is infection with the parasitic roundworm Ascaris lumbricoides. It is caused by consuming food or drink contaminated with roundworm eggs. Clinical manifestations of ascariasis are bloody sputum, persistent cough, and shortness of breath, wheezing, abdominal pain, nausea, vomiting, loose stools, bloody diarrhea, weight loss, and worm in stool or vomit. Diagnosis is usually established by stool ova and parasite exam. Treatment is by albendazole, or mebendanzole.

Entamoeba histolytica (amebiasis)

Amebiasis is an infection of the intestines caused by the

parasite Entamoeba histolytica. It can live in the large intestine (colon) without causing disease. However, sometimes, it invades the colon wall, causing colitis, acute dysentery, or chronic diarrhea and tenesmus. Diagnosis is established by examination of the blood for amebiasis. Treatment is by metronidazole for 10 days.

Necator americanus (hookworm)

Hookworm infection is caused by Ancylostoma duodenale and Necator americanus. They are ubiquitous organisms in rural, tropical, and subtropical locales. Humans are the only reservoir of infection. Hookworms flourish where soil is contaminated with human feces. Eggs hatch in soil in 1

to 2 days and become larvae. They may infect humans percutaneously, causing itching and burning of the skin, or may be ingested, causing pharyngitis and gastroenteritis. Diagnosis is made by finding eggs in stool, usually within 5 to 10 weeks after infection. Children who are infested with large worm burdens may exhibit failure to thrive, short stature, and anemia due to chronic blood loss in the intestine and inability to absorb iron. Treatment is by albendazole, mebendazole.

Taenia solium, Taenia saginata, Taenia asiatica

Cysticercosis is a major cause of seizures in countries where Taenia solium is endemic. Humans are infected either by ingestion of food contaminated with feces containing eggs, or by autoinfection. Treatment is by using praziquantel, albendazole and steroid.

Toxocara:

Toxocariasis is an infection caused by the ingestion of larvae of the dog roundworm Toxocara canis or the cat roundworm Toxocara cati. Clinical manifestions include covert toxcariasis, visceral or ocular larva migrans. Treatment is by using mebendazole, thiabendazole, and corticosteroids.

Emerging infectious diseases

SARS-associated coronavirus infection
Severe acute respiratory syndrome (SARS) burst upon the world scene in late 2002, early 2003. The initial cases appeared in the Guandong province of China and spread rapidly to Hong Kong and other areas of Southeast Asia. Through air travel, the disease subsequently spread

throughout the world, with a significant outbreak in Toronto, Canada. A number of cases also were reported in the United States.

Clinically, SARS is defined as a febrile illness with cough or difficulty breathing and close contact with a case of SARS or travel or residence in an area of local transmission of SARS within 10 days of symptom onset. Studies of affected patients led to the isolation of a new coronavirus (SARS-coronavirus).Treatment with corticosteroids and the antiviral drug ribavirin were tried during the SARS outbreak, but none of them were beneficial.

Avian influenza

Avian influenza H5N1 is a highly pathogenic strain in bird and poultry populations that originated in Southeast Asia in 1997. It is not a human strain, but it has demonstrated the capacity for transmission to humans who have close contact with infected birds or poultry. Avian influenza causes lower respiratory disease with greater than 50% mortality in confirmed cases.

Metabolic Disorders

Diagnosis 1 .General
Screening
Genetics

Genetic counseling is appropriate for the family of a child with a metabolic disease when that disease is a heritable condition. Testing parents and siblings may affect their future family planning.

Recognition by signs and symptoms

Signs and symptoms of inborn errors of carbohydrate metabolism are varied and depend on the defect. Clinical presentations include vomiting, lethargy, failure to thrive, hepatomegaly, hypoglycemia, metabolic acidosis, cardiomegaly, and myopathy.

Signs and symptoms of hyperinsulinism include neonatal hypoglycemia without maternal diabetes, elevated insulin levels during hypoglycemia, macrosomia, jitteriness, and seizures.

Lipoprotein disorders are most often asymptomatic and are found when serum lipid levels are taken on account of a family history of hypercholesterolemia. Familial hypercholesterolemia may present with xanthomas or corneal arcus in teenagers. Hypolipoproteinemias may present with diarrhea, fat malabsorption and failure to thrive. Severe variants present with impaired neurodevelopment.

Gaucher disease may present from infancy to adulthood. Symptoms include bruising, chronic fatigue, growth retardation and bone pain. Signs include thrombocytopenia, anemia, hepatosplenomegaly, elevated liver enzymes, Erlenmeyer flask deformity of distal femur, and Gaucher cells in bone marrow with wrinkled paper appearance. Presentation in adults is associated with Type 1 disease and Ashkenazi Jewish descent. Types 2 and 3 disease have a more rapid neurodegenerative course.

Urea cycle defects typically present within a few days of birth with the side effects of hypermmonemia – vomiting, dehydration, lethargy, coma, seizures, increased ICP. The clinical picture may resemble sepsis. Laboratory studies will typically be normal, including blood urea nitrogen and pH. Testing for the 6 enzymes in the urea cycle –Carbamyl phosphate synthetase (CPS), Ornithine transcarbamylase (OTC), Argininosuccinate synthetase (AS), Argininosuccinate lyase (AL), Arginase, and N-Actetylglutamate synthetase – will show absence or deficiency.

Organic acidemias have varied clinical presentations and laboratory findings. Common features include presentation of a sepsis-like illness in an apparently

healthy neonate, with high anion gap metabolic acidosis, pancytopenia, hypoglycemia, hyperammonemia, ketosis, vomiting, refusal to feed and lethargy.

Glycogen storage disease may present in the newborn period with hypoglycemia and lactic acidosis. More common presentation is at 3 months with hepatomegaly or hypoglycemic seizures. Appearance includes “doll facies,” with fat cheeks, thin extremities, short stature and protuberant abdomen due to hepatomegaly. Laboratory findings include hypoglycemia, lactic acidosis, hyperuricemia and hyperlipiemia. Later presentations may include polycystic ovaries, pancreatitis, pulmonary hypertension and renal disease.

Tay-Sachs disease typically presents with loss of motor skills at 4 months, increased startle reflex, retinal cherry red spots, and macrocephaly. In most cases, at least one parent will be of Ashkenazi Jewish descent. In early childhood, seizures unresponsive to antiepileptic therapy and further loss of skills occur. Death usually occurs by 5 years. Later onset forms typically present with ataxia and absence of the retinal cherry red spot.

Evaluation of comatose children with suspected metabolic disease should include electrolytes, calcium, magnesium, phosphorous, glucose, liver function tests, serum ammonia, plasma amino acids, and urine organic acids to evaluate for amino acid disorders, organic acidemias, urea cycle

disorders, fatty acid oxidation defects, mitochondrial disorders, and disorders of carbohydrate metabolism.

An important laboratory studies in a hypoglycemic child with suspected metabolic disease is serum ketones. Other studies include electrolytes, acylcarnatine, carnitine, creatinine kinase, uric acid, triglycerides and lactate to evaluate for glycogen storage disease fatty acid oxidation disorders, organic acidemias and mitochondrial disorders.

For an acidotic child with suspected metabolic disease, the anion gap, electrolytes, arterial blood gas, urine pH, plasma lactate, pyruvate, amino acids and urine organic acids should be obtained to evaluate for organic acidemias and urea cycle disorders.

Clinical features of disorders of fatty acid and carnitine metabolism include a typical presentation between 3 months and 5 years with an acute Reye's like encephalopathy triggered by prolonged fasting. Patients may have seizures, an acrid odor, vomiting, lethargy, hepatomegaly, proximal muscle weakness, cardiomyopathy, or arrhythmias.

Disorders of amino acid metabolism typically present in a well newborn that develops acute lethargy, poor feeding, encephalopathy or coma after feeding with protein. If undetected, developmental delay or regression may result. Laboratory studies may show metabolic acidosis, hyperammonemia,

hypoglycermia, elevated liver function tests and abnormal plasma amino acid and urine organic acid levels.

Symptoms of Mucopolysaccharidoses include neonatal inguinal hernias, coarse facial features, corneal clouding, large tongue, developmental delay, and hearing loss. Signs include heptosplenomegaly, short stature, skeletal dysplasia, cardiomyopathy, mitral and aortic valve incompetence, coronary artery narrowing, obstructive airway disease, and communicating hydrocephalus.

Symptoms of Mitochondrial diseases are varied and include skeletal malformations, hypotonia, and refusal to feed, vomiting, seizures, developmental delay, blindness or deafness. Signs include cardiomyopathy, increased liver function tests, metabolic acidosis, lactic acidosis and hypoglycemia.

Recognition by laboratory results

Laboratory findings with a disorder of fatty acid metabolism include dicarboxylic aciduria and non-ketotic hypoglycemia.

The laboratory findings in a disorder of carnitine metabolism include reduced carnitine levels in plasma and muscle tissue, non-ketotic hypoglycemia, hyperammoniemia, and increased liver enzymes.

Mucopolysaccharidosis laboratory findings include elevated urine excretion of glycoaminoglycan. Enzyme assay of serum, leukocytes or cultured fibroblasts will identify the deficient enzyme to diagnose subtype. Radiographs show dysostosis multiplex, with thickened ribs and ovoid vertebral bodies.

Before treating a child with a suspected metabolic disorder, the clinician should obtain a CBC, complete metabolic panel, urinalysis, arterial blood gas, serum ammonia, lactate, pyruvate, plasma amino acids, and urine organic acids.

reatment
General

With Phenylketonuria, untreated infants develop profound mental retardation. Only 2- 5% will have normal intelligence. Despite early and adequate treatment, some develop mental retardation, ataxia, and seizures due to neurotransmitter deficiency.

A positive neonatal screening test should be repeated to confirm a positive result, along with basic testing for the particular defect. If the screen was performed before adequate oral feedings, a repeat screen is especially important, as fasting or total parental nutrition can significantly alter results.

Diet – limit to safe and tolerable substrates.
Medication

Glycogen storage disease management is as varied as the subtypes. Most require treatment only during illness or strenuous activity. The exception is Type 1, Glucose-6- Phosphatase Deficiency, which is managed with a combination of dietary restriction of fructose and galactose as well as oral intake of uncooked cornstarch.

Hypoglycemia should be treated initially with 2mL/kg of D10W, followed by a glucose infusion rate of 6-8mg/kg/min, with the rate adjusted based on the serum glucose.

Prognosis and long-term care

Prognosis for Phenylketonuria has improved significantly since the advent of neonatal screening. Early and adequate treatment decreases the incidence neurodegenerative complications. Despite adequate treatment patients sometimes develop mental retardation, poor tone, balance and coordination, ataxia and seizures.

Management of Galactosemia is primarily through dietary avoidance of all forms of galactose. Long term prognosis is guarded, with many having cognitive delays, especially with speech, focal neurological deficits, ovarian dysfunction, and cataracts.

Treatment of Gaucher disease is with beta-glucosidase enzyme replacement. Type 1 disease is highly responsive, with reversal of skeletal symptoms. Types 2 and 3 have only a palliative effect from enzyme replacement, and still progress to neurodegeneration and death, typically from respiratory failure, in all cases.

Prognosis for urea cycle defects is variable, dependent on the particular deficiency, prompt recognition and treatment, and lifelong treatment compliance. Most patients have some degree of cognitive defect. Patients must be protein restricted and supplemented with alternatives and closely monitored for appropriate growth. There is an absolute contraindication to valproic acid for seizures, as it raises serum ammonia levels.

Management of the organic acidemias includes dietary protein restriction with supplementation of amino acid preparations that do not include the offending protein. Carnitine is also supplemented. Prognosis is variable. Many infants do not survive the initial presentation because of the severity of illness or misdiagnosis. Those who do survive may have varying degrees of cognitive delays.

Long term prognosis for Mucopolysaccharidosis is highly dependent on age of treatment. Most infants are asymptomatic. If hematopoietic stem cell transplantation is completed before age 24 months, neurological outcome is favorable. Enzymatic replacement is the mainstay of treatment. Airway obstruction, recurrent respiratory infections and cardiac complications are the most common causes of death, usually by 10 years of age.

The prognosis for Tay-Sachs disease is poor. Most children die by age 5 years. Chronic management involves only palliative care.

Acute treatment of symptomatic hypoglycemia due to hyperinsulinemia should include a 2mL/kg bolus of D10W, followed by glucose infusion rate starting at 6mg/kg/min.

Persistent hypoglycemia may be treated with diazoxide, octreoide and treatment of the underlying disorder. Asymptomatic infants with hyperinsulinism have a good long term-prognosis, with recurrence in only 10-15% after treatment. Those that develop ketotic hypoglycemia are at increased risk of persistent hyperinsulinemia and hypoglycemia of infancy. This is associated with poorer intellectual prognosis.

Chronic management of lipid disorders should always include a plant rich diet low in saturated fats and sugars as well as increased exercise. Pharmacologic therapy with lipid lowering medications should only be initiated in children under 10 years with severe primary hyperlipidemia. Children over 10 years may be initiated if lifestyle modification does not decrease cholesterol levels or other cardiovascular risk factors are present.

The prognosis for all forms of glycogen storage disease is good and has improved significantly. Glucose-6-Phosphatase Deficiency (G6PD) is the most concerning subtype, with the potential for short stature, cardiac disease from hyperlipidemia, polycystic ovaries, pancreatitis, pulmonary hypertension and renal disease. Those with uncontrolled disease may develop hepatic adenomas with risk for malignant transformation.

I. Endocrine Disorders

Sex differentiation

1. Normal Development

2. Ambiguous Genitalia

a. Congenital adrenal hyperplasia

The signs and symptoms of congenital adrenal hyperplasia depend on the enzymatic deficiency. In classic 21-hydroxylase deficiency females have ambiguous genitalia with virilization. Males may have no external features or may have over virilization with hyperpigmentation of external genitalia. Neonates can present at 1-2 weeks of age with salt wasting crisis, with vomiting and natriuesis. Labs would show hyponatremia, hyperkalemia and hypoglycemia. Non-classic 21-hydroxylase deficiency may be asymptomatic or show virilization in females which can present at any age. Both boys and girls with non-classic CAH can present with symptoms of precocious adrenarche. Those with 11-betahydroxylase deficiency have hypertension. Males have postnatal virilization, while females have ambiguous genitalia. In 3- betahydroxysteroid dehydrogenase deficiency both sexes exhibit ambiguous genitalia and disordered puberty. With 17-

alphahydroxylase deficiency, males have ambiguous genitalia, while both sexes show sexual infantilism.

Evaluation of suspected congenital adrenal hyperplasia includes serum electrolytes, plasma renin, 17-hydroxyprogesterone, 11-deoxycortisol, 17- hydroxypregnenolone, cortisol, androstenedione, and DHEA.

Congenital adrenal hyperplasia due to 21-hydroxylase deficiency may be diagnosed prenatally by examining the CYP21 gene via chorionic villus sampling or amniocentesis. This is done when parents already have an affected child.

Management of CAH includes glucocorticoid replacement with hydrocortisone and mineralocorticoid replacement with fludrocortisone. Infants less than a year of age will also need salt (sodium chloride 4 mEq/kg/day) added to their feeds. During mild illness, children will need triple their daily maintenance dose of Hydrocortisone.

For patients in adrenal crisis, the mainstay of treatment is stress dose hydrocortisone, approximately 50-100 mg/m2 which can be 5-10 times the daily maintenance dose, as well as a 20ml/kg bolus of D5 or D10 NS to replace fluid and sodium losses and correct hypoglycemia and hyponatremia. Other electrolyte imbalances, such as hyperkalemia, should be treated appropriately if present.

Neonatal screening programs analyze 17-hydroxyprogesterone levels in all newborns. Males with normal genitalia and salt wasting disease most often are detected by this routine blood work before an adrenal crisis. The screening, however, has a high false positive rate because the threshold is set so low and usually does not detect non-classic 21-hydroxylase deficiency.

b. Other intersex disorders

Intrauterine exposure to excess maternal androgens or progestins can cause virilization in female infants. This may also occur with ovotesticular DSD (Disorders of sexual differentiation) where there is some functioning testicular tissue present in the gonads.

B. Growth

1. Short stature (see also VII.D.1)

a. General

The most common causes of short stature are prenatal onset pathologic short stature, postnatal onset pathologic short stature, constitutional growth delay, and familial short stature. Both pre and postnatal pathologic short stature have varied causes, sometimes overlapping.

For a child whose height has decreased from the 20th to the 5th percentile, all other causes of short stature must be ruled out for idiopathic short stature to be diagnosed. A full history with special attention to familial growth history and physical should be undertaken. Laboratory evaluation includes CBC, complete metabolic profile, thyroid panel, inflammatory markers, IGF-1, tissue transglutaminase, karyotype (in girls) and bone age.

Children with chronic disease may exhibit decreased growth velocity. However, growth should not be stagnant. If so, this should prompt investigation to other causes.

Constitutional growth delay shows decreased growth velocity in late infancy with a steep increase during puberty, which is delayed. Charts for familial short stature show a curve below but parallel to normal growth velocity. Postnatal pathologic short stature, such as growth hormone or thyroid deficiency, shows normal stature and height velocity until the condition develops, then a sharp decrease in velocity that may be regained with treatment.

b. Familial

In familial short stature, both the infant and parents are below average size. Growth curves are parallel to but persistently below the mean from birth. Bone age is comparable to chronological age.

The natural history of familial short stature is for the patient to follow along a growth curve below but parallel to the mean, with a mature adult height two or more standard deviations below the mean.

c. Constitutional growth delay

In constitutional growth delay GH response testing is lower. IGF-1 levels are low for chronological age but normal for bone age. Bone age radiographs will show a delayed bone age as compared to chronological age.

Children with constitutional growth delay are likely to have a parent that exhibited the same growth pattern with delayed puberty. Most achieve normal adult stature.

Predictions of height based on bone age tend to overestimate, more so in boys than girls. Boys may benefit from a short course of testosterone if puberty is delayed beyond age 14 called a puberty "jump start".

On a growth chart for a patient with constitutional growth delay, weight and height decrease in early childhood, parallel the average growth curve through childhood, and accelerate after the average age of puberty.

d. Growth hormone deficiency

Congenital growth hormone deficiency presents with growth retardation, hypoglycemia and prolonged jaundice in the neonatal period, delayed milestones due to decreased muscle mass, and micropenis and scrotal hypoplasia when combed with gonadotropin deficiency. Acquired growth hormone deficiency presents with severe growth failure, delayed bone age, cherubic facies, micropenis and delayed puberty.

Growth hormone therapy results in improvement and subsequent normalization of linear growth velocity. Child exhibits catch up growth over time with height returning to a percentile that is within the range expected for genetic potential.

e. Tall stature

The most common cause of tall stature is familial. In patients with tall stature, a full history, particularly family history with relation to height and puberty, and physical should be performed. If there is familial tall stature, no work up may be needed. If not, bone age and chromosome studies are the most useful tests. Other less common causes include excessive growth hormone secretion, pituitary gigantism, prolactinomas and syndromic etiologies (Klinefelter, Beckwith-Wiedemann, Marfan).

C. Puberty

1. Normal

Pubertal changes are related to gonadarche (activation of the testes or ovaries by release of pituitary gonadotropins) and adrenarche (increase in adrenal androgens).

In girls, the first sign of true puberty is thelarche (increase in breast tissue) which can occur between 8 to 12 yrs of age. Average age of menarche is 12 years. In boys, the first sign of puberty is increase testicular volume which can occur between 9 to 13 years of age. Pubarche (development of thick, coarse and dark pubic hair) usually occurs around the time of gonadarche though in 15% of girls may be the first sign of puberty.

Precocious puberty is defined as the onset of these changes in females before age 8 and males before age 9. Studies have shown an earlier onset of puberty in recent years, particularly in African-American and Hispanic girls. The need for work up for pubertal changes in girls between the ages of 6 to 8 yrs, therefore depends on the rate of pubertal progression and family history of pubertal timing.

Normal gynecomastia in males occurs around the time of puberty, may be unilateral or bilateral and may be transiently tender. It is due to decreased ratio of testosterone to estradiol and excessive aromatase activity.

2. Precocious puberty

Precocious puberty is the appearance of pubertal changes in girls younger than 8 years and boys younger than 9 years. Signs include thelarche in girls and testicular enlargement in boys, with development of secondary sexual characteristics, in the same order as typical puberty. The rate of pubertal progression may be same as typical puberty or increased. Symptoms may include mood swings.

Precocious puberty may be central or peripheral. Central (gonadotropin-dependent) results from activation of the gonadal-pituitary- hypothalamic axis. Peripheral (gonadotropin independent) precocious puberty occurs with sex steroid production from the gonads without gonadotropic stimulation so LH and FSH are low/suppressed with elevated testosterone or estradiol. Peripheral activation of the gonads with sex steroid production may subsequently activate the central axis once the bone age is sufficiently advanced to pubertal age.

Premature thelarche is an isolated finding, common in girls 1-2 years old. This coincides with physiological “mini-puberty” which can occur up until 2 yrs in girls when gonadotropins and elevated. This most often regresses spontaneously and does not warrant treatment. A bone age may be obtained to help differentiate from central precocious puberty if progression is noted.

In a child of either gender with premature breast development, a history of medication use in the child or close contacts should be obtained, with attention to oral contraceptives or estrogen creams. Thelarche regresses after exogenous exposure ceases.

Exposure to testosterone creams, even from skin contact with another who has applied a cream, can result in virilization of both sexes. It regresses after exposure has ceased.

Central puberty is largely idiopathic in females. Seventy five percent of males with central precocious puberty have CNS tumors. Hypothyroidism can be a cause in both sexes. Hypothalamic hamartomas are the most common CNS lesion causing central precocious puberty. Astrocytomas, ependymomas, pineal and optic tract tumors are also implicated. Hepatoblastoma is another cause.

Peripheral precocious puberty may result from gonadal or adrenal tumors. Choriocarcinomas, teratocarcinomas, or teratomas in the mediastinum, gonads, or adrenal glands are a more common cause in boys. McCune Albright syndrome, exogenous androgen exposure and uncontrolled congenital adrenal hyperplasia (CAH) or non-classic CAH can also be possible etiologies of peripheral puberty in both boys and girls. Familial male-limited precocious puberty can be an etiology in males which occurs due to an activation of the LH receptor due to a genetic mutation.

3. Delayed puberty

a. General

Delayed puberty is the failure of development of thelarche in girls by age 13 and lack of testicular enlargement in boys by age 14.

Delayed puberty may be caused by constitutional pubertal delay, functional hypogonadotropic hypogonadism (due to GH deficiency, inadequate diet, excessive exercise), hypogonadotropic gonadism (GnRH deficiency), and hypergonadotropic hypogonadism (gonadal failure).

Work up in a child with delayed puberty would include laboratory evaluation of gonadotropins and sex steroid hormones. A bone age X-ray is also essential for evaluation which is usually delayed compared to chronological age in children. The severity of bone age delay may give an indication of when pubertal changes would be expected to occur.

b. Primary gonadal (gonadal dysgenesis/Turner syndrome)

Clinical features of Turner syndrome include short stature, congenital lymphedema, cubitus valgus, wide spaced nipples, shield-like chest, neck webbing, low posterior hairline, horseshoe kidney, cardiac abnormalities, and gonadal dysgenesis.

Turner syndrome is diagnosed by a karyotype showing either absence of or structural abnormalities in one of the X chromosomes.

Girls with Turner syndrome have a 40% incidence of cardiac abnormalities and 60% incidence of renal abnormalities. The most common cardiac manifestations are bicuspid aortic valve (20-30%) and coarctation (10%). The most common renal abnormality is horseshoe kidney. Cardiac assessment via Echocardiogram and 4 limb blood pressures are recommended at diagnosis. Cardiac MRI has also been used for accurate assessment of structural cardiovascular defects in these

patients. A baseline renal ultrasound is needed at diagnosis and yearly urinalysis (UA) and serum creatinine is advised.

Girls with Turner's syndrome may have initial breast development but most will not have pubertal progression due to premature ovarian failure. Some will have no thelarche. Laboratory evaluation reveals significant elevation of FSH with low/undetectable estradiol. Pelvic ultrasound may show an underdeveloped uterus and streak gonads. Management is with transdermal estrogen for approximately 2 years to allow for growth of breast and uterine tissue, followed by oral contraceptives (estrogen and progesterone) to initiate menses.

c. Constitutional

Family history of delayed puberty is important to determine in a child with delayed onset of puberty. Up to 75% of children with delayed puberty have a family history of a parent or sibling with history of pubertal delay. Research suggests that constitutional delay of puberty may be autosomal dominant with or without incomplete penetrance.

In constitutional delay of puberty, the growth chart will show slowed but not stalled growth. It may appear that the child has fallen off their linear growth curve because most other children of pubertal age will have an increase in linear growth velocity during the pubertal "growth spurt." Bone radiograph (bone age) will show a delayed bone age consistent with the current Tanner stage.

The natural history for constitutional delay puberty is for the child to start puberty at a later age than average, but attain full projected adult height and complete pubertal development.

Treatment for constitutional delayed puberty should only be used in boys older than 14 years and girls older than 13 years who show no signs of pubertal development and express significant anxiety about the delay. For boys monthly intramuscular testosterone is preferred and should only be used for a short course. For girls, oral or transdermal estrogen may be used and is given at half the adult dose and gradually increased. After two years of unopposed estrogen, progesterone therapy should be initiated.

D. Thyroid disorders

1. Hashimoto thyroiditis

The most common manifestations of Hashimoto thyroiditis are goiter (diffusely enlarged, firm, nontender) and growth retardation. Most children are asymptomatic at diagnosis. Few have clinical symptoms of hypothyroidism, and fewer have hyperthyroid symptoms.

Evaluation for hypothyroidism with concern for Hashimoto's includes TSH, Free T4 or total T4, thyroglobulin antibodies and thyroid peroxidase antibodies.

Thyroid function tests are often normal in Hashimoto thyroiditis, but TSH may be mildly elevated (subclinical hypothyroidism). Some children are positive for anti-thyroid peroxidase antibodies, while others are positive for thyroglobulin antibodies. Testing for both results in 95% sensitivity for autoimmune thyroiditis; Imaging is not warranted unless there are other symptoms that indicate an infectious etiology (fever, tenderness, and dysphagia).

The natural history of Hashimoto thyroiditis is variable. Most patients who are clinically euthyroid at presentation remain so. Approximately half of the patients with subclinical hypothyroidism develop overt hypothyroidism over time.

-Hashimoto thyroiditis is the most common cause of goiter in adolescents.

-Hashimoto thyroiditis may be associated with other autoimmune disorders.

2. Cyst, tumor, nodule

Most children with a thyroid cyst or tumor are asymptomatic with the lesion found incidentally on exam or imaging. Patients may have mild discomfort due to mass effect. On laboratory evaluation most have normal thyroid function studies.

Children with a thyroid mass should be referred to endocrinology or ENT for further evaluation due to the risk of malignancy. A solitary nodule at < 20 yrs old is more likely to be malignant c/f older person. About 1/3rd of nodules in children will be malignant. Thyroid cancer ~ 1 - 1.5% of all childhood cancers and usually has an indolent course.

History of previous irradiation to the head and neck significantly increases the risk of both benign and malignant thyroid nodules starting 3-5 years after treatment. Increased dose, younger age at treatment, and female sex increase the risk of malignancy.

Work up for a thyroid nodule starts with a thyroid/neck ultrasound. If concerning features are present, diagnosis by FNA will be necessary. Thyroid scan (I-123 or Tc99) may be used to evaluate hot/cold nodules.

Thyroglossal duct cysts are midline neck defects that may extend to the base of the tongue. They characteristically move vertically with swallowing. They often appear after an upper respiratory infection (URI) or as an infected midline neck mass. Surgical removal is usually indicated especially if infection has occurred.

3. Hypothyroidism

Signs and symptoms of congenital hypothyroidism include large tongue, macrocephaly with large fontanel, poor feeding, somnolence, prolonged jaundice, constipation, edema, developmental delay, heart murmurs, cardiomegaly, and low hairline. These are not commonly seen in countries where newborn screening tests for congenital hypothyroidism and the neonate is often asymptomatic at diagnosis. Untreated neonatal hypothyroidism may result in failure to thrive from feeding difficulties (choking spells, lethargy), respiratory difficulties (large tongue, respiratory distress syndrome [RDS]), constipation, cardiac issues (cardiomegaly, pericardial effusion), and anemia.

Signs and symptoms of acquired hypothyroidism include short stature, goiter, fluid retention, dry skin, cold intolerance, constipation, and bradycardia.

Congenital hypothyroidism is commonly due to a defect in thyroid development (thyroid dysgenesis) or hormone synthesis (dyshormonogenesis). Other etiologies include maternal medications, hypopituitarism or unresponsiveness to thyroid stimulating hormone (TSH). Acquired

hypothyroidism may be caused by an autoimmune process, an iatrogenic process, or hypothalamic-pituitary disease.

Congenital and acquired hypothyroidisms are treated with levothyroxine. Dosage depends on weight and age. TSH and T4 levels should be checked at regular intervals.

If congenital hypothyroidism is found on the neonatal screen and treated promptly, prognosis is good. Those more severely affected may still have abnormalities in muscle tone, speech, mentation or hearing even with adequate treatment. If treatment is delayed, profound mental and growth retardation will occur. Prognosis for acquired hypothyroidism is also good. There may be loss of potential height if treatment is delayed, but there is very rarely neurological sequela.

Thyroid-binding Globulin Deficiency typically presents with low levels of T4 and elevated resin thyroiiodine uptake (T3 uptake), but normal free T4 and TSH. It has no clinical manifestations and does not require treatment.

4. Hyperthyroidism

Hyperthyroidism may affect most body systems. Symptoms include irritability or anxiety, palpitations, heat intolerance, weight loss with increased appetite, increased stool frequency or diarrhea, polydipsia or amenorrhea. Signs include tachycardia, tremor, hyperreflexia, hair loss, moist skin, goiter or proptosis.

In hyperthyroidism history and physical may reveal many of the above symptoms or signs. Total and free T3 and T4 will be elevated, and TSH suppressed. Antithyroid antibodies may be elevated. TSI (thyroid stimulating immunoglobulin) and TSH receptor antibodies are commonly elevated in Grave's disease which is the most common cause of hyperthyroidism. Another autoimmune etiology is Hashitoxicosis where the same antibodies associated with Hashimoto's cause a brief period of hyperthyroidism and then hypothyroidism develops. TSI and TSH receptor antibodies are negative in Hashitoxicosis. Radioactive iodine or Tc uptake can help differentiate Grave's disease from Hashitoxicosis and subacute thyroiditis.

Hyperthyroidism due to subacute thyroiditis usually follows a respiratory tract infection and is transient. This is also associated with pain in the thyroid/neck region.

Antithyroid drugs, primarily Methimazole, are common for treatment of hyperthyroidism in young children. Radioactive iodine therapy or excisional surgery are definitive therapies. These may be used if the patient and family does not wish to continue Methimazole due to risk of side effects, or if medical management is not practical or has failed after more than 2 years on therapy. Beta blockers may be useful for treating symptomatic patients on an emergent basis until thyroid hormone levels normalize on therapy.

Neonatal hyperthyroidism can present with an irritable neonate with exophthalmos, tachycardia, tachypnea and hyperthermia. Severely affected neonates develop weight loss, hepatosplenomegaly, jaundice and hypertension. T4 is markedly elevated with low TSH. This is caused by transplacental passage of TSH receptor antibodies/TSI from the mother to the fetus; remits spontaneously by 6-12 weeks though babies will need Methimazole and beta-blockers for management during this time.

E. Adrenal gland disorders

General

2. Addison disease

Signs of Addison disease include hypoglycemia, decreased cardiac output, orthostatic hypotension, hyperpigmentation, hyponatremia and hyperkalemia. Symptoms include nausea, vomiting, anorexia, weakness, dizziness, and seizures.

Laboratory testing in Addison disease will show hypoglycemia, ketosis, hyponatremia, and hyperkalemia. Cortisol and aldosterone levels can be low but can be inappropriately normal as compared to the requirements of acute illness. ACTH and renin levels are elevated. The most definitive test is an ACTH stimulation test. Cortisol levels are drawn before administration of Cosyntropin (ACTH) and 30 and 60 minutes after. A peak Cortisol of 18 or above would rule out adrenal insufficiency.

Patients with Addison Disease in Adrenal Crisis should be treated with D5 or D10 NS to correct hypoglycemia, hypovolemia, and hyponatremia. Hyperkalemia may require treatment with calcium, bicarbonate, kayexalate, or glucose and insulin. A hydrocortisone bolus should be given IV, with the dose dependent on age and size (stress dose of 100 mg/m2 may be needed in shock).

For maintenance therapy in Addison's disease, glucocorticoid replacement with Hydrocortisone at about 10 mg/m2/day is given in 2 to 3 divided doses and mineralocorticoid replacement is with Fludrocortisone daily. Triple the dose of maintenance Hydrocortisone is needed during periods of acute illness.

The most common cause of Addison disease is autoimmune destruction of the adrenal gland with antiadrenal antibodies, most commonly 21-hydroxylase autoantibodies.

After discontinuation of exogenous corticosteroid therapy, signs and symptoms of adrenal insufficiency include dizziness and weakness due to orthostatic hypotension, hypoglycemia and hyponatremia. Aldosterone levels will be normal.

Sudden withdrawal of corticosteroids in patients who have been on long term steroid treatment may result in Adrenal Crisis, with hypotension, hypoglycemia, hyponatremia, and hypercalcemia resulting in vomiting, diarrhea, dehydration, syncope, seizures, confusion and lethargy. Therefore a steroid wean may be needed to allow for recovery of the hypothalamic-pituitary-adrenal axis to avoid signs and symptoms of adrenal insufficiency.

Hyponatremia may be a feature of both adrenal insufficiency and SIADH. However, in SIADH, hypoglycemia and hypotension would not be expected whereas these are usually seen with adrenal insufficiency. In adrenal insufficiency due to Addison's disease, hyperkalemia would also be seen but this is not usually a feature of central adrenal insufficiency or SIADH.

3. Cushing syndrome

In younger children signs and symptoms of Cushing syndrome include moon facies, obesity, masculinization, impaired linear growth, hypertension and increased susceptibility to infections. Older children may have short stature, truncal obesity, moon facies, purple striae, delayed puberty, hypertension, hyperglycemia and osteoporosis.

Cushing syndrome in children may occur due to long term exposure to exogenous corticosteroids, including topical and inhaled preparations.

Laboratory work up in Cushing syndrome will show elevated midnight cortisol levels, which can be accomplished by saliva testing at home. Urinary cortisol levels are elevated in a 24 hour urine sample. Dexamethasone suppression test may be used, and protocols vary. A positive test will not show morning suppression of cortisol levels following a dose of Dexamethasone the night before. Once diagnosis is established, etiology can be determined with ACTH levels which would help differentiate ACTH-dependent from ACTH-independent Cushings.. Corticotropin Releasing Hormone tests and a two-step Dexamethasone suppression test are helpful to identify the source of ACTH (central versus ectopic) in ACTH-dependant Cushings. CT of the adrenal glands may also be useful particularly in ACTH-independent Cushings.

F. Pituitary gland disorders

Most children with hypopituitarism are of normal size and weight at birth, but may be as low as 4 standard deviations below the mean by 1 year. In the neonatal period they may present with apnea, cyanosis, hypoglycemia, seizures, microphallus, or prolonged jaundice. Appearance may include a short, broad face with a prominent frontal bone and flattened nasal bridge, small mandible and chin, bulging eyes, small hands and feet, pudgy appearance and delayed sexual maturation.

Craniopharyngioma may present with failure to thrive, linear growth deceleration, delayed sexual maturity, or visual changes. Children may have symptoms associated with multiple hormone deficiencies including hypothyroidism, growth hormone deficiency, adrenal insufficiency and diabetes insipidus.

Diabetes insipidus typically presents with polyuria and polydipsia. Signs of dehydration may be present. Rarely children may have a fever, nausea and vomiting.

Labs: BMP, serum osmolality, urine osmolality, urinalysis. Diagnosis of DI is clear if serum osm > 300 mOsm/kg, with a urine osm < 300 mOsm/kg. DI is unlikely if serum osm < 270 mOsm/kg, Urine osm > 600 mOsm/kg. Laboratory values of serum glucose, bicarbonate and calcium will be normal in DI. There may be hypernatremia from dehydration. Urinalysis will show low specific gravity. A water deprivation test may be needed if serum osmolality is borderline. With a water deprivation test, urine osmolality will be below 280 Osm/kg even with serum osm rising to over 300 mOsm/kg

G. Diabetes

1. General

a. Natural history of Type 1 DM:

Predisposition for T1DM begins at birth. The following factors may come into play:

Inheritance of genetic risk factors

Major histocompatibility complex on chr 6

HLA-DR3 and DR4

Triggering environmental factor is likely what induces progression to diabetes due to autoimmune activation. This may occur because of cross-reactivity against antigens on the beta cells being induced. There is then production of proinflammatory cytokines that injure islet tissue.

> The initiation of autoimmunity and loss of beta cell function precedes clinical disease. Once patients are on an insulin regimen, some experience a "honeymoon" phase, with apparent remission; this is due to variable amounts of insulin still produced by the pancreas working in tandem with exogenous insulin, causing intermittent hypoglycemia. Established disease follows this transient phase, which may progress to complications if not managed properly.

b. Clinical features: As Type 1 diabetes develops, children may experience intermittent polyuria or nocturia and/or enuresis. As it progresses, polyphagia develops to compensate for the calories lost due to glucosuria. As ketones build up, abdominal pain, nausea and vomiting develop, followed by dehydration. Signs include Kussmaul breathing, acetone breath, decreased cognition and prolonged QTc.

c. Diagnostic evaluation:

In new onset Type 1 diabetes, initial evaluation includes a BMP to assess for DKA, insulin level, C-peptide, Hemoglobin A1c and urinalysis. Antibodies associated with Type 1 Diabetes (GAD-65, insulin antibodies and islet antibodies may also be tested. If symptoms suggest, thyroid function tests and celiac screening may also be done due to the predisposition for these conditions in patients with Type 1 DM.

d. Management:

Glycemic control in Type 1 diabetes is achieved through a combination of insulin, attention to diet, exercise and disease acceptance. Exogenous insulin administration initially would include both long and short acting preparations. Dietary education includes carb counting with approximately 55% of calories from carbohydrates, preferably complex. Exercise is encouraged with possible adjustments in insulin dosing to avoid hypoglycemia. Acceptance of the disease is perhaps most important, because other components cannot be achieved without it. Insulin pump therapy, which utilizes only short acting insulin in a basal bolus pattern, is increasingly being used in patients with Type 1 DM once they have developed diabetes self-management skills.

When patients with Type 1 diabetes are sick, they may develop significant hyperglycemia. This puts them at greater risk for DKA. On sick days urine ketones should be monitored and corrected for with increased insulin. Patients who do not respond to the insulin or have emesis should be seen in the emergency department. Patients should always be in close contact with their physicians during sick days in order to avoid clinical deterioration.

The long term complications of Type 1 diabetes fall into three categories: microvascular (nephropathy, retinopathy), macrovascular (coronary artery disease, cerebrovascular disease, peripheral vascular disease) and neuropathies.

Intensive blood glucose management significantly reduces the incidence of long term sequelae; maintaining glucose levels near normal decreases incidence of retinopathy, nephropathy and neuropathy nearly 50%.

Children with Type 1 diabetes have a higher incidence of other autoimmune disorders. Celiac disease occurs in 5-10%, and Autoimmune Thyroiditis occurs in up to 20%. All patients should be screened for these diseases at diagnosis.

In a child with hypoglycemic symptoms, documentation via finger stick glucose should be attempted before treatment. For treatment, 15 g of glucose should be given orally and the blood glucose checked 15 minutes later. Intramuscular glucagon can be given when the patient is unable to take oral glucose due to repeated emesis, loss of consciousness or altered mental status.

Ketotic hypoglycemia is the most common type of hypoglycemia in early childhood, presenting between 18 months and 5 years, with spontaneous remission by age 9 years.

Ketotic hypoglycemia typically presents as a child who has a seizure or is comatose with a history of skipping dinner or decreased PO intake due to illness. The child is difficult to rouse in the morning, and skips breakfast. Hypoglycemia would occur within 12 – 18 hrs of fasting if susceptible. It also may involve a child whose overnight fast is prolonged by a parent sleeping late.

2. Diabetic ketoacidosis

The pathophysiology of Diabetic Ketoacidosis has three pathways. The first is increased serum glucose causing osmotic diuresis, dehydration and activation of the renin angiotensin-aldosterone system. The second pathway is increased catabolic processes leading to electrolyte depletion. The final pathway is release of free fatty acids to act as an energy source, which depletes the buffer and results in metabolic acidosis.

Patients initially have the same classic symptoms of DM (polyuria, polydipsia, polyphagia, weight loss), which become more severe. There is appetite loss with nausea, vomiting, and abdominal pain. Hyperpnea (Kussmaul respirations) develops and neurologic status progressively worsens from drowsiness to lethargy to possible obtundation.

Biochemical criteria

Blood glucose > 200 mg/dL

Venous pH < 7.30 or bicarbonate level <15 mmol/L

Treatment of DKA is by continuous insulin drip and fluid replacement with a solution containing dextrose, potassium and sodium. Concentrations vary by protocol. Glucose should be monitored hourly and ketones, electrolytes and pH regularly. Once stable, patients can be transferred to subcutaneous insulin and labs performed less frequently. A child with mild to moderate Diabetic Ketoacidosis should be admitted to a Pediatric ICU with an insulin drip, rehydration with a solution containing dextrose, sodium and potassium with frequent monitoring of glucose, ketones and electrolytes, magnesium, calcium and phosphate. Patients must be monitored closely for the development of hypoglycemia, hypokalemia due to hyperglycemia as well as dilutional effect, cerebral edema and shock.

Cerebral edema is a complication of the treatment of Diabetic Ketoacidosis for which the etiology is unknown. The incidence remains stable over the past two decades despite slow rehydration

protocols; occurs in 0.5 to 1% of pediatric DKA. Most cases occur 4 to 12 hours after treatment initiation.

Using sodium bicarbonate is not routinely recommended as a treatment for Diabetic Ketoacidosis as it may cause hypernatremia, volume overload, and over correction of acidosis leading to hypokalemia and hypophosphatemia. It is typically only used in children with extremely severe DKA.

Noncompliance is a frequent cause of recurrent episodes of diabetic ketoacidosis. In a child with frequent episodes, psychological issues such as depression or anxiety or familial conflict should be investigated.

3. Type 2 Diabetes

Type 1 Diabetes results from an autoimmune destruction of the pancreatic beta islet cells and subsequent insulin deficiency. Type 2 Diabetes is due to insulin resistance at the end organ level with various degrees of beta cell impairment.

Treatment of Type 2 Diabetes should always include lifestyle modification of diet, exercise and weight loss. In children, this often involves changing habits for the entire family. If those strategies do not work, Metformin may be used. In patients with significant elevation of HBA1C, insulin therapy may be necessary.

Screening by fasting serum glucose should be performed in any child 10 years or older who is overweight and has two of the following risk factors: family history of Type 2 Diabetes, mother with gestational diabetes, minority ethnicity, or signs of insulin resistance (acanthosis, hypertension, dyslipidemia, polycystic ovarian syndrome).

Testing should begin at 10 yrs of age or at age of pubertal onset. This should be repeated every 3 years. Either a fasting plasma glucose or 2 hour OGTT may be used.

Criteria for diagnosis are either a HBA1C of 6.5% OR Fasting plasma glucose of 126 mg/dl, OR a glucose level of > or = 200 on a 2 hour OGTT, OR a random glucose of 200 or above with symptoms of hyperglycemia.

Long term complications of Type 2 Diabetes include hypertension, fatty liver, polycystic ovarian syndrome, dyslipidemia, microalbuminuria, sleep apnea and retinopathy.

Acanthosis nigricans is a clinical sign of insulin resistance.

Complications of Type 2 diabetes may be present at diagnosis. Some, such as hypertension, sleep apnea, and Polycystic Ovarian syndrome, may be the presenting symptom for which the child was initially evaluated.

H. Metabolic syndrome

The screening for metabolic syndrome includes measures of obesity, blood pressure and serum lipids and glucose. The following features characterize metabolic syndrome:

Fasting hyperglycemia (fasting glucose 100 mg)/impaired glucose tolerance

Elevated blood pressure (95th percentile for age, sex, and height percentile)

Generalized or central obesity

Decreased HDL cholesterol concentrations (5th percentile for age)

Elevated triglyceride values (95th percentile)

Children should have their height, weight, and blood pressure measured at each visit. Those children over 10 years with central obesity >90th percentile, hypertension, clinical signs of insulin resistance, or family history of dyslipidemia should have fasting glucose and lipid panels.

Pediatric patients with metabolic syndrome should be managed with lifestyle modification such as physical activity and diet for weight loss and to prevent progression of the syndrome.

Little is known of the natural history of metabolic syndrome in children. If not managed properly or reversed with lifestyle modification, however, the syndrome can continue into adulthood. Adults with metabolic syndrome are at significantly increased risk of Type II Diabetes Mellitus and Cardiovascular Disease.

I. Disorders of PTH, calcium, and phosphate metabolism (see II.C.)

1. Hypocalcemia

Hypocalcemia may present similarly to Rickets, with failure to thrive, muscle weakness and bone manifestations (bowed long bones, delayed fontanel closure, rachitic rosary, and scoliosis). Severe cases may present with seizures or tetany. Patients often present after 15 months once weaned off formula or breast milk. Children who are breast fed without Vitamin D supplementation may present at less than a year of age. Serum calcium is low, with increased alkaline phosphatase, PTH and 1, 25 – dihyroxy vitamin D. Laboratory findings in vitamin D deficiency are hypocalcemia with hypophosphatemia and low 25-hydroxy Vitamin D.

Hypocalcemia in neonates may be caused by prematurity and very low birth weight, along with inadequate dietary sources (unfortified breast milk, non-premie formulas, and prolonged parenteral nutrition), diuretic use and inadequate Vitamin D or phosphorus.

Hypercalcemia

Hypercalcemia may present with muscle weakness, fatigue, constipation, weight loss or skeletal deformations. Calcium precipitation in the renal parenchyma can lead to hematuria. Laboratory findings include elevated serum and ionized calcium. If hypercalcemia is due to hyperparathyroidism, PTH will be elevated with low phosphorus and magnesium. Radiographs of long bones may show reabsorption of subperiosteal bone. In hypercalcemia due to malignancy, PTH will be appropriately suppressed but PTH-related peptide (PTH-rP) will be elevated.

Prolonged immobilization may lead to hypercalcemia, resulting in renal dysfunction, hypertension and encephalopathy.

Hypophosphatemia

Patients with familial hypophosphatemic rickets have poor growth, bone disease predominantly in the lower extremities and poor dentition. Serum phosphate is low and alkaline phosphatase is elevated. PTH and calcium are normal, with low 1, 25-D levels.

Familial hypophosphatemic rickets should be treated with a combination of oral phosphate and 1, 25-Vit D supplementation. Frequent dosing is needed to maintain steady serum levels. Care must be taken to balance phosphorus and 1, 25-D, avoiding excess phosphorus and hyperparathyroidism.

Rickets (see II.C)
Parathyroid disorders
Laboratory findings in hypoparathyroidism include low serum calcium and elevated phosphorous levels. PTH may be low or inappropriately normal. Typically alkaline phosphatase and 1,25 dihydroxy Vitamin D levels are low, while magnesium is normal. Plain films show increased density at the metaphyses of long bones, and CT scans of the skull can show calcifications of the basal ganglia. QT interval is prolonged on EKG due to decreased calcium levels.
Laboratory findings in hyperparathyroidism include hypercalcemia and hypophosphatemia, urine calcium is also low.
DiGeorge Syndrome is one cause of hypoparathyroidism due to congenital absence of the parathyroid gland. Albright's hereditary osteodystrophy is characterized by resistance to PTH (pseudohypoparathyroidism) so patients will also have low calcium, elevated phosphorus and low 1.25-dihyroxy Vitamin D but elevated PTH.

Gastrointestinal Disorders

Abdominal pain
Acute
General

Visceral pain receptors usually respond to stretch as these are located in the muscles, serosal and mucosal surface of the hollow organs and in the mesentery. Visceral pain may be associated with sweating, nausea, vomiting, pallor and anxiety due to secondary involvement of the autonomic nerves. The three major areas of association include epigastric area, umbilical area and the left lower quadrant. Any pathology that causes pain in the stomach, lower esophagus, and duodenum is perceived in the epigastric area. Visceral pain from the small intestine is perceived in the umbilical area and pain in colon is felt in the lower abdomen.

Evaluation should start with good history taking followed by physical exam. Laboratory work should include CBC with diff, basic metabolic panel, amylase and lipase. Also urinalysis may be done along with stool studies if indicated. Imaging may include X ray, ultrasound and even a CT scan.

Up to 1 year- pyloric stenosis (usually between 2 and 5 weeks of age), intestinal malrotation (with or without volvulus), incarcerated hernia, intussusception (two third of the total cases occur before 1 year of age). But between 6 months to 2 years incidence of intussusception is high.

Appendicitis, gastroenteritis and constipation can cause abdominal pain at any age. Children 1 year to 10 years old may have abdominal pain from hemolytic uremic syndrome. In children age ten years and older inflammatory bowel disease can cause abdominal pain.

Nonsteroidal anti-inflammatory medications inhibit cyclooxygenase and prostaglandin formation causing direct irritation to the mucosa. Lack of prostaglandin decreases resistance to injury.

Appendicitis

Appendicitis is a cause of acute abdominal pain.

Laboratory evaluations of appendicitis include CBC, CRP, and urinalysis. Electrolytes, liver chemistries with amylase, lipase and liver enzymes are done to rule out other causes of abdominal pain.

Cholecystitis, cholelithiasis

Presentation of chronic and acute cholecystitis is somewhat different. Children with chronic cholecystitis may have a history of intolerance of fatty foods and episodic pain in the upper abdominal area. Children with acute cholecystitis may present with nausea, vomiting, jaundice and low grade fever. Physical examination may reveal right upper quadrant pain, tenderness, guarding and positive Murphy sign. Laboratory results in acute cholecystitis include elevated white blood cell count with left shift. Other laboratory values may include elevated levels of bilirubin (direct hyperbilirubinemia), alkaline phosphatase and gamma-glutamyl transferase and often mildly elevated transaminases and amylase. These laboratory values may or may not be elevated in chronic cholecystitis.

The risk factor associated with the development of cholelithiasis are sepsis, hemolytic disease like sickle cell disease, abdominal surgery, malabsorption, prolong exposure to parenteral nutrition, bronchopulmonary dysplasia, necrotizing enterocolitis, cystic fibrosis, chemotherapy and heart valve transplant. The risk increases with age and obesity. Pregnancy and use of oral contraception may play a role in developing cholelithiasis. There is higher incidence of cholelithiasis in ethnic groups like Pima Indians, Scandinavians and Hispanics.

Pancreatitis

Abdominal pain, nausea, vomiting, anorexia and low grade fever are typical presentation of pancreatitis. A child with pancreatitis is ill appearing and often sitting upright, or lying on the side with knees flexed. Pain is located in the epigastric area or upper quadrant, periumbilical area, back or lower chest. The pain is usually sharp and sudden or constant in nature. On physical exam a distended abdomen with palpable mass may be appreciated with decreased bowel sounds. Child may seem icteric and have hypotension and tachycardia. In severe acute pancreatitis, ascites, hypocalcaemia, and pleural effusion may be appreciated. Blue discoloration of the flanks (Grey Turner sign) or around the umbilicus (Cullen) sign may be seen with necrosis or hemorrhage.

Laboratory findings include increased levels of serum amylase and lipase. However, normal levels of concentration do not exclude diagnosis of pancreatitis. Hemoconcentration, coagulopathy, leukocytosis, hyperglycemia, glucosuria, hypocalcemia, elevated gamma-glutamyl transpeptidase and hyperbilirubinemia may be present.

Differential diagnoses for chronic or recurrent pancreatitis in children are biliary colic, cholecystitis, gastroenteritis, and peptic ulcer.

Intussusception, volvulus, malrotation

Signs and symptoms of intussusception include abdominal pain, vomiting, pallor with a playful child in between attacks of pain. Pain is typically sudden in onset and recurrent paroxysmal colicky in nature. If the pain is prolonged then child may become lethargic and may go into a shock like state and have abdominal distention and bleeding per rectum. The classic triad of intussusception consists of abdominal pain, palpable sausage shaped mass and currant jelly stools.

Neonates in their first week of life usually present with bilious emesis whereas older infants may present with recurrent colicky abdominal pain. Among adolescent about 25% are asymptomatic and those who are symptomatic may have symptoms of acute intestinal obstruction, history of recurrent abdominal pain but less frequently diarrhea or emesis. Imaging may show gasless abdomen, or a double bubble sign. Upper gastrointestinal series may show bird's beak appearance of the duodenum. A superior mesenteric vein located to the left of superior mesenteric artery picked up by ultrasound is diagnostic of malrotation.

Volvulus may present with dull aching pain, bile stained emesis and rectal bleeding which is an indication of vascular webs. Duodenal obstruction, thickened bowel loops located to the right of spine with free peritoneal fluid are suggestive of volvulus.

Trauma

The likely sites of injury following blunt abdominal trauma are spleen, liver and kidney; injury to bowel, pancreas and lower genitourinary tract may occur.

Obstruction

Postoperative intestinal obstruction due to adhesion may be treated with initial nasogastric decompression followed by surgical intervention.

Chronic
Functional

Differential diagnosis of recurrent abdominal pain in an 11-year-old girl include peptic ulcer disease, carbohydrate intolerance, constipation and bowel dysmotility, irritable bowel syndrome , non-ulcer dyspepsia and psychological issues.

Chronic recurrent abdominal pain may present as a paroxysmal episodes, usually lasting less than 1hour and occur in clusters. Pain is nonradiating, dull, crampy or sharp in nature. Pain is not associated with activity, meals, stress, or bowel habits. Pain interferes with normal activities but resolve fully between episodes. Physical exam and laboratory test results are within normal limit.

Management of chronic recurrent pain should be directed by pertinent history, physical exam finding and laboratory tests. Parents should be reassured and child in concern should be followed closely. Patient is to be encouraged to continue normal

activities at home and school. In case of peptic ulcer disease non-steroidal anti- inflammatory drugs (NSAIDS) and salicylate (ASA) is contraindicated. Dietary fiber, anticholinergic and antispasmodic therapies are controversial.

The evaluation of a patient with chronic recurrent abdominal pain should start with taking a complete history and a comprehensive physical exam. History should include character of the pain and in the context in which the pain occurs, associated symptoms, patient's diet, sleep routine, and bowel habits. A good history will explore psychosocial aspects of patient's life. Common laboratory

investigations should be done with an aim to screen for occult disease such as inflammatory diseases, urinary tract infection, parasitic infection and carbohydrate intolerance. With this in mind a CBC, ESR, stool occult blood, urinalysis and urine culture need to be done. Other tests like stool ova and parasite and giardia, lactose breath test, and abdominal radiograph may be ordered based on the history and physical exam.

Irritable bowel syndrome

Key features of irritable bowel syndrome are abdominal pain relieved by stooling, increased frequency of bowel movements at the onset of the pain, along with bloating or abdominal distention, change in form of stool (hard or loose or watery), and passage of mucus. Management involves managing pain, diarrhea and constipation. Pain may be controlled by peppermints, antispasmodics and tricyclic antidepressant. Also, patients may be offered cognitive behavioral therapy and referral to pain clinic. Drugs for diarrhea are Loperamide, oral non absorbable antibiotics alosetron. Treatment for constipation includes fiber, psyllium, increasing fluid intake, lactulose, tegaserod and selective C2 chloride channel activating agents.

Acid-peptic disorder

Acid peptic disorder manifests in variety of ways depending on the age of the child. In neonates the presenting symptoms may be perforation. In infants and older children symptoms are vomiting, difficulty feeding, crying spells hematemesis or malena. Older children tend to have nocturnal awaking from pain. Common presenting symptoms in school aged children are epigastric pain and nausea. Older children are likely to complain of abdominal pain or fullness in epigastric area with dyspepsia.

Abdominal pain due to acid peptic disorder is dull or aching in nature and poorly localized and may be periumbilical. Duration of pain could be variable, lasting minutes to hours with frequent remissions and exacerbations.

Other

Patients with lactose intolerance get bloating, loose stools, and cramping abdominal pain when they ingest lactose containing food. It is reasonable to use lactase-treated milk products or a complete restriction of milk products for several weeks as a therapeutic trial.

Abdominal migraine is characterized by episodic vague pain located in the midline or periumbilical area that last for hours. Features of abdominal migraine overlap with those of migraine without aura and is seen in school aged children who have a history of chronic, recurrent abdominal pain.

Abdominal mass

Neonates may have abdominal mass due to hydronephrosis, multicystic kidney, renal vein thrombosis, mesoblastic nephroma, polycystic kidney disease, wilms tumor, rhabdoid tumor. Pelvic pathologies that cause mass are ovarian cyst, hydrocolpos and hydrometrocolpos. Gastrointestinal duplication can present with an abdominal mass in neonates. Infants and children may have abdominal masses from retroperitoneal neuroblastoma, Wilms' tumor and lymphoma. Hepatic causes of abdominal mass in this age group are hepatoblastoma and embryonal sarcoma. Gastrointestinal causes are duplication, Meckel's diverticulum and fecal mass. Ovarian cysts, teratomas and other omental or mesenteric cyst can present as abdominal mass in children.

Vomiting
Gastrointestinal and nongastrointestinal causes of vomiting

Differential diagnoses for vomiting in newborn are intestinal atresia, intestinal web, meconium ileus, hirschsprung disease, necrotizing enterocolitis and inborn errors of metabolism. From birth to 3 months, infants may present with vomiting due to pyloric stenosis, malrotation with midgut volvulus, inborn errors of metabolism, milk or soy protein allergy, gastroesophageal reflux and child abuse. In infants age 3 months to 12 months, the differentials for vomiting include gastroenteritis, intussusception, and child abuse. Older children with gastroenteritis, meningitis, elevated intracranial pressure, cyclic vomiting syndrome, or abdominal mass may present with vomiting. Other differentials for children presenting with emesis are migraine, superior mesenteric artery syndrome, and rumination.

Bilious emesis is an indication of congenital obstructive gastrointestinal malformations, like duodenal and jejunal atresias, malrotation with midgut volvulus, meconium ileus or plugs, and Hirschsprung disease. Nonsurgical causes of bilious emesis include necrotizing enterocolitis and gastroesophageal reflux.

Evaluation and management of a 3-week-old infant with projectile non-bilious vomiting- Likely diagnosis is pyloric stenosis. Evaluate for detail history, basic metabolic panel, imaging (ultrasound of abdomen, upper GI). Management includes making patient NPO, rehydration, electrolyte correction and surgical intervention (pylorotomy).

Selective 5-hydroxy tryptamine (5HT3) receptor antagonist block vagal afferent neurons that release serotonin and thus inhibit initiation of vomiting reflex.

Vomiting from infectious and noninfectious causes
may be a symptom of a systemic illness
Acute gastroenteritis is the most common cause of vomiting in children
Structural causes of vomiting

Clinical features of pyloric stenosis include nonbilious vomiting immediately after feeds with infant being eager to eat; vomiting is progressive and may or may not be projectile in nature; Other clinical findings are hypochloremic metabolic alkalosis, olive shaped mass in pyloric area and a peristaltic wave may be seen. Hyperbilirubinemia has high association rate with pyloic stenosis. Ultrasound will show pyloric channel length = > 15 – 19 mm, pyloric diameter = > 1 - 15 mm, pyloric muscle length – 19 – 21 mm, and pyloric muscular wall thickness = 73 – 5 mm. Contrast study may show string sign, shoulder sign and double tract sign.

Duodenal atresia results from failed recanalization of the intestinal lumen after 7th week of gestation. The extrinsic causes of duodenal atresia are annular pancreas, preduodenal portal vein, duplication cysts, and congenital bands with malrotation.

Evaluation and management of a 2-year-old child with the acute onset of vomiting with obstruction include symptomatic care and surgical relief of the obstruction.

Management of an infant with duodenal atresia starts with nasogastric decompression and intravenous fluid replacement. Surgical intervention is deodenoduodenostomy. Associated anomalies should be ruled out by doing echocardiography, renal ultrasound, and radiograph of chest and spine.

Disorders associated with chronic vomiting

Evaluation of cyclic vomiting syndrome (CVS) include excluding underlying gastrointestinal, neurologic, renal, metabolic, and endocrine causes. But currently there is no specific test for diagnosing cyclic vomiting syndrome (CVS). Evaluation includes thorough history taking and physical exam. In certain cases endoscopy, contrast GI imaging studies, brain MRI and metabolic tests may be performed. Metabolic tests may include lactate (lactic acid level), ammonia, amino acid and organic acid levels..

Esophageal disorders
Motility

Regurgitation is physiologic in a significant number of infants.

Rumination is effortless and passive regurgitation of gastric contents into mouth whereas regurgitation results from laxity of the distal esophageal sphincter and results in the return of gastric content into the esophagus.

Trauma
General

Signs and symptoms of esophageal perforation can include pain in neck, chest and abdomen, subcutaneous emphysema, dyspnea, and sore throat. Late diagnosis and treatment may lead to respiratory distress or shock. Plain radiograph of chest and soft tissue of neck may show subcutaneous emphysema, pneumomediastinum, and mediastinal widening.

Caustic ingestion

Corrosive agents like alkali may cause esophageal burns in the absence of mouth burns.

Foreign body

Symptoms of an esophageal foreign body are refusal to take oral feedings, excessive salivation, drooling, emesis, odynophagia, gagging, sensation of foreign body, and pain radiating to sternum or back.

Treatment of an esophageal foreign body is prompt removal by urgent endoscopic method, especially if the object is sharp, disc button battery or causing respiratory symptoms. In case of meat impaction with patient handling secretion nicely, removal may be delayed up to 12 hours and if the foreign body is blunt or a coin then removal may await up to 24 hours. Alternate or adjunct therapeutic options following coin ingestion are not recommended. Drinking carbonated beverages to dilate the esophagus and facilitate passage of esophageal coins (previously studied in adult subjects), parenteral administration of glucagen, treatment with prokinetic agents to enhance gastrointestinal motility, and the use of cathartics have not proved to be effective.

Gastroesophageal reflux

Symptoms of complications of gastroesophageal reflux are poor growth, pain and irritability from esophagitis, anemia, and dystonic movements (Sandifer syndrome).

Evaluation starts with careful history taking and physical exam to rule out other causes of chronic vomiting. Contrast imaging is done to rule out differentials of vomiting. In order to confirm acidic

reflux episodes, pH monitoring may be done, but impedance study is useful for non acidic reflux. Endoscopic evaluation could be done to identify esophagitis and treat reflux induced strictures. Laryngotracheobronchoscopy is used to evaluate signs of extraesophageal GERD. In older children proton pump inhibitor may be used for a time limited trial to make diagnosis.

Gastroesophageal reflux management may include conservative drug therapy using histamine – 2 receptor blocker, proton pump inhibitor or surgical intervention, such as fundoplication. Conservative management focuses on feeding technique, thickening of milk, and positioning for infants. In older children acidic and reflux inducing foods and beverages should be avoided.

Most children with uncomplicated GER usually outgrow it between ages 7 months to 1 year. The likelihood of developing GERD or its complications are high among children with comorbid conditions such as obesity, neurologic impairment, interstitial lung disease, anatomic gastrointestinal abnormalities, malrotation, hiatal hernia, and prematurity.

Children with gastroesophageal reflux may have respiratory symptoms such as hoarseness or chronic cough. Laryngoscopic findings may show upper airway edema, erythema, cobblestoning, and granulomas.

Diarrhea
Diarrhea caused by infectious mechanisms

Common etiologic agents of infectious diarrhea in children are E. coli, v. cholerae, salmonella, shigella, clostridium difficile, clostridium perfringens, staph aureus, aeromonas hydrophila, vibrio parahaemolyticus, bacillus cereus, yersinia enterocolitica, giardia lamblia, entameba histolytica, rotavirus, Norwalk agent, cryptosporidium.

Enteropathogenic Escherichia coli infection presents with acute watery diarrhea without dysentery.

Campylobacter diarrhea presents with fever, severe abdominal pain which may be located in the right lower quadrant, diarrhea with or without blood or mucous.

Salmonella frequently causes bacteremia and is common in immunocompromised individuals. Shigella infection is well responsive to antimicrobial therapy.

Cryptosporidium can be a cause of chronic diarrhea in a nonimmunocompromised host.

Pseudomembranous colitis may be a complication of antibiotic therapy.

Clinical manifestations of Giardia lamblia (giardiasis) are greasy or foul smelling stool reflecting the malabsorptive nature of the diarrhea although most cases are asymptomatic with chronic infections causing failure to thrive and impaired cognitive function. Acute infection presents with watery diarrhea with abdominal cramping, nausea, vomiting, flatulence and weight loss.

Treatment of Escherichia coli diarrhea is mainly supportive care with rehydration and correction of electrolyte abnormalities.

Clinical signs - 24 - 48 hours of non-bloody diarrhea that is followed by bloody diarrhea. There is a self-limited fever, intermittent spasmodic abdominal pain, headache, myalgias, vomiting and irritability before onset of diarrhea). Laboratory findings associated with Escherichia coli 0157:H7 infection are fecal leukocytes, microangiopathic hemolytic anemia with schistocytes or helmet cells on blood smear, thrombocytopenia and high serum urea nitrogen and serumcreatinine level reflecting renal dysfunction.

Antidiarrheal medications are not recommended for children.

Diarrhea caused by noninfectious mechanisms

Secondary lactose intolerance occurs due to small bowel mucosal damage and resolves with mucosal healing. Congenital lactase deficiency is a rare condition. Primary adult type hypolactasia is a physiologic process where there is decrease production of lactase with increase in age. Treatment is milk free diet. Infants can have lactose free formula. Older children can have their calcium need met by taking yogurt, cheese and low lactose milk.

Incidence of lactase deficiency may occur as early as 2 years in Thai population, between ages 3 to 5 years in American Indians and African Americans and after 5 years in whites.

Sucrase isomaltase deficiency in different ethnic groups - sucrose isomaltase deficiency occurs in about 0.2% of North Americans.

Milk-protein intolerance- clinical presentation of milk protein allergy is variable. Gastrointestinal symptoms are diarrhea, emesis, edema of lips and pharynx.

Dermatologic symptoms are atopic dermatitis, urticarial rash. Respiratory symptoms are rhinitis, otitis media, chronic cough; wheezing and even hemosiderosis. These children can also have occult blood in stool and iron deficiency anemia. Diagnosis is made based on age of onset, good history, physical exam and elimination and oral challenge. Treatment is avoidance of cow's milk.

Allergens in the mother's diet may cause colitis in a beast-fed infant.

Differential diagnoses of noninfectious intractable diarrhea in infancy are disaccharidase deficiency, protein intolerance, enteric infection and malabsorption.

Chronic nonspecific diarrhea

Diagnosis is made with good history, physical exam and minimum laboratory investigations. Usual presentation of chronic nonspecific diarrhea is between age 12 to 30 months in a toddler who has frequent, large, watery stools in absence of physical or laboratory findings of malabsorption or infections and has normal growth and development. Toddler's diarrhea has excellent prognosis as it usually spontaneously resolves between ages 2 years and 4 years. There is no association with any gastrointestinal disease later in life and no effect on growth unless hypo caloric elimination diet is used.

Poor growth, fever, and melena are incompatible with the diagnosis of chronic nonspecific diarrhea.

Extremely low fat diets, sorbitol, fruit juices, and excessive water consumption may produce chronic nonspecific diarrhea.

Protracted diarrhea

Malnutrition, chronic infection, systemic disease, and immunodeficiency are predisposing factors to the development of diarrhea.

Infants with protracted diarrhea require a thorough history and careful physical examination with serial weights followed by laboratory investigation. Stool studies should include reducing substance, guaiac, pH, fat and culture. Other tests include CBC with differentials, urinalysis, ESR,

basic metabolic panel, albumin, total protein and liver function test. Serial stool exams, D-Xylose test, lactose tolerance test or breath hydrogen (H2) test, sweat chloride, immunoglobulins.

Imaging should include upper gastrointestinal studies, barium enema, sigmoidoscopy with small bowel biopsy and nutritional monitoring.

Parenteral nutrition is used to provide nutrition and calories in treating protracted diarrhea.

Constipation

In case of long segment Hirschsprung disease where there is extensive involvement of the small bowel, patients have to be on prolong period of parenteral nutrition and may have anastomotic stricture from reconstructive surgery. These patients can have recurrent obstruction and explosive diarrhea. While 90% patients with Hirschprung after surgery enjoy normal life, 10% of patients may have constipation and incontinence. Rare complications are urinary incontinence and impotence.

In constipation onset is usually after age 2 years, encopresis is a common finding with history of forced bowel training. Whereas the history of Hirschprung is distinct as symptoms begin in newborn period patients may have failure to thrive and/or enterocolitis. In hirschsprung common physical findings are abdominal distention, poor weight gain and rectal exam reveals empty rectum explosive passage of stool. In functional constipation rectum is filled with stool.

In neonates Hirschspung disease usually presents with distended abdomen, failure to pass meconium, feeding intolerance and or bilious emesis or aspirates. Infants with hirschsprung disease have delayed passage of stool or chronic constipation. Undiagnosed children may present with failure to thrive with hypoprotinemia, enterocolitis with associated diarrhea, abdominal distension, tenderness, and sepsis.

Children with fecal overflow incontinence tend to show sign of stool holding behavior. These children tend to contract their pelvic and gluteal muscles by standing upright supporting on furniture, holding their legs firmly together and sometimes lie on the floor and “stiffen up”.

Laxatives helps to promote bowel movements, stool softeners and osmotic agents increase the fluid content of the stool, and lubricants (e.g. mineral oil) help to ease passage.

Differential diagnosis for constipation in a young child could be approached in broad categories – Functional: lack of dietary fiber, excessive calcium intake, dehydration, malnutrition, Structural: anorectal malformations like ectopic anus, anal stenosis, imperforate anus; colonic stricture, meningocele, teratoma, Endocrine: hypothyroidism, hyperparathyroidism, cystic fibrosis, Metabolic : hypercalcemia, hypokalemia, uremia Neurologic: cerebral palsy, hypotonia, spina bifida, spinal tumor, tethered cord Neuromuscular: hirschsprung disease, infant botulism Connective tissue disorder scleoderma, SLE Drugs antacids, anticholenergics, tricyclic antidepressants, opiates, phenobarb, sympathomimetics.

Rectal suction biopsy is the confirmatory test for diagnosis of Hirschprung disease which shows large number of nerve bundles with absent ganglionic cells.

Jaundice
Neonatal and infancy
Bilirubin metabolism

Diagnostic studies are needed to detect hemolytic diseases in a full term infant that become clinically jaundiced during the first day after birth.

Diagnostic tests to establish the cause of unconjugated hyperbilirubinemia-blood types and a screen for unusual isoimmune antibodies should be checked in all pregnant women. If mother is Rh-negative, the infant's cord blood should be evaluated for a direct antibody (Coombs) test, blood type, and Rh determination. A complete blood count with smear and direct bilirubin concentration, haptoglobin, hemoglobin electrophoresis, red cell enzyme assay and test for spherocyte should be done.

Evaluation of a 2-day-old infant with jaundice - blood types and a screen for unusual isoimmune antibodies should be checked in all pregnant women. If mother is Rh- negative, the infant's cord blood should be evaluated for a direct antibody (Coombs) test, blood type, and Rh determination. A complete blood count with smear and direct bilirubin concentration, haptoglobin, hemoglobin electrophoresis, red cell enzyme assay and test for spherocyte should be done.

Bilirubin metabolism is increased due to erythrocyte turnover and decreased intracellular metabolism and excretion in the newborn infant.

Gilbert syndrome is an autosomal recessive disease causing unconjugated hyperbilirubinemia and a modest rise in bilirubin level. Jaundice is appreciated when patient is under oxielative stress, fasting or has an infection. Patient is otherwise healthy with normal liver function.

Breast-milk jaundice

Breast-feeding is the most common cause of exaggerated unconjugated hyperbilirubinemia in the neonatal period.

Breast-feeding does not cause conjugated hyperbilirubinemia.

Management of the infant with breast-milk jaundice in an infant with bilirubin level 20mg/dl is temporary interruption of breastfeeding and phototherapy. In full term infants with bili level 12-17mg/dl mother should increase breast

feeding to 8-12 times per day or supplement with formula and remeasure bilirubin in 24 hours.

Sepsis, galactosemia, and endocrine disorders can be easily diagnosed in the neonate with conjugated hyperbilirubinemia.

Infectious and noninfectious causes of jaundice

Cholecystitis in children manifests as jaundice, periumbilical or right upper quadrant abdominal pain, vomiting, and fever.

Biliary Atresia: Signs and symptoms - Persistence of jaundice or the presentation of jaundice after the tenth postnatal day, and acholic stools. There may be weight loss and change in appetite. If discovered later, a firm enlarged liver may be palpable with or without splenomegaly. Laboratory test will show conjugated hyperbilirubinemia.

Diagnostic tests for biliary atresia are fractionated bilirubin, aspartate aminotransferase and alanine aminotransferase, gamma glutamyl transpeptidase, serum albumin, prothrombin time, viral and bacterial cultures, viral serology (hepatitis B surface antigen, TORCH, human immunodeficiency virus) VDRL titers, alpha-1-antitrypsin and phenotype and level, thyroxine,

thyroid-stimulating hormone, sweat chloride, metabolic screen (urine and serum amino acids, urine reducing substances, and succinyl acetonetyrosinemia); ferritin, ultrasonography, nuclear medicine imaging (DISIDA or other hepatobiliary scan), operative cholangiography and exploration with liver biopsy.

Management of biliary atresia isoperative intervention, kasai portoenterostomy, followed by orthotpic liver tansplant.

Metabolic diseases that can lead to conjugated hyperbilirubinemia in the neonatal period are Wilson disease, alpha 1 antitrypsin deficiency, and cystic fibrosis.

Signs and symptoms of a choledochal cyst are epigastric pain, fever, and jaundice in a patient of any age.

Differential diagnoses of infectious causes of jaundice in an infant are hepatitis viruses, Epstein Barr virus, cytomagalovirus and herpes simplex virus.

Management of a patient who has obstructive jaundice - fulfill caloric need, medium chain fatty acids are useful to provide additional calories, give nasogastric feeds if anorexia is present, provide oral fat soluble vitamins, if indicated ursodeoxycholic acid may be used to reduce cholestasis, protect

hepatocytes and increase bile flow, antihistamine or rifampin for pruritus. Surgical intervention depending on cause of obstruction.

Children and adolescents

A.Infectious and noninfectious causes of jaundice

Evaluation of a child with conjugated hyperbilirubinemia - History, physical examination and laboratory work which includes measuring of serum aminotransferases, GGT, and bilirubin (including conjugated, or direct bilirubin), as well as performing tests of liver synthetic function (prothrombin time and serum albumin level). Ultrasound of abdomen to revaluate the biliary tree should be done.

Laboratory evaluation of hepatitis - serologic markers for hepatitis A, B, and C virus, serum AST and ALT levels, serum conjugated bilirubin level, identifying other organism that may cause hepatitis - Epstein Barr Virus (EBV), cytomegalovirus (CMV), adeno virus.

Immediate and long-term complications of hepatitis-

Signs and symptoms of Wilson disease- The younger the child the higher likelihood of hepatic involvement. Asymptomatic hepatomegaly with or without splenomegaly, subacute or chronic hepatitis, hepatic failure with or without hemolytic anemia, cryptogenic cirrhosis, portal hypertension, ascites, edema, variceal bleeding, or other effects of hepatic dysfunction (coagulation defect, delayed puberty, amenorrhea).

In young children symptoms of acute hepatitis are similar to gastroenteritis and could be anicteric. In older children and adolescents there is acute onset of febrile illness, anorexia, nausea, vomiting, malaise and jaundice. There could be regional lymph node enlargement, splenomegaly, ulceration in the GI tract, bone marrow hypoplasia and aplastic anemia. Rarely there is pancreatitis, myocarditis, nephritis, arthritis, vasculitis and cryoglobulinemia.

Signs and symptoms of chronic hepatitis are elevated serum aminotransferase levels , progressive hepatic fibrosis, cirrhosis, hepatic failure. With hepatitis C there may be primary hepatocellular carcinoma, and extrahepatic manifestations are small vessel vasculitis, mixed cryoglobinuria, peripheral neuropathy, cerbritis, membranoproliferative glomerulonephritis and nephrotic syndrome with antibodies to smooth muscle, ANA and low thyroid hormone levels.

Signs and symptoms of liver disease due to alpha-1 antitrypsin deficiency

Jaundice, acholic stools and hepatomegaly are present in 1st week of life with jaundice clearing at 2nd to 4th month. Infants with jaundice have similar presentation as infants with idiopathic neonatal hepatitis. Older children may

have asymptomatic hepatomegaly, chronic liver disease or cirrhosis with portal hypertension.

B.Obstructive jaundice

Signs and symptoms of cholelithiasis and choledocholithiasis -Children are mostly asymptomatic, with incidental cholelithiasis diagnosis. . Infants may present with obstructive symptoms, such as irritability, cholestatic jaundice, and acholic stools. In children presentation include intermittent abdominal pain of variable intensity. Diagnosis is made by ultrasonography of the liver and gallbladder. Liver function tests usually are within normal range.

astrointestinal bleeding
Upper versus lower gastrointestinal bleeding

Esophageal varices may initially present with upper gastrointestinal bleeding. Evaluate a patient with upper gastrointestinal bleeding.

Plan the appropriate evaluation for a patient who has blood in vomitus or stool- upper gastrointestinal bleeding (esophagus, stomach, duodenum)is investigated with esophagogastroduodenoscopy and lower bleeding is evaluated with colonoscopy. A tagged red blood cell scan can be used in case of small intestinal bleed with unknown location. A guaiac test may be done to detect occult bleeding.

Importance of alcohol-induced gastritis in adolescents provides the opportunity to identify use of alcohol which leads to interventions to decrease the morbidity and mortality associated with alcohol use and abuse. Adolescents should be asked about alcohol use and counseling on its dangers especially drinking and driving must be provided.

Rectal bleeding and malena may be seen in neonatal period due to swallowed blood. Most common causes of rectal bleeding in infancy are allergic colitis and anorectal fissures. Vascular malformations, intestinal duplication, intussusception, necrotizing enterocolitis or Hirschsprung disease also may present with rectal bleeding. Among young children, common etiologies of rectal bleeding are anorectal fissures, infectious gastroenteritis, and intussusception. Inflammatory bowel diseases, e.g., Crohn disease and ulcerative colitis, vasculitic diseases such as Henoch-Schonlein Purpura (HSP) and hemolytic uremic syndrome (HUS).

Differential diagnosis of vomiting bright red blood include swallowed blood (epistaxis, sore throat, or breastfeeding or may follow dental work or tonsillectomy), upper GI

mucosal lesions (esophagitis, Mallory-Weiss tear, reactive gastritis, stress ulcer, and peptic ulcer), variceal bleeding, or rarely, hemobilia.

Etiologies of occult blood include esophagitis, reactive gastritis, acid peptic disease, eosinophilic gastroenteritis, colitis, celiac disease, inflammatory bowel disease, polyposis, Meckel's diverticulum, vascular malformation. Bright red blood per the rectum could be due to anal fissure, streptococcal cryptitis, ulcerative proctitis, rectal prolapse, solitary rectal ulcer, and internal hemorrhoids.

It is important to do an anal examination in the evaluation of rectal bleeding in order to rule out anal fissure, juvenile polyp, painless bleed in Meckel's diverticulum and perianal streptococcal cellulitis.

Vomiting coffee-ground material is usually due to localized gastritis which is usually associated with nonsteroidal anti-inflammatory drugs (NSAID gastropathy), alcoholic gastritis, cocaine ingestion, ingestion of caustic substances, Helicobacter pylori infection, viral infection, Crohn disease, vasculitis (Henoch-Schonlein Purpura), radiation exposure, bile reflux, bezoar, hiatal hernia, prolapse of the gastroesophageal junction, or congestive gastropathy (associated with portal hypertension). Reactive gastritis may coexist with duodenal erosive lesions.

Vomitus that tests positive for occult blood may be due to swallowing maternal blood during birth, from fissures in the nipple during nursing.

Nasogastric tube is helpful in assessing the source and extent of bleeding but cannot clearly identify upper or lower gastrointestinal origin. It is also helpful for gastric lavage, endoscopic evaluation, prevent hyperammonemia in liver disease and to stop further bleeding.

A child with melena and hemodynamically significant blood loss merits urgent volume resuscitation and prompt diagnostic evaluation. Management should start with careful physical examination and vital signs monitoring.

Polyps

A solitary juvenile polyp is not associated with adenocarcinoma of the colon.

Familial polyposis is an inherited polyposis syndrome that carries a risk of colon cancer. Patients usually become symptomatic after 10 years of age; multiple adenomatous polyps are seen in colon and rectum with rarely any involvement of gastric and small intestine.

Signs and symptoms of juvenile polyposis- Usually cause blood streaking on the outside of the stool and rarely causes massive rectal bleeding. Polyps are present throughout the colon where individual polyps have adenomatous and hamartomatous elements.

Meckel diverticulum

Signs and symptoms of Meckel diverticulum- intermittent painless rectal bleeding, brick red or currant jelly color or melanotic stool, anemia, hypovolemia and partial or complete bowel obstruction.

Meckel diverticulum- treatment is surgical excision.

Ulcer disease

Evaluation of a child with suspected ulcer disease includes taking focused history, careful physical examination and confirmation by endoscopy.

H2 receptor antagonists inhibit the binding of histamine at the H2 subtype receptor of the gastric parietal cell. Proton pump inhibits the H+/K+ ATPase pump in a dose dependent fashion thus reducing basal and stimulated gastric acid secretion.

Gastritis is a clinical manifestation of Helicobacter pylori infection

Risk factors for ulcer disease in childhood- stress (shock, respiratory distress, life threatening illness, hypoglycemia, dehydration, burns, trauma, vasculitis, renal failure, intracranial lesions), use of non-steroidal-anti-inflammatory-medications.

Methods for diagnosing Helicobacter pylori infection- biopsy of the greater curvature of antrum of the stomach, culture of antral or duodenal tissue, breath test.

Recognize the symptoms of dyspepsia in a child with recurrent abdominal pain

Hepatomegaly

Simultaneous splenomegaly and hepatomegaly in neonate may indicate TORCH infections, sepsis, metabolic disease, a child older than 1 year who has both splenomegaly and hepatomegaly may have leukemia, lymphoma, congestive heart failure portal hypertension or parasitic infections.

Hepatomegaly in a 1-month-old infant is suspicious of cystic fibrosis and alpha-1-ntitrypsin deficiency.

A liver edge greater than 3.5 cm in newborns and greater than 2 cm in children below the right costal margin indicates hepatomegaly; the normal range for liver span by percussion at 1 week of age is 4.5 to 5 cm. At 12 years of age the normal liver span for boys is 7 to 8 cm and 6 to 6.5 cm in girls.

Evaluation of hepatomegaly include checking excretory and synthetic function of liver by investigating AST, ALT, bilirubin, alkalaine phosphate, albumin and prothrombin time; performing liver biopsy, imaging studies like abdominal radiograph, ultrasound, CT or MRI, cholangiography, radionucleotide scan, ERCP.

Signs, symptoms of portal hypertension are hepatomegaly, splenomegaly, ascites or a prominent abdominal venous pattern.

alabsorption
General

Appropriate laboratory work for malabsorption - Stool culture and antibody tests for parasites, stool microscopy for ova and parasites and stool occult blood to exclude inflammatory disorders. Stool pH and reducing substances for carbohydrate malabsorption, and quantitative stool fat and alpha-1-antitrypsin to show fat and protein malabsorption should be ordered. Fecal stool elastase to rule out exocrine pancreatic insufficiency should be ordered. A complete blood count with peripheral smear should be ordered to check for anemia, lymphopenia (lymphangiectasia), neutropenia (Shwachman syndrome) and acanthosis (abetalipoproteinemia) are useful tests. Serum IgA and tissue transglutaminase antibody levels can be done if celiac disease is suspected. Further laboratory work may be pursued depending on initial laboratory finding.

Differential diagnosis for malabsorption at various ages - In infants, malabsorption occurs due to congenital disorders of nutrient digestion or transport. In older children the reason for malabsorption is mostly due to congenital or acquired mucosal villous disorders.

Clinical conditions associated with a rectal prolapse-intestinal parasites, malnutrition, diarrhea, ulcerative colitis, pertussis, Ehlers-Danlos syndrome, meningocele, cystic fibrosis and chronic constipation.

Mucosal disease (celiac disease)

Diagnosis of celiac disease depends on characteristic small intestinal histopathology findings and response to a gluten-free diet.

Clinical manifestations of celiac disease are: failure to thrive, chronic diarrhea, vomiting, and abdominal distention, muscle wasting, anorexia, and irritability are seen among children diagnosed within 24 months of life. In older children extra intestinal manifestations (iron deficiency anemia, osteoporosis, short statue, endocrinopathies, arthritis and arthralgias, epilepsy with bilateral occipital calcifications, peripheral neuropathies, cardiomyopathy, chronic lung disease, isolated hypertransaminasemia,

dental enamel hypoplasia, aphthous stomatitis and alopecia) are common even in absence of digestive symptoms.

In cystic fibrosis malabsorption occurs due to pancreatic insufficiency and inability to absorb fat and protein. In celiac disease there is malabsorption due to gliadin induced villous injury.

Foods in which gluten can be found are wheat, barley and rye.

Pancreatic insufficiency present in cystic fibrosis and Shwachman syndrome.

Routine use of supplemental pancreatic enzyme preparations prior to meals allow for normal or near-normal absorption of dietary fat and protein in most patients who have pancreatic insufficiency.

Age-related gastrointestinal signs and symptoms of cystic fibrosis - Pancreatic insufficiency is common in both neonatal period and early childhood. Newborns present with meconium ileus, intestinal obstruction and visceral perforation;.adolescent: cholelithiasis without history of hemolytic anemia, or hepatic disease.

Hepatobiliary diseases like hepatic cirrhosis and gallbladder disease with or without cholelithiasis may result from cystic fibrosis as thick secretions cause ductal obstruction.

Shwachman-Diamond syndrome is a familial disorder that is the second most common cause of pancreatic insufficiency. Symptoms include diarrhea, failure to thrive, bone marrow hypoplasia, neutropenia and metaphyseal dysostosis. Anemia, thrombocytopenia and intermittent galactosuria may be seen.

Chemotherapeutic agents can cause alteration of the mucosal barrier and cause diarrhea.

Symptoms of malabsorption as a result of pancreatic insufficiency are malodorous stool, abdominal distension and growth failure.

Enzyme deficiency (lactase, sucrase-isomaltase)

Symptoms of a carbohydrate malabsorption disorder are loose watery diarrhea, flatulence, abdominal distention and pain.

Short-gut syndrome, including bacterial overgrowth

Clinical situations where gastric pH or small bowel motility is altered can give rise to bacterial overgrowth. Partial bowel obstruction, diverticula, short bowel, intestinal duplications, diabetes mellitus, idiopathic intestinal pseudo-obstruction syndrome,

scleroderma, prematurity, immunodeficiency, and malnutrition can lead to bacterial overgrowth.

Fat malabsorption and chronic liver disease (biliary atresia, cystic fibrosis)

Medium-chain triglyceride oil partially soluble in water and compared to long chain triglyceride MCT is hydrolyzed more effectively by lipase. So there is less reliance on bile acid micelle formation and much of it can bypass the lymphatics and be transported to the portal vein.

Inflammatory bowel disease (IBD)

Children with IBD often experience abdominal pain and diarrhea after eating resulting in decreased oral intake and less caloric consumption. Anorexia and growth hormone resistance may be caused by elevated level of proinflammatory cytokines. Decrease absorption in the small intestine results in protein losing enteropathy. Patients have fat malabsorption which causes lack of caloric intake and deficiency of fat soluble vitamins. The complications of IBD could further decrease nutrient absorption due to internal fistulae, surgical bowel resections, or diverting ostomies. Use of corticosteroids during treatment plays a role in growth failure too.

Clinical manifestations of Crohn disease are- diffuse crampy abdominal pain or pain located in right lower quadrant, diarrhea, and weight loss. Chronic perianal disease, including tags, fissures, fistulae, and abscesses, may be seen. Recurrent aphthous-stomatitis , poor appetite, fevers, and iron deficiency anemia, growth failure could be initial manifestations of Crohn disease. Most children with Crohn disease present with classic symptoms like diffuse crampy abdominal pain or pain located in right lower quadrant, diarrhea, and weight loss. Chronic perianal disease, including tags, fissures, fistulae, and abscesses, may be seen. Recurrent aphthous-stomatitis, poor appetite, fevers, and iron deficiency anemia, growth failure with could be all initial manifestations of Crohn disease. Endoscopic exam shows skip lesions with transmural involvement and granulomas not usually seen in UC.

Chronic perianal lesions can often be an early sign of Crohn disease

Children with Crohn disease may have growth failure due decreased caloric intake.

The main symptoms of ulcerative colitis (UC) are diarrhea, rectal bleeding, and abdominal pain. One third present with moderate symptoms, including hematochezia, abdominal cramping associated with defecation urgency, malaise, low grade or intermittent fevers, anorexia with weight loss, mild anemia, and hypoalbuminemia. Colitis, profound anemia and hypoalbuminemia are seen in only 10% of patients. Tachycardia, diffusely tender or

distended abdomen may be the initial presentation. Children who have UC may develop symptoms of reflux or dyspepsia associated with inflammation of the upper gastrointestinal tract. Endoscopic examination demonstrates crypt abscess and linear ulcerations which are uncommon findings in Crohn disease. Initial evaluation of a patient with suspected inflammatory bowel disease starts with

getting excellent history and physical exam. Laboratory includes getting stool studies to rule out infectious causes, complete blood count with differentials, ESR and albumin. Imaging studies may be done -upper GI, CT and MRI of abdomen. In case of abnormal laboratory finding and or suspicion for IBD Gastroenterologist must be consulted who conducts endoscopic evaluations and biopsy to confirm diagnosis.

Management of a patient with severe colitis (fever, hypoalbuminemia, and anemia)- Infliximab an immunemodulator (colitis chimeric monoclonal antibody directed against the cytokine TNF-alpha that acts by inducing apoptosis of active T lymphocytes) is the drug of choice for management of severe colitis. For moderate to severe colitis corticosteroids are used to inhibit inflammation.

Differential diagnoses of acute colitis in an adolescent with inflammatory bowel disease: pseudomembranous enterocolitis, lymphocytic colitis, eosinophilic enterocolitis, Henoch- Schonlein Purpura, and hemolytic-uremic syndrome. Non Hodgkin lymphoma, periodic fevers, intestinal tuberculosis, rheumatologic disorders like juvenile idiopathic arthritis, ankylosing spondylitis, and systemic lupus erythematosus should be considered.

Refeeding syndrome

Refeeding syndrome may present with weakness, acute rhabdomyolysis, neutrophil dysfunction, cardiorespiratory failure, arrhythmia, seizure, altered level of consciousness, acute heart failure, or sudden death. Laboratory abnormalities in refeeding syndrome are low serum phosphorus, magnesium and low potassium.

Respiratory Disorders

General signs and symptoms
Stridor

The differential diagnosis of congenital stridor includes: laryngomalacia, papillomas, tumors, cysts, laryngeal webs, bilateral abductor paralysis of the vocal cords, tracheomalacia, subglottic tracheal webs, endotracheal tumors, endobronchial tumors, subglottic tracheal stenosis, mediastinal masses, vascular ring, lobar emphysema, bronchogenic cyst, thyroid enlargement, macroglossia, Pierre Robin syndrome, cri-du-chat syndrome.

The etiology of stridor: congenital versus acquired, presents in children of different ages.

Endoscopy is the diagnostic tool of choice for laryngeal and vocal cord disorders.

The appropriate approach to the evaluation of congenital stridor includes an evaluation of the history including: previous intubation, and congenital cardiac disease. Other symptoms such as difficulty feeding, gastroesophageal reflux disease (GERD), and changes in sound of stridor with position may help to localize the lesion. Physical examination may reveal findings such as dysmorphic features and hemangiomas. Neck and Chest radiographs may be obtained. Bronchoscopy should be performed.

Know that vocal cord dysfunction may mimic asthma.

Respiratory failure

Respiratory failure is an inability of the respiratory system to fulfill the gas exchange needs of the body. It is traditionally defined by abnormalities in PO2 and PCO2.

The manifestations of chronic hypoxemia include: polycythemia, pulmonary hypertension, and cor pulmonale.

The clinical manifestations of acute hypercapnia include: flushing, agitation, confusion, tachycardia, and headache.

The combination of arterial blood gas values that indicate chronic carbon dioxide retention are: increased PCO2, normal pH, increased serum bicarbonate concentration, and increased base excess.

Children with chronic respiratory failure benefit from oxygen therapy. However, inspired oxygen at high concentrations may cause pulmonary capillary and epithelial cell injury.

Oxygen therapy should be provided to any patient who is hypoxemic or at risk for acute respiratory failure even in the initial absence of evidence of hypoxemia. Patients at risk for respiratory failure should be tried on non-invasive ventilatory support. Patients with impending respiratory failure should be intubated and started on mechanical ventilation. This is often indicated by worsening hypoxemia and hypercarbia; however dysfunction in gas exchange may also be due to other causes such as cardiovascular shock when decreased blood flow can cause a secondary respiratory failure.

Cough

Differential diagnosis of chronic cough in children of different ages:

Inflammatory: athsma.
Chronic pulmonary disease: BPD, chronic lung disease, post infectious bronchiectasis, cystic fibrosis, tracheomalacia, bronchomalacia, ciliary abnormalities.
Chronic and congenital disease: laryngeal cleft, swallowing disorders, gastroesophageal (GE) reflux, airway compression (vascular ring etc.), congenital heart disease.
Infectious and Immune: immunodeficiency, TB, allergies, sinusitis, tonsillitis, adenoiditis, Chamydia and Ureoplasma,Bordetella pertussis, Mycoplasma pneumonia.
Acquired: Foreign body aspiration

The clinical manifestations of psychogenic cough versus those of cough caused by organic etiology are:

Psychogenic cough often begins with an upper respiratory infection (URI), then the cough lasts several weeks, is refractory to treatment, and disappears with sleep or distraction. The cough is typically loud and abrupt with a barking quality. The child is often quiet when the physician stands outside the examination room and the cough immediately begins when attention is given to the child upon entry to the examination room.

Cough is a major manifestation of asthma and may be the only symptom.

There are limited indications for cough suppressants.

The initial screening evaluation of a chronic cough should include: chest radiograph, a sweat chloride test, tuberculin skin test, and pulmonary function tests.

Conditions which can impair the effectiveness of cough include: cerebral palsy, muscle weakness, vocal cord dysfunction, central nervous system (CNS) disease, thoracic deformities, and pain.

Exercise intolerance

Know that exercise intolerance may reflect etiologies other than pulmonary disease: anemia, muscle weakness, deconditioning, cardiac disease, and psychogenic causes.

Know that exercise intolerance may be a presenting symptom of chronic lung diseases such as asthma, interstitial lung disease or vocal cord dysfunction.

Apnea

Apnea – pause in breathing. Serious apnea is a pause in breathing for greater than 20 second.

Periodic breathing – intermittent cessation of breathing lasting 5-10sec followed by rapid respiration for 10-15 sec.

Central apnea – Apnea occurring because of decreased CNS stimuli, frequently due to immaturity of the brainstem respiratory center.

Obstructive apnea – Apnea occurring as a result of airway obstruction including: pharyngeal instability, neck flexion, and nasal occlusion. It is characterized by absent airflow with chest wall motion.

Polysomnography is used to evaluate the presence and degree of obstructive apnea in older children. Other helpful tests include chest radiograph, lateral neck radiograph, bronchoscopy, complete blood count, EKG, Echocardiogram.

The differential diagnosis of central apnea in infancy includes: immaturity of respiratory center, sleep state, intraventricular hemorrhage, drugs, seizures, hypoxic injury, herniation, neuromuscular disorders, Leigh syndrome, brainstem infarction, brainstem anomalies, general anesthesia, sepsis, necrotizing enterocolitis, meningitis, hypoglycemia, hypocalcemia, hyponatremia, hypernatremia, hyperammonimia, increased organic acids, hyperthermia, hypothermia, hypotension, hypertension, heart failure, anemia, hypovolemia, vagal tone.

Caffeine is the treatment of idiopathic recurrent apnea in premature infants. There is an association between apnea and anemia in premature infants.

Wheezing

The differential diagnosis of recurrent wheezing includes: viral infection, bacterial infection, fungal infection, papillomatosis, asthma or reactive airway disease, anatomic abnormalities, immunodeficiency, mucocilliary clearance disorders, bronchopulmonary dysplasia, aspiration syndromes, interstitial lung disease, heart failure, anaphylaxis, inhalation injury.

Consider a foreign body in the differential diagnosis of wheezing. 7.Tachypnea

Respiratory rates vary with age, with normal variations occurring with sleep, eating, and activity.

Tachypnea is a sensitive indicator of respiratory disease.

Hemoptysis

The differential diagnosis of hemoptysis in children includes:

Focal hemorrhage: bronchitis, bronchiectasis, infection, tuberculosis, trauma, pulmonary arteriovenous malformation, foreign body, neoplasm, pulmonary embolism.

Diffuse hemorrhage: idiopathic, congenital heart disease, prematurity, cow's milk sensitivity, Goodpasture's syndrome, collagen vascular disease, Hepoch-Schonein Purpura (HSP)vasculitic disorders, Granulomatous disease, Celiac disease, coagulopathy, malignancy, immunodeficiency, exogenous toxins, pulmonary hemosiderosis, tuberous sclerosis, lymphangiomyomatosis, physical injury and abuse.

The initial management of hemoptysis in children and adolescents includes volume resuscitation and transfusion of blood products if needed. Adequate ventilation and circulation should be maintained. Broncoscopy may be used to remove foreign debris or apply topical vasoconstrictive agents. Treatment consists of addressing the cause of the hemoptysis.

Cyanosis

Cyanosis is not a sensitive indicator of oxyhemoglobin desaturation.

Know the common extrapulmonary causes of cyanosis especially right-to-left shunt and methemoglobinemia.

Know how to validate and quantitate a clinical observation of cyanosis: arterial blood gases, oxyhemoglobin saturation.

Clubbing
Disorders commonly associated with digital clubbing include: chronic pulmonary disease, cyanotic congenital heart disease, subacute bacterial endocarditis, chronic congestive heart failure, and thalassemia.
Other causes include: congenital methmoglobinemia, Crohn disease, ulcerative colitis, chronic dysentery and sprue, polyposis coli, GI hemorrhage, small bowel lymphoma, liver cirrhosis, thyroid deficiency, chronic pyelonephritis, toxic, lymphomatoid granulomatosis, Fabry disease, Raynaud disease and scleroderma, vascular disorders, shoulder subluxation, median nerve injury, and local trauma.
Upper airway
General

Upper respiratory tract infection and airway obstruction in young infants lead to respiratory distress.

Croup
Viral croup is always associated with a viral infection of the respiratory tract.
Noninfectious croup is also called spasmodic or recurrent croup. It usually occurs in children between 1and 3 years of age and is thought to have an allergic component. There is no prodrome, no fever and usually resolves quickly without treatment.

Airway management is the mainstay of treatment for croup. Nebulized epinephrine has been used to temporarily relieve symptoms of moderate to severe croup. Oral corticosteroids are well established as a treatment for croup. There is no evidence that cool mist therapy is an effective treatment for croup.

Clinical manifestations of laryngotracheobronchitis (croup) are a "barking" cough, hoarseness, and inspiratory stridor preceded by URI symptoms of rhinorrhea, pharyngitis, mild cough, and fever for 1 to 3 days. Symptoms are usually worse at night and recur with total resolution within a week.

Epiglottitis

Epiglottitis is a medical emergency and patients require immediate artificial airway management. Parentral antibiotic treatment should be initiated after obtaining cultures from blood, and epiglottic surfaces.

Viral croup: characterized by URI symptoms, "barky" cough, hoarseness.

Radiograph shows subglottic narrowing.

Epiglottitis: characterized by high fever, pharyngitis, dyspnea, and rapidly progressing respiratory obstruction. The patient appears toxic, may be in the

tripod position, and may be unable to handle secretions. Laryngoscopy shows a "cherry red" epiglottis. Lateral radiograph of the neck shows the "thumb sign." Intubation is required.

Bacterial tracheitis: characterized by brassy cough, fever, toxic appearance, and respiratory distress. Purulent material can cause pseudomembranes. Radiograph classically shows pseudomembrane detatchment in the trachea. Intubation is often required. Treatment with antibiotics is indicated.

Patients with suspected epiglottitis are at risk from death due to complete airway obstruction. These patients should have a secured airway before placing the patient in the supine position, directly inspecting the oral cavity, or obtaining IV access.

Lower airway
Vascular anomalies

Vascular anomalies affecting the airway have variable presentations.

Aberrant vasculature, commonly of the innominate artery, is the most common cause of secondary tracheomalacia. This is often self-limited.
Vascular rings most commonly result from a double aortic arch which surrounds both the trachea and esophagus. Children are symptomatic by 3 months of age, predominantly with respiratory symptoms although dysphagia may be present as well.
Other anomalies include pulmonary artery sling, and aberrant right subclavian. Any condition which causes pulmonary hypertension may cause an increase in pulmonary vasculature which can compress the left mainstem bronchus.

Barium esophagram identifies posterior indentation of the esophagus by a vascular ring. CT angiogram with contrast or MRI shows the anatomy of vascular anomalies obstructing the airway.

Congenital malformations

Congenital malformations of the lung such as hypoplastic lung, and cystic adenomatoid malformation may cause respiratory signs and symptoms.

Bronchiolitis

Bronchiolitis is an acute illness which initially presents with upper respiratory infection (URI) symptoms of sneezing, rhinorrea and fever. It is characterized by obstruction, edema with mucous production and cellular debris. This causes airway obstruction from increased airway resistance.

Patients present with cough, wheezing, dyspnea, tachypnea, and decreased feeding. It is caused by a viral infection most commonly Respiratory syncytial virus which causes more than 50% of cases.

Hospital admission for a child with bronchiolitis is warranted if the patient is in respiratory distress.

Bronchiolitis is managed with supportive care. Nebulized albuterol and racemic epinephrine should be administered and can be continued if the patient demonstrates a therapeutic response. Ipatroprium bromide may be beneficial especially in infants with tracheal and bronchialmalacia. Oral steroids are not indicated in first time wheezing although it may be used in atopic patients. Inhaled steroids may be beneficial in patients who have responded to oral steroids.

Aspiration syndromes

There is often no history of foreign body aspiration. Signs and symptoms of foreign body aspiration can present in three stages.

First – violent paroxysms of cough may occur with the initial obstruction of the airway.
Second – there may be an asymptomatic period after tiring of reflexes.

Potentially the most dangerous.

Third – marked by complications such as obstruction, erosion or infection in the airway. The child may develop fever, cough, hemoptysis, pneumonia, and atelectasis.

Chest radiograph and lateral neck films may be useful in evaluation for foreign bodies. The object is radiopaque 10-25% of the time, however radiograph is normal and the patient asymptomatic 15-30% of the time. CT or MRI may reveal radiolucent objects; however rigid bronchoscopy should be performed if there is a high index of suspicion.

Management of a patient with aspiration of a foreign body is removal by rigid bronchoscopy.

Gastroesophageal reflux can predisopose a child to recurrent respiratory disease secondary to recurrent aspiration. However, recurrent aspiration can recur with swallowing disorders independent of gastroesophageal reflux and can occur despite the presence of a tracheostomy.

Hydrocarbon pneumonitis may cause acute and chronic lung disease. Management of hydrocarbon pneumonitis involves supportive care with oxygen, fluids and ventillatory support if needed. Gastric emptying is contraindicated unless there has been an ingestion of >30ml as in an intentional overdose. Compounds which have greater systemic toxicity are: camphor, halogenated carbons, aromatic hydrocarbons, metals and pesticides.

Bronchiectasis

Bronchiectasis is characterized by irreversible dilatation of the bronchial tree as the end result of various disease processes. The differential diagnosis of bronchiectasis includes: cystic fibrosis, ciliary dyskinesia, immune deficiency, infection especially pertussis, measles, tuberculosis, right middle lobe syndrome, and yellow nail syndrome. Congenital causes include: Williams-Campbell syndrome (absence of annular cartilage), Marnier-Kuhn syndrome (congenital tracheobronchomegaly.) High-resolution CT of the chest is useful to diagnose bronchiectasis in a child.

Tracheomalacia

Tracheomalacia can occur as a complication of chronic mechanical ventilation in children.

Tracheoesophageal fistula may result in tracheomalacia.

Tracheomalacia and laryngomalacia present with persistant wheezing often with respiratory congestion in the absence of infection.

Tracheitis

Bacterial tracheitis is an acute infection of the upper airway which does not involve the epiglottis but can cause significant airway obstruction. It presents with fever, brassy cough, and respiratory distress.

The typical clinical course includes biphasic illness with precipitous worsening which can occur after several days of apparent improvement. Respiratory distress may lead to a requirement for intubation which can be relatively prolonged.

Bacterial tracheitis is typically caused by Staphylococcus aureus, Moraxella catarrhalis, non-typable Hemophilus influenza (H. Flu), and anerobic organisms. Treatment is with intravenous antibiotics.

Hemosiderosis

Hemosiderosis is associated with hemoptysis.

Parenchymal
Pneumonias

Pneumonia that is suspected to be of bacterial etiology should be treated with antibiotics based on the presumptive cause. Antibiotics should be withheld in cases where viral pneumonia is suspected.

Pneumonia typically presents following a few days of URI symptoms and fever. It is often marked by tachypnea, and retractions. Diminished breath sounds, crackles and wheezing can be heard on auscultation.

Bacterial pneumonia begins with a shaking chill, high fever, cough, and chest pain. Patients may exhibit drowsiness, restlessness, anxiety, splinting, and occasionally delirium.
Viral pneumonia presents with fever although it usually is not as high as in bacterial pneumonia.

Laboratory tests for pneumonia include: chest radiograph, blood culture, complete blood count, urine antigen detection studies, serology for Mycoplasma, and sputum culture. Patients with acute pneumonia may require invasive studies such as bronchoscopy, lung aspiration, and open lung biopsy.

Viral pneumonia is most common in children less than 5 years of age with the highest frequency between 2 and 3 years of age. The most common pathogens in this age group are influenza and RSV.

The most common bacterial causes are Streptococcus pneumoniae, Chlamydia pneuoniae, and Mycoplasma pneumonia. Other bacterial causes include: Streptococcus pyogenes and Staphylococcus aureus.

The organisms likely to cause the pleural and parenchymal complications of pneumonia are: Staphylococcus aureus, Streptococcus pneumoniae, and Streptococcus pyogenes.

The major complications of pneumonia include empyema, sepsis, pneumothorax, bronchopleural fistula, and pneumatoceles. Ultrasound and CT scan can assist in evaluation of the stage of empyema. Treatment includes: antibiotic therapy, drainage, fibrinolytic therapy, video-assisted thorascopic surgery (VATS), and occasionally thoracotomy and debridement.

Vaccinations against H. Flu and Pneumococcus, along with antibiotic treatment have led to an increase in prevention and control of pneumonia.

The differential diagnosis of recurrent pneumonia includes: Hereditary: cystic fibrosis, sickle cell disease, immunological disorders: AIDS, Bruton agammaglobulinemia, IgG deficiency, common variable immunodeficiency syndrome, severe combined immunodeficiency syndrome, chronic granulomatous disease, hyperimmunoglobulin E syndrome, leukocyte adhesion defect, Cilia disorders: immotile cilia syndrome, Kartagener syndrome, Anatomic disorders: pulmonary sequestration, lobar emphysema, esophageal reflux, foreign body,

tracheoesophageal fistula, gastroesophageal reflux, bronchiectasis, aspiration. Congenital lesions of the lung may mimic pneumonia.

Respiratory insufficiency is the most common cause of morbidity and mortality in progressive neuromuscular disorders such as: Duchenne muscular dystrophy, spinal muscular atrophy, congenital myotonic dystrophy, and myasthenia gravis. It is also significant in acquired conditions such as high level spinal cord injury, poliomyelitis, Guillain-Barre syndrome, and botulism. Treatment involves respiratory support in addition to antibiotics when infection is present.

Diaphragmatic hernia

Diaphragmatic hernia can be diagnosed on prenatal ultrasound in 50% of cases. At birth, respiratory distress is the cardinal sign characterized by tachypnea, grunting, accessory muscle use, and cyanosis. Physical examination reveals a scaphoid abdomen, increased chest diameter, breath sounds are decreased and bowel sounds are heard in the chest. Diagnosis can be made with nasogastric tube placement and chest radiograph.

Initial therapy for a diaphragmatic hernia involves respiratory support including: intubation with sedation and paralysis. Initial stabilization maneuvers include: nasogastric tube placement and arterial and venous lines should be placed. To prevent enlargement of the stomach, mask ventilation should be avoided. Infants may require high frequency oscillation ventilation or extracorporal membrane oxygenation. Surgical repair is usually made after 48 hours if the infant is on conventional mechanical ventilation.

Diaphragmatic hernia is associated with persistent pulmonary hypertension and subsequent abnormalities including poor growth, tracheomalacia, and developmental delay.

Trauma

Know how to evaluate a child with respiratory symptoms following chest wall trauma

- Following chest trauma, a child with respiratory symptoms should be assessed for airway, breathing, circulation, neurologic deficit, and exposure. Chest radiograph, CBC, electrolytes, glucose, BUN, Creatinine, amylase, liver function tests, prothrombin, partial thromboplastin, type and cross match, urinalysis, and blood gas should be obtained. Secondary survey should include an evaluation for thoracic trauma. In children blunt force to the chest is transferred mostly to the lungs instead of

the chest wall. It is important to observe the patient because respiratory symptoms may not develop immediately.

-Children with respiratory symptoms following chest trauma should be stabilized by ensuring the airway by jaw-thrust, suction, and endotracheal intubation if necessary. Surgical intervention may be needed.

Drowning, near drowning, acute respiratory distress syndrome

Clinical manifestations of acute respiratory distress syndrome include: tachypnea, decreased lung compliance, hypoxemia, and respiratory fatigue. Diagnostic signs are a triad of: noncardiogenic pulmonary edema, impaired oxygenation, and bilateral pulmonary infiltrates on chest radiograph.

The natural history of acute respiratory distress syndrome consists of three phases. First is the Exudative stage marked by increased capillary endothelial vascular permeability. Patients present with rapid development of respiratory failure. Second is the Inflammatory stage with fibrosing alveolitis and surfactant deficiency. The third stage is Resolution.

Acute respiratory distress syndrome has multiple etiologies including: pneumonia, drowning, aspiration, lung contusion, smoke inhalation, blood product transfusion, and sepsis. It may result from a near- drowning after a period of initial recovery. Patients with minimal symptoms following a near drowning may later develop symptoms that require hospitalization.

Acute respiratory distress syndrome often resolves with restoration of normal lung function; however there may be residual fibrosis.

The major causes of death in children with acute respiratory distress syndrome are: sepsis, extrapulmonary multiorgan failure, and air leaks.

Newborn infants
Bronchopulmonary dysplasia (chronic lung disease of infancy)

Bronchopulmonary dysplasia can develop in a newborn infant, regardless of gestational age, who has been treated with artificial ventilation and an enriched oxygen concentration. These infants are prone to corpulmonale and recurrent wheezing with severe respiratory infections.

Infants with BPD commonly have failure to thrive which may be a sign of insufficient oxygenation. Home oxygen therapy may be required.

Feeding methods are limited in infants with BPD due to aversive oral motor behavior. They often require greater than 100% of the recommended dietary allowance for calories in order to grow.

BPD is commonly associated with gastroesophageal reflux and may worsen their respiratory status.

Other - not BPD

Radiographic findings in idiopathic neonatal respiratory distress syndrome typically show fine reticular granularity of the lung parenchyma and air bronchograms. Early on, the air bronchograms are most prominent in the left lower lobe due to superimposition of the cardiac shadow.

The radiographic patterns that reflect meconium aspiration show patchy infiltrates, bilateral coarse streaking, increased AP diameter and flattening of the diaphragms. Meconium aspiration may result in chronic lung disease.

The radiographic patterns that reflect pneumonia in the newborn are often indistinguishable from the radiographic patterns of RDS.

Subglottic stenosis is a complication of endotracheal intubation.

The hyperoxia test may be used to distinguish between pulmonary disease and cyanotic congenital heart disease as a cause of hypoxemia and metabolic acidosis in a neonate. Arterial PaO2 is evaluated after administration of 100% oxygen. In pulmonary disease, the PaO2 typically increases significantly with oxygen administration. The PaO2 is usually not as low as that seen in congenital heart disease, and also varies with time and changes in ventilator management. In cyanotic congenital cardiac lesions, the PaO2 does not increase in response to oxygen administration.

Asthma (see VIII.C)
Cystic fibrosis

The common microbial pathogens involved in the pulmonary complications of cystic fibrosis are: Staphylococcus aureus, Haemophilus influenza, Pseudomonas aeruginosa, and Burkholderia cepacia.

Extrapulmonary complications of cystic fibrosis include:

Upper airway: Sinusitis and nasal polyps.
Pancreatic disease secondary to pancreatic duct obstruction. This usually leads to CF related diabetes.
Intestinal obstruction: manifested as mecomium ileus in newborns, distal intestinal obstruction syndrome (DIOS), and rectal prolapse. Rectal prolapse is associated with cystic fibrosis.
Hepatobiliary disease: gallbladder disease, liver dysfunction which can lead to liver failure.

Fertility: absent Vas Deferens in males, mucous obstruction of cervix in females.

In CF, bacterial colonization of the respiratory tract requires aggressive management with antimicrobial therapy to suppress bacterial infection. Therapy is tailored to microbial culture and sensitivity results from the lower respiratory tract. Scheduled antibiotic therapy with inhaled tobramycin may be used in patients colonized with Pseudomonas aeruginosa.

The inheritance of cystic fibrosis is autosomal recessive with the most common defect being delta F508.

Cystic fibrosis is diagnosed by history, typical clinical features, and two sweat chloride tests obtained on separate days. The test is positive if chloride is greater than 60mEq/L. DNA testing is available for the most common CFTR mutations and can identify greater than 90% of individuals with two CF mutations. Other diagnostic testing include: loss of increased potential differences across nasal epithelium with the application of amiloride, and absent voltage response to beta adrenergic agonist. This is particularly useful in patients with equivocal sweat chloride results. Many infants are identified on newborn screen which is 95% sensitive. The newborn screen should be confirmed by sweat chloride test.

The common non-pulmonary manifestations of cystic fibrosis in the neonate are: meconium ileus, meconium peritonitis, and prolonged jaundice.

Supplemental calories, pancreatic enzymes, and fat-soluble vitamins are needed in patients with cystic fibrosis.

In infancy, the manifestations of cystic fibrosis are: hypoproteinemia, anemia, steatorrhea, recurrent pulmonary symptoms, hypochloremic alkalosis.

Exocrine pancreatic insufficiency in infants can be noted clinically with increased stooling and failure to thrive. Stool can be evaluated for elastase-1 activity. Elevated serum trypsinogen levels are an indication of pancreatic dysfunction.

The average lifespan of patients with CF is 37 years therefore, it is important to plan for survival into adulthood.

Hemoptysis and pneumothorax can be potentially life- threatening complications of cystic fibrosis.

The management of pulmonary disease includes: airway clearance with chest physiotherapy, use of inhaled hypertonic saline and DNAse to aid in mucous clearance,

and aggressive antibiotic treatment parenterally and with inhaled tobramycin. Pulmonary function is monitored. Patients may undergo lung transplantation.

Management of extrapulmonary complications of cystic fibrosis include:

Upper airway: antibiotic treatment of sinusitis, removal of obstructive nasal polyps.
Pancreatic disease: supplement fat soluble vitamins, pancreatic enzymes, caloric intake, monitor for development of diabetes.
Intestinal obstruction: management of distal intestinal obstruction syndrome (DIOS), and rectal prolapse.
Hepatobiliary disease: monitor liver function and gallbladder.
Primary ciliary dyskinesia (dysmotile cilia syndrome)

Otitis media, dextrocardia, and/or bronchiectasis may be due to primary ciliary dyskinesia.

Ciliary biopsy is required to diagnose primary ciliary dyskinesia.

Extrapulmonary
Pleural fluid

Pleural effusion or exudate can be seen on radiograph of the chest as diffuse haziness at the pleural surface, or a dense, sharp shadow. Although radiograph can be negative, ultrasound or CT will be positive.

Accumulations of pleural fluid may be caused by: bacterial pneumonia, heart failure, rheumatologic disease, and metastasis.

Pleural fluid associated with empyema shows bacteria on gram stain, has a pH less than 7.20, and greater than 100,000 neutrophils.

Chylothorax is a collection of fluid formed by chyle from the thoracic duct or lymphatics. The fluid usually appears milky. Triglyceride level is greater than 110mg/dL, pleural fluid to serum triglyceride ratio is greater than 1, pleural fluid to serum cholesterol level is less than 1, and chylomicrons are seen on lipoanalysis. Cells present in the fluid are usually T lymphocytes.

Drainage of the purulent material is an essential component of treatment of empyema..

Pneumothorax, pneumomediastinum

The signs and symptoms of pneumothorax include: pain, dyspnea and cyanosis. Breath sounds are decreased over the affected side and respiratory distress and retractions are usually present. Percussion over the affected side is tympanic. On

,chest radiograph the airway may be shifted to the unaffected side.

Therapy for a child with pneumothorax includes: drainage via needle thoracostomy, chest tube placement, video-assisted thorascopic surgery, and open thoracostomy.

Spontaneus pneumothoraces occur most frequently in tall, thin, males and may recur in young asthenic boys. Patients with collagen synthesis defects are prone to pneumothorax development.

Asthma may be associated with pneumothorax and pneumomediastinum. Pneumothorax may be a complication of resuscitation and mechanical ventilation.

Thoracic deformities

Scoliosis can lead to restrictive pulmonary diseases because of chest wall restriction.

Restrictive pulmonary disease can result from severe progressive neuromuscular disease of any etiology.

Pectus excavatum is not usually associated with pulmonary disease or exercise limitation.

Pulmonary hypertension and cor pulmonale

Oxygenation may decrease during abnormal sleep, which may cause pulmonary hypertension or exacerbate existing cor pulmonale.

Pulmonary hypertension is potentially reversible.

Pulmonary hypertension may be idiopathic (primary) or secondary.

-Primary: familial, pulmonary artery involvement, venous or capillary involvement, persistent pulmonary hypertension of the newborn, pulmonary venous hypertension, associated with hypexemia, chronic thrombotic disease, chronic embolic disease. Corpulmonale is right ventricular failure resulting from pulmonary pathology.

-Secondary: cystic fibrosis and bronchopulonary dysplasia.

Sleep disorders

In broad categories, sleep disorders in children may be caused by: insomnia, sleep- related breathing disorders, hypersomnia not due to a breathing related sleep disorder,

circadian rhythm sleep disorder, parasomnia, sleep related movement disorder, isolated symptoms, normal variants, and unresolved issues.

Symptoms that reflect poor sleep quality in children include: excessive daytime sleepiness, and decreased daytime alertness. In younger children other symptoms may be present such as: mood disturbances, behavioral problems, hyperactivity, poor impulse control, neurocognitive dysfunction, inattention, impaired vigilance, and compromised higher level cognitive skills. Poor sleep quality can result in problems with learning, ability to function in school, and social functioning.

Adenoid tonsillectomy is the first line treatment for most children diagnosed with obstructive sleep apnea.

The gold standard for evaluation of suspected obstructive sleep apnea in children is an overnight polysomnogram. Other studies can include a lateral neck radiograph to evaluate adenoid size, complete blood count to evaluate for polycythemia, and a bicarbonate level may suggest compensation for hypoventilation. Chest radiograph, EKG, and echocardiogram may show evidence of right ventricular hypertrophy or cor pulmonale.

The differential diagnosis of obstructive sleep apnea in children includes: primary snoring, upper airway resistance syndrome, obstructive hypoventilation, and central sleep apnea.

Children with severe obstructive apnea due to upper airway obstruction are at significant risk for respiratory distress postoperatively (e.g., due to postoperative airway swelling, postoperative obstructive pulmonary edema). They should be monitored in an inpatient setting postoperatively.

Night terrors are a partial arousal parasomnia occurring during the transition out of stage 4 or slow wave sleep. The tendency for them to occur during the first two hours of sleep is due to the predominance of slow wave sleep which occurs during the first third of the night. They occur more frequently in young children because of a higher percentage of slow wave sleep compared to older children and adults. There is amnesia of the event upon waking.

Nightmares are far more common than night terrors and occur during Rapid Eye Movement (REM) sleep. They occur during the last third of the night.

Common causes of somnolence in adolescents include: insufficient sleep, delayed sleep phase, narcolepsy, restless legs syndrome and periodic limb movement disorder.

Sleep may be affected by both prescribed and over-the-counter medications.

Sudden infant death syndrome

An apparent life-threatening event (ALTE) is defined as “an episode that is frightening to the observer and that is characterized by some combination of apnea, color change, marked change in muscle tone, choking, or gagging.”

Appropriate management of an infant with ALTE includes: 23 hours of observation with cardiopulmonary monitoring. Laboratory studies may include: complete blood count with differential, electrolytes, C-reactive protein, magnesium, ammonia, lactate, pyruvate, glucose, calcium, arterial blood gas, and testing for Bordatella pertussis and Respiratory Syncytial Virus during the appropriate season.

Risk factors for sudden infant death syndrome include: age (2-4 months), male gender, race (African American, Native American), growth failure, no pacifier, prematurity, prone and side sleeping position, recent febrile illness, smoking exposure, soft sleeping surface, infant-parent bed sharing, overheating, cold season, no central heating.

There is no evidence of a relationship between apnea and sudden infant death syndrome.

The differential diagnosis of apparent life-threatening events (ALTE) in infancy includes: infection, metabolic abnormality, gastroesophageal reflux, aspiration, cardiac dysrhythmia, seizures, nonaccidental trauma, and apnea of infancy.

There is no evidence that the use of cardiorespiratory ("apnea") monitors at home in infants following ALTE can prevent SIDS.

Diagnostic testing

Pulmonary function testing

Spirometry measures lung volumes.

Oximetry

Know the correlation between PaO2 and oxyhemoglobin saturation.

-The oxygen dissociation curve relates PaO2 to hemoglobin saturation. Factors which cause a shift in the curve will change the relationship between PaO2 and SaO2.

Pulse oxymetry is a simple non-invasive method for assessing oxygen saturation in children with acute pulmonary disease.

Common causes of inaccurate measurement of SpO2 are: carboxyhemoglobin, methemoglobin, motion artifact, oxygen-hemoglobin dissociation curve shifts, poor tissue perfusion, severe anemia, dark skin pigmentation, external light, and dyes.

Blood gas analysis

Analysis of an arterial blood gas is important during the assessment of pulmonary function. There are limitations of obtaining capillary blood gas.

Arterial blood gas findings in a patient who has acute respiratory failure (or opiate overdose) typically show an early increase in PCO2, proportionate decrease in PO2, and an excellent response to supplemental oxygen.

Imaging

On chest radiograph, intrathoracic airway obstruction is best appreciated on expiratory film. Air trapping can be seen with persistent inflation of the obstructed side and the mediastinum is shifted to the opposite side.

Atelectasis can be recognized by volume loss and fissure displacement on chest radiograph.

Environmental tobacco smoke exposure

Side effects of environmental tobacco smoke exposure include: chronic cough, increased sputum production, wheezing, decreased birth weight, drug interactions especially with oral contraceptives, phenacetin, theophylline and imipramine.

Household fumes, such as those from cooking and cleaning, may be harmful to children.

Cardiovascular Disorders

A) General aspects

Blood pressure (see III.B.2)

Many prescription medications (amphetamines, corticosteroids, cyclosporine, erythropoietin, estrogens containing birth control pills and migraine medications- ergotamine), over-the-counter medications (cough and cold medications, nasal decongestants) and illicit drugs (alcohol, ecstasy and cocaine) likely will elevate the blood pressure.

Treatment of hypertension in children should address the cause and correct it. (e.g. Coarctation of aorta, renal artery stenosis, pheochromocytoma, hyperaldosteronism, renal disorder); Lifestyle interventions should be initiated in all hypertensive children. When lifestyle modifications are inadequate for BP control or are unsuccessful in patients with more elevated BP, pharmacologic therapy must be considered.

Coarctation of the aorta causes upper extremity hypertension and low BP or low to absent pulses in lower extremity.

Chest pain

Chest pain in healthy children is generally not cardiopulmonary in origin. Cardiovascular causes of chest pain:

Inflammatory: Pericarditis, Myocarditis
Infective: viruses, bacteria
Noninfective: SLE, Crohn disease
Postpericardiotomy syndrome
Increased Myocardial Demand or Decreased Supply
Cardiomyopathy: dilated or hypertrophic
Arrhythmias
LVOT obstruction: aortic stenosis, subaortic stenosis, supravalvar aortic stenosis
Coronary Artery Abnormalities
Congenital: ALCAPA (anomalous origin of the left coronary artery from pulmonary artery), ALCA(anomalous left coronary artery) from right coronary sinus, coronary fistula
Acquired: Kawasaki disease, postsurgical, posttransplant coronary vasculopathy, familial hypercholesterolemia
Miscellaneous
Aortic dissection, Rupture of aortic aneurysm, Pulmonary hypertension, Mitral valve prolapsed, Atrial myxomas.
Cardiac device/stent complications
Drugs
Cocaine, Sympathomimetic overdose
Syncope

Description of a syncopal episode usually directs the evaluation- Cardiac disease is suggested when syncope accompanies exercise. Cardiac syncope is potentially fatal and always deserves careful evaluation and treatment.

Cardiovascular evaluation is needed in patients with syncopal or pre-syncopal episodes with exercise.

Cardiac causes of syncope: Arrhythmias (SVT, WPW syndrome, VT, Vfib), Obstructive cardiac lesion (Aortic stenosis, mitral stenosis, atrial myxoma) Structural cardiopulmonary disease (hypertrophic

cardiomyopathy, acute aortic dissection, pericardial tamponade, pulmonary embolism, aortic stenosis, and pulmonary hypertension) Others (Sick sinus syndrome, Adams-Stokes syndrome , Aortic dissection and cardiomyopathy)

4.Murmur

Qualities of innocent heart murmurs: Early systolic ejection, Short duration, low intensity (grade 2 or 1), vibrating (or musical) quality

Provide appropriate counseling - Heart murmurs are heard commonly in infants, children, and adolescents. Approximately 50% to 70% of individuals seen for school or sports preparticipation examinations have a heart murmur. Indeed, a murmur is heard in most children at one or more of their examinations. Most murmurs are innocent (i.e., normal).

Child with an innocent murmur requires no further evaluation.

ongestive heart failure (CHF)
Diagnosis

Symptoms of congestive heart failure (HF) in infants are irritability, dyspnea during feeding, and decreased volume with each feeding.

Symptoms of congestive heart failure in older children are early fatigue, exercise intolerance, anorexia, and cough.

Signs and symptoms of congestive heart failure:

In neonates, the earliest clinical manifestations may be subtle. Most commonly infants have feeding difficulties due to dyspnea, increased fatigability, and secretion of anorexic hormones that limit the volume of feedings. Ultimately, affected babies fail to thrive. Physical findings in infants who have HF include mild-to-severe retractions, tachypnea or dyspnea with grunting (a form of positive end-expiratory pressure), tachycardia, a gallop rhythm (S3, S4), and hepatomegaly.

Older children manifest exercise intolerance, somnolence, anorexia, or more “adultlike” symptoms such as cough, wheezing, or crackles (rales). As with younger children, the physical examination may reveal a gallop rhythm and hepatomegaly as well as peripheral edema and jugular venous distention.

Important physical findings in congestive heart failure in older children are edema, hepatomegaly, jugular vein distention, cardiomegaly, gallop rhythm.

Imaging study of the chest may help diagnose congestive heart failure- e.g. chest radiograph and Echocardiography.

There is an association between systemic arteriovenous malformation and congestive heart failure in a newborn infant.

Common causes of congestive heart failure in infants and children:

Cardiac – congenital and structural heart disease, cardiomyopathy, myocarditis, myocardial infarction, acquired valve disorders, hypertension, Kawasaki syndrome, dysrhythmia (bradycardia or tachycardia)

Non-Cardiac-Anemia, Sepsis, Hypoglycemia, Diabetic ketoacidosis, Hypothyroidism, Other endocrinopathies, Arteriovenous fistula, Renal failure, Muscular dystrophies

Role of the pulmonary vascular bed in the presentation of congestive heart failure in infants with large volume left-to-right shunts: Ventricular-level shunts include ventricular septal defect and patent ductus arteriosus. The left heart volume overload causes increased cardiac filling pressure and pulmonary edema. Typically, affected patients present during the first 2 to 3 postnatal months, after the natural decrease in the pulmonary vascular resistance occurs. The lower pulmonary vascular resistance relative to the systemic vascular resistance leads to a significant increase in the pulmonary blood flow relative to the systemic blood flow.

Management

Treatment of congestive heart failure:

Recognition and Treatment of Underlying Systemic Disease or structural anomalies.
Afterload Reduction: Angiotensin-converting enzyme inhibitors, Angiotensin receptor blockers, Milrinone

Nitrates, Brain natriuretic peptide (BNP:Nesiritide, a recombinant form of BNP)

Preload reduction: Diuretics, BNP
Sympathetic Inhibition: Beta blockers, BNP, Digoxin
Cardiac Remodeling Prevention: Mineralocorticoid inhibitors (spironolactone)
Inotropy: Digoxin, milrinone
Heart transplantation
ongenital heart disease
General

Recognize the increased risk and plan appropriate evaluation of congenital heart disease in a newborn infant with congenital anomalies, e.g.trisomy 21, trisomy 18, fetal alcohol syndrome, 22q11 microdeletion, 45,XO.

Electrocardiogram and echocardiography should be part of the evaluation of a patient with possible cardiogenic shock.

Cardiogenic shock may be the initial finding in a newborn infant with congenital heart disease.

Findings of cardiogenic shock in the newborn infant are decreased spontaneous movements, low thready to absent peripheral pulse, mottled, blue or pale skin, low

BP, prolonged capillary refill, tachycardia, hepatomegaly, rales on lung exam, gallop rhythm on cardiac examinations, cardiomegaly on chest radiograph, low valtage QRS complexes and T wave abnormality on EKG.

Treatment of cardiogenic shock in the newborn infant: Immediate management: ABC, O2 if needed, IV access, IV fluids, vasopresors and Inotropes; Cause specific therapy: EKG to rule out arrhythmia and treat if needed, rule out sepsis, ductal dependent CHD- start Prostaglandin E1, chest radiograph to rule out pneumothorax and pneumomediastinum. Unresponsive to Inotropes – transvenous cardiac pacing.

Important lesions associated with the shock-like presentation in a newborn infant are:

congenital heart diseases Such as Hypoplastic left heart syndrome (HLHS), Congenital aortic stenosis, Coarctation of aorta and Interrupted aortic arch, Anomalous origin of left coronary artery from pulmonary artery (ALCAPA).
Cardiac Muscle disorder: cardiogenic shock secondary to intra-partum asphyxia, Myocarditis secondary to viral, bacterial or protozoal cause.
Dysrhythmias: SVT / VT /VF/ Complete AV block, Long QT syndrome.
Metabolic: severe hypoglycemia, adrenal insufficiency.

(e)rare other causes(pyruvate dehydrogenase deficiency, carnitine deficiency, myotonic dystrophy).

Central cyanosis is usually pathological; affected infants have bluish discoloration of the lips, tongue, mucosa, and nail beds. Neonatal cyanosis is due to either a right-to- left intracardiac shunt as a result of congenital heart disease or to respiratory compromise resulting in a ventilation-perfusion mismatch. Other causes include hypoventilation due to neurologic disorders or pharmacologic agents or methemoglobinemia. Acrocyanosis is the transient blue discoloration of the hands and feet (Item C264) in response to vasomotor instability or a cool environment. The perioral region (sparing the lips and mucous membranes) can also be involved. Extremities may be cool to the touch. Acrocyanosis is believed to be caused by vasoconstriction of the small arterioles. It typically resolves in the first few weeks or months after birth.

Cyanotic disease A Diagnosis

Distinguish between central cyanosis and acrocyanosis: Central cyanosis is usually pathological; affected infants have bluish discoloration of the lips, tongue, mucosa, and nail beds. Neonatal cyanosis is due to either a right-to-left

intracardiac shunt as a result of congenital heart disease or to respiratory compromise resulting in a ventilation-perfusion mismatch. Other causes include hypoventilation due to neurologic disorders or pharmacologic agents or methemoglobinemia.

Clinical characteristics of a tetralogy spell: A decreased or absent pulmonary stenosis murmur, respiratory distress, crying, inconsolability, hyperpnea, and increasing cyanosis. Cyanotic spell frequently occur in the morning or at times of dehydration (e.g., fever, gastroenteritis). If not treated quickly, cyanotic spells can lead to serious morbidity and even death.

Cardiac causes of cyanosis in the newborn infant: Hypoplastic left heart syndrome, complete transposition of the great arteries, single ventricle lesions such as tricuspid atresia, pulmonary atresia, severe tetralogy of Fallot, truncus arteriosus, interrupted aortic arch, and total anomalous pulmonary venous return.

Absence of improvement in arterial oxygen content with 100% oxygen in comparison with room air is compatible with the diagnosis of cyanotic congenital heart disease. (Hyperoxia test)

Clinical features of transposition of the great arteries: (TGA) should be suspected in the infant exhibiting profound cyanosis without respiratory distress. Affected neonates have oxygen saturations that average 50% to 70%. Chest radiograph typically demonstrates a narrow mediastinal shadow resulting from the anteroposterior alignment of the aorta and pulmonary artery.

b. Management

Complications of polycythemia in a patient with cyanotic congenital heart disease: Cerebrovascular accident (CVA) or stroke especially if patient has also iron deficiency anemia.

Prognosis for a patient with tetralogy of Fallot: Most cases can be corrected with surgery. Babies who have surgery usually do well. More than 90% survive to adulthood and live active, healthy, and productive lives. Without surgery, death usually occurs by the time the person reaches age 20.

Prognosis for cognitive development in patients with cyanotic congenital heart disease: Overall outcomes for the arterial switch operation (for TGA) demonstrate that 55% of patients have some degree of impaired cognitive, behavioral, motor, or speech function, after surgery with an average IQ score of 93. After Norwood procedure fro HLHS patient has IQ score between 71-90.

A relative anemia can be associated with a stroke in a patient with cyanotic congenital heart disease.

Immediate management of a child with a hypoxic episode: Depends on hemoglobin concentration (if low transfuse PRBC), cardiac output (if low increase cardiac output by increase intravascular volume - IV fluids or diuretics, inotropes) and the amount of hemoglobin that is saturated with oxygen, fever control with antipyretics which decreases oxygen.

Role of ductus arteriosus in cyanotic congenital heart disease and the use of prostaglandin E1 in treatment: The ability to maintain or re-establish patency of the ductus arteriosus with prostaglandin E1 allows for effective stabilization of the severely cyanotic neonate, which provides a period of circulatory control while metabolic derangements are corrected and controlled surgical planning can ensue. Prostaglandin E1 is used in ductal dependent cyanotic congenital heart lesions such as interrupted aortic arch, hypoplastic left heart syndrome, critical aortic valve stenosis, and coarctation of the aorta to maintain systemic circulation. In other ductal-dependent lesions, such as pulmonary valve atresia, severe tetralogy of Fallot, and critical pulmonary valve stenosis, prostaglandin allows for left-to-right flow to maintain pulmonary artery circulation.

Acyanotic disease
Diagnosis

Major clinical findings in patients with cardiac anomalies such as

Ventricular Septal Defect (VSD): asymptomatic or symptoms related to heart failure. The most common symptoms include shortness of breath, fast breathing, paleness, failure to gain weight, fast heart rate, sweating while feeding, frequent respiratory infections. On examinations holo-systolic murmur if small defect; no murmur if large defect.

Atrial Septal Defect (ASD): Asymptomatic or easy fatigability, recurrent respiratory infections, or exertional dyspnea. On examination wide fixed split of S2, ejection systolic murmur due to increase pulmonary flow.

Patent Ductus Arteriosus (PDA): small PDA may not cause any symptoms. However, some infants may have symptoms such as: fast breathing, poor feeding habits, rapid pulse, shortness of breath, sweating while feeding, tiring very easily, poor growth. On examination continuous musical murmur; Murmur may be absent if wide open PDA and no resistance to blood flow.

Aortic Stenosis: easily tired with exertion (in mild cases), failure to gain weight, poor feeding, serious breathing problems that develop within days or weeks of birth bin severe cases. Children

with mild or moderate aortic stenosis may get worse as they get older. They also run the risk of developing a bacterial endocarditis.

Pulmonic Stenosis: mild cases do not cause symptoms. An ejection systolic murmur on physical examination. In moderate to severe cases, patient become symptomatic: cyanosis, shortness of breath, abdominal distention, chest pain, fainting, fatigue, poor weight gain or failure to thrive in infants with severe obstruction, sudden death in severe case. Symptoms may get worse with exercise or activity.

Know the importance of patent ductus arteriosus in the presentation of hypoplastic left heart syndrome and in coarctation of the aorta: In HLHS right to left shunt at PDA provide adequate systemic blood flow and also retrograde blood flow to hypoplastic aorta thus to coronary arteries. In coarctation of aorta (COA), right to left shunt at PDA provide adequate systemic blood flow.

Management

Initial management of a premature infant with patent ductus arteriosus: Judicious fluid management (<150 mL/kg per day) in the first week after birth may reduce the likelihood of clinically significant PDA. However, once the PDA is diagnosed, fluid restriction is a mainstay of treatment. Diuretic therapy has been recommended also for the treatment of symptomatic neonates, although no rigorously collected data support this approach. The use of positive end-expiratory pressure (PEEP) also may help reduce pulmonary overcirculation in the ventilated patient. Pharmacologic management consists of intravenous indomethacin or ibuprofen-lysine, usually administered in three successive doses. If the PDA fails to close with these pharmacologic measures, surgical ligation may be warranted.

Importance of immediate referral and long-term frequent BP measurements, management in a patient with coarctation of the aorta: Coarctation that presents in the symptomatic neonate should be repaired surgically. However, even with aggressive and excellent surgical repair, recoarctation can occur as the child grows. In addition, patients who undergo surgical repair of aortic coarctation have a higher incidence of hypertension at long-term follow-up and should be followed closely for this complication.

Expected natural history of ventricular septal defect : The natural history of a VSD depends on the magnitude of the left-to-right shunt. Smaller defects are

restrictive, resulting in a high-grade murmur due to the pressure gradient but the magnitude of the left-to-right shunt is minimal and, therefore, pulmonary overcirculation and associated congestive heart failure do not occur. However, moderate-to-large ventricular septal defects are associated with a lower-grade murmur due to the lack of a pressure gradient between the two ventricles. Pulmonary artery pressure will be at systemic levels. Therefore, congestive heart failure evolves over the first 1 to 2 postnatal months, as the pulmonary vascular resistance naturally falls. Surgical intervention typically is performed at 4 to 6 months of age.

Expected natural history of a bicuspid aortic valve: bicuspid (two-leaflet) aortic valve is the most common congenital heart defect, present in approximately 1% of the general population. Most of these valves function well with minimal, if any, aortic valve stenosis or insufficiency. Patients who have bicuspid aortic valves and, at most, mild aortic stenosis or insufficiency are evaluated every 2 to 3 years because as many as 70% develop some degree of stenosis or insufficiency by the age of

30 years; In most reports, up to 10% of first-degree relatives of an individual who has a bicuspid aortic valve also show evidence of a congenital anomaly of the aortic valve.

Management of severe pulmonary valve stenosis: Prostaglandin E1 to keep PDA open and maintain left to right shunt. The current best practice is interventional cardiac catheterization with balloon pulmonary valvuloplasty. Rarely, balloon pulmonary valvuloplasty is unsuccessful. This is more common when the valve disorder is associated with specific genetic conditions (e.g., Noonan syndrome) in which the valve leaflets are much thickened, "rubbery," and have a tendency to recoil to their native state, even after aggressive attempts at valvuloplasty. Other patients may have not only valvular but also subvalvular or supravalvular pulmonary stenosis (the latter commonly seen in Williams syndrome), which are not amenable to balloon valvuloplasty. In these cases, surgical valvotomy at times accompanied by infundibular muscle resection or patch augmentation of the right ventricular outflow tract and main pulmonary artery is the therapeutic intervention of choice.

Patients with untreated large left-to-right shunt lesions with pulmonary hypertension (e.g., large VSD, AV septal defect, large PDA) are at risks for pulmonary vascular obstructive disease (Eisenmenger complex) due to gradual muscularization of the pulmonary arterial tree, and eventual shunt reversal that yields right-to-left circulation and cyanosis. Increasingly uncommon in this era in which most children that have congenital heart disease obtain timely diagnosis and surgical repair.

Infectious and postinfectious diseases
Infective endocarditis (IE)

Know the indications for antibiotic prophylaxis in children with congenital heart lesions:

Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
Previous infective endocarditis
Congenital heart disease (CHD)

Unrepaired cyanotic CHD, including palliative shunts and conduits

Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure while endothelialization occurs (i.e. ASD devices, PDA coils, etc.)

Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization) (e.g., following repair of VSD with patch leak)

Cardiac transplantation recipients who develop cardiac valvulopathy.

Know the drugs of choice for the prophylaxis of infective endocarditis: oral amoxicillin, 50 mg/kg can be administered approximately 30 to 60 minutes prior to the procedure for patients who require IE prophylaxis.

Clinical manifestations of infective endocarditis: Fever, malaise, endurance fatigue a new or changing heart murmur, weight loss, and coughing. Vascular phenomena: septic embolism (causing thromboembolic problems such as stroke in the parietal lobe of the brain or gangrene of fingers), Janeway lesions (painless hemorrhagic cutaneous lesions on the palms and soles), intracranial hemorrhage, conjunctival hemorrhage, splinter hemorrhages, renal infarcts, and splenic infarcts. Immunologic phenomena: Glomerulonephritis which allows for blood and albumin

to enter the urine, Osler's nodes (painful subcutaneous lesions in the distal fingers), Roth's spots on the retina, positive serum rheumatoid factor. Other signs may include; night sweats, rigors, anemia, splenomegaly.

A common mnemonic for the signs and symptoms of endocarditis is “FROM JANE”:

Fever
Roth's spots
Osler's nodes
Murmur
Janeway lesions
Anemia
Nail hemorrhage (splinter hemorrhages)
Emboli

Know the management of infective endocarditis: Diagnosed by modified Duke criteria.

Major Criteria

Positive blood culture
Positive echocardiogram (vegetation, paravalvular abscess, or valve dehiscence after surgery)
New valvular regurgitation (by auscultation, not echocardiogram) Minor Criteria
Predisposing heart condition (including prior IE)
Injection drug use
Fever (temperature >100.4°F [38°C])
Major arterial emboli
Septic pulmonary infarcts
Mycotic aneurysm
Intracranial hemorrhage
Conjunctival hemorrhage
Janeway lesions (painless hemorrhagic lesions on palms and soles)
Glomerulonephritis
Osler nodes (painful lesions at fingertips)
Roth spots (retinal hemorrhages)
Positive rheumatoid factor
Single positive blood culture
Serologic evidence of active infection with an “organism consistent with IE”

Note: Splinter hemorrhages and erythrocyte sedimention rate are not criteria. Also, there is no “minor” echo criteria, i.e., valvular regurgitation alone is not a criterion.

Know the microbiology of infective endocarditis: Typical microorganism consistent with IE from two separate blood cultures, as noted below: Viridans-group streptococci, or Streptococcus bovis including nutritional variant strains, or HACEK group, or Staphylococcus aureus, or Community-acquired Enterococci, in the absence of a primary focus. Staphylococcus aureus followed by Streptococci of the viridans group and Coagulase negative Staphylococci are the three most common organisms responsible for infective endocarditis.

Know the epidemiology of infective endocarditis, including risk factors: Epidemiology of endocarditis has changed in the modern era. Although it remains a rare diagnosis, the rate of IE may be increasing, and it is a frequent concern among pediatricians. The prevalence of rheumatic heart disease has decreased, and a population of patients who survive beyond infancy with severe congenital cardiac malformations has emerged. Importantly, 90% of IE cases occur in individuals who have heart disease, usually congenital. In fact, IE may be the presenting sign of a bicuspid aortic valve. The increased use of invasive procedures in neonatal and pediatric intensive care units, however, has placed individuals whose hearts are structurally normal at risk.

Blood culture is the most important test for the diagnosis of infective endocarditis.Rheumatic fever:

Clinical manifestations of rheumatic fever: Major criteria:

J- joints/migratory polyarthritis

O- obvious reason for diagnosing and treating this patient as cardiac involvement; carditis endocarditis, valvulitis, myocarditis, paricarditis.

N- nodes, subcutaneous nodule.

E- erythema marginatum, serpentine/slightly raised red or pink rash over trunk and extremities.

S- sydenham chorea, A characteristic series of rapid movements without purpose of the face and arms

Minor criteria:

Fever of 38.2–38.9 °C (101–102 °F).

Arthralgia: Joint pain without swelling (cannot be included if polyarthritis is present as a major symptom).

Raised erythrocyte sedimentation rate or C reactive protein. Leukocytosis.

ECG showing features of heart block, such as a prolonged PR interval (Cannot be included if carditis is present as a major symptom).

Previous episode of rheumatic fever or inactive heart disease.

Other sign and symptoms: Abdominal pain, Nose bleeds, Preceding streptococcal infection: recent scarlet fever, raised antistreptolysin O or other streptococcal antibody titre, or positive throat culture.

According to revised Jones criteria, the diagnosis of rheumatic fever can be made when two of the major criteria, or one major criterion plus two minor criteria, are present along with evidence of streptococcal infection: elevated or rising antistreptolysin O titrer or DNAase. Exceptions are chorea and indolent carditis, each of which by itself can indicate rheumatic fever.

Laboratory findings of rheumatic fever: Raised erythrocyte sedimentation rate or C reactive protein, raised antistreptolysin O or other streptococcal antibody titer, or positive throat culture.

Murmurs of mitral insufficiency and aortic insufficiency are the most common murmurs in rheumatic fever.

Epidemiology of rheumatic fever: In developing areas of the world, acute rheumatic fever (ARF) and rheumatic heart disease are estimated to affect nearly 20 million people. Rheumatic fever can

occur at any age, although most cases occur in children 5 to 15 years of age. Worldwide, there are 470,000 new cases of rheumatic fever and 233,000 deaths attributable to rheumatic fever or rheumatic heart disease each year. The mean incidence of ARF is 19 per 100,000. In the United States and other developed countries, the incidence of ARF is much lower at 2 to 14 cases per 100,000; this is probably due to improved hygienic standards and routine use of antibiotics for acute pharyngitis.

Echocardiography should be done in a patient with rheumatic fever.

Plan the initial management of acute rheumatic fever: Eradicate streptococcal organisms and bacterial antigen with Penicillin (single parenteral injection of benzathine benzylpenicillin). Also anti-inflammatory medications play an important role: High dose aspirin. Corticosteroids should be reserved for the treatment of severe carditis.

Myocarditis

Clinical manifestations of myocarditis: In newborn present with fever, URI symptoms and then progress to symptoms of cardiac failure and poor perfusion. An S3 gallop is common. In older child present with fever, chest pain and symptoms of cardiac failure (fatigue, shortness of breath, palpitation).

Know the laboratory evaluation of myocarditis: CBC with differential, blood culture, ESR, CRP, Nasopharyngeal and rectal viral culture, cardiac enzymes (CKMB, troponin I). Other tests: chest radiograph, EKG, cardiac echocardiography.

Know the microbiology of myocarditis: Most likely viral pathogens- enterovirus, coxsackievirus, and adenovirus, parvovirus B19, HIV and CMV. Bacterial causes are rare, except for immunodeficient patients. Bacterial pathogens include Brucella, corynebacterium diphtheriae, Hemophilus influenzae and Borrelia burgdorferi (Lyme disease). Fungal causes are rare except in immunocompromised patients.

Pericarditis

Causes/microbiology - most commonly infection(viral- Coxsackievirus, echovirus, adenovirus, Epstein-Barr virus, influenza virus, and human immunodeficiency virus; Bacterial(Staphylococcus aureus and Haemophilus influenzae), Tuberculous pericarditis in immunocompromised, , autoimmune disease (juvenile idiopathic arthritis and systemic lupus erythematosus) , rheumatic fever, uremia, malignancy, as a reaction to a drug, or after cardiac surgery. Up to one third of cases have no identifiable cause and are considered idiopathic.

Clinical Feature: chest pain, which tends to be substernal, sharp, worse with inspiration, and relieved by sitting upright and leaning forward. The pain typically radiates to the scapular ridge due to irritation of the phrenic nerves. More specific symptoms of pericarditis depend on the cause. On physical examination, the pathognomonic finding is the pericardial friction rub, loudest when the patient is upright and leaning forward, the sound is heard best in the second to fourth intercostal spaces along the left sternal border or the midclavicular line.

Laboratory evaluation: elevations of the WBC, ESR, CRP, plasma troponin. To determine the specific cause, blood cultures, viral cultures, tuberculin skin testing, gastric cultures for Mycobacterium, and measurement of rheumatoid factor and antinuclear antibody may be helpful. EKG: diffuse ST elevation, PR depression.

Pathogenesis of pericarditis: In most cases of acute pericarditis, the pericardium is acutely inflamed and has an infiltration of polymorphonuclear (PMN) leukocytes and pericardial vascularization. Often, the pericardium manifests a fibrinous reaction with exudates and adhesions. The pericardium may develop a serous or hemorrhagic effusion. A granulomatous pericarditis occurs with tuberculosis, fungal infections, rheumatoid arthritis (RA), and sarcoidosis.

Treatment of pericarditis, including the importance of surgical drainage: Treat causes. Nonsteroidal anti-inflammatory drugs (NSAIDs) are the mainstay of therapy, and symptoms resolve within days for most patients. If chest pain persists beyond two weeks or recurrent pericarditis develops, colchicine can be added as another effective treatment. The use of corticosteroids in acute pericarditis remains controversial. Pericardiocentesis is indicated in idiopathic pericarditis with hemodynamic compromise, cardiac tamponade, purulent pericarditis, and suspected neoplastic pericarditis. In resistant cases, a pericardial window (surgical incision in the pericardium) or pericardiectomy (surgical removal of part or all of the pericardium) may be necessary.

Kawasaki disease (KD) (see also XXI.B.2.)

Use of echocardiography in the evaluation and management of patients with Kawasaki disease: Initial echocardiography should be performed as soon as the diagnosis is suspected, but treatment should not be delayed by the timing of the study. Echo should focus on maximal efforts to see all major coronary segment, most common sites of coronary aneurysms include the proximal left anterior descending coronary artery (LAD) and proximal right coronary artery (RCA). In addition, assessment of left ventricular function should be a part of the echocardiographic evaluation of all patients in whom KD is suspected because some degree of myocarditis is almost universal. Echocardiography should be performed at the time of diagnosis and subsequently usually at 2 and 6 weeks after disease onset.

Cardiac complications of Kawasaki disease and the timing of onset - Coronary artery aneurysms occur in 20% to 25% of untreated children. Other cardiovascular complications include myocarditis, pericarditis with effusion, and valvulitis, the latter occurring in about 1% of patients, most commonly involving the mitral valve. Resolution of aneurysms 1 to 2 years after disease onset has been observed in approximately 50% of vessels that have coronary aneurysms. Vessels in which

resolution does not occur are at risk for developing stenosis or occlusion by thrombus or myointimal proliferation. A coronary aneurysm can rupture within the first few months after KD, but this complication is exceedingly rare. The worst prognosis is noted in children who have giant aneurysms. Myocardial Infarction (MI

) caused by thrombotic occlusion of an abnormal coronary artery is the principal cause of death from KD. The greatest risk of MI occurs in the first year after disease onset.

Understand their prevention and treatment and the importance of follow-up evaluation:

Patients who have KD should be treated with IVIG (2 g/kg in a single infusion) together with high-dose aspirin. Whenever possible, this therapy should be started within the first 10 days of illness, with the first day of fever considered the first day of illness.

Most clinicians continue high-dose aspirin until day 14 of illness if the child remains afebrile. When high-dose aspirin is discontinued, therapy is continued with low-dose aspirin at 3 to 5 mg/kg per

day until the patient shows no evidence of coronary changes by 6 to 8 weeks after the onset of illness.

Patients who have small solitary aneurysms should take long-term aspirin therapy until the aneurysms regress and be followed by a pediatric cardiologist. Patients who have giant aneurysms or multiple complex aneurysms should be considered candidates for long-term antiplatelet therapy and anticoagulation therapy. The choice of regimen depends on the degree of coronary enlargement. Regimens include combinations of aspirin, clopidogrel, heparin, and warfarin; commonly combination of aspirin and warfarin, aiming for an International Normalized Ratio of 2.0 to 2.5.

The primary surgical management of KD is coronary artery bypass grafts for obstructive lesions. Catheter interventions such as balloon angioplasty, rotational ablation, and stent placement have been performed in a relatively small number of children who have KD.

Cardiac transplantation should be considered for individuals who have severe, irreversible myocardial dysfunction and coronary lesions for which interventional catheterization procedures or coronary bypass are not feasible.

Follow up with cardiologist is important to identify and prevent the grave cardiac consequences of the Kawasaki disease.

Rate and rhythm disorders, ischemia

Identify the clinical manifestations of common cardiac arrhythmias: In infant – pale, listless and feels limp, child – rapid heartbeat, palor, fatigue, weakness, dizziness, fainting, syncope and chest pain.

Understand the clinical significance of a prolonged corrected QT interval: QT interval represents the period of activation and recovery of the ventricular myocardium. When recovery is prolonged from electrical/excitation, some part of the myocardium might be refractory to subsequent depolarization. This physiologic state predisposes to circus reentrant rhythm and polymorphic ventricular tachycardia (torsade de pointes), leading to ventricular fibrillation and ineffective ventricular contraction. This abnormal rhythm may resolve spontaneously but also can progress to sudden death.

Electrocardiographic patterns:

Premature atrial contractions(PAC): premature P waves/ ectopic P wave (often called P') is often hidden in the ST-T wave of the preceding beat, irregular P-P interval.
Premature ventricular contractions (PVC): Wide QRS, unrelated to P wave, Supraventricular tachycardia (SVT): different types (sinoatrial, atrial, atrioventricular (junction); regular rhythm, usually normal QRS complex, tachycardia- HR > 140 to 220. P wave buried in previous T wave.
Atrial Flutter: P-wave saw-tooth appearance, The atrial rate is usually about 300/min, but may be as slow as 150-200/min or as fast as 400- 450/min. Ventricular rate usually 110/min.
Atrial Fibrillation: P wave is poorly defined, may seem coarse or fine baseline undulations (wiggles/flutter) or no atrial activity at all. Ventricular response (RR intervals) is irregularly irregular and may be fast (HR 100-160 bpm).

Ventricular tachycardia: regular rhythm, HR 180-190/min, wide QRS complexes, P wave not seen.

Treatment of supraventricular tachycardia:

Hemodynamically stable: Vagal maneuver (infant – ice on face , rectal thermometer; child- bearing down, valsalva maneuver) , Antiarrhythmic drugs: adenosine (1st line), if no response after repeated dose, 2nd line- amidarone, procainamide.
Hemodynamically unstable- synchronized cardioversion.
Systemic diseases affecting the heart

Hyperthyroidism should be considered in the evaluation of a patient with persistent sinus tachycardia.

Patients with Marfan syndrome may have associated cardiac disease(aortic root dilation, aortic aneurysm, dissection) that precludes participation in sports.

Cardiovascular conditions associated with Turner syndrome: Bicuspid aortic valve(most common 16%), coarctation of aorta (11%) , partial anomalous pulmonary venous connection(13%), persistent left superior vena cava, generalized dilation of the aorta and other major vessels, such as the brachial and carotid arteries also has been demonstrated.

Superior vena cava syndrome: Symptoms- Dyspnea is the most common symptom, followed by trunk or extremity swelling, facial swelling, cough, orthopnea, headache, nasal stuffiness, light-headedness, neurologic symptoms, such as dizziness and confusion, are late findings as cerebral edema occurs. Signs: facial or upper extremity edema, jugular venous distention, stridor if airway narrowing.

Importance of cardiovascular evaluation when there is a family history of hypertrophic cardiomyopathy, muscular dystrophy or Marfan syndrome: - Familial hypertrophic cardiomyopathy is inherited as an autosomal dominant trait and is attributed to mutations in one of a number of genes that encode for one of the sarcomere proteins, significant cause of sudden unexpected cardiac death in any age group. Marfan syndrome is a genetic disorder (autosomal dominant) of the connective tissue due to mutation in FBN1 gene, which leads to aortic dilation and may progress to dissection. Muscular dystrophy is a group of inherited disorders that involve muscle weakness and loss of muscle tissue, which get worse over time. Duchenne muscular dystrophy (DMD) is the most common childhood form of muscular dystrophy; it generally affects only boys becoming clinically evident when a child begins walking. DMD gene mutation affects dystrophin protein.

Importance of a family history of cardiovascular disease; hypertension, sudden death, MI in young age, familial hyperlipidemia and hypercholesterolemia in children. Evaluate appropriately - childhood hyperlipidemia is concerning as childhood obesity and the subsequent increasing risk of type 2 diabetes mellitus, hypertension, and cardiovascular disease in adolescents and young adults are becoming increasingly common.

Identify the cardiovascular risk factors in children, and evaluate appropriately: Risk factor: obesity, hypertension, diabetes mellitus, smoking, high cholesterol, family history of premature cardiovascular disease or high blood concentrations of cholesterol. Screening fasting lipid profile and evaluation for diabetes is appropriate.

Initial management of a child with a positive family history of hyperlipidemia: Rule out risk factors such as obesity, hypertension, diabetes mellitus, smoking, high

cholesterol in a child. Screening fasting lipid profile and evaluation for diabetes is appropriate. A healthy diet for all children older than 2 years that includes low-fat dairy products; For children between 12 months and 2 years of age for whom obesity is a concern or who have a family history of obesity, dyslipidemia, or cardiovascular disease, the use of reduced-fat milk is appropriate. For pediatric patients who are overweight or obese and have high triglyceride or low high-density lipoprotein cholesterol concentrations, weight management is the primary treatment, which includes improvement of diet with nutritional counseling and increased physical activity to produce improved energy balance. Pharmacologic intervention should be considered for patients 10 years and older who meet the criteria. Criteria for starting antilipemic drugs ---no risk factor: cut off LDL persistently > 190 mg/dl; other risk factors like smoking, obesity, hypertension, family history of premature cardiovascular problems: cut off LDL persistently > 160 mg/dl; for a child with diabetes mellitus : LDL persistently > 130 mg/dl is the lipid criterion.

Blood and Neoplastic Disorders

A.General aspects

Recognition by history

Jaundice, dark urine, and a sudden change in exercise tolerance may indicate a hemolytic anemia.

Recurrent bacterial infections are a manifestation of quantitative or qualitative leukocyte disorders.

Children with severe neutropenia may become infected with their own skin and bowel flora.

Children with a family history of hematologic disorders (e.g., hemophilia, bleeding complications, hemoglobinopathy, hemolytic disease) may also be at risk and require screening or evaluation.

Children with a family history of excessive cancers may also be at risk and require screening or evaluation.

Recognition by physical examination

Mucosal ulcerations can be a sign of neutropenia.

Distinguish between bruising due to thrombocytopenia and normal bruising in an active child: Patient with thrombocytopenia present with bruising with little or no trauma, nosebleeds, blood in the urine or stool, gum bleeding, bleeding with surgical or dental procedures, or excessive menstrual bleeding, petechiae on exam. In active child, bruising is usually seen over shin or below knee area.

Child abuse may be a cause of bruising in a child with a normal platelet count.

Vasculitic disorders may be a cause of bruising or purpura in a child with a normal (or increased) platelet count.

Thrombocytopenia or functional platelet disorders may cause bruising, petechiae, epistaxis, or gastrointestinal bleeding but rarely cause deep muscle or joint bleeding.

Palpable bruises or bruises in areas not exposed to trauma are distinctly abnormal. Differential diagnosis of a patient with a purpuric rash:

Disruptions in vascular integrity: Trauma, Infection, Henoch-Schönlein purpura, Drug-induced vasculitis, Neonatal purpura fulminans, Vitamin C deficiency, Ehlers- Danlos syndrome

Disorders of hemostasis: Platelet disorder – Thrombocytopenia, Immune/idiopathic thrombocytopenic purpura, Hemolytic uremic syndrome, Thrombotic thrombocytopenic purpura, Disseminated intravascular coagulation (DIC), Bone marrow infiltration, Neonatal alloimmune thrombocytopenia, Inherited thrombocytopenias, Drug induced thrombocytopenia, Bone marrow failure, Sequestration, Inherited platelet function abnormalities, Acquired platelet function abnormalities; Clotting factor deficiencies – Von Willebrand disease, Hemophilia, Other congenital factor deficiencies. Vitamin K deficiency, Liver disease

Children with hemihypertrophy and somatic overgrowth syndromes should be periodically evaluated for the development of associated embryonal tumors: Beckwith-Wiedemann syndrome (hemihypertrophy, macroglossia, umbilical hernia, omphalocele, ear lobe creases, neonatal hypoglycemia) – screening for neuroblastoma by periodic chest radiograph and urinary catecholamines, screening for Wilms tumor by renal ultrasound, screening for hepatoblastoma by alpha fetoprotein in first few years after birth.

Isolated hemihyperplasia – screening for Wilms tumor by renal ultrasound every 3 month until 8 years to detect Wilms tumor early before it become clinically apparent.

Klippel-Trenaunay-Weber syndrome (KTWS): presence of vascular malformation, high risk for hemihyperplasia due to increase vascular supply to affected body region caused by localized capillary-lymphatic-venous malformation. They are not associated with Wilms tumour or any embryonal tumor so no screening needed.

Interpretation of laboratory results

Physiologic anemia of infancy: Within the first week after delivery, hemoglobin and hematocrit values drop in response to the higher ambient oxygen concentration ex- utero. One theory is that this increase in oxygen concentration, combined with a rising percentage of adult hemoglobin, down-regulates production of erythropoietin in the neonate leading to a decrease in hemoglobin. The hemoglobin and hematocrit values continue to decrease until they reach the physiologic nadir, usually at 8 to 12 weeks after birth in the term neonate. Normal values for hemoglobin range from 9 to 11 g/dL. The nadir is termed “physiologic anemia of infancy,” although it is not truly anemia defined by a deviation from age-specific norms. Understand that further laboratory evaluation is unnecessary.

Iron deficiency and thalassemia minor are the most common causes of a microcytic anemia.

Reticulocyte count usually distinguishes between disorders of erythrocyte production and those of erythrocyte destruction.

Total leukocyte count and a leukocyte differential count are needed to diagnose neutropenia.

Neutropenia is usually defined as an absolute neutrophil count <1000/mm3. Bleeding time is a test of platelet and blood vessel function.

Thrombocytopenia is defined as a platelet count <150,000/mm3.

Bone marrow aspirate is necessary in the evaluation of a child with multiple pancytopenias.

Normal variation in hemoglobin concentration and mean corpuscular volume during childhood – The normal hemoglobin concentration for a term newborn is 19.3± 2.2 g/dL, values that continue to rise until they reach a maximum at about 2 hours after birth. After 1 week hemoglobin begins decreasing reaching the physiologic nadir at 8 to 12 weeks of age. Normal values for hemoglobin at this point range from 9 to 11 g/dl.

Normal developmental changes in MCV occur in children after 6 months of age. Lower limit of normal for MCV is 70 fl between 10 and 17 months of age and there is a gradual increase of MCV with age; the lower limit is 74 between 1.5 and 4 years and 76 between 4 and 7 years.

Erythrocyte disorders

Nutritional anemias

Iron deficiency

Iron deficiency causes non-hematologic effects such as behavior and learning disturbances.

Most common cause of iron deficiency anemia in young children is dietary deficiency.

Cow milk contains very little bio-available iron and an infant with iron deficiency often drinks large amounts of cow milk.

Look for bleeding as a cause of iron deficiency in the child with a normal diet.

Diagnosis of iron deficiency anemia: Low hemoglobin, low hematocrit, low MCV, low MCH, High RDW, elevated free erythrocyte protoporphyrin (normal RDW and free erythrocyte protoporphyrin usually present in thalassemia trait); Iron studies – low iron, low ferritin, normal CRP (since serum ferritin is an acute phase reactant, it can be falsely elevated in inflammatory states. A normal CRP is therefore necessary to ensure the serum ferritin level is truly reflective of the child's iron status), high TIBC, increased transferrin, low % iron saturation;

Peripheral smear – microcytosis, hypochromia, poikilocytosis, anisocytosis. Reticulocyte and hemoglobin concentration (CHr) also may be used to diagnose iron deficiency. Standard values for children have been determined, and a low CHr is a strong predictor of iron deficiency.

Treatment of iron deficiency with ferrous sulfate should continue after the hemoglobin concentration has returned to normal.

Intramuscular iron injections or erythrocyte infusions are not appropriate for the child with routine nutritional iron deficiency.

Vitamin B12, folic acid deficiency

Vitamin B12 or folate deficiency is a cause of macrocytic anemia.

Document the diagnosis of B12 or folate deficiency with specific measurement of serum B12 concentration, or serum or erythrocyte folic acid concentrations before beginning replacement therapy.

B12 deficiency may occur following small bowel resection or as a result of a maternal vegan diet in a child who is breast-fed exclusively.

Ingestion of fresh goat milk as a principal source of nutrition in infancy is a cause of folate deficiency.

Hemolytic anemias a.Membrane disorders

Jaundice and splenomegaly can be findings of hereditary spherocytosis.

Increasing pallor in a child with hereditary spherocytosis may be a sign of an aplastic crisis that warrants monitoring of the hemoglobin concentration and reticulocyte count.

Sudden increase in jaundice in a child with hereditary spherocytosis may be a sign of increasing hemolysis warranting monitoring of the hemoglobin concentration.

Child with hereditary spherocytosis or other erythrocyte membrane disorders should receive pneumococcal, meningococcal, and Hemophilus influenzae vaccines before splenectomy and prophylactic penicillin after splenectomy.

Parvovirus B19 is the most common cause of an aplastic crisis in patients with hereditary spherocytosis.

Enzyme abnormalities

G6PD deficiency is a common X-linked disorder, making the disorder much more common in males.

Sudden onset of pallor and anemia may be a manifestation of G6PD deficiency.

Causes of hemolysis in patients with G6PD deficiency, and manage appropriately: Medications, particularly sulfonamides, some antimalarial drugs (primaquine, chloroquine), and nitrofurantoin – avoid oxidative stress agents; Those of Asian descent commonly have a more pronounced deficiency and also should avoid aspirin, acetaminophen, and ciprofloxacin.

Viral and bacterial infection – treat infection.

Fava beans and Naphthalene balls – avoid offending agents.

Phototherapy and exchange transfusion are the treatment for severely jaundiced newborn.

It is estimated that 10% of the African-American population has G6PD deficiency.

Clinical differences in the form of G6PD deficiency seen in patients of different ethnic backgrounds: G6PD deficiency should be suspected in any newborn with pronounced jaundice and is of African, Asian, or Mediterranean heritage. Other clinical indicators in the neonatal period include a falling hematocrit in the patient who has hyperbilirubinemia (without ABO incompatibility) or a bilirubin level that is poorly responsive to phototherapy. A repeat bilirubin measurement after 6 hours of phototherapy may help identify these patients.

Hemoglobinopathies

Sickle cell disease can be diagnosed at birth, usually by newborn screen.

Children with sickle cell disease are particularly susceptible to death from overwhelming bacterial sepsis and require early evaluation and treatment when febrile.

Enlarged spleen and worsening anemia is indicative of a sequestration crisis in sickle cell disease.

Immediate intervention with intravenous fluids with or without blood is the treatment for acute sequestration crises.

Painful swelling of the hands and/or feet (dactylitis) is a manifestation of sickle cell disease in young children.

Use of prophylactic penicillin in children with sickle cell disease: usually

penicillin VK 125 mg po twice daily start by 3 month, continue until 3 years, then increase dose to 250 mg po bid at 3 years of age. After 5 years of age, discontinue penicillin VK.

Increasing pallor, decreased hemoglobin concentration, and a decreased reticulocyte count are findings in an aplastic crisis in sickle cell disease.

Acute chest syndrome and painful crises are common manifestations of sickle cell disease.

Thalassemia major usually presents as a severe hypochromic, microcytic anemia with enlargement of the liver and spleen.

Association of cholelithiasis in a patient with Sickle Cell Disease (SCD), Cholelithiasis is seen in 42% of sickle cell disease by adolescence. Although cholelithiasis is a common finding in patients who have SCD, cholecystectomy is indicated only in those suffering from abdominal pain or cholestatic jaundice due to common duct obstruction. Patients who present with cholecystitis should be treated with antibiotics and supportive care and be referred for elective cholecystectomy weeks after the acute event.

Priapism is a common manifestation of sickle cell disease. Presenting signs and diagnostic evaluation for suspected thalassemia:

Signs: Patients have microcytic, hypochromic anemia and hepatosplenomegaly, and also may have bony abnormalities resulting from marrow expansion, as well as cholelithiasis and icterus. Hydrops fetalis occurs when all four alpha-globin genes are defective or absent; Hemoglobin Bart is the predominant hemoglobin – affected fetus has profound anemia, hepatosplenomegaly, and anasarca, and usually is stillborn or dies shortly after birth.

Labs: CBC (low Hgb, low MCV, low MCH, low MCHC), Hemoglobin electrophoresis (alpha-thalassemia can be diagnosed by identification of Bart hemoglobin on newborn screening. b-Thalassemia is diagnosed when hemoglobin electrophoresis done in early childhood demonstrates elevated levels of hemoplobin A2).

A normal Red blood cell Distribution Width (RDW) and free erythrocyte protoporphyrin usually are present in thalassemia trait.

Peripheral smear: microcytic, hypochromic, anisocytosis, and poikilocytosis, with occasional macrocytes, target cells, basophilic stippling, and polychromasia.

Abnormal liver function tests (LFT) results indicate hemolysis and liver dysfunction.

Immune-mediated anemias

Pallor, jaundice, and splenomegaly are the signs of autoimmune hemolytic anemia in children, and manage appropriately.

Direct and indirect Coombs tests are a necessary part of the evaluation of a child with acute-onset anemia.

Complications of an erythrocyte transfusion in a child with autoimmune hemolytic anemia: Children who experience autoimmune hemolytic anemia (AIHA) can develop cardiac and respiratory compromise quickly from rapid destruction of RBCs.

Corticosteroids are useful in treating autoimmune hemolytic anemia (for warm- reactive antibodies) & plasmapheresis (for cold-reactive antibodies).

ABO incompatibility may cause anemia in a first-born child, but Rh incompatibility rarely does.

Progressive and severe anemia may occur at 4 to 8 weeks of age in infants with ABO or Rh incompatibility.

Aplastic and hypoplastic erythrocyte disorders

Diamond-Blackfan syndrome

Clinical characteristics of Diamond-Blackfan syndrome and transient erythroblastopenia of childhood:

Diamond-Blackfan syndrome – caused by insensitivity to erythropoietin, usually present in infant < 1 year of age with pallor and fatigue. Other features, such as facial abnormalities, thumb abnormalities and congenital heart disease, are present in up to 25% of affected patients. Labs: macrocytic anemia, reticulocytopenia, red cell adenosine deaminase (ADA) activity and I antigen expression are increased. Treatment includes steroids, red blood cell transfusion and bone marrow transplant.

Transient erythroblastopenia of childhood: Unknown causes that result in transient failure of the erythroid cell line. Possible etiology may be viral illness. Average affected age is 2 years. Physical examination includes marked pallor, tachycardia, tachypnea, fatigue. Labs: Normocytic anemia, red cell ADA activity is normal and I antigen expression can be normal or increased.

Transient erythroblastopenia of childhood

Signs, symptoms, and laboratory findings of transient erythroblastopenia of childhood: The average age of affected children is 2 years; rarely occurs in the first year after birth. Normal physical exam except marked pallor and other signs

of anemia, such as tachycardia, tachypnea, fatigue; Labs: Normocytic anemia, reticulocytopenia, red cell ADA activity is normal, and I antigen expression can be normal or increased.

Role of erythrocyte transfusions in transient erythroblastopenia of childhood: RBC transfusion in those who have Hgb < 5 mg/dl, those who have Hgb > 5 mg/dl recover without transfusion. Prognosis: excellent.

Therapeutic approaches

Risk of transmitting infectious diseases during blood transfusion(s): Although transmission of HIV, hepatitis B virus (HBV), or hepatitis C virus (HCV) remains a primary concern of many patients and parents, advances in donor screening and in the testing of donated blood over the past 2 decades have lessened significantly the risks of acquiring these infections via transfusion. One recent estimate of the risk of HIV transmission was 1 in 2,135,000, for HCV was 1 in 1,935,000, and for HBV was 1 in 205,000; these risks are expected to decrease further with advancement of nucleic acid amplification testing (NAT) screening. Thirty cases of transfusion associated West Nile virus

(WNV) infection were reported in the United States between 2002 and 2004. Variant Creutzfeld-Jacob disease (vCJD) has not been reported in the United States. CMV is transmitted via transfusion from a CMV- positive donor to a CMV-negative recipient (or reactivated in a CMV-positive recipient).

Erythrocyte transfusions may be associated with hemolytic, febrile, and urticarial reactions.

Role of erythrocyte transfusions in the management of anemia: Blood transfusions are used frequently in the management of surgical or traumatic hemorrhage, in the care of preterm (anemia of prematurity) or sick newborns, and in the management of a variety of congenital and acquired anemias. Children who have inherited bone marrow failure syndromes such as Fanconi anemia, dyskeratosis congenita, and Pearson syndrome frequently require regular RBC and platelet transfusions, as do children afflicted with severe acquired aplastic anemia. Hemoglobinopathies, such as sickle cell disease and its complications need frequent packed red blood cell (PRBC) transfusion.

Erythropoietin is useful in management of anemia of renal failure and chronic inflammatory disease. Use of recombinant human erythropoietin (rHuEPO) has been shown to reduce the number of transfusions needed by very low birth-weight (VLBW) infants, but not the number of donor exposures.

Leukocyte disorders

Quantitative leukocyte disorders

A.Congenital and immune-mediated neutropenia

Congenital neutropenia may be persistent or cyclical. Cyclic neutropenia is characterized by approximately 21-day cycles of changing neutrophil counts, with neutropenia spanning 3 to 6 days. The nadir of the neutrophil count may be in the severe range. Fever and oral ulcerations usually are seen during the nadir. Patients also may develop gingivitis, pharyngitis, and skin infections. However, by the time the patient comes to medical attention, the neutrophil count may be recovering. Therefore, diagnosing cyclic neutropenia may require obtaining blood counts two to three times per week for 4 to 6 weeks in an effort to observe the periodicity of the cycle. Severe congenital neutropenia may be inherited as an autosomal recessive condition (Kostmann syndrome) involving mutations in the HAX1 gene.

b.Acquired, non-immune neutropenia (1). Sepsis

Neutropenia is a sign of overwhelming bacterial sepsis.

(2). Drugs

Neutropenia may be drug-induced and, if so, should lead to discontinuation of the drug.

Common viral infections Ebstein Barr virus (EBV), parvovirus, Human Herpes virus-6 (HHV6) and other viruses may cause transient neutropenia that does not require specific treatment.

Qualitative leukocyte disorders

Child with recurrent bacterial infections and a normal neutrophil count may have abnormal neutrophil function.

Clinical signs of abnormal leukocyte function include periodontal disease, perirectal ulceration, delayed umbilical cord separation. .

Therapeutic approaches

Role of growth factors in the treatment of neutropenia: granulocyte colony stimulating factor (GCSF) promotes expansion of the neutrophil pool by stimulating the precursors' entry into the cell-cycle and inhibiting apoptosis. In addition, GCSF directly increases neutrophil chemotaxis and microbicidal ability. GCSF can reduce infection related morbidity and mortality for patients who have cyclic neutropenia and recurrent severe infections, severe congenital neutropenia, or bone marrow failure syndromes.

Platelet disorders

Quantitative platelet disorders
Decreased platelet production

Thrombocytopenia in a newborn infant may be a sign of bacterial sepsis and, in an ill child, should lead to appropriate culture and antibiotic therapy.

History of medications should be part of the evaluation of a child with thrombocytopenia.

Presence of thrombocytopenia in a newborn infant with microcephaly or other congenital abnormalities may be due to a congenital viral infection such as CMV.

Increased platelet destruction (1). Congenital

Thrombocytopenia, eczematoid rash and recurrent infections (low IgM, elevated IgA & IgE) are signs of Wiskott-Aldrich syndrome.

A rapidly enlarging hemangioma should lead to a platelet count to check for thrombocytopenia.

Manage the thrombocytopenia associated with TAR syndrome (Thrombocytopenia Absent Radius syndrome): Thrombocytopenia is associated with a hyporegenerative marrow that is unresponsive to corticosteroids and IVIG. Avoid trauma and elective surgery, keep platelet count > 15000/mcl using platelet transfusion. Thrombocytopenia is self- limited and normal platelet counts are established by 2 years.

(2). Acquired

Idiopathic Thrombocytopenic Purpura (ITP) is characterized by a low platelet count and normal or increased platelet production in the bone marrow.

Most children with ITP will recover in less than one year without treatment. Most common presenting symptom of ITP is increased bruising.

Corticosteroids and IVIG usually increase the platelet count in children with ITP but do not alter the natural course (i.e. length of disease).

Aspirin or other drugs that interfere with platelet function should not be given to children with ITP or other quantitative or qualitative platelet disorders.

Persistent or severe headache is a symptom of intracranial hemorrhage in ITP.

Splenectomy is not an appropriate therapy at the onset of ITP in a child who is not having major bleeding problems.

A mother with ITP (Maternal Idiopathic Thrombocytopenic Purpura) may have an infant with thrombocytopenia, and management of the infant is

usually unnecessary; transfusion of IVIG if platelet count <10,000 to 20,000/mcl.

Multiple siblings with neonatal thrombocytopenia suggestisoimmune thrombocytopenia.

Thrombocytopenia due to maternal ITP or isoimmune thrombocytopenia usually resolves within 6 to 12 weeks.

Appropriate management of a patient with ITP: Most children with ITP will recover in less than one year without treatment.

Neonatal Alloimmune Thrombocytopenia – treat with maternal platelets or IVIG if platelet count <20 to 30,000/mcl.

Thrombocytopenia due to maternal ITP – treatment usually unnecessary; IVIG if platelet count <10,000 to 20,000/mcl.

Acute ITP – avoidance of aspirin and ibuprofen; avoidance of contact sports and limitation of activities, if possible, and most especially, reassurance of the parents regarding the child's recovery and the rarity of serious or life-threatening hemorrhage. No treatment alters the natural history of acute ITP. However, several drugs are effective in raising the platelet count temporarily by blocking the destruction of antibody-coated platelets by mononuclear phagocyte. The three treatments employed most commonly are prednisone or other corticosteroids, IVIG, and anti-D immunoglobulin (WinRho). The easiest and least expensive of these agents is oral prednisone at a dose of 1 to 2 mg/kg per day.

Chronic ITP – The only proven "curative" treatment for chronic ITP is splenectomy.

Qualitative platelet disorders

Hereditary platelet function disorders can be divided into five groups: Disorders of platelet adhesion (e.g. Bernard-Soulier syndrome):

Rare. Caused by a deficiency on the surface of the platelet in an area called Glycoprotein 1b/1X leading to failure of platelets to stick and clump together. First noticed during childhood. Presents as nosebleeds, bleedings in mouth and gums, and occasionally gastrointestinal bleeding. Females may experience heavy menstrual bleeding.

Disorders of platelet aggregation:

This rare disorder is called Glanzmann Throinbasthemia. It can be life-threatening. It is caused by a deficiency of protein on the surface of the platelet, called Glycoprotein 11b/111a. Platelets fail to form a plug at the site of an injury. Presentation is similar to Bernard Soulier syndrome.

Disorders of platelet secretion:

Platelets fail to secret granules which slows down platelet adhesion, aggregation and repair of blood vessels following injury.

Disorders of platelet Procoagulant activity:

Rare disorder, called Scott syndrome. Bleeding time and aggregation are normal. Platelets fail to promote activation of the blood clotting proteins.

Combined abnormalities of number and function:

Some hereditary disorders of platelet production show low platelets and platelets of abnormal size. For example:

May-Hegglin Anomaly Alport Syndrome

Wiskott-Aldrich Syndrome Acquired platelet function disorders:

Platelet function can be affected by common drugs: Salicylate, Non- steroidal Anti-inflammatory drugs (NSAIDS), ticlopidine, etc.

Some medication conditions can cause abnormal platelet function:

- Chronic kidney disease
- Heart bypass surgery
- Some forms of leukemia

Therapeutic approaches

- Pancytopenia
- Decreased production
- Congenital (Fanconi anemia)
- Acquired aplastic anemia

Aplastic anemia and childhood leukemia may both present with purpura, pallor, and fever.

Absence of blasts in the peripheral blood of a patient with pancytopenia does not rule out the diagnosis of leukemia.

Increased destruction

Pancytopenia may result from autoimmune destruction of erythrocytes, leukocytes, and platelets (Evans syndrome). Management includes:

First-line therapy is usually corticosteroids and/or intravenous immunoglobulin (IVIG) – most patients respond; however, relapse is frequent.

Second-line therapy includes immunosuppressive drugs like ciclosporin or mycophenolate, vincristine, danazol or a combination of these agents.

A small number of patients have been treated with rituximab, which induces remission in the majority although the long-term effects in children are unclear.

Splenectomy may also be considered although long-term remissions are less frequent than in uncomplicated ITP.

For very severe and refractory cases, stem cell transplantation (SCT) offers the only chance of long-term cure.

Therapeutic approaches

Coagulation disorders
Congenital bleeding and thrombotic disorders

Excessive bleeding after circumcision may be the first sign of a congenital coagulation factor deficiency.

Prothrombin time and partial thromboplastin time are the important parts of the evaluation of a patient with increased bruising.

Child born to the daughter of a person with hemophilia should be tested for that particular bleeding disorder as hemophilia is an X-linked disorder.

For a woman who is a carrier of hemophilia, there is a 50% chance that a male offspring will have that bleeding disorder.

Headache is an important symptom of intracranial bleeding and requires early assessment and treatment.

Serious head trauma in a person with hemophilia requires careful assessment and early replacement therapy even in the absence of neurologic abnormalities.

Bleeding in the forearm of a person with hemophilia is an emergency because of the danger of nerve compression.

Femoral or jugular venipunctures should be avoided in the person with hemophilia who has not received replacement treatment.

Some children with hemophilia have a negative family history for bleeding disorders. Only two thirds of patients who have hemophilia have a positive family history of the condition. Therefore, a high degree of suspicion is required, even with a negative family history.

Partial thromboplastin time (PTT) is often normal in patients with von Willebrand disease (VWD), but the bleeding time is commonly prolonged.

First manifestation of von Willebrand disease in girls may be heavy menstrual bleeding.

Strong family history of pulmonary emboli or deep vein thrombosis may suggest a congenital hypercoagulable disorder.

Desmopression (DDAVP) is useful in the treatment of a patient with hemophilia or von Willebrand disease.

Von Willebrand disease is a common inherited bleeding disorder: VWD is diagnosed most often if the patient satisfies three criteria:

a positive bleeding history,
Reduced levels of Van Willebrand Factor (VWF) activity (ristocetin cofactor [RCo] activity), and 3. a positive family history suggestive of vWD.

The VWF:RCo assay is the most useful tool for screening, but other assays are required to distinguish among the various subtypes. These tests include VWF antigen, FVIII activity, VWF multimer analysis, ristocetin-induced platelet aggregation at low concentrations of ristocetin (seen with Type 2B), and blood typing (Type 1, 2A, 2B, 2N, 2M, 3).

Management: Two primary options include desmopressin (DDAVP) and transfusion therapy with plasma-derived vWF products.

DDAVP is the treatment of choice for most patients who have type 1 VWD and a subset of patients who have types 2A and 2M VWD. Patients who have the more severe type 3 and in most patients who have type 2 disease, DDAVP is ineffective or is contraindicated, necessitating the use of plasma concentrates containing both FVIII and VWF.

Acquired bleeding and thrombotic disorders

Measure prothrombin time, partial thromboplastin time, and platelet count as part of the evaluation for disseminated intravascular coagulation in a child with sepsis and purpura.

Purpura is an indication of bacterial sepsis in a febrile child.

Therapeutic approaches

Role of coagulation factor replacement for hemophilia: most pediatric patients in the developed world now are treated with recombinant-derived Factor-VIII (FVIII) and Factor-IX (FIX) concentrates.

Management of coagulopathy due to hepatic dysfunction should include replacement of coagulation factors with fresh frozen plasma (FFP) or cryoprecipitate, vitamin K replacement, and platelet transfusions.

Role of thrombolysis and anticoagulation in the management of thrombotic disorders: Anti-coagulants:

Heparin (Binds to antithrombin III and accelerates inhibition of thrombin and factor Xa), LMWH (Inhibits factor Xa)

Warfarin (Reduces the concentrations of vitamin K-dependent factors: II, VII, IX, and X)

Thrombolytic agents:

Tissue plasminogen activator (TPA) - promotes clot breakdown by activating plasminogen to plasmin, thus tipping hemostatic balance in favor of increased fibrinolysis.

Neoplastic disorders

Hematologic malignancies
Leukemias

Bone pain can be a symptom of leukemia.

Most patients with acute lymphoblastic leukemia will be cured of their disease using current treatment strategies.

Central nervous system and testes are the important sites of relapse of acute lymphoblastic leukemia (ALL).

Down syndrome is a disease with an increased risk of leukemia.

Different childhood leukemias (ALL, AML, chronic leukemias) have distinct therapies and outcomes:

Acute lymphocytic leukemia (ALL) accounts for 3 of 4 cases of leukemia in children. Treatment is divided into 4 categories (usually last 30 to 36 months): Induction (vincristine, glucocorticoids, L-asparaginase and the 4th agent, depending on initial risk, is doxorubicin); Consolidation for 6 to 12 months (cytarabine, methotrexate, cyclophosphamide, anthracyclines & epipodophylotoxins); Maintenance therapy for 18-24 months(oral vincrisitine and weekly methotrexate with intermittent pulses of vincristine and glucocorticoid, Central Nervous System (CNS) prophylaxis. Sseries of lumbar puncture with single agent intrathecal methotrexate or triple agent chemotherapy is given to all regardless of initial CNS finding. Five-year survival rates for children receiving new diagnoses of ALL in the United

States and other developed countries are now in excess of 80% for standard risk and 60-65% survival rate at 4 years for high risk patient.

Acute myelogenous leukemia (AML) accounts for 20% of leukemia in children. Treatment: Induction with anthracyclines and Ara-C requires 6 week or longer intense therapy, CNS prophylaxis. After remission, most children with HLA-matched donor should undergo stem cell transplantation, which can result in 70% cure rate. Exception- AML M3 (promyelocytic leukemia) – should receive retinoic acid with chemotherapy and not undergo transplant. Also children with Down syndrome do well with chemotherapy alone. For those who response to chemotherapy followed by bone marrow transplant, prognosis is good. For those who don't respond to chemotherapy and relapse, the prognosis is very poor.

Chronic leukemias are more common in adults than in children, and although they tend to grow more slowly than acute leukemias, they are harder to treat. These chronic leukemias are divided into two types: chronic myelogenous leukemia (CML) and chronic lymphocytic leukemia (CLL). CML is rare in children, but does occur and is treatable in children the same as in adults. CL. Allogenic BM transplant during the 1st year of chronic phase is the best treatment and result in cure rates as high as 80-90%.

Lymphomas

A palpable lymph node in the supraclavicular fossa is worrisome and should prompt a thorough evaluation for a cause, with malignancy high in the differential diagnosis, creating a low threshold for excisional biopsy.

Chest radiograph is an important part of the initial evaluation of the patient with an unexplained lymphadenopathy.

Overwhelming sepsis is a serious complication in patients with Hodgkin disease who have undergone splenectomy, and know that such patients should be evaluated thoroughly if fever develops.

Renal function and serum electrolytes should be assessed in patients who have suspected leukemia or lymphoma to exclude tumor lysis syndrome.

Solid tumors

Neuroblastoma

Neuroblastoma usually presents as a non-tender abdominal mass.

Urinary catecholamine excretion is increased in most patients with a neuroblastoma and tests of urine for VMA and HVA are appropriate screening tests for the tumor.

Wilms tumor

Wilms tumor is associated with hemihypertrophy and aniridia, somatic overgrowth, and/or genitourinary abnormalities

Wilms tumor usually presents as an abdominal mass and may cause hypertension and/or hematuria.

Prognosis of Wilms tumor: Between 85% and 90% of children diagnosed with Wilms tumor are cured and become long-term survivors.

Brain tumors

Signs and symptoms of craniopharyngioma: most common intracranial tumors of non-glial origin in children constituting 6% to 9% of pediatric brain tumors. The peak incidence of presentation is 5 to 10 years of age, but these lesions can manifest at any age, including infancy and the neonatal period. When confined to the sella turcica, clinical manifestations result from pituitary- hypothalamic involvement; Short stature due to growth hormone deficiency and visual field defects due to pressure on the optic chiasm are typical complications. Other manifestations of pituitary dysfunction may occur, including diabetes insipidus, hypothyroidism, and precocious puberty. If the tumor extends through the diaphragma sella, it may compress the third ventricle, causing hydrocephalus and clinical features of increased intracranial pressure (ICP), such as headaches, vomiting, and papilledema. Alterations of personality are frequent symptoms of both supra- and infratentorial tumors. Lethargy, hyperactivity, irritability, and forgetfulness may result from increased ICP, the focal lesion, or both.

Presenting signs of brain tumor include early morning headaches, deteriorating school performance, ataxia, emesis, vision changes, nystagmus.

Bone and soft tissue tumors

Presenting symptom of osteosarcoma is usually bone pain or swelling.

Clinical and laboratory features of osteoid osteoma: usual age 5 to 20 years old. The most common sites of involvement are the proximal femur and tibia, but any bone can be involved, including the posterior elements of the spine. Pain is the symptom that causes the patient to seek care. Classically presents with gradually increasing sharp focal pain that is worse at the end of the day or at night. Pain from osteoid osteomas located in bones of the leg usually is not related to activity, which helps differentiate this condition from a stress fracture, and the pain responds dramatically to NSAIDs, including aspirin. Constitutional symptoms usually are absent. Physical examination may reveal tenderness to touch or pressure. Patients also may have disuse atrophy, painful scoliosis, or limb-length discrepancy. Laboratory findings: a CBC, ESR, and

radiographs. A radiograph of the normal side is recommended for comparison. It can appear as a small spherical or oval lytic lesion surrounded by soft-tissue edema and reactive bone formation on plain radiographs. CT is used for precise localization of the lesion and as a guide for percutaneous biopsy and ablation. A biopsy confirms the diagnosis.

Ewing sarcoma and osteosarcoma are the most common malignant bone tumors in children, and both may metastasize to the lungs.

Biopsy of a malignant bone tumor should be done at a pediatric oncology center with attention to subsequent safe tumor resection.

Histiocytosis syndromes of childhood

Clinical manifestations of the histiocytosis syndromes of childhood: In Langerhans cell histiocytosis (LCH), the most common form of histiocytosis, the presentation varies. Lytic bone lesions, which can involve the skull, vertebral bodies, ribs, scapula, and femur, occur in approximately 80% of patients. Most of these bone lesions are asymptomatic or can present as raised, soft, tender areas. Lesions in the periorbital region may cause proptosis. Skin involvement can be generalized or localized, manifesting as seborrheic dermatitis of the scalp, purpuric or necrotic lesions, or diffuse candidal diaper dermatitis. Patients also can have gingival hypertrophy; ulcers of the palate, buccal mucosa, tongue, or lips; chronic otitis externa; and pancytopenia. Localized or disseminated lymphadenopathy is seen in 33% of patients. Approximately 20% of patients have hepatosplenomegaly, with increased liver enzyme values, hypoalbuminemia, hyperbilirubinemia, clotting factor deficiencies, and hypersplenism. Pulmonary infiltrates, diffuse fibrosis, and disseminated nodular infiltrates can be seen on imaging studies. Encroachment on the pituitary gland, especially the posterior fossa (reported in 5% to 50% of patients) causes endocrine abnormalities, such as growth retardation and diabetes insipidus. CNS findings due to infiltration by the proliferating Langerhans cells or pressure on surrounding structures include seizures, nystagmus, paresis, ataxia, and headache.

Other tumors (eg, germ cell, liver, retinoblastoma)

Serum alpha-fetoprotein and beta-human chorionic gonadotropin may be markers of germ cell tumor and hepatoblastoma.

Hereditary retinoblastoma frequently involves both eyes and presents at a younger age than sporadic retinoblastoma.

Genetics of retinoblastoma and the importance of frequent ocular examinations in children with a positive family history: Bilateral

retinoblastoma is much more likely to result from an inherited mutation, and patients who have bilateral retinoblastoma have a greater chance of having affected children than patients who have unilateral retinoblastoma and no family history. Subsequently, the gene for retinoblastoma (RB1) was identified on the long arm of chromosome 13 at band 13q14. Although some patients who have retinoblastoma have a cytogenetically visible deletion of this area of one of their two copies of chromosome 13, a smaller deletion or even a point mutation of the RB1 gene is more common.

Retinoblastoma can occur sporadically (with no family history of affected relatives) or as an autosomal dominant trait with incomplete penetrance. About nine in ten cases of retinoblastoma occur in children who have no family history of affected relatives; these children are more likely to have unilateral disease (60%) than bilateral disease. When there is a family history, children are more likely to have bilateral disease (85%). Although it is usually one of the parents who is affected, it is possible that neither is affected but instead a more distant relative.

Children who have unilateral retinoblastoma need to be followed for the development of tumor in the other eye. All children who have a family history of retinoblastoma, particularly involving a parent or sibling, need to be evaluated prospectively by an ophthalmologist from early infancy to detect any signs of the disease (leukocoria).

Children found to have the RB1 germline should have an ophthalmologic examination every 3 to 4 weeks until 1 year of age and continued close follow-up until 3 years of age. The family also should be referred for genetic counseling and possible molecular testing.

Oncologic emergencies

Spinal cord compression

Differentiate the clinical manifestations of spinal cord compression (eg, from a tumor) from those of other myelopathies, and evaluate appropriately: For a patient presenting with neurologic symptoms attributable to the spinal cord (lower extremity weakness, bowel and bladder dysfunction, abnormal deep tendon or plantar reflexes), the cause can be either compressive or non-compressive. Although the timing of the appearance of signs and symptoms and accompanying features, such as a preceding viral illness, can suggest one cause over another, there is enough clinical overlap that a compressive lesion cannot be excluded reliably without neuroimaging. Delaying potentially definitive therapy (laminectomy, radiotherapy, chemotherapy) for compressive lesions contributes directly to adverse neurologic outcomes. Transverse Myelitis (TM) frequently is difficult to distinguish from Guillain-Barre´ syndrome (GBS), which classically presents as an acute, ascending flaccid paralysis following a viral infection.

However, TM is more likely to present as pure lower extremity paraplegia or paraparesis, whereas GBS more commonly causes a gradient of weakness affecting the lower extremities more severely than the upper extremities. Patients who have TM generally have brisk deep tendon reflexes, whereas reflexes typically are absent in GBS. Bladder involvement is common with TM but uncommon with GBS, and cranial nerve VII (facial nerve) involvement is common with GBS. MRI demonstrates spinal cord inflammation in TM but not in GBS. Finally, lumbar puncture demonstrates a lymphocytic pleocytosis in TM, whereas typically there are few cells and a high protein concentration (albuminocytologic dissociation) in GBS.

Evaluation: Once compression is excluded, lumbar puncture should be performed and CSF sent for routine studies, as well as IgG index, oligoclonal bands, viral polymerase chain reaction studies, Lyme antibodies, Mycoplasma antibodies, and venereal disease research laboratory (VDRL) testing for syphilis, as indicated. Serologic evaluation can include HIV studies, Lyme titers, Mycoplasma titers, hepatitis serology, VDRL, and an evaluation for underlying autoimmune disease, as indicated by history and physical examination. Electromyography and evoked potentials may demonstrate the extent of neural injury and provide prognostic information.

There is a need for immediate evaluation of children complaining of back pain, lower extremity weakness, and/or bowel and bladder dysfunction to evaluate for spinal cord compression.

Mediastinal mass

Infection and sepsis

Varicella is a life-threatening illness in a patient receiving chemotherapy, and varicella-zoster immune globulin should be given immediately after exposure to varicella.

Immediate evaluation of a febrile child who is neutropenic as a result of chemotherapy is required.

Wheezing, positional dyspnea, and a chest mass as an indication for immediate evaluation and management due to the risk of acute respiratory failure.

Children receiving chemotherapy require prophylaxis against Pneumocystis jiroveci (carinii) pneumonia such as bactrim or pentamidine.

Therapeutic considerations

Live-virus vaccines should not be given during chemotherapy.

Gonadal effects associated with the use of chemotherapeutic drugs:

Chemotherapy, particularly the alkylating agents, is known to cause gonadal toxicity. Damage depends on agent, regimen and total dosage. Patient's sex and gonadal development during therapy also influence the toxicity. The germ cells of postpubertal males generally are more vulnerable than are those of prepubertal males.

Cranial irradiation can lead to growth hormone deficiency as late effect. Monitor height and weight and measure Growth Hormone (GH),Insulin-like growth factor 1 (IGF-1), and bone age for child who is poorly growing.

Late sequelae of cancer and cancer therapy:

Late effect of radiation therapy:

Late Effect of chemotherapy:

Most vaccinations (even inactivated) are withheld during chemotherapy since adequate immune response is compromised.

Renal Disorders

General
Normal function

Neonates have a glomerular filtration rate of 20-30mL/min/1.73m2 due to immature nephrons, which increases until 2 years of age, after which adult GFR numbers can be used. Creatinine generation increases from 0.3-0.4mg/dL for young infants to 1.0 in adolescent females and 1.3 in adolescent males. The Schwartz formula can be used to estimate GFR in children ages 1 to 16 based on creatinine, height and age.

kinfant <1 year=0.45 kchild=0.55 kadolescent male=0.70

24 hour urine collections are difficult to perform in young patients who may not be toilet-trained, may still have physiologic daytime or nighttime incontinence or may not consistently remember to void in the collection container.

Proteinuria

A patient with proteinuria on random sample should have a first morning urine specimen tested for protein to exclude orthostatic proteinuria, and only if it is positive should further evaluation of renal function, serum albumin, and quantitative urine protein excretion testing be done.

Fever and exercise can cause transient proteinuria.

If urine is alkaline, dipstick testing may cause a false-positive result for proteinuria.

Hematuria

Differential diagnosis for child with gross hematuria:

Tumor (bladder or renal)
Nephrolithiasis
Postinfectious glomerulonephritis
Hemolytic uremic syndrome
IgA nephropathy (synpharyngitic hematuria)
Sickle cell disease
Alport syndrome
Pulmonary-renal syndrome (Wegener's granulomatosis, microscopic polyangiitis, SLE, Goodpasture's)
Urinary tract infection
Trauma
Coagulopathy
Hypercalciuria
Membranoproliferative glomerulonephritis
Benign familial hematuria
Shistosomiasis

Persistent microscopic hematuria

Initial workup for microscopic hematuria: blood pressure, urine microscopy (for white blood cells, red blood cell morphology, crystals and casts), and urine culture should be performed.

Next step: serum electrolytes, blood urea nitrogen, creatinine can be measured if renal dysfunction is suspected. If glomerulonephritis is suspected, antistreptococcal antibody panel (including antistreptolysin O and anti-DNAse B) and C3 can be done, as well as other antibodies for infectious and autoimmune causes. Hemoglobin electrophoresis may also be considered. If structural abnormalities or nephrolithiasis are suspected, renal ultrasound, abdominal radiographs or abdominal spiral helical CT scan can be performed.

If nonglomerular causes are excluded or suspicion for glomerular disease is high:

Renal biopsy may be necessary.

Microscopic hematuria with proteinuria which persists indicates higher likelihood of renal disease, especially glomerulonephritis. Isolated hematuria generally suggests lack of significant renal disease and therefore generally better prognosis. Isolated hematuria may be due to benign familial hematuria or hypercalciuria (idiopathic or due to immobilization, diuretic use, intoxication with vitamin D, hyperparathyroidism or sarcoidosis).

Inquire about family history in a patient with persistent microscopic hematuria, as family members with chronic kidney disease may lead to diagnosis of hereditary nephritis, such as Alport syndrome.

Abnormalities in urinary tract structure, such as trauma, mass lesions or urinary tract stones should be ruled out in the patient presenting with gross hematuria.

Urinalysis can falsely demonstrate hematuria in a patient with myoglobin, but no blood, in the urine.

Causes of gross and microscopic hematuria

The patient with sickle cell trait or disease who presents with hematuria should have urinalysis with microscopy done to look for proteinuria or the sloughed papillae from papillary necrosis. Delayed hemolytic transfusion reactions and renal cell carcinomas (with abdominal or flank pain or weight loss) are other differentials to keep in mind.

One cause of microscopic hematuria is hypercalciuria, and random urine calcium- to-creatinine ratio is an initial screening test.

Nonhematogenous etiology of red urine

Some causes of red urine other than hematuria, which can be identified by history include:

Foods: beets, blackberries, rhubarb, paprika, coloring Toxins: lead, benzene

Drugs: sulfonamides, nitrofurantoin, salicylates, phenazopyridine, phenolphthalein

Urate crystals

Microscopic urinalysis showing 5 or more RBCs per high power field in 3 fresh, centrifuged specimens acquired consecutively over several weeks confirms hematuria.

Dysuria

In different age groups, the etiology of dysuria may be different. Besides urinary tract infections (UTIs), other etiologies include urethritis, nephrolithiasis, dysfunctional voiding, vaginitis, cervicitis, pelvic inflammatory disease, balanitis, inflammatory diseases, trauma, pinworms, exposure to irritants and a variety of skin conditions.

Obtaining an accurate sexual history, including risks for sexually transmitted infections and sexual assault, is essential in trying to determine the cause for abdominal pain and dysuria in an adolescent.

Perineal inspection in girls with dysuria can show irritation, rashes, pigment changes and signs of trauma.

Incontinence

Nocturnal

Enuresis in some children may be a result of a functional reduction in bladder capacity or bladder contractions which are not inhibited.

Risk of underlying serious disease in the child with secondary nocturnal enuresis is generally low.

Organic

Incontinence in females can be caused by an abnormally placed urethral opening.

Some of the causes of nocturnal incontinence Urinary tract infection

Urinary tract anomaly Lumbosacral anomaly Diabetes

Epilepsy

Obstructive sleep apnea

Examining the sacral area for dimples or hair tufts is an important etiological factor in the patient with nocturnal incontinence.

Functional, daytime incontinence

In the child with daytime incontinence evaluate with:

History about voiding: age when toilet trained, number of daily voids, pattern and volume of wetting, number of times per day child wets, whether child has been dry before, if there is a certain associated time or activity, urinary tract infection (UTI) history, nighttime wetting, presence of abuse, attitudes about wetting and other psychosocial factors.

History about bowel function: number of weekly stools, stool consistency, encopresis, abdominal pain.

Physical examination for:

Genitourinary (GU) abnormalities: hypospadias, meatal stenosis, phimosis, female epispadias, labial adhesions, intralabial masses, ureterocele or ectopic ureter

Back/sacral abnormalities: asymmetric gluteal crease, sacral dimple, hairy patch or vascular malformation

Digital rectal examination for tone and fecal material Perineal skin maceration

Other investigations: urinalysis, urine culture, bladder ultrasound for post-void residual, bladder wall thickening/trabeculation (sign of detrusor over-contraction), uroflow testing.

Females with constant dribbling should have magnetic resonance urography to look for ectopic ureter.

When tethered cord is suspected, MRI of lumbosacral spinal cord is indicated. Management:

- Voiding every 2 hours
- Double voiding (urinating a second time 10-15 minutes after the first)
- Avoid products known to irritate bladder (caffeine, carbonated beverages, artificial red dyes, beverages with high citrus content)
- Sit on toilet 30 minutes after large meal for 10 minutes with feet supported
- High fiber foods and/or polyethylene glycol for constipation
- Emptying problems, detrusor sphincter dyssynergia: biofeedback (pelvic muscle physical therapy)
- Bladder storage problem with frequency, suprapubic or penile pain

Anticholinergic medications (oxybutynin, tolterodine)

Wiggle incontinence, methylphenidate One factor in urinary incontinence is constipation.

Infrequent voiding may contribute to urinary incontinence.

Voiding dysfunction

Children with urinary frequency but negative cultures may have voiding dysfunction, either pollakiuria (often triggered by psychosocial stressors, presenting as many small daytime voids) or unstable bladder (frequent daytime and nighttime enuresis).

Congenital

Renal dysplasia
Unilateral multicystic dysplastic kidney

Often multicystic dysplastic kidney will present in infancy as a unilateral flank mass.

Autosomal-Dominant Polycystic Kidney Disease

Children with ADPKD can have elevated systemic blood pressure.

Intracranial aneurysms are a rare but significant associated finding in patients with ADPKD.

For the patient suspected of having ADPKD, abdominal ultrasound should be performed for diagnosis.

Autosomal-recessive polycystic kidney disease

Autosomal-recessive polycystic kidney disease either presents with severe renal failure in the neonatal period, with respiratory symptoms in patients with lung hypoplasia, or symptoms of portal hypertension secondary to congenital hepatic fibrosis.

Renal agenesis

Pulmonary hypoplasia can be associated with bilateral renal aplasia or severe dysplasia.

Abnormalities of the collecting system, kidney and bladder

General

Differential diagnosis for urinary tract obstruction:

Ureteropelvic junction obstruction
Ureterocele
Posterior urethral valves
Neurogenic bladder
Duplicated collecting system

Evaluation of an infant with anuria more than 48 hours after birth:

History: Important to ascertain history of prenatal oligohydramnios or polyhydramnios on fetal ultrasounds. Fetal hydrops may occur with congenital nephrotic syndrome. Oligohydramnios occurs with severe urinary tract obstruction or renal agenesis, which is associated with pulmonary hypoplasia (Potter's syndrome). In neonates, history of umbilical artery catheterization implies renal artery thrombosis.

Physical: Look for dysmorphic features like low-set ears, single umbilical artery (2-vessel cord), hypospadias, anorectal abnormalities, polythelia, vertebral anomalies, tracheoesophageal fistula, palpable suprapubic bladder suggesting obstruction, sacral tuft of hair or myelomeningocele (neurogenic bladder), palpable renal mass (Wilms tumor, polycystic kidney), abdominal wall defects ("prune belly syndrome", exstrophy of bladder.

Laboratory Findings: Urinalysis can reveal if prerenal, renal or postrenal etiology. Ultrasonography provides important information regarding kidney size and echogenicity, renal blood flow, collecting system, and urinary bladder, congenital renal disorders such as polycystic kidney disease and multicystic dysplastic kidney. Doppler examination of the renal blood flow is helpful in diagnosing renal vascular thrombosis.

Hydronephrosis

One reason for abdominal masses in infants is hydronephrosis.

Hydroureter and megaureter

Prune belly syndrome is associated with bilateral hydronephrosis.

Ureterocele

Outpouching of the distal ureter (ureterocele) can cause urinary tract obstruction

Vesicoureteral Reflux (VUR)

Vesicoureteral reflux can be caused by anatomic deformity of the ureterovesical junction or as a result of posterior urethral valves or other urinary tract abnormalities leading to misplaced ureteral insertion. There is a familial association, with 30-35% risk of VUR with an affected sibling and 67% risk with an affected parent. VUR is more common in the first two years of life and often spontaneously resolves over time, with above 80% of grade I and II resolved at a five-year follow-up and lower rates of spontaneous resolution for higher grade reflux.

Abnormalities of the urethra

Posterior urethral valves

A newborn or infant boy with weak urinary stream and a distended bladder detected by palpation suggests the possibility of posterior urethral valves.

Even if posterior urethral valves are repaired, renal failure may still develop.

Kidney and bladder function must be followed long term in patients with history of posterior urethral valve due to possibility of renal dysfunction and urinary incontinence.

Urethral stricture

Frequently urethral trauma is the cause of urethral strictures in boys. Females with narrow urethras do not need treatment.

Hereditary nephropathy (eg, familial nephritis)

Alport syndrome is a hereditary condition that causes nephritis and bilateral sensorineural hearing loss.

Acquired

Infection of the urinary tract
Pyelonephritis

The most common organisms causing urinary tract infections (UTIs) in children are Escherichia coli, as well as less commonly Pseudomonas aeruginosa, Enterococcus faecalis, Klebsiella pneumonia, group B Streptococcus, Staphylococcus aureus, Proteus mirabilis and coagulase negative Staphylococcus.

Reflux nephropathy often produces no symptoms.

A child who is not toilet-trained should have a renal ultrasound to look for hydronephrosis or other signs of urinary tract abnormalities following the first urinary tract infection. According to 2011 AAP UTI guidelines, VCUG is no longer routinely recommended for healthy 2 to 24 month children until after the second UTI - a change from the previous position which recommended VCUG following first UTI. This remains a controversial policy among the medical community.

Long-term antibiotic prophylaxis against urinary tract infection remains a controversial subject, and is currently being studied in the RIVUR study. AAP UTI guidelines from 1999 did recommend routine antibiotic prophylaxis after first UTI. Antibiotic prophylaxis has been recommended for all grades of VUR. The Randomized Intervention for Children with Vesicoureteral Reflux (RIVUR) study aims to clarify the role of antibiotic prophylaxis in UTI prevention.

Before culture results are reported, suggested empiric therapy for acute pyelonephritis includes trimethoprim-sulfamethoxazole, amoxicillin-clavulanate,

cephalexin, cefixime, cefpodoxime, ciprofloxacin, or nitrofurantoin. In severe pyelonephritis, gentamicin plus ampicillin, ampicillin plus cefotaxime, ceftriaxone or ciprofloxacin are choices for empiric therapy. Ceprafloxacin is recommended only when there is no alternative choice in children.

Hypertension is an uncommon complication of VUR, but occurs at higher rates in patients with a greater degree of renal injury.

Urinary tract infection is an important cause of fever without an obvious source in young infants.

Antibiotic susceptibility testing may be helpful to identify resistant pathogens and tailor antimicrobial therapy in cases of acute pyelonephritis.

Urinary tract infections are more common in females than males, and occur most frequently in infants and adolescents.

Urinalysis is not sufficient to diagnose UTI, as pyuria has other causes and nitrites are not always present, as they are only produced by gram negative bacteriaand in infants.

Constipation can lead to incomplete urinary bladder emptying which can lead to urinary tract infections.

Cystitis

Cystitis can be treated with a 3 to 7 day course of trimethoprim-sulfamethoxazole, amoxicillin, amoxicillin-clavulanate, or cephalexin.

A patient with cystitis is at increased risk of cystitis with Staphylococcus saprophyticus if they have been sexually active.

Cystitis in the sexually active patient should be treated with antibiotics that cover Staphylococcus saprophyticus and Escherichia coli. Generally, in non-pregnant patients, trimethoprim-sulfamethoxazoe is first line, with ciprofloxacin if there is an allergy. Nitrofurantoin or cephalexin should be given if patient is pregnant.

Prophylactic antibiotics (quarter to half daily dose of TMP-SMX, trimethoprim, cephalexin, nitrofurantoin or ciprofloxacin) can be considered in the case of recurrent cystitis.

Cystitis can cause secondary enuresis.

Acute glomerulonephritis

To evaluate possible acute post-streptococcal nephritis, serum electrolytes, BUN and creatinine should be done to assess renal status, streptococcal antibodies, (ASO, streptozyme) and complement levels should be measured, and urinalysis should be

done with microscopy to look for red blood cells and casts.

Chronic renal failure is generally not the outcome of acute post-streptococcal glomerulonephritis.

Post-streptococcal nephritis may immediately cause:

Hypertension
Fluid overload
Pulmonary edema

In post-streptococcal glomerulonephritis, kidney function improves about 3 weeks after onset of symptoms, while complement levels take 8-12 weeks to resolve and microscopic hematuria may take 6 to 24 months to resolve.

Management of acute post-streptococcal glomerulonephritis is supportive care based on symptoms. Fluid overload should be avoided and managed appropriately if it occurs (with fluid and sodium restriction, as well as diuretic therapy as needed), electrolytes monitored and abnormalities managed accordingly, blood pressure monitored and antihypertensives administered as needed.

Acute post-streptococcal glomerulonephritis (APSGN) occurs 10-14 days after streptococcal throat infection, unlike IgA nephropathy which occurs within a few days of upper respiratory infection (URI). APSGN is associated with elevated antistreptococcal titers and decreased complement levels.

Nephrotic syndrome

Initially, edema occurs in the periorbital, scrotal and labial areas, which have low tissue retention. The edema is periorbital in the morning, and it is often initially attributed to allergies. As day progresses, edema shifts to lower extremities. Generalized edema can ultimately develop. Other common symptoms include anorexia, irritability, fatigue, abdominal discomfort and diarrhea.

Laboratory findings in children with minimal-change nephrotic syndrome: Blood:

Albumin less than 2.5 g/dL (because albumin is lost through urine)

Elevated blood cholesterol, triglycerides and lipoproteins (because low albumin stimulates liver to synthesize lipids)

Decreased serum sodium partially as a response to hyperlipidemia and partially due to water retention caused by intervascular hypovolemia and increased antidiuretic hormone (ADH) secretion

Low total calcium due to hypoalbuminemia, normal ionized calcium

Urine:

Positive urine dipstick for protein, which may be affected by urine concentration.

Elevated urine protein:urine creatinine ratio, greater than 3.0; first void

specimen is most accurate.

Initial treatment of first episode of nephrotic syndrome should be prednisone 60mg/m2 body surface area per day based on ideal weight for height, not to exceed 80mg/day divided into three doses. Administer daily for 4-6 weeks, then 40mg/m2 BSA per day as a daily morning dose given on alternate days for an additional 4-6 weeks.

Differential diagnosis of nephrotic syndrome

With hematuria: Focal segmental glomerulonephritis (sometimes includes hematuria; this heterogenous condition includes heroin-induced nephropathy, AIDS, multiple myeloma, Alport syndrome, reflux nephropathy, diabetic nephropathy and obesity, as well as genetic forms), membranous nephritis (sometimes includes hematuria; this disease may be primary or due to hepatitis B, malaria, syphilis, SLE, Crohn disease, penicillamime, nephroblastoma, neuroblastoma), membranoproliferative glomerulonephritis, post-streptococcal glomerulonephritis, Henoch-Schönlein purpura, lupus nephritis, IgA nephritis (microscopic hematuria alternating with macroscopic hematuria).

Without hematuria: Minimal change nephrotic syndrome, congenital nephrotic syndrome, focal segmental glomerulonephritis, membranous glomerulonephritis.

Sixty percent of patients who initially respond to steroids will relapse. Most who relapse will stop having relapses after an average of 3 years. Relapses are treated with prednisone 60mg/m2 BSA daily in two divided doses until protein free for 3 days, then 40mg/m2 BSA per day as single morning dose on alternate days for 6 weeks if relapse was more than 3 months after initial response and for 12 weeks if relapse was within 3 months of initial response.

Aggressive diuresis may induce hypovolemia and secondary renal failure, thromboembolism or electrolyte abnormalities, some of which may be offset by giving salt-poor albumin (0.5-1 gram/kg body weight up to 25g in a 25% solution administered IV over 30 to 60 minutes).

Serum sodium decreases as a result of hyperlipidemia (pseudohyponatremia), hypovolemia and high ADH levels.

Complications of nephrotic syndrome:

Acute renal failure: caused by hypovolemia and reversed by intravascular volume expansion with salt-poor albumin or caused by renal vein thrombosis.

Thromboses: caused by loss of antithrombin III and protein S in the urine and increase in fibrinogen; antiphospholipid antibody syndrome has been a cause of nephrotic syndrome, with

persistently elevated antibodies against membrane anionic phospholipids; renal vein thrombosis, extremity deep venous thrombosis

and thromboembolic events (pulmonary embolism, stroke, cerebral venous sinus thrombosis).

Infections: caused by urinary loss of factor B [factor B contributes to opsonization of bacteria], decreased IgG synthesis, and impaired T-cell function

Peritonitis: most common; (S. pneumonia is most common cause); characterized by fever, chills, ascites, abdominal pain, ileus, diarrhea.

Cellulitis, meningitis and pneumonitis can also occur.Complications of prolonged steroid use: growth stunting, reduced mineral bone density.

Prognosis of nephrotic syndrome can be predicted by: By diagnosis:

Congenital nephrotic syndrome: survival beyond several months only with dialysis and kidney transplantation

Focal Segmental Glaminilo Sclerosis (FSGS): only 20% have remission of proteinuria

Minimal Change Nephrotic Syndrome (MCNS): about 50% of children have 1 or 2 relapses within 5 years, approximately 20% continue to relapse 10 years after diagnosis

By initial symptoms:

Hematuria and hypertension are signs of increased likelihood of being steroid resistant and having poorer prognosis

10% of patients with nephrotic syndrome are steroid-resistant and their renal function deteriorates to the point of needing dialysis or renal transplantation. These patients may require treatment with other immunosuppressive medications, such as cyclophosphamide, cyclosporine and mycophenolate mofetil, which have significant side-effects.

Decreased C3 complement levels are associated with membranoproliferative glomerulonephritis, post-streptococcal glomerulonephritis, and lupus nephritis.

Hemolytic uremic syndrome (HUS)

Children with HUS may have hemoglobin less than 10g/dL, negative Direct Coombs test, schistocytes on peripheral smear, increased bilirubin, decreased haptoglobin, increased lactate dehydrogenase, low platelets (40x103/mcL), elevated BUN, elevated creatinine, hypoalbuminemia, hematuria, and proteinuria.

Children with HUS should receive supportive care, with fluid to cover insensible water loss of 500mL/m2 per day plus replacement of ml per ml stool and urine to maintain euvolemia. Electrolyte abnormalities should be avoided. Packed red blood cells can be given for anemia, but platelets should not be given due to the potential for worsening the microangiopathic thrombosis.

Most cases of HUS come after a diarrheal illness, most frequently with strains of

Escherichia coli.

Enterohemorrhagic E. coli O157:H7 is responsible for about two-third of the cases of HUS.

One study showed an increased risk of HUS in children under 10years with E coli O157:H7 that were treated with antibiotics, although metanalysis did not confirm increased risk. However, antibiotics do not have proven benefit in the management of E coli O157:H7.

Henoch-Schonlein purpura (HSP)

When immune complexes deposit in the renal mesangium, glomerulonephritis can occur, ranging from a mild proliferative glomerulonephritis to a severe crescentic glomerulonephritis. Microscopic hematuria, red cell casts and proteinuria may occur as late complications of HSP.

The prognosis of HSP is worse if nephrotic syndrome develops, with 19.5 percent of patients with nephritic or nephrotic symptoms going on to develop chronic renal impairment.

Patients with HSP progress to chronic renal insufficiency 1 to 5 percent of the time.

IgA Nephropathy

IgA nephropathy presents as cola-colored urine just after an upper respiratory infection, generally without any other symptoms.

Other renal conditions

Renal failure
Acute renal failure

Children with prerenal failure should initially be managed with fluid repletion with isotonic fluids. Electrolyte abnormalities should be managed appropriately, with rapid correction of serum sodium less than 120mEq/L and aggressive management of serum potassium over 6mmol/L. Hypocalcemia can be managed with oral or intravenous calcium, and hyperphosphatemia can be managed with phosphate binders such as calcium carbonate or calcium acetate. Severe anemia (hematocrit less than 25%) can be treated with transfusion, and hypertension can be controlled with diuretics or dialysis.

Fractional excretion of sodium can help differentiate between prerenal and intrinsic renal failure in the oliguric patient, with values <1% for the older child and <2.5% for the newborn representing prerenal failure and >3% representing intrinsic renal failure.

Acute prerenal failure can be caused by extracellular fluid deficits (gastrointestinal losses, urinary losses, blood losses, redistribution of fluid, skin losses, and vasodilation) or cardiac dysfunction (resulting from congenital heart disease,

cardiomyopathy, arrhythmia, acquired valvular disease or tamponade).

Intrinsic renal failure can be caused by vascular spasm, intravascular coagulation or microvascular injury.

Postrenal failure can be caused by congenital abnormalities, such as posterior urethral valves or ureteropelvic junction defects or acquired obstruction from urolithiasis, clots or tumors.

Intrinsic renal failure

Fluid should be watched closely in children with acute renal failure to avoid fluid overload and sodium abnormalities, and hyperkalemia and severe acidosis should be identified and corrected.

Bicarbonate, glucose, and insulin are temporizing measures to lower serum potassium levels by shifting potassium intracellularly, and calcium can serve to stabilize cardiac myocyte membranes during hyperkalemia, but these do not have a lasting effect on the hyperkalemia resulting from renal failure.

Complications of acute renal failure include multisystem organ failure, electrolyte abnormalities, anemia, hypertension and progression to chronic renal failure.

In acute renal failure, renal excretion of drugs is altered, and therefore many drug dosages require modification.

A child with acute renal failure is in a catabolic state, so malnutrition is a common consequence. Phosphorous intake should be limited, since persons with acute renal failure often already have hyperphosphatemia.

When a patient has hypocalcemia, his or her risk of tetany during bicarbonate administration is increased because bicarbonate lowers serum calcium levels.

Chronic kidney disease (chronic renal failure)

Major complications of chronic kidney disease include decreased linear growth, nutritional deficiencies (especially folic acid, trace minerals and vitamin B complexes), metabolic acidosis, anemia, hypertension, and decreased immune response to vaccinations.

An infant with renal dysplasia or hydronephrosis may develop diuresis with water and electrolyte depletion.

Children with chronic kidney disease commonly have growth failure, which may be attenuated by growth hormone therapy.

One contributor to growth failure in chronic kidney disease is metabolic acidosis.

Chronic kidney disease leads to hypocalcemia, low vitamin D levels (and low active 1,25-dihydroxyvitamin D3 levels), with increased parathyroid hormone (PTH) and increased phosphate. This constellation of abnormalities is termed renal osteodystrophy, and lead to abnormal bone mineralization with consequential fractures and osteitis fibrosa.

End stage kidney disease and transplantation

Some infants with congenital chronic kidney disease may benefit from renal transplantation, which generally increases survival in comparison to dialysis.

Before renal transplantation, patients should receive all immunizations, because live vaccines are contraindicated while on immunosuppressive therapy and immune response to inactivated vaccines will be lessened once immunosuppressive therapy is initiated.

Patients with end-stage kidney disease either need to attend school or have home- schooling.

Trauma

Renal injuries

The child with blunt abdominal trauma needs appropriate history and physical examination, complete blood count, AST, ALT and urinalysis. Generally abdominal CT is performed, although it could potentially be avoided in the older child if physical examination and laboratory studies are all

normal. Renal injury is possible both with and without hematuria, so index of suspicion must be high regardless of urinalysis results.

Urethral injury

If urethral bleeding is grossly present after acute trauma, urinary catheterization is contraindicated.

Toxins

Antibiotics, NSAIDs, chemotherapeutic agents, cyclosporine, and tacrolimus can cause renal toxicity.

Urinary tract stones

Urinary tract stones present with variable abnormalities in the urinalysis (microscopic or macroscopic hematuria, pyuria), with or without flank or pelvic pain.

The child with suspected urinary tract stones should be evaluated for signs of poor growth, spina bifida or metabolic disease. Initial laboratories include serum electrolytes, BUN, creatinine, uric acid, calcium, phosphorus, UA with microscopy, spot urine calcium and creatinine. After initial evaluation, two 24 hour urine specimens should be gathered for stone profile, a three day food diary should be recorded, a renal ultrasound or CT without radiographic contrast performed, and stone analysis on any captured stone. If there is hypercalcemia, parathyroid hormones (PTH) should also be measured.

Loop diuretics such as furosemide can cause hypercalciuria and nephrocalcinosis.

Low fluid intake is a contributing factor to urinary tract stones, so increasing fluid intake can protect against urolithiasis.

Calcium excretion is increased with furosemide treatment, because this medication blocks the active transport of sodium chloride in the thick ascending loop of Henle, and reabsorption of calcium by a passive process is dependent upon this. Thiazides, however, act in the distal tubule to inhibit chloride reuptake, but this does not have an effect on calcium reabsorption. In fact, thiazides are thought to possibly increase distal tubule calcium reabsorption.

Hypercalciuria is found in up to 50% of the children who form urinary tract stones.

Recurrent urinary infections, which may be the result of urinary stasis, predispose to developing struvite stones. Urinary stasis also may directly contribute to urolithiasis.

Renal tubular acidosis

Growth failure is a way for any type of Renal Tubular Acidosis (RTA) to present.

In the patient with suspected renal tubular acidosis, a variety of primary and secondary causes must be considered.

Proximal RTA (type II) involves the loss of sodium, potassium and bicarbonate into the urine, and can either be an isolated anomaly or related to Fanconi syndrome, where there is global proximal tubular dysfunction. Fanconi syndrome can be due to cystinosis, Lowe syndrome, galactosemia, tyrosinemia, fructosemia, Fanconi-Bickel syndrome, Wilson disease, mitochondrial diseases, x-linked nephrolithiasis, or be secondary to heavy metals, outdated tetracycline, gentamicin, ifosfamide, and cyclosporine/tacrolimus.

Distal RTA (type I) patients have hyperchloremic non-anion gap metabolic acidosis and growth failure. Distal RTA can be hereditary, sporadic or secondary to interstitial nephritis, obstructive uropathy, Vesico Uretral Reflux (VUR), pyelonephritis, transplant rejection, sickle cell nephropathy, lupus nephritis, nephrocalcinosis, medullary sponge kidney, hepatic cirrhosis or due to amphotericin B, lithium, glue sniffing or cisplatin.

Hyperkalemic RTA (type IV) results in the failure of renal tubule to secrete hydrogen ions and potassium ions. It can be hereditary, sporadic, or secondary to hypoaldosteronism (from Addison's disease or congenital adrenal hyperplasia), pseudohypoaldosteronism, obstructive uropathy, pyelonephritis, interstitial nephritis, diabetes mellitus, sickle-cell nephropathy, trimethoprim-sulfamethoxazole, angiotensin converting enzymes inhibitors, or cyclosporine.

Other tubular disorders, such as Bartter syndrome, Gitelman syndrome and Liddle

syndrome result in metabolic alkalosis.

Hereditary conditions with renal manifestations (eg nephrogenic Diabetes Insipidus [DI])

Signs and symptoms of nephrogenic DI include failure to thrive, polydipsia with preference for water, polyuria with dilute urine, irritability, constipation, vomiting and seizures. Patients may present with signs of dehydration, poor perfusion, and abdominal masses (hydronephrosis or fecaliths).

Urine and serum osmolality should be measured during a water deprivation test in a child suspected of having DI. They will not be able to achieve normal response to water restriction, which includes urine osmolality less than or equal to 450mOsm/kg water and urine to plasma osmolar ratio >1.5.

Diabetes insipidus can be inherited, or it can be a result of cranial injury or surgery – Central Diabetes Insipidus.

Hypertension

General

Symptoms of hypertensive encephalopathy include stupor, seizures and vomiting.

For suspected hypertension, the diagnosis should be confirmed by repeat blood pressures with an appropriate sized cuff at three or more office visits about one week apart. Medications should be evaluated. Obesity and its complications, including obstructive sleep apnea, should be evaluated and managed. Neonatal history should be taken for risk factors. Family history should be taken to identify possible inherited causes. Signs and symptoms of endocrine and renal disease should be evaluated.

Most children do not have symptoms at the time they are diagnosed with hypertension.

Differential diagnosis of hypertension in children and adolescents includes the following:

edications: albuterol, atomoxetine, amphetamines, antidepressants, antipsychotics, caffeine, carbamazepine, cocaine, cyclosporine, dexedrine, ephedrine, epogen, ethanol, fentayl, gentamicin, ketoconazole, methylphenidate, metoclopramide, NSAIDS, oral contraceptives, phenylephrine, pseudoephedrine, steroids, tacrolimus

enal/urologic: obstructive uropathy, cystic renal disease, parenchymal renal disease (acute tubular necrosis, interstitial nephritis), renal vein thrombosis, history of umbilical artery catheter, SLE/lupus nephritis, diabetic nephropathy
aternal substance abuse
eonatal asphyxia, bronchopulmonary dysplasia, obstructive sleep apnea
ndocrine: pheochromocytoma, Cushing's syndrome, hyperaldosteronism
ardiac: coarctation of the aorta
ystemic disease: Neurofibromatosis

Medications for hypertension and their mechanism of action:

Angiotensin converting enzyme inhibitors (captopril, enalapril, benazepril, lisinopril, fosinopril, quinapril) – inhibit conversion of angiotensin I to angiotensin II (a vasoconstrictor), inhibit kininase (which plays a role in increasing vasodilatory bradykinins)

Angiotensin receptor blockers (irbesartan, losartan) – block angiotensin II receptors to inhibit its actions on blood pressure

Calcium channel blockers (amlodipine, felodipine, isradipine, extended release nifedipine) – directly affect blood pressure by inhibiting the influx of calcium into the vascular smooth muscle, thus inhibiting their constriction

Alpha and beta blocker (labetalol) and beta blockers (atenolol, metoprolol, propranolol) – possible mechanisms include lowering cardiac output, lowering peripheral vascular resistance, inhibiting the secretion of renin, decreasing the volume of plasma and inhibiting the sympathetic activity of the central nervous systems

Central alpha agonist (clonidine) – acting on central alpha adrenoceptors to decrease presynaptic calcium, thus inhibiting secretion of norepinephrine and decrease sympathetic input in blood pressure regulation

Vasodilator (hydralazine, minoxidil) – dilates the arteriolar resistance vessels Diuretics (hydrochlorothiazide, furosemide, spironolactone, triamterene) – inhibit water reabsorption in the kidney thus reducing intravascular volume and therefore reducing blood pressure

Blood pressure may be falsely elevated if the cuff size is too small for the child or the patient is agitated during the measurement. Variation can also occur based on patient positioning, the type of equipment used, and the experience and training of the observer.

When "white coat" hypertension is suspected based on history and physical, ambulatory blood pressure monitoring is the preferred method of obtaining repeated measurements in the patient's natural environment.

Renal

Renal causes of hypertension include:

Renovascular disease: (fibromuscular dysplasia (may be related to Williams syndrome or neurofibromatosis 1), arteritis from Kawasaki, Takayasu or Moyamoya disease, renal transplant artery stenosis, umbilical catheters leading to thrombosis or emboli, renal trauma; compression of renal artery from neoplasia, perirenal hematoma, retroperitoneal fibrosis or congenital fibrous bands.

enal parenchymal disease: glomerulonephritis, systemic vasculitis, hemolytic uremic syndrome, interstitial nephritis, hereditary diseases (polycystic kidney disease, medullary cystic disease, Denys-Drash syndrome, sickle cell disease, liddle syndrome), congenital anomalies (vesicoureteral reflux [VUR], obstruction, multicystic dysplastic kidney, horseshoe kidney, segmental hypoplasia).

Vascular

Neurofibromatosis can lead to fibromuscular dysplasia of the renal arteries and therefore cause consequential hypertension.

Williams's syndrome can lead to renal artery stenosis, which can then cause hypertension.

Adrenal

Pheochromocytoma

In children, hypertension from pheochromocytoma may present as encephalopathy or cardiac failure from having sustained hypertension, rather than the presentation of intermittent tachycardia, headache and diaphoresis, which is more common in adults.

A rare complication of prolonged glucocorticoid use is pheochromocytoma, which has only been reported in adults in this clinical context.

Pheochromocytoma may occur as a part of Von-Hippel-Lindau disease, neurofibromatosis, or multiple endocrine neoplasias IIA or IIB.

Cushing syndrome

If a child who has hypertension is short and obese, Cushing syndrome should be considered.

Miscellaneous causes

Essential hypertension

Differential diagnosis of essential hypertension includes secondary causes, such as medications, obesity, hyperlipidemia, diabetes mellitus, pre-eclampsia, renal disease, neurofibromatosis, SLE, tumors, and other endocrine or renal etiologies.

Essential hypertension should first be managed with lifestyle modification, including reduction of sodium intake to 2-3g/day and increasing physical activity to reduce obesity and decrease blood pressure. If these measures are inadequate, then pharmacologic therapy should be considered.

Administration of drugs

Albuterol, contraceptives, corticosteroids, decongestants, and illicit drugs can cause hypertension in children.

Genital System Disorders

General
Pain
Infection

Pelvic inflammatory disease (PID): caused by C. trachomatis with a concomitant infection of N. gonorrhea and usually presents with the following symptoms:

Vaginal discharge, cervical motion tenderness, fever, elevated CRP and ESR, Gram-negative intracellular diplococci in cervical secretions, and purulent material from peritoneal cavity; Usually seen following onset or cessation of menses.

Laparoscopy is the gold standard for diagnosing PID. Diagnosis can be made by history and physical examination alone.

Treatment: outpatient antibiotic therapy or hospitalization and inpatient antibiotic therapy for women severely ill, non-compliant, unable to tolerate outpatient treatment, or with complications like suspected abscess.

Adolescents should be advised to delay the onset of sexual activity until age 16 years or older, as they are at an increased risk for PID.

Complications: endometritis, salpingitis and peritonitis. Once the disease spreads to the peritoneum, gonococci can seed into the liver capsule causing perihepatitis with RUQ pain (Fitz-Hugh-Curtis syndrome). C. trachomatis can also cause perihepatitis.

Interpret slides of wet preparations of vaginal discharge:

Clue cells – Epithelial cells covered with adherent bacteria from Gardnerella vaginalis.

Leukocytes – WBCs normally absent or in very low quantity. Trichomonads – Oval-bodied, flagellated trichomonads.

Trauma (accidental or self-induced)

Consider instrumentation of the penis or insertion of a foreign body into the vagina as a cause of genital pain and malodorous discharge.

Male genital trauma or instrumentation:

Sexually active adolescent males are considered among the high-risk groups for sexually transmitted infections (STIs) and should be screened even if asymptomatic. Noninvasive tests such as urine-based nucleic acid amplification tests (NAATs) should be obtained to screen for chlamydia and gonorrhea.

Female vaginal pain and discharge:

In pre-pubertal girls, vaginal pain is often due to accidental injuries, but physical and sexual abuse must be ruled out. In post-pubertal girls, good possibility of sexual activity exists and STIs must be suspected.

Pre-pubertal vaginal mucosa is thin and breaks down easily causing bleeding. One of the most common foreign substances found is residual or wadded toilet paper.

Straddle injuries may present with bruising, hematomas, and lacerations of the mons and labia. Usually vagina and hymen are spared.

Both sexes should get testing for Syphilis and HPV if sexually active. HIV screening should be discussed with all adolescents and encouraged for those who are sexually active.

Parental involvement in an adolescent's health care is usually desirable; however, it is not required when the adolescent consents to HIV testing.

Abnormal masses

Hydrocele, inguinal hernia

Differential diagnoses of mass in the inguinal area in an infant: congenital anomalies, non-congenital hernias, vascular conditions, infections, inflammatory processes, trauma, and neoplasms.

Know how to diagnose an inguinal hernia

In girls, the mass is typically found in the upper portion of the labia majora. The bulge is best visualized with increased intra-abdominal pressure (Valsalva maneuver), i.e. crying, straining, blowing a balloon, or coughing.

In boys, "silk glove sign" is feeling of the layers of the hernia sac (processus vaginalis) as they slide over the spermatic cord structures, with rolling of the spermatic cord beneath the index finger at the pubic tubercle.

A femoral hernia should be suspected if the bulge is located below the inguinal canal on the medial aspect of the thigh.

Know how to diagnose a hydrocele

Hydroceles are smooth and non-tender. Transillumination of the scrotum confirms the fluid-filled nature of the mass. Palpation of the testis is necessary as hydrocele can develop in association with a testicular tumor.

Plan the evaluation of a patient with a mass in the inguinal area

Diagnostics: In incarcerated inguinal hernia, radiographs typically demonstrate multiple air-fluid levels. Transillumination can be misleading as the thin intestine wall can mimic that of a hydrocele wall and both transilluminate. Scrotal ultrasound can distinguish between a hernia and a

hydrocele. Diagnostic laparoscopy has emerged as an effective and reliable tool; however, it requires general anesthesia.

Bartholin gland cyst: can occur due to obstruction of ductal outflow, a change in mucus consistency, mechanical trauma, or congenitally narrowed ducts. People at high risk for sexually transmitted infections have an increased risk for Bartholin gland abscess formation. Most cysts are unilateral, round or ovoid, and tense, and located in the posterior labia majora or lower vestibule causing vulvar pain.
Varicocele: is a congenital condition with abnormal dilation of the pampiniform plexus in the scrotum (painless "bag of worms") from valvular incompetence of the internal spermatic vein. Varicocele is the most common surgically correctable cause of subfertility in males. Examine an adolescent for a varicocele in the standing position while he is performing a Valsalva maneuver.

Discharge

Vaginal

Maternal estrogens can influence vaginal discharge in an infant during the first three weeks of life.

Neonates and peripubertal girls can present with a white discharge, which is known as physiologic leukorrhea. Management: reassurance to mother.

Evaluate and recognize the cause of a vaginal discharge in a pre-adolescent girl:

Urethral

Gram-negative intracellular diplococci in urethral discharge supports the diagnosis of gonococcal disease in males.

Bleeding

Vaginal

Differential Diagnoses of Vaginal bleeding in preadolescent and adolescent females:

Uterine

Estrogen withdrawal is an etiology of uterine bleeding in a newborn infant.

Management initially is to provide reassurance to the parents. Bleeding that is excessive or persistent beyond 1mo of life requires additional evaluation.

Male

Congenital abnormalities
Hypospadias: Incidence is increased in disorders of sexual differentiation (intersex), anorectal malformation, renal anomalies, and congenital heart disease. Chordee, a ventral penile curvature that occurs during erection, can be seen with proximal hypospadias.

Complications: Approximately 10% of boys with hypospadias have an undescended testis; inguinal hernias also are common.

A karyotype should be obtained in patients with hypospadias and cryptorchidism to rule out genetic conditions.

Complications of untreated hypospadias: ventral deflection or severe splaying; sexual dysfunction secondary to penile curvature; infertility if the urethral meatus is proximal; meatal stenosis (congenital, uncommon); and cosmetic appearance.

Circumcision should be delayed until after hypospadias repair because the foreskin is used in the repair in most cases. The most common complications post-surgery include urethrocutaneous fistula, meatal stenosis, deformed urinary stream, persistent penile curvature, and dehiscence of the hypospadias repair.

Cryptorchidism

Undescended (cryptorchid) testis is the most common disorder of sexual differentiation in males. Testicular descent occurs at 32 to 36 weeks gestation; thus, premature male infants can have undescended testes.

Majority of cryptorchid testes descend spontaneously during the first 3 months of life. If undescended by 4 months of age, it will remain undescended.

Complications: infertility, increased incidence of testicular tumors, associated hernia, torsion of the cryptorchid testis, and possible psychologic effects of an empty scrotum.

Plan the appropriate management of a patient with undescended testes.

Congenital cryptorchidism should be treated surgically no later than 9 to 15 months of age. Orchiopexy involves an inguinal incision, mobilization of the testis and spermatic cord, and correction of the indirect inguinal hernia.

Failure to find the testis in the scrotum indicates that the testis is undescended, absent, or retractile.

Distinguish between undescended testes and retractile testes:

Retractile testes: Males should be examined with their legs in a relaxed frog- leg position, and if the testis can be manipulated into the scrotum comfortably, it is probably retractile. Monitor every 6 to 12 months with follow-up physical examinations.

Hypospadias with bilateral cryptorchidism is an indication to evaluate for intersex disorders.

Micropenis

Know the significance of hypoglycemia in a patient with micropenis.

Micropenis is failure of the hypothalamus to produce adequate GnRH that occurs after 14 weeks of gestation. If growth hormone deficiency is also present, neonatal hypoglycemia can occur. This can occur in Kallmann syndrome, Prader- Willi syndrome, and Lawrence-Moon-Biedl syndrome.

Know how to diagnose micropenis by measurement in a newborn boy.

Micropenis = normally formed penis that is at least 2.5 standard deviations below the mean in size. The pertinent measurement is the stretched penile length, measured by stretching the penis and assessing the distance from the penile base under the pubic symphysis to the tip of the glans. The mean length of a term newborn penis is 3.5 ± 0.7 cm with a diameter of 1.1 ± 0.2 cm. The diagnosis of micropenis is made if the stretched length is <1.9 cm.

Management involves referral to pediatric endocrinologist and pediatric urologist, karyotype, assessment of anterior pituitary function and testicular function, and MRI of the brain to determine the anatomic integrity of the hypothalamus, the anterior pituitary gland, and the midline structures of the brain.

The significance of the suprapubic fat pad in evaluating penile size:

The concealed or buried penis is a normally developed penis that is camouflaged by the suprapubic fat pad, which may be congenital, iatrogenic after circumcision, or a result of obesity.

Phimosis

The accumulation of smegma (epithelial debris) beneath the infantile prepuce is not pathologic, but physiologic and does not require circumcision.

Differentiate between phimosis and paraphimosis:

Phimosis is the inability to retract the prepuce, physiologic at birth. In 90% of uncircumcised boys the prepuce becomes fully retractable by 3 years of age. Application of corticosteroid cream to the foreskin may loosen the phimotic ring.

Paraphimosis occurs when the foreskin is retracted past the coronal sulcus and the prepuce cannot be pulled back over the glans. Treatment includes lubricating the foreskin and glans and simultaneously compressing the glans and placing distal traction on the foreskin to try to push the phimotic ring past the coronal sulcus.

Acquired abnormalities

Testicular torsion

Inadequate fixation of the testis within the scrotum, resulting from a redundant tunica vaginalis, allows excessive mobility of the testis. The abnormal attachment is termed a bell clapper deformity and often is bilateral.

Signs and symptoms of testicular torsion: it can often be bilateral: there is acute pain and swelling of the scrotum, often difficult to examine; Cremasteric reflex is usually absent.

Prompt referral for surgical exploration of testicular torsion is crucial to salvage the testis. The contralateral testis should also be fixed in the scrotum as the condition is often bilateral.

Early ultrasonography with Doppler flow is important for the diagnosis of testicular torsion.

Infection

Orchitis

Diagnose orchitis based on signs and symptoms:

Orchitis is the inflammation and swelling of one or both testes. It may be caused by sexually transmitted infections (STIs) such as gonorrhea and chlamydia. The rate of sexually transmitted orchitis or epididymitis is higher in men ages 19 - 35.

Symptoms can include penile discharge, blood in the semen, fever, groin pain, dysuria, testicular pain, scrotal swelling, and inguinal lymphadenopathy.

Treatment includes anti-inflammatory medications, antibiotics if STI is present, bed rest with scrotal elevation and ice packs to the area.

Identify the common causes of orchitis:

Testicular torsion, torsion of appendix testis, epididymitis, trauma with hematocele, incarcerated inguinal hernia, and mumps orchitis.

Epididymitis

Know the signs and symptoms of epididymitis:

This is an acute ascending infection from the urethra through the vas deferens into the epididymis. In toddlers, Escherichia coli often causes it due to potty-training. Treatment usually consists of bed rest and antibiotics. Differentiating epididymitis from testicular torsion can be difficult and may require surgical exploration.

The most common cause of epididymitis in adolescents:

After puberty, bacterial epididymitis becomes more common and is usually the cause of acute painful scrotal swelling in young sexually active men. Urinalysis reveals pyuria.

Urethritis

Signs and symptoms of urethritis:

Symptoms: urethral discharge, urethral itching, dysuria, or urinary frequency, erythema of the urethral meatus, and scrotal pain. Most patients are asymptomatic.

Signs of urethritis: (1) mucoid or purulent urethral discharge, (2) ≥5 WBC/high-power field (HPF) on Gram stain, (3) ≥10 WBC per HPF on microscopic examination of first-void urine specimen, or (4) positive leukocyte esterase test. Presence of gram-negative intracellular diplococci on microscopy confirms the diagnosis of gonococcal urethritis.

An important cause of urethritis in adolescents is chlamydial infection.

Alternative non-culture methods for identifying urethritis include nucleic acid amplification test(NAAT), which are more specific than culture for Chlamydia trachomatis.

Treatment of urethritis

Ceftriaxone 250mg IM as a single dose or Cefixime 400mg PO as a single dose + Azithromycin 1g PO as a single dose or Doxycycline 100mg PO BID x 7days for gonococcal/chlamydial urethritis.

Trauma

Urethral stricture can be a complication of bladder catheterization.

Testicular masses

Methods of evaluation for testicular masses

Scrotal ultrasound, serum tumor markers including lactate dehydrogenase, α- fetoprotein (produced by yolk sac cells) and β-HCG (elevated in choriocarcinoma and seminoma), CT of the chest, abdomen, and pelvis, and referral to a Pediatric Oncologist.

Surgical exploration through an inguinal incision with a radical orchiectomy, consisting of removal of the entire testis and spermatic cord, is usually performed.

In a prepubertal boy, if the ultrasound study or surgical exploration suggests that the tumor is localized and benign, such as a teratoma or epidermoid cyst, testis-sparing surgery with removal only of the mass may be appropriate.

Risks for testicular cancer:

Approximately 35% of prepubertal testis tumors are malignant; Differential Diagnosis include yolk sac tumors, rhabdomyosarcoma and leukemia also can occur in this age group.

In adolescents, 98% of painless solid testicular mass are malignant manifesting as a painless, hard testicular mass that does not transilluminate. Primary germ cell tumors (GCTs) of the testis constitute 95% of all testicular neoplasms.

25% of patients with seminomas and 60% to 70% of those with nonseminomatous germ cell tumors present with metastatic disease. Hence, the history and physical examination should include testes, back or abdominal pain, unexplained weight loss, dyspnea (pulmonary metastases), gynecomastia, supraclavicular adenopathy, urinary obstruction, or a “heavy” or “dragging” sensation in the groin.

Female

Congenital abnormalities
Imperforate hymen

Signs and symptoms of an imperforate hymen:

The hymen perforates normally during embryonic life to establish a connection between the lumen of the vaginal canal and the vestibule. If this perforation does not take place, the hymen is imperforate. The incidence is approximately 1 in 1000 live-born females.

Symptoms include hydrocolpos or mucocolpos in childhood, urinary tract obstruction, and primary amenorrhea with cyclic cramping. A hematocolpos and a hematometrium may develop. Many patients are free of symptoms.

Recognize the clinical manifestations of hydrometrocolpos

Hydrometrocolpos is the accumulation of fluid in the uterine cavity and vagina. It may present prenatally at ultrasound screening with a pelvic cystic mass, and if large enough, may cause partial obstruction of the urinary system with hydroureters and hydronephrosis.

Vaginal obstructions like imperforate hymen, complete transverse vaginal septum, or partial atretic vagina may cause hydrocolpos.

Diagnosis is made by pelvic ultrasound. If hydronephrosis exists, an intravenous pyelogram might be needed to evaluate for renal pathology.

Drainage of the fluid should be the immediate intervention, postponing the final surgical procedure at puberty.

Labial adhesions

Recognize labial adhesions

Re-epithelialization of labial skin occurs on both sides resulting in chronic irritation. It is important to differentiate this condition from congenital absence of the vagina.

Know the natural history of labial adhesions and the appropriate therapy

Labial adhesion is common in pre-pubertal children. The etiology is likely due to low estrogen levels. Symptoms can include dysuria and recurrent vulvovaginitis. Treatment includes sitz baths and emollients for the underlying vulvovaginitis. Topical estrogen creams and low-potency steroid creams may help to mature mucosa and reduce inflammation, respectively. Blunt separation should be avoided as it can rescar over time.

Acquired abnormalities

Ovarian torsion

Know the signs and symptoms of ovarian torsion.

In early puberty, ovaries drop from their prepubertal position into the pelvis under the influence of LH and FSH at puberty, and may cause longer supportive ligaments, predisposing them to twisting. Ovarian torsion usually occurs on the right side with similar presentation to acute appendicitis. The sigmoid colon in the left lower quadrant helps prevent the left ovary from twisting.

Symptoms: acute pain and rebound tenderness, nausea and emesis, low-grade fever, leukocytosis, and the development of peritoneal signs.

In children and adolescents, the differentiation between torsion and appendicitis is a common dilemma. Pelvic ultrasound can help. Early diagnosis and surgery are necessary to save the ovary and fallopian tube.

Ovarian cyst

Plan an appropriate diagnostic evaluation for a patient with an ovarian cyst: Pelvic ultrasound to rule out other causes of lower abdominal pain;

CT Abdomen to visualize appendix.

Recognize that small ovarian cysts are part of normal development.

Vulvovaginitis

Differential diagnosis of vulvovaginitis:

In premenarcheal children: nonspecific vulvovaginitis, poor perianal hygiene, urethral prolapse, labial adhesions, lichen sclerosus, genital injuries, neoplasms, pinworm infestation, foreign body, and sexual abuse.

In menarcheal adolescents: nonspecific vulvovaginitis, STIs such as HSV infection or trichomoniasis and candidiasis. Pregnancy and cervical cancer can also present thus.

Dermatologic disorders

Contact dermatitis

Causative agents of perineal contact dermatitis in various age groups: clothes (nickel allergy from metal buttons), diapers, topical powders or ointments in younger children; perfumes, leotards, sprays, tampons, and douches in adolescents.

Parasites

Signs and symptoms of pediculosis pubis, Phthirus pubis: prefers the pubic area, but other areas with hair can be affected such as the axilla, eyelashes, or scalp hair. Saliva of the lice causes a maculopapular reaction and results in intense itching. Bluish-gray macular lesions due to deep dermal hemosiderin deposition from louse bites, maculae cerulean, may be noted.

Treatment regimens for pediculosis pubis: Permethrin cream rinse (1%).

Sexual partners within the past 30 days should also be treated. Clothing or bed linen should be washed and dried by machine on a hot cycle.

Warts

Signs and symptoms of condylomata acuminata:

Lesions can occur on the lips, gingivae, tongue, and conjunctiva. HPV types 6 and 11 are most commonly associated with genital warts.

Other types (eg, 16, 18, 31, 33, and 35) have a strong association with cervical and other anogenital cancers.

Symptoms include burning, itching, pain, and fullness (urethra, vagina, or anus); however, many patients are asymptomatic.

Presence of condylomata acuminata in a prepubertal child raises the suspicion of sexual abuse. Majority of venereal warts in children less than 2 years of age are acquired by nonsexual transmission.

Treatment:

Warts smaller than 1 cm at the base can be successfully treated by topical therapy alone; In contrast, diffuse and large lesions may require surgical intervention.

Topical treatments include Imiquimod (Aldara) 5% cream or Podofilox 0.5% gel or solution. Cryotherapy using liquid nitrogen, Podophylin resin 25%, or Trichloroacetic acid 80% can be applied to the lesion.

Quadrivalent HPV vaccine (Gardasil) against types 6, 11, 16, and 18 is now available for use in males and females aged 9 to 26 years. There is also a bivalent vaccine (Cervarix) available for girls aged 10 to 24 years.

Implications of condylomata acuminata in an adolescent: Genital HPV infection occurs in sexually active adolescents.

Herpes simplex (see also IX.H.3)

Diagnosing a genital herpes simplex virus infection: Usually diagnosis is made by clinical examination. However, herpes should be confirmed by isolating the virus or viral DNA by polymerase chain reaction (PCR), which is highly sensitive and specific. Virus culture remains the gold standard for diagnosing HSV infections.

In the evaluation of the neonate with suspected HSV infection cultures of suspicious lesions, eye and mouth swabs, and PCR of CSF should be included.

As HSV diagnostic tests take a few days to complete, treatment should not be withheld but promptly initiated to ensure the maximum therapeutic benefit.

Management of a genital HSV infection:

Acyclovir, valacyclovir, and famciclovir are the three drugs available for treatment of HSV infections. Early initiation of therapy results in the maximal therapeutic benefit.

Patients with genital herpes also require counseling to address psychosocial issues, including possible stigma, and to help them understand the natural history and management of this chronic infection.

Neurologic Disorders

Signs and symptoms of neurologic dysfunction
Headache
Differentiation

Know the elements of history that characterize a headache due to increased intracranial pressure:

Normal intracranial pressure (ICP) in adults is less than 10 to 15 mm Hg.

Presence of tumor or blood, hydrocephalus, cerebral edema, or increased cerebral blood flow due to head injury may raise the ICP. Increased ICP results in decreased cerebral perfusion and ischemia or mass effect with uncal herniation.

Immediate signs: altered mental status, headache, vomiting, and papilledema and retinal hemorrhages on fundoscopic examination.

In infants, before the cranial sutures and fontanels close, the head enlarges in response to increasing pressure.

Cushing triad = hypertension, bradycardia, and respiratory irregularity

(Kussmaul breathing).

Late findings: obtundation, 3rd or 6th cranial nerve compression (anisocoria or diplopia, respectively), and hemodynamic instability.

Physical characteristics of a headache due to increased intracranial pressure: Not well-characterized, usually bifrontal, worse in the morning, aggravated by straining or exercises, and often followed by vomiting and dehydration. Rehydration, even in small amounts, can cause a sudden rise in ICP and deterioration in patient's condition.

Elements of history that characterize a migraine:

Recurrent, episodic headaches with symptom-free intervals, intense frontal or temporal headaches that last 1 to 48 hours, with nausea, vomiting, photophobia, and phonophobia; occasionally, children can have neurologic signs along with the migraine.

Neurologic defects that can be associated with a migraine include hemiparesis, language disturbances or altered mental status, visual disorders, or oculomotor dysfunction.

Elements of history that characterize a headache due to stress, tension oremotion:

Mild to moderate "squeezing" or "bandlike" sensation with non-pulsatile quality and usually bilateral - bi-frontal or bi-occipital

Pain radiates to the posterior neck or causes tightness of the jaw muscles. Nausea, photophobia, and phonophobia are absent. Tension headaches are not exacerbated by routine physical activity and do not have prodromal or focal neurological symptoms unlike migraine.

These patients are often aware of stressful emotional triggers that precipitate their headaches.

Headaches can be caused by depression and is seen in up to one third of patients with persistent tension headaches.

Evaluation

Signs and symptoms of a headache that indicate a need for follow-up with MRI or CT scan:

Recent onset of severe headache, change in the type of headache, or neurologic dysfunction.

Children with abnormal neurologic examinations including focal findings, signs of increased ICP, altered consciousness, or seizure history.

Values and limitations of ancillary neurodiagnostic tests in the evaluation of a headache: Neuroimaging may be considered for children who have recurrent headache based on clues extracted from the medical history or findings on the neurologic examination.

Treatment

Abortive treatment of an acute migraine

Includes aggressive oral or intravenous hydration, sedation, and an antiemetic agent; Nasal or subcutaneous Sumitriptan may be used to abort the migraine. Anticonvulsants such as Valproic acid or Topiramate may be used as an abortive or prophylactic agent. Dihydroergotamine (DHE) can be used as abortive therapy; however, there is risk of rebound headaches.

Treatment of a headache due to stress, tension, or emotion: OTC pain medications such as Advil, Naproxyn or Ibuprofen. Tylenol may be less effective and can cause overuse headaches. Combinations medications such as Excedrin (ASA, acetaminophen and caffeine) can be more effective than single-ingredient pain relievers.

Potential complications of using narcotics, sedatives, and NSAIDs to treat a chronic or recurrent headache include loss of effectiveness, dependence issues, peptic ulcers if not taken with PPI, perforations and prolonged bleeding.

Prophylactic treatment for recurrent migraine:

Initiation of a prophylactic agent such as antihistamine Cyproheptadine (Periactin), tricyclic antidepressants (TCA) such as Amitriptyline, anticonvulsants such as Valproate or Topiramate, or a beta-blocker like Propranolol can be used to prevent headache occurrence.

Improvement of quality of life and reduction of stressors. Avoidance of acute headache medication escalation.

Avoidance of triggers such as foods like chocolate, caffeine, aged cheese, dairy products or stress, etc.

Take the medication as soon as headache begins.

Altered level of consciousness

General

Measure ammonia concentration and organic acid concentrations in neonatal coma.

Common causes of an altered level of consciousness (AEIOU TIPS): Alcohol, Abuse of substances

Epilepsy, Encephalitis, Electrolyte abnormalities, Endocrine disorder Insulin, Intussusception

Overdose, Oxygen deficiency (anoxia, hypoxia) Uremia (hemolytic-uremic syndrome)

Trauma, Temperature abnormality, Tumor, Toxins Infection, Inborn errors of metabolism

Poisoning, Psychiatric conditions

Shock, Stroke, Space-occupying lesion (intracranial – hydrocephalus, bleed, tumor, empyema)

Initial phase of evaluation for an altered level of consciousness Initially, Airway, Breathing & Circulation (ABCs)

Initially 100% oxygen is given by non-rebreather facemask if oxygenation is required. If respiratory depression or distress is observed, an oral airway or endotracheal intubation is necessary. Immediate intravenous access is required for fluids or medications. Next steps to determine etiology include laboratory testing such as CBC with differential, blood culture, BMP or CMP, serum and urine toxicology screens, ammonia, lactate, lumbar puncture, urinalysis, urine culture and other diagnostic studies such as EKG, CT head or MRI brain. If hypoglycemia is suspected, intramuscular glucagon should be administered emergently, plus intravenous glucose or dextrose.

Metabolic disorders of of the, liver, kidneys, lungs or heart can be manifested as encephalopathy.

Ingestions and intoxications

Ingestions that are likely to result in neurologic toxicity include: Amphetamines, Anticholinergics, Anticonvulsants Barbiturates, Benzodiazepines

Clonidine, Cocaine Ethanol Haloperidol Narcotics Phenothiazines

Salicylates, selective serotonin reuptake inhibitors (SSRIs)

TCAs

Child abuse

Historic findings that lead to consideration of child abuse as a cause of an altered level of consciousness include:

(1) the child presents with significant injuries and history of trauma is denied or does not explain the identified injuries, (2) the history changes over time or between caregivers, (3) a history of self-inflicted trauma that is beyond the child's developmental stage, (4) delay in seeking medical attention, (5) injuries in different stages of healing, (6) failure to thrive, and (7) any injury pathognomonic for child abuse.

Physical findings that lead to consideration of child abuse include:

Bruises, skeletal trauma, burns, head injuries, abdominal injuries including intramural hematomas of the small bowel, splenic or hepatic lacerations, traumatic pancreatitis, and renal contusions.

Ataxia

Common causes of acute ataxia

Encephalitis, Viral syndrome (transient myositis), Drug ingestion, Brain tumor, Conversion reaction, Genetic disorders, Migraine (basilar), Benign paroxysmal vertigo. Post-infectious/Immune causes are ADEM, acute post-infectious cerebellitis, Guillain-Barre and Miller Fisher syndrome. Other causes include epilepsy, trauma and vascular disorders.

Common causes of vertigo

Labyrinthitis, Benign paroxysmal vertigo, Vestibular neuritis, Herpes zoster oticus (Ramsay-Hunt Syndrome), Meniere disease, Acoustic neuroma, Aminoglycoside toxicity, Otitis media, Migraine, Chiari malformation, Multiple sclerosis.

Evaluation of a child with ataxia:

History includes recent illness, prior vaccinations, head trauma, toxic substance ingestion, altered mental status, fever, meningismus, onset of symptoms – sudden or insidious, family history.

Laboratory tests: CBC with differential, CMP, Serum and urine toxicology screen, Ammonia, Lumbar puncture, MRI Brain, CT head.

Prognosis of childhood acute cerebellar ataxia: Usually resolves without sequelae within 2-3 weeks of presentation. Approximately 10% have long-term neurologic sequelae.

Movement disorders (involuntary, paroxysmal)

Etiology, clinical features, prognosis, and treatment of Sydenham chorea (SC): Etiology: Sydenham chorea (SC) begins several weeks to several months after a Group-A Beta-hemolytic streptococcus (GABHS) infection with a 2:1 female predominance. Choreic movements become more obvious over a week and typically become generalized. Chorea is asymmetric and, in rare cases, unilateral.

Clinical features: insidious onset with progressive clumsiness and hypotonia, and behavior change including emotional lability. “Spooning sign” = hyperextension at the metacarpophalangeal joints; “milkmaid’s grasp” = chorea that is detected when the patient squeezes the examiner’s fingers in a sustained manner; and “darting tongue” = due to choreic movements of the tongue, “hung-up” tendon jerk reflexes = brisk reflex followed by a slowed return to the neutral position. Behavioral changes may include impulsivity, aggression, and obsessive- compulsive behaviors.

Diagnosis: Most children who have SC have positive ASO titers and anti-DNase B antibodies; more than 25% are serologically negative. Other laboratory tests include throat culture, thyroid function tests, ANA, ESR, CRP, serum ceruloplasmin level, antiphospholipid/anticardiolipin antibodies, urine and serum drug screens, and urine beta-HCG. MRI brain scans can show signal abnormalities in the basal ganglia; however, no sensitive or specific tests exist for SC.

Treatment: consists of secondary prevention, symptomatic treatment of chorea, and immune modulation. All children diagnosed as having SC should be treated with penicillin. Antiepileptic medications such as carbamazepine or valproate can be given.

Drugs that reverse the symptoms of drug-induced movement disorders include anticholinergic agents, beta blockers, or benzodiazepines. Antimuscarinic agents (Benztropin) or diphenhydramine can also reverse Extra Pyramidal Syndrome (EPS.)

Differentiate between tic disorder and Tourette syndrome:

Tics are repetitive, non-rhythmic, and intermittent muscle movements. When these movements involve laryngeal-pharyngeal muscles and produce noise, they are “phonic” or “vocal” tics. All other tics are “motor”. Tourette’s disorder requires the presence of both motor and phonic tics.

Tourette syndrome is associated with other co-morbidities including Attention Deficit Hyperactivity Disorder (ADHD) (50% of children with Attention Deficit Hyperactivity Disorder (ADHD) have Tourette sundrome), Obsessive-compulsive Disorder (OCD) behaviors, learning disabilities, anxiety or mood disorders, oppositional defiant disorder, speech and language disorders, and self-injurious behaviors.

Distinguish between tics and other involuntary movements:

Myoclonus is characterized by brief, abrupt, involuntary, non-suppressible, jerky contractions involving a single muscle or muscle group. Myoclonus can be rhythmic, in which case it often appears tremor-like.

Tremor is a rhythmic oscillation about a central point or position involving one or more body parts. Tremor is classified by when it occurs: with rest, intention, or action.

Common causes of chorea:

Primary chorea, uncommon in childhood, includes benign familial (hereditary) chorea and Huntington disease. Secondary chorea in childhood can be due to acute rheumatic fever (ARF), systemic lupus erythematosus (SLE), cardiac surgery, and perinatal hypoxia-ischemia.

Drugs that can cause movement disorders:

Dopamine antagonists such as Haloperidol, Chlorpromazine, Metoclopramide, Risperidone; Antiepileptic agents such as Phenytoin, Carbamazepine, and Valproate; Beta-adrenergic agents such as Albuterol, Metaproterenol; Amphetamines; Cocaine; and Lithium.

Increased intracranial pressure

Exclude a mass lesion by brain imaging in the presence of increased intracranial pressure.

Common causes of pseudotumor cerebri:

Drugs (sulfonamides, corticosteroids, nitrofurantoin, phenytoin, cytarabine, cyclosporine), Infections: acute sinusitis, otitis media, mastoiditis, measles, varicella), Renal (nephrotic syndrome, chronic renal insufficiency, malnutrition. Connective tissue disorders: antiphospholipid antibody syndrome, SLE, Behcet disease. Endocrine: menarche, polycystic ovarian syndrome, parathyroid dysfunction, CAH, Addison disease. Other: obesity, head trauma, superior vena syndrome, sleep apnea, Guillain-Barre syndrome, Turner syndrome, cystic fibrosis (CF), Down syndrome, (trisomy 21).

Need to measure the opening pressure at lumbar puncture.

Signs and symptoms of increased intracranial pressure in infants:

Bulging fontanel, suture diastasis, distended scalp veins, a persistent downward gaze or “sun-setting” of the eyes, rapid growth of head circumference. Infants do not develop papilledema or abducens palsy.

Signs and symptoms of increased intracranial pressure in children:

Major specific sign of increased ICP is dilation and poor reactivity of one or both pupils.

Most sensitive sign of increased ICP is depressed consciousness.

Headache, vomiting, lethargy, irritability, abducens palsy, strabismus, diplopia, and papilledema are seen in older children.

Contraindications of immediate examination of the cerebrospinal fluid is uncal herniation.

Plan the medical management of a patient who has increased intracranial pressure LP is contraindicated in patients with ICP until a CT head is done. Goal of management is to maintain adequate cerebral perfusion pressure. Management includes sedation, drainage of CSF after CT Head, and mannitol or hypertonic saline. For refractory causes, barbiturate coma, hypothermia, or decompressive craniectomy should be considered.

Weakness and hypotonia

Distinguish among acute and chronic causes of weakness

Acute causes: Cerebral cortex (intracranial hemorrhage, trauma, stroke, tumor, Todd paralysis, hemiplegic migraine); Spinal cord (trauma, tumor, discitis, epidural abscess, transverse myelitis); Anterior horn cell (poliomyelitis); Peripheral nerve (GBS, heavy metal poisoning, acute intermittent porphyria); Neuromuscular junction (botulism, myasthenia gravis, organophosphate poisoning, tick paralysis); Muscle (rhabdomyolysis, viral myositis, periodic paralysis); Electrolyte abnormality; Drug-related; and Conversion disorder.

Chronic causes: Cerebral cortex (CP, tumor); Spinal cord (myelomeningocele, tethered spinal cord, Chiari malformations, ALS); Anterior horn cell (spinal muscular atrophy); Peripheral nerve; Neuromuscular junction; Muscle; Electrolyte abnormality; Drug-related; and Conversion disorder.

Know the benefits and limitations of ancillary neurodiagnostic tests in the evaluation of weakness (eg, serum creatine kinase activity, electromyography).

Know the differential diagnosis of hypotonia in infants: Botulism, inborn errors of metabolism, cerebral palsy, chromosome abnormalities (eg. Down syndrome, Prader- Willi Syndrome).

Know how to evaluate hypotonia in infants: Brain imaging, laboratory tests like urine organic acids and serum amino acids, ammonia level, karyotype, thyroid function tests, creatine phosphokinase, and specific genetic tests.

Infection

Meningitis
Pathophysiology

Common acute complications of meningitis:

Seizures, subdural effusions, increased ICP, cranial nerve palsy, monoparesis, hemiparesis, gaze preference, aphasia, and ataxia can occur. SIADH is seen in some patients with bacterial meningitis.

Know the etiologies of neonatal meningitis

Group B Streptococcus, Listeria monocytogenes, Escherichia coli, H. influenza, HSV.

Etiologies of meningitis in children

Pneumococcus, Meningococcal disease (Neisseria meningitidis), and viral meningitides. In certain populations, H. influenza and Salmonella sp. are also causative agents.

Causes of meningitis when no bacteria are isolated (eg, partially treated, parameningeal focus, Borrelia, spirochete, M. tuberculosis). Aseptic meningitis is caused by infectious and non-infectious agents including enteroviruses

Diagnosis

Distinguish among cerebrospinal fluid findings in bacterial, fungal, and viral meningitis.

Clinical manifestations of bacterial meningitis:

Sudden onset with rapid progression to shock, purpura, DIC, and obtundation, coma or death within 24 hours.

More often, meningitis is preceded by fever due to upper respiratory tract or gastrointestinal symptoms, followed by poor feeding, headache, myalgias, arthralgias, hypotension, petechiae, purpura, or an erythematous macular rash.

Patients present with nuchal rigidity, back pain, positive Kernig sign and positive Brudzinski sign.

Seizures due to cerebritis, infarction, or electrolyte disturbances occur in 20- 30% of patients.

Differential diagnosis of fever and petechiae or purpura in bacterial meningitis must include Rocky Mountain spotted fever, meningococcemia, and ehrlichiosis.

Clinical manifestations of aseptic meningitis:

Present with a nonspecific febrile illness. Meningoencephalitis or encephalitis presentations often predominate with pathogens such as HSV, Mycoplasma, arboviruses, Epstein-Barr virus, rabies virus, human herpesvirus-6, Ehrlichia sp, and Rickettsia rickettsii.

Laboratory diagnosis of aseptic meningitis:

On CSF: Pancytopenia may be present. Neutrophils predominate early in the course of infection, and shifts to lymphocytic predominance during the illness. Glucose and protein concentrations frequently are normal. Gram stain is negative. Enteroviral PCR can confirm the diagnosis. Viral cultures can be sent from mucous membranes and CSF.

Management

Manage subdural effusion associated with bacterial meningitis

Signs of increased intracranial pressure in the presence of a subdural effusion indicate the need for neurosurgical drainage.

Acceptable treatments of meningitis include:

Ampicillin (300 mg/kg per day divided every 6 hours) and cefotaxime (200 to 300 mg/kg per day divided every 6 hours) is appropriate. Acyclovir (60 mg/kg per day divided every 8 hours) should be added if HSV infection is a concern. In the young infant, if the Gram stain suggests pneumococcus, vancomycin (60 mg/kg per day given every 6 hours) should be added. An alternative therapy for children who have had anaphylactic reactions to penicillin or cephalosporins is a carbapenem or a quinolone in addition to vancomycin. Consultation with an infectious disease specialist may be helpful in such cases.

Know the management of cerebral edema in meningitis:

Treatment depends on the severity and begins with fluid restriction. These patients require diuretics, mannitol, or hypertonic saline. Invasive measurement of intracranial pressure and serial imaging should be considered.

Importance of monitoring fluid balance and electrolyte concentrations in meningitis:

SIADH in children who have bacterial meningitis can occur with rates between 7% and 89%. Initial moderate fluid restriction with isotonic fluid, especially if the serum sodium value is less than 130 mEq/L (130 mmol/L).

Indications for diagnostic imaging in patients with meningitis:

Lumbar puncture should be deferred until CT scan is performed. If a mass lesion, hemorrhage, midline shift, effacement of the basilar cisterns, or effacement of the sulci is noted, lumbar puncture should be deferred and antimicrobial therapy started promptly. MRI may be a more appropriate study to assess complicated meningitis.

Auditory testing should be obtained prior to discharge following meningitis to rule out hearing loss. Hearing loss can occur in 30% of patients with pneumococcal meningitis.

Potential long-term sequelae of meningitis:

For neonates, mortality rates of 10% in GBS meningitis and up to 20% in E coli meningitis, and neurologic sequelae are common. Long-term sequelae occur in 30% of those who have GBS disease and 50% of those who have gram-negative meningitis. The prognosis for patients with TB meningitis and neonatal HSV disease is extremely guarded.

Encephalitis

Pathophysiology

Common causes of encephalitis:

Viral (herpes, enteroviruses, adenoviruses, togaviruses), Bacterial (Listeria monocytogenes, Francisella tularensis, Rickettsia species, Mycoplasma pneumonia, Chlamydia pneumoniae), Fungal (Cryptococcus neoformans, Blastomyces dermatitidis, Histoplasma capsulatum, Paracoccidioides brasiliensis), Parasitic infections, Immune-mediated response (ADEM, acute hemorrhagic leukoencephalitis, Mycoplasma encephalitis), SLE, and Malignancies

Diagnosis

Signs and symptoms of herpes encephalitis:

HSV-1 causes rapid development of fever, headache, confusion, and seizures in the child; progressive obtundation with focal disturbances (eg. aphasia or hemiparesis) may develop. Untreated, the mortality approaches 70%.

Recognize the cerebrospinal fluid findings in herpes encephalitis: CSF shows pleocytosis (early neutrophils, late lymphocytes) and elevated protein due to

brain necrosis of the frontal and temporal lobes. Thus, the lumbar puncture is bloody. HSV can be detected in CSF by PCR.

Clinical symptoms of encephalitis: altered mental status, seizures, weakness, sensory disturbances, non-epileptic movement disorders, delirium, autonomic instability, and respiratory depression.

Role of neurodiagnostic testing in the evaluation of a child with encephalitis: Neuroimaging, including MRI, early in the course. CT imaging for substantial cerebral edema, midline shift or hemorrhage; not sufficient for diagnosis.

Appropriate microbiologic, serologic, and molecular diagnostic tests in a child with encephalitis: CSF for pressure (elevated), cell count (WBCs elevated), protein (elevated, especially IgG fraction), glucose (normal), RBCs (suggestive of herpesvirus or other necrotizing virus). CSF cultures should be sent for viruses, bacteria, fungi, and mycobacteria (low yield). CSF PCR for CMV, HSV, VZV, enterovirus and West Nile virus.

Management

Know how to manage encephalitis:

Treatment of acute symptoms often takes priority including respiratory, cardiac, and neurologic support. Seizures are treated with benzodiazepines, followed by loading dose of fosphenytoin, phenobarbital, or phenytoin, but newer agents such as divalproex or levetiracetam are also successful. Sedatives are the next most likely treatment, including the use of midazolam, pentobarbital, propofol, or ketamine drips, if seizures are refractory to first- line treatments. When symptomatic intracranial hypertension exists, mannitol or hypertonic saline can be used on a limited basis.

Cerebral perfusion pressure (CPP) should be generally maintained as 70 mm Hg or higher over 2 years of age. Preserving CPP by managing intracranial pressure is an important element of treatment.

Infectious encephalitis is treated with intravenous antibiotics and acyclovir while awaiting lumbar puncture culture results or for laboratory results, including HSV PCR.

ADEM is the most common cause of non-infectious encephalitis. High-dose corticosteroids are a first-line treatment, followed by IVIG or plasmapheresis in refractory cases.

Know the common sequelae of encephalitis:

Sequelae can include behavioral problems such as ADHD and conduct disorders and persistent seizures.

Abscess

Signs and symptoms

Know the clinical manifestations of brain abscess: characterized by nystagmus, ipsilateral ataxia and dysmetria, vomiting, and headache. If the abscess ruptures into the ventricular cavity, overwhelming shock and death usually ensue.

Pathophysiology

10% of brain abscesses contain multiple organisms.

Most common bacteria include Streptococci (Streptococcus pyogenes group A or B, Streptococcus pneumoniae, Enterococcus faecalis), gram-negative aerobic bacilli (H. influenzae, Enterobacter, E. coli, Proteus spp.), and anaerobes (Bacteroides spp., Fusobacterium spp., Prevotella spp., Actinomyces spp.).

Fungal abscesses (Aspergillus, Candida) are more common in immunosuppressed patients.

Diagnosis

CT Head should be done before lumbar tap in a suspected brain abscess.

Imaging techniques for diagnosing brain abscess: CT with contrast and MRI are the most reliable methods of demonstrating cerebritis and abscess formation. An abscess cavity shows a ring-enhancing lesion by contrast CT, and MRI demonstrates an abscess capsule with gadolinium administration.

Management

Treatment of brain abscess:

Combination of Vancomycin, a 3rd-generation Cephalosporin, and Metronidazole is commonly used. Meropenem has good activity against gram- negative bacilli, anaerobes, staphylococci (except MRSA), and streptococci (except penicillin-resistant strains of S. pneumonia). In immunocompromised patients, amphotericin B should be added to the broad-spectrum antibiotic regimen.

An encapsulated abscess should be treated with a combination of antibiotics and aspiration. Surgery is indicated when the abscess is >2.5 cm in diameter, gas is present in the abscess, the lesion is multiloculated, located in the posterior fossa, or a fungus is identified.

The duration of antibiotic therapy is usually 4 to 6 weeks.

There is a propensity for brain abscess to complicate cyanotic heart disease, sinusitis, and pulmonary disease due to embolization.

Myelitis

Signs and symptoms

Clinical manifestations of myelitis:

Bilateral sensorimotor and autonomic spinal cord dysfunction

Demonstration of spinal cord inflammation with CSF pleocytosis or elevated IgG index, or MRI revealing a gadolinium-enhancing cord lesion

Exclusion of compressive, post-radiation, neoplastic, and vascular causes

The IgG index is a measure of intrathecal synthesis of immunoglobulin and is calculated with the use of the following formula: (CSF IgG ÷ serum IgG) ÷ (CSF albumin ÷ serum albumin).

Diagnosis

Evaluation of a patient in whom post-infectious myelitis is suspected: MRI with and without contrast is essential to rule out mass-enhancing lesion requiring neurosurgical intervention. Lumbar puncture can be performed if no spinal cord compression. CSF should be evaluated for myelin basic proteins and immunoglobulin levels, which are elevated in transverse myelitis. The presence of inflammatory cells confirms the diagnosis of transverse myelitis. Serum studies should be sent for autoimmune disorders such as SLE.

Degenerative conditions

Signs and symptoms

Historical features indicative of a degenerative CNS disorder include:

Progressive loss of previously acquired abilities where infants and young children show a deceleration in the rate of development and subsequently lose previously acquired milestones.

Acquisition of new developmental milestones does not exclude a degenerative disorder, eg. Healthy infants develop sporadically, with a child often appearing to have reached a plateau for several weeks. Prolongation of plateau state may be only indication to degenerative condition.

Signs and symptoms of degenerative CNS disorders include:

Gray-matter disease leads to early onset of dementia, progressive loss of cognitive abilities, and seizures, frequently myoclonic. Extrapyramidal and cerebellar signs, such as ataxia, are common. Ganglion cells of the retina are affected in many of the gray-matter disorders, producing pigmentary degeneration of the retina.

White-matter disease results in abnormal myelin that breaks down rapidly. The earliest sign is spasticity. Dementia and seizures can occur, but usually later in the clinical course. Extrapyramidal signs are rare, but involvement of cerebellar pathways can cause ataxia. Optic atrophy is the most characteristic ocular change seen in white-matter disease. Some patients even have cortical blindness from demyelination of the optic pathways.

Clinical presentation and course of Rett syndrome (RS):

Usually the antenatal and birth histories are normal, but most girls affected with RS pass through four predictable stages:

Stage 1 (birth to 18 months): Deceleration of head growth, beginning between 2 and 4 months of age, results in an acquired microcephaly. These babies show disinterest in their environment and become markedly hypotonic.

Stage 2 (1 to 2 years): The loss of both expressive language and gross motor milestones, and the onset of abnormal hand movements, seizures, irritability, and insomnia.

Stage 3 (2 to 10 years): Severe mental retardation, seizures, and persistent stereotypic hand wringing movements persist. Most patients develop ataxic and wide-based gaits, but many affected girls never walk. Tremors and breath- holding spells are common.

Stage 4 (older than 10 years): As girls who have RS approach adolescence, they develop progressive scoliosis, muscle wasting, and can become wheelchair-bound. By the early adolescent years, they reach a plateau in their neurologic regression, but death usually occurs during late adolescence from infection or cardiac arrhythmia.

Diagnosis

Initial evaluation of a patient with suspected CNS degenerative disease:

Laboratory test: CBC, urine and serum toxicology screens, serum ammonia, chromosomal studies.

Diagnostics: Magnetic resonance imaging (MRI) with gadolinium enhancement can reveal plaques on the brain and spinal cord. A lumbar puncture can be done to measure levels of immune proteins, which are usually elevated in the CSF of a person with multiple sclerosis (MS).

Developmental malformation, static neurologic deficit

Malformations
Spinal dysraphism

Myelomeningocele is usually associated with hydrocephalus.

Most common orthopedic problems associated with a myelomeningocele and their relative significance:

Usually present with a high incidence of lower extremity deformities (clubfeet, ankle and/or knee contractures, and subluxation of the hips). Infants with myelomeningocele typically have an increasing neurologic deficit as the myelomeningocele extends higher into the thoracic region. These infants sometimes have an associated kyphotic gibbus that requires neonatal orthopedic correction.

Understand the evaluation and fundamental long-term management of neurogenic bladder:

A low sacral lesion leads to bowel and bladder incontinence associated with saddle anesthesia but with no impairment of motor function.

Careful evaluation and reassessment of the genitourinary system are some of the most important components of the management. Regular catheterizations of a neurogenic bladder is a crucial step in maintaining a low residual volume and bladder pressure that prevents UTIs and reflux leading to pyelonephritis, hydronephrosis, and bladder damage.

Periodic urine cultures and assessment of renal function, including serum electrolytes and creatinine, renal scans, vesicourethrograms (VCUGs), renal ultrasonography, and cystometrograms (CMGs), are obtained according to the risk status and progress of the patient and the results of the physical examination. Some children can become continent with surgical implantation of an artificial urinary sphincter (these are used less often) or bladder augmentation at a later age.

Know the fundamental long-term management of a neurogenic bowel: Incontinence of fecal matter is common and can occasionally cause fecal impaction and/or megacolon. Many children can be bowel-trained with a regimen of timed enemas or suppositories that allows evacuation at a predetermined time once or twice a day. Special attention to low anorectal tone and enema administration and retention is often required.

Know the clinical and radiographic features and prognosis of spina bifida occulta: Clinical features: midline defect of the vertebral bodies without protrusion of the spinal cord or meninges. Most patients are asymptomatic and lack neurologic signs. This simple defect does not have an associated spinal cord malformation. Occult spinal dysraphism (OSD)can have cutaneous manifestations such as a hemangioma, discoloration of the skin, pit, lump, dermal sinus, or hairy patch.

Diagnostic: Spinal radiograph shows a defect in closure of the posterior vertebral arches and laminae, typically involving L5 and S1; there is no abnormality of the meninges, spinal cord, or nerve roots. Occult spinal dysraphism is often associated with developmental abnormalities of the spinal cord, including syringomyelia, diastematomyelia, and/or a tethered cord. A spine radiograph in these cases might show bone defects or may be normal. All cases of occult spinal dysraphism are best investigated with MRI. Initial screening in the neonate may include ultrasonography.

Prognosis: For a child who is born with a myelomeningocele, the mortality rate is 10-15%, and most deaths occur before age 4 years. At least 70% of survivors have normal intelligence, but learning problems and seizure disorders are common. Multidisciplinary follow-up is required for life. Renal dysfunction is one of the most important determinants of mortality.

Know the differential diagnosis of acute neurologic deterioration in a child with myelomeningocele:

Tethered spinal cord, Chiari II malformation, Syringomyelia

Identify the clinical manifestations and plan the diagnostic evaluation of occult spinal dysraphism:

Clinical manifestations: Most forms of Occult Spinal Dysraphism OSD have an associated overlying cutaneous abnormality such as hypertrichosis, capillary hemangioma, atretic meningocele, subcutaneous mass (eg, lipoma), or a caudal appendage. Gluteal cleft anomalies have a weak association with milder forms of OSD and warrant further evaluation. Criteria to differentiate and define dimples includes: multiple dimples, dimple diameter larger than 5 mm, location greater than 2.5 cm above the anal verge, and association of the dimple with other cutaneous markers.

Diagnostic evaluation: Not all dimples are associated with a spinal dysraphism; distinction between cutaneous stigmata associated with OSD and innocent skin dimples can be difficult and may lead to unnecessary tests or referrals. Either ultrasonography or MRI can be used to evaluate OSD. Ultrasonography is useful in children younger than 3 months of age because ossification of the vertebral arches has not yet occurred. Recognition of a suspicious skin dimple with prompt radiographic evaluation and neurosurgical referral is crucial.

Importance of folic acid supplementation in the prevention of neural tube defects: Folate is intricately involved in the prevention and etiology of Neural Tuber Defects (NTDs). Folate coenzymes are involved in DNA synthesis, purine synthesis, generation of formate into the formate pool, and amino acid interconversion; the conversion of homocysteine to methionine provides methionine for the synthesis of S-adenosyl-methionine (SAM-e, an agent important for in vivo methylation). Maternal periconceptional use of folic acid supplementation reduces the incidence of NTDs in pregnancies at risk by at least 50%. To be effective, folic acid supplementation should be initiated before conception and continued until at least the 12th wk of gestation, when neurulation is complete. The mechanisms by which folic acid prevents NTDs remain poorly understood.

Head size and shape

Clinical features of hydrocephalus:

In an infant, an accelerated rate of enlargement of the head is the most prominent sign. The anterior fontanel is wide open and bulging, and the scalp veins are dilated. The eyes might deviate downward because of impingement of the dilated suprapineal recess, producing the “setting-sun” eye sign. Brisk tendon reflexes, spasticity, clonus (particularly in the lower extremities), and Babinski sign are common owing to stretching and disruption of the corticospinal fibers from the leg region of the motor cortex. In an older child, the cranial sutures are partially closed so that the signs of hydrocephalus may be subtler. Irritability, lethargy, poor appetite, and vomiting are common, and

headache is a prominent symptom in older patients. A gradual change in personality and deterioration in academic productivity suggest a slowly progressive form of hydrocephalus.

Signs and symptoms of shunt malfunction in hydrocephalus Headache, irritability, lethargy

Vomiting, anorexia

Increasing head circumference, bulging anterior fontanel Esotropia, diplopia, paralysis of upward gaze

Cognitive change, such as decline in school performance Moodiness, change in personality

Signs of Chiari malformation (dysphagia, stridor, aspiration, apnea)

Signs of tethered spinal cord (weakness in lower extremities, deterioration of gait, pain in back/legs, sensory loss in lower extremities, local swelling in the back, new decubitus ulcer)

Differential diagnosis of microcephaly

Familial microcephaly, Down syndrome, Trisomy 18, Cri-du-chat (5p-), Rubenstein Taybi syndrome, Congenital infections (TORCH), Drugs (Fetal alcohol, fetal hydantoin), in utero radiation exposure, meningitis, encephalitis, metabolic syndromes, hypoxic-ischemic encephalopathy (HIE), Rett syndrome.

Differential diagnosis of macrocephaly

Familial megalencephaly, non-communicating hydrocephaly, pseudotumor cerebri, IVH, hygromas, neurocutaneous syndromes (neurofibromatosis), genetic conditions (Sotos syndrome), infant of diabetic mother, Chiari II malformation, aqueductal stenosis, lysosomal storage disorders (Hurler's MPS, Tay-Sachs disease), Cretinism.

Know the causes and management of abnormal head shape:

Causes: head might appear enlarged and can be confused with hydrocephalus secondary to a thickened cranium resulting from chronic anemia, rickets, osteogenesis imperfecta, and epiphyseal dysplasia. Chronic subdural collections can produce bilateral parietal bone prominence. Various metabolic and degenerative CNS disorders produce megalencephaly including lysosomal diseases (Tay-Sachs, gangliosidosis, mucopolysaccharidoses), the aminoacidurias (maple syrup urine disease), and the leukodystrophies (metachromatic leukodystrophy, Alexander disease, Canavan disease). Familial megalencephaly is inherited as an autosomal dominant trait and is characterized by delayed motor milestones and hypotonia but normal or near- normal intelligence.

Management: Measurement of parents' head circumferences is necessary to establish the diagnosis. Diagnostics such as MRI of the brain can be helpful in determining the diagnosis. Neurosurgical intervention may be necessary.

Cerebral palsy (CP)

Recognize the clinical features of cerebral palsy, including classifications

Clinical features: CP is commonly associated with a spectrum of developmental disabilities, including mental retardation, epilepsy, and visual, hearing, speech, cognitive, and behavioral abnormalities.

Risk factors associated with cerebral palsy:

The prevalence of CP has increased somewhat due to the enhanced survival of very premature infants weighing <1,000 g, who go on to develop CP at a rate of approximately 15/100. The major lesions are intracerebral hemorrhage and periventricular leukomalacia (PVL).

Disabilities associated with cerebral palsy: cognitive, visual, communication, auditory, motor, seizure activity, behavioral, oral function, nutritional imbalances.

Principles of management for children with cerebral palsy (eg, feeding, spasticity, mobility, activities of daily living, education).

Multi-specialty physicians, occupational and physical therapists, speech pathologists, social workers, educators, and developmental psychologists provide important contributions to the treatment of those children who develop CP.

Several drugs have been used to treat spasticity, including the benzodiazepines and baclofen. These medications have beneficial effects in some patients, but can also cause side effects such as sedation for benzodiazepines and lowered seizure threshold for baclofen.

Botulinum toxin injected into specific muscle groups for the management of spasticity shows a very positive response in many patients. Botulism toxin injected into salivary glands may also help reduce the severity of drooling, which is seen in 10-30% of patients with CP and has been traditionally treated with anticholinergic agents.

Patients with rigidity, dystonia, and spastic quadriparesis sometimes respond to levodopa, and children with dystonia may benefit from carbamazepine or trihexyphenidyl.

Seizures

General
Pathophysiology

Metabolic causes: Electrolyte imbalance, Hypercarbia, Hypoxia, Hypoglycemia, Liver failure, DKA, Hypoxic-ischemic encephalopathy, inborn errors of metabolism, Drug ingestion, Uremia, Pyridoxine deficiency.

Drugs that may precipitate or exacerbate seizures

Alcohol, amphetamines, antihistamines, anticholinergics

Cocaine, carbon monoxide Isoniazid

Lead, lithium, lindane

Oral hypoglycemics, organophosphates Phencyclidine, phenothiazines Salicylates, sympathomimetics

Tricyclic antidepressants, theophylline, topical anesthetics Withdrawals (alcohol, anticonvulsants)

Most common causes of acute seizures

Infectious (brain abscess, encephalitis, meningitis), Neurologic (birth injury, congenital anomalies, degenerative CNS disease, HIE, neurocutaneous syndromes, VP shunt malfunction), Metabolic (hypercarbia, hypocalcemia, hypoglycemia, hypoxia), trauma, cerebral contusion, cerebrovascular accident (CVA), child abuse, head trauma,intracranial hemorrhage (ICH), Toxicologic (alcohol, amphetamines, cocaine, opiates, lead, lithium, TCAs, salicylates, withdrawal symtpoms), oncologic, obstetric (eclampsia), and idiopathic.

Diagnosis

Distinguish among epileptic seizures and paroxysmal non-epileptic events (eg, breath-holding spells, tics, self-stimulation, syncope, gastroesophageal reflux, psychogenic seizures, sleep disturbances).

Factors associated with an increased risk of seizure disorder:

SGA infant, genetic abnormality, brain injury, complex febrile seizure, inborn errors of metabolism, diabetes mellitus, nutritional deficiencies, meningitis encephalitis.

Etiologic and therapeutic implications of partial versus generalized seizures Generalized seizures: involve both cerebral hemispheres. Types of generalized seizures include tonic-clonic (grand mal), tonic, clonic, myoclonic, atonic- akinetic (drop attacks) or absence (petit mal).

Partial seizures: may be simple, with no impairment of consciousness, or complex, with altered mental status. Both simple and complex partial seizures may progress to secondarily generalized seizures in up to 30% of children.

Many anti-epileptic medications are available to control seizures. Lamotrigine has been shown to be effective for partial seizures, and valproate has been shown to be effective for generalized and unclassified epilepsies.

Know how to manage a child following a first seizure:

According to the AAP, a CT or MRI is not recommended in evaluating the child after a first simple febrile seizure. The work-up of children with complex febrile seizures needs to be individualized.

Lumbar puncture is recommended in children less than 12 months of age after their first febrile seizure to rule out meningitis. An EEG would not predict the future recurrence of febrile seizures or epilepsy even if the result is abnormal. Blood glucose should be determined only in children with prolonged postictal obtundation or those with poor oral intake (prolonged fasting).

Know how to manage a child with recurring seizures.

Testing in infants and children with recurrent seizures can include measure- ment of serum lactate, pyruvate, acyl carnitine profile, creatine, very long chain fatty acids, and guanidino-acetic acid. CSF glucose testing looks for glucose transporter deficiency, and CSF can be examined for cells and proteins (for parainfectious and postinfectious syndromes). Urine is tested for urinary sulfite indicating molybdenum cofactor deficiency and for oligosaccharides and mucopolysaccharides. Muscle biopsy can be performed for mitochondrial enzymes, and skin biopsy for inclusion bodies seen in neuronal ceroid lipofuschinosis is sometimes needed.

Management includes trial of anti-epileptic medications to control seizure activity and neurologic evaluation and follow-up.

Management plan for a patient with psychogenic nonepileptic seizure (PNES) After EEG shows non-epileptiform activity, and other causes of seizures have been ruled out, psychiatric evaluation is also initiated to manage psychogenic seizures. Age appropriate patient education is crucial.

Treatment

Drug selection is based on seizure type.

At present, the understanding of the pathophysiology of epilepsy does not allow specific choice of Antiepileptic Drugs (AEDs) based on the assumed pathophysiology of the epilepsy. However, in general, it is believed that it is better to avoid combining medications that have similar mechanisms of action, such as phenytoin and carbamazepine (both work on sodium channels). A number of medications, such as lamotrigine and valproate or topiramate and lamotrigine, have been reported to have synergistic effects, possibly because they have different mechanisms of action.

Understand the indications for discontinuing anticonvulsant therapy: Discontinuation of AEDs is indicated when children are seizure-free for at least 2 years.

Factors to consider before discontinuing medication include the likelihood of remaining seizure-free after drug withdrawal based on the epilepsy type and etiology, the risk of injury in case of seizure recurrence (e.g., if the patient drives), and the adverse effects of AED therapy.

AED therapy should be discontinued over a period of 3 to 6 months. Abrupt discontinuation can result in withdrawal seizures or status epilepticus, especially with phenobarbital and benzodiazepines.

Indications for initiating anticonvulsant therapy include generalized seizures or partial seizures that are affecting child's ability to function normally.

Relationship between etiology and prognosis in seizures: Based on the etiology, prognosis can determine duration of treatment.

Know the side effects and toxicities of anticonvulsants.

Valproate, carbamazepine, phenobarbital, and phenytoin, are associated with teratogenic effects. ~10% of patients develop reversible dose-related leukopenia on carbamazepine or phenytoin. Gum hyperplasia seen with phenytoin necessitates good oral hygiene. Allergic rash, and Stevens Johnson reactions, is common with lamotrigine, carbamazepine, and phenytoin.

Irreversible hepatic injury and death are feared in young children (less than 2 years old) who are on valproate in combination with other AEDs, particularly those with inborn errors of metabolism such as acidopathies and mitochondrial disease.

Know the value, limitations, and timing of serum drug concentration determinations during the management of seizures.

Serum levels of many AEDs should be determined after initiation to ensure compliance and therapeutic concentrations. Monitoring is helpful for phenytoin, carbamazepine, valproate, phenobarbital, and ethosuximide after starting maintenance dosage or any change in the dosage. A steady state is not reached until 5 half-lives have elapsed which, for most AEDs, is 2 to 7 days.

Know the laboratory abnormalities caused by anticonvulsants.

Carbamazepine or phenytoin can cause aplastic anemia or agranulocytosis. Frequent (even weekly) monitoring of liver function and of blood counts throughout the therapy is required for felbamate, due to the high incidence of liver and hematologic toxicity (1:500).

Laboratory monitoring is more relevant early on, because adverse effects such as allergic hepatitis and agranulocytosis are more likely to occur in the first 3 to 6 months of therapy.

Know the interactions of anticonvulsants with other drugs, including other anticonvulsants.

Understand the cognitive/behavioral consequences of treatment with anticonvulsants.

AEDs may be associated with an increased risk of suicidal ideation and requires counseling of adolescents and adults and those on AEDs for purposes other than the treatment of epilepsy (e.g., chronic pain) about this side effect before starting these medications.

Understand the cognitive and behavioral problems associated with seizure disorders: are associated with increased incidence of ADHD/ADD.

Management and monitoring

Provide appropriate counseling regarding activities and behavior of a child with a seizure disorder (eg, athletics, school, driving, medications) and psychosocial effects of epilepsy.

Know the effects of epilepsy and anticonvulsant therapy on reproductive health (eg, contraception) and the fetus, especially with Valproate, carbamazepine, phenobarbital, and phenytoin.

Neonatal

Clinical manifestations of neonatal seizures:

Five main neonatal seizure types: subtle, clonic, tonic, spasms, and myoclonic.

Subtle seizures include transient eye deviations, nystagmus, blinking, mouthing, and abnormal extremity movement; More common in premature than in full-term infants.

Clonic seizures can be focal or multifocal. Generalized bilateral clonic seizures are uncommon in the neonatal period.

Tonic seizures can be focal or generalized (generalized are more common).

Spasms are sudden generalized jerks lasting 1-2 sec that are short duration and usually associated with a single, very brief, generalized discharge.

Myoclonic seizures can be distinguished from clonic seizures by the rapidity of the jerks and by their lack of rhythmicity.

Although neonatal EEG interpretation is very difficult, an abnormal background in a premature or newborn infant is a powerful predictor of less- favorable later outcome. The underlying etiology of the seizures is the main determinant of outcome.

Febrile

General

Natural history of febrile seizures: occur between ages 6 months and 5 years with a temperature of 38°C or higher, that are not due to CNS infection or metabolic imbalances, and that occur in the absence of prior afebrile seizures.

Simple febrile seizure: primary generalized, usually tonic-clonic, attack associated with fever, lasting for a maximum of 15 minutes, and not recurrent within a 24-hour period.

Complex febrile seizure: more prolonged (>15 minutes), is focal, and may recur within 24 hours.

Febrile status epilepticus: febrile seizure lasting longer than 30 minutes.

Risk factors associated with febrile seizures related to later epilepsy include neurodevelopmental abnormalities (33%), focal complex febrile seizures (29%), family history of epilepsy (18%), fever less than one hour before febrile seizure (11%), complex febrile seizure any type (6%), or recurrent febrile seizures (4%).

Having no risk factors carries a recurrence risk of about 12%; 1 risk factor, 25-50%; 2 risk factors, 50-59%; 3 or more, 73-100%.

Diagnosis

Appropriate evaluation of a child with febrile seizures:

History, physical examination, manage acute febrile seizure and acute illness (first-aid, midazolam, diazepam, diagnostic tests) as needed. Determine risk factors for recurrence and estimate recurrence risk. Counsel parents about recurrence risk and how to provide first-aid and manage fever. Determine risk factors for later epilepsy -- Low risk: No therapy or investigations are necessary. Intermediate or high risk: 1. Consider EEG and imaging 2. Consider intermittent oral diazepam or, in exceptional cases that recur, continuous anti-epileptic therapy.

Treatment

Management of febrile seizures: antiepileptic therapy, continuous or intermittent, is not recommended for children with one or more simple febrile seizures. If the seizure lasts for longer than 5 minutes, then acute treatment with diazepam, lorazepam, or midazolam is needed. Rectal diazepam is often prescribed to be given at the time of recurrence of febrile seizure lasting longer than 5 minutes. Intravenous benzodiazepines, phenobarbital, phenytoin, or valproate may be needed in the case of complex febrile seizures or febrile status epilepticus.

Infantile spasms

Signs and symptoms

Recognize the characteristic clinical features of infantile spasms (or West syndrome).

Presents typically between 4 and 18 months of age, with males affected more commonly than females.

Symptoms include sudden jerking movements of the extremities, head, and trunk, which is spasmodic and often occurs in clusters. Episodes rarely occur during sleep. Up to 25% of patients have tuberous sclerosis.

Pathophysiology:

The EEG shows hypsarrhythmia (random high-voltage slow waves with multifocal spikes).

Treatment

Adrenocorticotropic hormone (ACTH) and prednisone have been used with some success. Valproic acid, topiramate, lamotrigine, vigabatrin, and zonisamide have also shown some effectiveness.

Prognosis for children with infantile spasms:

95% of affected children are mentally retarded, and there is a 20% mortality rate.

Absence epilepsies (petit mal)

Drugs used to treat absence epilepsy: Ethosuximide is the drug of choice. Side-effects:

GI upset, weight gain, headache, rarely erythema multiforme, lupus-like syndrome.

Recognize the characteristics of absence epilepsy:

Generalized seizures with staring, unresponsiveness, eye flutter of few seconds duration.

Typical absences are associated with 3 Hz spike and slow wave discharges and with petit mal epilepsy, which has a good prognosis.

Atypical absences are associated with 1-2 Hz spike and slow wave discharges, with head atony and myoclonus during the seizures.

Complex partial seizures

Recognize the clinical features of complex partial epilepsy:

Usually last 1 to 2 minutes and are often preceded by an aura, such as a rising abdominal feeling, déjà vu or déjà vécu, a sense of fear, complex visual hallucinations, micropsia or macropsia (temporal lobe), generalized difficult-to- characterize sensations (frontal lobe), focal sensations (parietal lobe), or simple visual experiences (occipital lobe).

Presents with decreased responsiveness, staring, and automatisms; often there is salivation, dilation of the pupils, and flushing or color change.

Understand the drugs used to treat complex partial seizures:

Lamotrigine: interferes with other anticonvulsant agents and dosage should be adjusted in conjunction with other antiepileptic medications. Lamotrigine is generally well-tolerated, with side-effects including GI upset, somnolence, dizziness, headache, and diplopia. Adverse effect is the development of Stevens- Johnson syndrome, common in patients who are also taking Valproate.

Topimarate: interaction with other anticonvulsant agents is minor. Several adverse effects can occur including nephrolithiasis, behavioral problems, anorexia, weight loss, sleep problems, fatigue, headache, diplopia, speech problems, and confusion. Its use should be carefully considered in patients who have a history of kidney stones or those on a ketogenic diet.

Status epilepticus

Measure serum glucose, electrolyte, calcium, and magnesium concentrations in a patient with status epilepticus

Management requires securing airway, breathing, and circulation (with continuous monitoring of vital signs including ECG) and determination and management of the underlying etiology (e.g., hypoglycemia).

EEG is helpful in ruling out pseudo–status epilepticus (psychological conversion reaction) and in identifying the type of status epilepticus (generalized versus focal), which can guide further testing for the underlying etiology and further therapy.

Neuroimaging should include CT and/or MRI brain once the child is stabilized, especially if it is indicated by the clinical manifestations.

Medications that can be administered rectally to treat status epilepticus:

Initial therapy is IV lorazepam (Ativan), effective with fewer side-effects than IV Diazepam. If no IV access, then buccal midazolam or intranasal lorazepam are two effective options. Nasal midazolam and rectal diazepam have also been used, but less evidence to support use in status epilepticus.

Patient should be monitored and managed for respiratory depression.

Fosphenytoin can be used with loading dose usually 15 to 20 mg/kg. Other medications include Phenytoin, Phenobarbital, Valproate.

For refractory status epilepticus, IV bolus of midazolam, propofol, pentobarbital, or thiopental is initially used with maintenance of a corresponding continuous IV infusion.

Possible etiologies of status epilepticus:

infection, toxin, electrolyte imbalance, drug withdrawal.

Epilepsy syndromes

Identify the clinical manifestations of rolandic epilepsy:

Children 3 to 13 years of age with benign rolandic epilepsy experience nighttime seizures during sleep. An autosomal dominant disorder, the initial phase of the seizure involves clonic activity of the face, including grimacing and vocalizations, which often wake the child from sleep. Unless these seizures are frequent, no

therapy is needed as patients outgrow them by early adulthood. Carbamazepine is used to treat frequent rolandic seizures.

Recognize the clinical manifestations of juvenile myoclonic epilepsy:

It is inherited as an autosomal dominant trait that manifests in early adolescence (onset 12 to 18 years of age). Patients experience myoclonic jerks, typically on awakening, but may also have tonic-clonic or absence seizures. Provoking factors include stress, alcohol, hormonal changes, or lack of sleep. EEG shows a pattern of fast spike-and-wave discharges. Valproic acid is the drug of choice, with lamotrigine, topiramate, felbamate, and zonisamide as alternate options.

Tuberous sclerosis

Recognize the clinical manifestations of tuberous sclerosis, and manage appropriately:

An autosomal dominant neurocutaneous syndrome, it has multiple small tumors found in multiple organs, which can cause associated symptoms. More than 95% of affected infants have at least one hypopigmented macule or “ash-leaf spot” at birth usually on the trunk and extremities. The characteristic skin finding of adenoma sebaceum, or benign facial angiofibromas, usually does not appear until 2 to 5 years of age. Patients can also present with shagreen patches, raised patches of skin with an orange-peel texture. These patients have developmental delays, and can present with seizures and mental retardation.

Vigabatrin (Sabril) is effective for treating tuberous sclerosis. Side effects include weight gain, hyperactivity, and behavioral changes.

Cerebrovascular disease

Stroke

Signs and symptoms

Clinical features of childhood stroke: The acute onset of a focal neurologic deficit in a child is stroke until proven otherwise. Most common focal presentation is hemiparesis, but acute visual, speech,

sensory, or balance deficits also occur. Urgent neuroimaging and consultation with a child neurologist are emergency interventions.

Pathophysiology

Causes of stroke in children:

Arterial ischemic stroke: 3 main categories of etiology should be considered

– arteriopathic (disorders of the cerebral arteries), cardiac (embolism occurring either spontaneously, or during catheterization or surgical repair), and hematological (iron deficiency anemia, sickle cell anemia, coagulation disorders like factor V Leiden disorder, antiphospholipid antibodies, prothrombotic states, and medications like oral contraceptives and asparaginase chemotherapy).

Cerebral sinovenosis thrombosis (CVST): Cerebral venous drainage occurs via superficial (cortical veins, superior sagittal sinus) and deep (internal cerebral veins, straight sinus) systems that converge at the torcula to exit via the paired transverse and sigmoid sinuses and jugular veins. In CSVT, thrombotic occlusion of these venous structures can create increased intracranial pressure, cerebral edema, and, in 50% of cases, venous infarction (stroke).

Hemorrhagic strokes: Hemorrhagic stroke (HS) includes non-traumatic intracranial hemorrhage and is classified by the intracranial compartment containing the hemorrhage. Intraparenchymal hemorrhage may occur in any location within the brain. Intraventricular hemorrhage may be primary or an extension of intraparenchymal hemorrhage. Bleeding outside the brain may

occur in the subarachnoid, subdural, or epidural spaces.

Vascular anomalies

Clinical features of arteriovenous malformations of childhood: Arteriovenous malformations (AVM) are the most common cause of childhood subarachnoid and intraparenchymal hemorrhagic stroke, and may occur anywhere in the brain. Physical findings for an infant include nonspecific flow murmur, hepatomegaly, loud heart sounds, and bounding pulses.

Spinal cord disease

Signs and symptoms

Clinical manifestations of an acute spinal cord lesion (SCL):

Location of the injury on the spinal cord will determine how severe the injury will be. Thus, a cervical spine injury can cause tetraplegia often requiring mechanical breathing assistance, as with a ventilator, due to weakened chest muscles. An injury to a lower part of the spinal cord causing paraplegia with loss of function in the legs and lower body.

Association between atlanto-axial instability (AAI) in Down syndrome and potential neurologic complications: AAI, reported in 10-30% individuals with Down syndrome, occurs due to the laxity of the transverse ligament which holds the dens against the posterior border of the anterior arch. Symptomatic AAI occurs when subluxation or dislocation causes the odontoid process to impinge on the spinal cord and cause neurologic manifestations.

Significance of bladder and bowel dysfunction in spinal cord disease: cauda equina syndrome requiring prompt neurosurgical intervention.

Diagnosis

Plan the initial neurodiagnostic evaluation in a patient with acute spinal cord dysfunction: check patient's airway, breathing and circulation. Then assess spinal injury site. Spinal radiographs, CT Spine and MRI are tests that can help diagnose the cause.

Peripheral nerve and nerve roots

Guillain-Barre syndrome (GBS)
Signs and symptoms

Presenting signs and symptoms of Guillain-Barre syndrome: Onset is gradual over days or weeks. Affected individuals present with ascending paralysis and involvement of proximal and distal muscles. Affected children are irritable. Weakness can progress to inability or refusal to walk and later to flaccid tetraplegia. Paresthesias occur in some cases.

Respiratory compromise may occur rapidly in GBS.

Risk factors associated with GBS include recent immunization against rabies, influenza and oral poliomyelitis, and following administration of conjugated meningococcal vaccine, or varicella infection.

The original infection might have caused only gastrointestinal (Campylobacter jejuni or Helicobacter pylori) or respiratory tract (Mycoplasma pneumoniae) symptoms.

Diagnosis

Differential diagnosis of GBS includes:

Expected results of laboratory procedures such as examination of the cerebrospinal fluid, nerve conduction studies, and electromyography in Guillain- Barre syndrome.

CSF protein is elevated to more than twice the upper limit of normal, glucose level is normal, and there is no pleocytosis.

Cranial nerves may be affected in GBS. Autonomic dysfunction may be prominent and dangerous.

Treatment

Treatment includes IVIG and plasmapheresis.

Neuropathy

Clinical manifestations of childhood peripheral neuropathy:

Usually a gradual and slowly progressive course; Acute presentation may occur with toxic exposure or inflammatory conditions.

Mostly peripheral neuropathies present with bilateral, symmetric distal involvement, and the injury is related directly to axon length. Most have combined sensory and motor involvement.

Most common motor symptom is weakness. May present as clumsiness, difficulty with gross motor skills like climbing stairs, or impaired fine motor skills such as writing, buttoning clothes, or tying shoes; Sensory symptoms may include numbness, dysesthesias such as burning pain, or ataxia.

Common causes of peripheral neuropathy in childhood include hereditary sensory and motor neuropathy, GBS, Diabetes mellitus, rheumatologic disorders (JIA, HSP, Polyarteritis nodosa, Sarcoidosis, SLE, Sjogren's syndrome), infectious causes (Chagas disease, Rabies, Lyme disease, Leprosy, Diphtheria, Tick paralysis), Drugs (alcohol, chemotherapy agents, Phenytoin, Thalidomide), Toxins (arsenic, lead, mercury, thallium), Factitious, Bell's palsy, brachial nerve palsy.

Signs and symptoms of and treatment for Bell palsy: Upper and lower portions of the face are paretic and corner of the mouth droops, ipsilateral ptosis with possible development of keratitis. Taste on the anterior 2/3 of the tongue is lost on the involved side in about 50% of cases. Ipsilateral facial numbness is reported and probably is due to viral (especially herpes) or postviral immunologic impairment of the trigeminal and the facial nerves. Treatment involves corticosteroids, antivirals (association with herpes virus), physiotherapy and/ or surgery.

Brachial plexus injuries at birth

Clinical manifestations of neonatal brachial plexus injuries: Injury to the brachial plexus may cause paralysis of the upper part of the arm with or without paralysis of the forearm or hand or, more commonly, paralysis of the entire arm. Occur in macrosomic infants and when lateral traction is exerted on the head and neck during

delivery of the shoulder in a vertex presentation, or when the arms are extended over the head in a breech presentation, or when excessive traction is placed on the shoulders.

Management and prognosis of neonatal brachial plexus injuries includes initial conservative management with monthly follow-up and a decision for surgical intervention by three months if function has not improved.

Partial immobilization and appropriate positioning are used to prevent the development of contractures.

In upper arm paralysis, the arm should be abducted 90 degrees with external rotation at the shoulder, full supination of the forearm, and slight extension at the wrist with the palm turned toward the face with a brace or splint during the 1st 1 to 2 weeks. Immobilization should be intermittent throughout the day.

In lower arm or hand paralysis, the wrist should be splinted in a neutral position, and padding placed in the fist.

When the entire arm is paralyzed, the same treatment principles should be followed. Gentle massage and range-of-motion exercises may be started by 7-10 days of age. Infants should be closely monitored with active and passive corrective exercises.

If the paralysis persists without improvement for 3 months, neuroplasty, neurolysis, end-to-end anastomosis, and nerve grafting may be done.

Neuromuscular junction

Ticks may cause paralysis.

Signs and symptoms of myasthenia gravis (MG):

Three clinical varieties are found in childhood: juvenile myasthenia gravis in late infancy and childhood, congenital myasthenia, and transient neonatal myasthenia.

In the juvenile form, ptosis and some degree of extraocular muscle weakness are early signs. Older children might complain of diplopia, dysphagia and facial weakness, and in early infancy, feeding difficulty is often the cardinal sign. Poor head control from weakness of the neck flexors is also prominent. Muscle fasciculations, myalgias, and sensory symptoms do not occur. Tendon stretch reflexes may be diminished but rarely are lost.

Rapid fatigue of muscles is a characteristic feature. Repetitive opening and closing of the fists produces fatigue of hand muscles, and patients cannot elevate their arms for more than 1 to 2 minutes because of fatigue of the deltoids. Patients are symptomatic late in the day or when tired.

Left untreated, MG is usually progressive and can become life threatening from involvement of respiratory muscles and risk of aspiration.

Laboratory and electrophysiologic studies to evaluate children with MG: Electromyography (EMG) is more specifically diagnostic than a muscle biopsy.

Estimating number of Acetylcholine (Ach) receptors per endplate and in vitro study of endplate function are required in the classification of congenital myasthenic syndromes.

Anti-ACh antibodies should be assayed in the plasma. Other serologic tests of autoimmune disease, such as ANA and abnormal immune complexes, should also be sought. Thyroid profile should always be examined. Serum creatine kinase (CK) level is normal in MG.

A clinical test for myasthenia gravis is administration of a short-acting cholinesterase inhibitor, usually edrophonium chloride. Ptosis and ophthalmoplegia improve within a few seconds, and fatigability of other muscles decreases.

Appropriate management for a patient with myasthenia gravis:

Cholinesterase-inhibiting drugs are the primary therapeutic agents – Neostigmine methylsulfate. If dysphagia is a major problem, the drug should be given 30 minutes before meals to improve swallowing. Pyridostigmine is an alternative; it may be slightly longer acting. Overdoses of cholinesterase inhibitors produce cholinergic crises; atropine blocks the muscarinic effects but does not block the nicotinic effects that produce additional skeletal muscle weakness.

Long-term steroid treatment with prednisone may be effective. Thymectomy should be considered and might provide a cure. Thymectomy is most effective in patients who have high titers of anti-ACh receptor antibodies in the plasma and who have been symptomatic for less than two years.

Plasmapheresis is effective in some children, particularly if no response to steroids, but it provides only temporary remission. IVIG is beneficial and should be tried before plasmapheresis because it is less invasive. Plasmapheresis and IVIG appear to be most effective in patients with high circulating levels of anti-ACh receptor antibodies. Refractory patients might respond to rituximab, a monoclonal antibody to the B-cell CD20 antigen.

Neonates with transient maternally transmitted myasthenia gravis require cholinesterase inhibitors for only a few days or occasionally for a few weeks, especially to allow feeding. No other treatment is usually necessary.

Muscle diseases

Signs and symptoms

Clinical features of dystrophinopathy (Duchenne Muscular DystrophyBecker muscular dystrophy [DMD]):

Early gross motor skills, such as rolling over, sitting, and standing, are usually achieved at the appropriate ages or may be mildly delayed. Poor head control in infancy may be the first sign of weakness. Walking is often accomplished at the normal age of about 12 months, but hip girdle weakness may be seen in subtle form as early as the 2nd year. Toddlers might assume a lordotic posture when standing to compensate for gluteal weakness. An early Gowers sign, showing proximal muscle weakness, is often evident by age 3 years and is fully expressed by age 5 or 6 years. A Trendelenburg gait, or hip waddle, appears at this time. Common presentations in toddlers include delayed walking, falling, toe walking and trouble running or walking upstairs, developmental delay, and, less often, malignant hyperthermia after anesthesia. Contractures most often involve the ankles, knees, hips, and elbows. Scoliosis is common. The thoracic deformity further compromises pulmonary capacity and compresses the heart.

The length of time a patient remains ambulatory varies.

Natural history and late complications of the muscular dystrophies:

A muscular dystrophy is distinguished from all other neuromuscular diseases by four obligatory criteria: It is a primary myopathy, it has a genetic basis, the course is progressive, and degeneration and death of muscle fibers occur at some stage in the disease. Some muscular dystrophies are severe diseases at birth that lead to early death; others follow very slow progressive courses over many decades, may be compatible with normal longevity, and might not even become symptomatic until late adult life. Some categories of dystrophies, such as limb-girdle muscular dystrophy (LGMD), are not homogeneous diseases but rather syndromes encompassing several distinct myopathies.

In Becker muscular dystrophy, boys remain ambulatory until late adolescence or early adult life. Calf pseudohypertrophy, cardiomyopathy, and elevated serum levels of creatine kinase (CK) are similar to those of patients with DMD. Learning disabilities are less common. The onset of weakness is later in Becker than in DMD. Death often occurs in the mid to late 20s; fewer than half of patients are still alive by age 40 years; these survivors are severely disabled.

Differential diagnosis for a patient who has weakness and an increased serum creatine kinase activity:

Myotonia congenita

Calcium, Sodium or Chloride channelopathies Thomsen disease

Mytonias Becker disease

Anderson syndrome Malignant hyperthermia

Hyperkalemic periodic paralysis

Hypokalemic periodic paralysis

Pathophysiology

Genetics of dystrophinopathy (Duchenne/Becker muscular dystrophy): mutations in the DMD gene with decreased dystrophin production leading to weakness of skeletal and cardiac muscles.

Differential diagnosis for a patient who has an acquired muscle disorder (eg, inflammatory, infectious, toxic): Endocirine myopathies (hyper- and hypothyroidism, hyperparathyroidism) steroid-induced myopathy, toxic myopathies with certain medications, metabolic myopathies (glycogen metabolism, lipid metabolism disorders, mitochondrial myopathies, purine metabolism disorders), Guillain-Barre syndrome, Bell palsy.

Diagnosis

Laboratory studies available to diagnose muscle disease of childhood: serum CK, DNA markers of common myopathic diseases, muscle biopsy, nerve biopsy.

Central nervous system trauma

Pathophysiology

Outcome of a head injury is related to the duration and degree of coma.

A linear skull fracture in an infant younger than 1 year of age is a sign of possible child abuse.

Signs and symptoms of spinal trauma: Spine of a small child is very mobile, and fractures of the spine are exceedingly rare. However, spinal distortion can maintain the structural integrity of the spine but lead to significant injuries of the spinal cord.

Complete spinal cord injury will lead to spinal shock with early areflexia. Paradoxical respiration can occur when the intercostal musculature innervated by the thoracic spinal cord is paralyzed. In this situation, inspiration fails to expand the chest wall but distends the abdomen.

Very minor injury to the spinal cord is transient quadriparesis evident for seconds or minutes with complete recovery in 24 hours. This injury follows a concussion of the cord.

A transverse injury in the high cervical cord level (C1-C2) causes respiratory arrest and death in the absence of ventilatory support. Fracture dislocations at the C5-C6 level are characterized by flaccid quadriparesis, loss of sphincter function, and a sensory level corresponding to the upper sternum. Fractures or dislocations in the low thoracic (T12-L1) region may produce the conus medullaris syndrome, a loss of urinary and rectal sphincter control, flaccid weakness, and sensory disturbances of the legs. A central cord lesion may result from contusion

and hemorrhage and typically involves the upper extremities to a greater degree than the legs.

Thoracolumbar injuries, usually fracture-dislocations, occur in severe motor vehicle accidents when children are wearing lap belts but not shoulder harnesses. These injuries lead to a conus medullaris syndrome.

Clinical features of epidural hematoma: Patient may be alert at presentation but may deteriorate after a period of hours. Bleeding between the dura and the skull; commonly occurs with skull fracture and middle meningeal artery laceration.

Clinical features of subdural hematoma: Subdural hematomas produce mass lesion that can compress the underlying brain. Bleeding between the dura and the arachnoid space; occurs with disruption of bridging veins connecting cerebral cortex and dural sinuses.

Management

Long-term neurologic and behavioral consequences of head trauma include seizures, behavior disorders like ODD, PDD, Depression, mental retardation & developmental delays, respiratory distress requiring tracheostomy, neuromuscular defects or delays.

Neuroendocrine complications of a head injury: hypothalamic-pituitary dysfunction. Cerebral edema is a consequence of head trauma.

Clinical course and management of epidural hematoma: These hemorrhages are often traumatic in origin and usually present with headache or altered consciousness. Seizures and retinal hemorrhages with increased ICP is typical. Two-thirds of epidural hemorrhages are associated with skull fracture. In children approximately half epidural hematomas originate from venous injuries.

Association of cervical cord injury with head trauma

Intracranial hematomas can occur in the absence of a skull fracture.

Clinical course and management of subdural hematoma: Subdural hematomas may be acute, subacute, or chronic. In acute hematomas, symptoms occur in the first 48 hours after injury. In subacute subdural hematoma, symptoms occur between 3 and 21 days after injury, whereas chronic hematomas cause symptoms after 21 days. Symptoms may include chronic emesis, seizures, hypertonicity, irritability, personality changes, inattention, poor weight gain, fever, and anemia.

Role of pharmacologic therapy in acute spinal cord or craniocerebral trauma

Neurodiagnostic testing

Value and limitations of neurodiagnostic techniques such as magnetic resonance imaging, computed tomography, and ultrasonography

MRI: Neurological indications include diagnosis of brain and spinal cord tumors, eye disease, inflammation, infection, stroke, multiple sclerosis, and traumatic brain injury. MRI does not use ionizing radiation to produce images. The test is painless. Patients with implanted medical devices, such as pacemakers, should avoid the test due to the strong magnetic field generated by an MRI.

CT: Noninvasive, painless process to produce two-dimensional images of organs, bones, and tissues. Neurological CT scans are used to detect bone and vascular irregularities, certain brain tumors and cysts, herniated discs, epilepsy focus, encephalitis, spinal stenosis, blood clot or intracranial bleeding in stroke patients, brain damage from head injury, and other disorders. Limitations include radiation exposure. Pregnant women should avoid the test.

Ultrasound: Neurosonography (ultrasound of the brain and spinal column) analyzes blood flow in the brain and can diagnose stroke, brain tumors, hydrocephalus, and vascular problems. Transcranial Doppler ultrasound is used to view arteries and blood vessels in the neck and determine blood flow and risk of stroke. Ultrasound is painless, noninvasive, and risk-free.

Value and limitations of neurodiagnostic techniques such as evoked potentials, electromyography, and electroencephalography

Evoked potentials: used to assess sensory nerve problems and confirm neurological conditions including multiple sclerosis, brain tumor, acoustic neuroma (small tumors of the inner ear), and

spinal cord injury. Evoked potentials are also used to test sight and hearing (especially in infants and young children), monitor brain activity among coma patients, and confirm brain death.

Electromyography: Nerve and muscle dysfunction and spinal cord disease can be diagnosed using EMG. Patients who are preparing to take an EMG or NCV test may be asked to avoid caffeine and not smoke for 2 to 3 hours prior to the test, as well as to avoid aspirin and non-steroidal anti-inflammatory drugs for 24 hours before the EMG.

Electroencephalography: EEG can help diagnose certain seizure disorders EEGs are also used to evaluate sleep disorders, monitor brain activity when a patient has been fully anesthetized or loses consciousness, and confirm brain death.

Musculoskeletal Disorders

Congenital
General body
Osteogenesis imperfecta

Clinical features of osteogenesis imperfecta include

triangular facies, low nasal bridge, blue or gray sclerae, pectus excavatum or carinatum, joint laxity, dentinogenisis imperfecta,

bone abnormalities: fractures as a neonate, bowing of the extremities, vertebral compressions, small bones in the skull sutures known as wormian bones, cardiac abnormalities: aortic valve disease, mitral valve prolapse

Hearing loss, which is initially conductive and then later becomes sensorineural as well, is associated with some types of osteogenesis imperfecta.

Chondrodysplasias (see also VII.D.5)

Achondroplasia has an autosomal dominant pattern of inheritance with complete penetrance. Affected individuals often have a spontaneous mutation in the FGFR3 gene, and advanced paternal age is a risk factor for this.

Clinical features of achondroplasia include rhizomelic long bone shortening (most proximal part of limbs), macrocephaly, frontal bossing, contracted skull base, square-shaped long bones, trident hands, radiolucency of the proximal femur, hypotonia, and initial delay in meeting motor milestones. Complications include craniocervical junction narrowing with spinal cord compression at the cervical medullary junction, hydrocephalus, lumbar spinal stenosis, thoracolumbar gibbus deformity and obesity.

In a person with achondroplasia, apnea may be central in origin, from the brainstem compression from narrow craniocervical junction as well as obstructive in origin from midface hypoplasia.

Figure: Proximal segment shortening in achondroplasia

Arthrogryposis

Arthrogryposis is characterized by multiple joint contractures present at birth which may become progressively worse, but which does not develop in new joints after birth. Muscle mass is decreased

and limbs may appear shaped as cones, cylinders or tubes. There are sometimes deep dimples over the joints, as well as decreased subcutaneous fat and skin creases. Scoliosis or joint dislocation may occur.

Head and neck
Torticollis

Congenital torticollis is due to dense, fibrotic sternocleidomastoid muscle, the etiology of which is not yet determined, but possibly due to intrauterine position, ischemia to the muscle or trauma sustained during the birth process.

Physical therapy, by means of stretching the neck by either a pediatric physical therapist or a parent can help reduce torticollis

Head tilt could also be due to cervical spine malformations, vision problems, or a tumor in the posterior fossa.

Congenital torticollis is present from early infancy and consistently produces head tilt, while paroxysmal torticollis is intermittent and usually resolves by 3 years of age.

Klippel-Feil syndrome

Clinical features of Klippel-Feil syndrome include short neck with limited range of motion due to fused cervical vertebral segments, a low posterior hairline, and in half of the affected individuals, a Sprengel deformity (upward displacement of the scapula.) Abnormalities in other systems, such as genitourinary system, may be involved in certain syndromes. Radiographs will show fusion of at least two vertebrae.

Trunk and spine (eg tethered cord, occult spina bifida)

Congenital scoliosis is associated with genitourinary anomalies, cardiac anomalies and spinal dysraphism

Lower extremities
Clubfoot

Generally the initial management of congenital talipes eequinovarus is serial casting by the Ponseti method.

Of the four possible components of clubfoot (equinus positioning, hindfoot varus, cavus positioning and metatarsus adductus), the most common component is the equinovarus deformity.

Early treatment of clubfoot seems to be associated with better outcomes, so early referral is important.

Metatarsus valgus, varus

When the forefoot can be abducted past the midline, metatarsus varus and metatarsus valgus can often be managed with exercise and massage.

Coxa valgus, vara

Coxa valga deformity is characterized by an increased angle between the femoral head and shaft (greater than 135 degrees), and it may present with gait abnormality and limb-length discrepancy.

Coxa vara deformity is characterized by a decreased angle between the femoral head and shaft, and appears clinically as a waddling gait with limited internal rotation and abduction on exam.

valgus

Generally flat feet do not require any treatment in childhood. If plano valgus becomes painful, which occasionally occurs during adolescence, longitudinal arch support can be helpful.

Femoral anteversion, tibial torsion

Femoral anteversion generally resolves by 12 years of age

Physical exam is sufficient for diagnosis of femoral anteversion, and additional studies, such as radiographs, are not necessary unless patient is undergoing surgical correction

To examine a child older than 3 or 4 years of age with persistent intoing for femoral anteversion, hip rotation should be examined in prone rotation with expected findings of internal rotation beyond 60-65 degrees and decreased external hip rotation.

Toe-walking may be a normal stage in gait development or may be a sign of underlying neuromuscular disease.

Polydactyly

The supernumerary digit should be removed, either with suture ligation for poorly formed extra digits or with reconstructive procedures for more well-formed extra digits.

Leg length discrepancy

In patients with leg length discrepancy, there may also be abnormal hip abduction.

Acquired
Infections
Osteomyelitis

Bacteremia usually precedes osteomyelitis.

Staphylococcus aureus is the most common cause of bacterial osteomyelitis.

Although it is less common than hematogenous spread, local extension can lead to osteomyelitis.

Early signs of osteomyelitis include pain with weight bearing and tenderness which is localized over the metaphysis.

The most accurate way to confirm diagnosis of osteomyelitis is to send metaphyseal aspirate for culture and sensitivity.

There is generally a lag time of 10 to 14 days before plain radiograph findings are present in osteomyelitis.

Three phase bone scanning is useful in establishing diagnosis in osteomyelitis if the infection has been present for at least three days, but MRI is able to detect diagnosis earlier and provide greater anatomical detail, allowing one to distinguish discitis from vertebral osteomyelitis and defining any adjacent soft tissue infection

Management of osteomyelitis includes identification of the causative organism (ideally through culture of a direct specimen), debridement of related abscess, and intravenous antimicrobial therapy after cultures obtained.

The most common cause of osteomyelitis is Staphylococcus aureus in all age groups.

Salmonella is an important cause in patients with hemoglobin SS or SC. In the neonatal age group, Streptococcus agalactiae, coagulase negative staphylococci, and Enterobacteraceae are also possible causes of osteomyelitis.

Antimicrobial treatment for osteomyelitis includes initiation of intravenous therapy with empiric choice based on local Staphylococcus aureus susceptibilities, then modifying antibiotic choice if needed, switching to oral therapy after improvement has occurred. Total antibiotic duration should be 4 to 6 weeks. If methicillin resistance is low, first generation cephalosporins or semisynthetics are indicated.

Penicillins are good choices. If methicillin resistance is high, clindamycin is an effective agent which has good penetration of bone. Vancomycin, daptomycin or linezolid can be used if there is clindamycin resistance. Neonatal osteomyelitis should be treated with antibiotic coverage that includes Enterobacteriaceae, such as a third generation cephalosporin.

Pelvic osteomyelitis may present with pain in the hip, thigh or buttock, as well as limp. Flexion, external rotation or abduction of the hip can localize pain to sacroiliac joint if this area is involved. Leukocyte count, ESR and CRP will be elevated

Arthritis

Staphylococcus aureus has been the most common cause of pyogenic arthritis since the introduction of Haemophilus influenzae type B immunization. Streptococcus pneumonia, Streptococcus pyogenes, and Kingella kingae are also childhood causes. Sexually active adolescents may have Neisseria gonorrhea pyogenic arthritis. Neonates may get Streptococcus agalactiae.

Spread of osteomyelitis to the adjacent joint is a common cause of pyogenic arthritis

Toxic synovitis classically occurs after a recent upper respiratory infection and may present with limp, unilateral hip pain, groin pain or knee pain, and may show pain with abduction and internal rotation on exam. White blood cell count and inflammatory markers may be mildly elevated. Pyogenic arthritis typically has fever, non-weight bearing, and more significantly elevated inflammatory markers and leukocyte count.

In neonates, pyogenic arthritis may have a more subtle presentation, with lack of fever, only mild elevation in leukocyte count and inflammatory markers, with irritability and mild swelling of the affected joint as the main clinical manifestations

The most helpful evaluation for pyogenic arthritis is aspiration of the joint fluid with analysis

Prompt surgical drainage of purulent fluid from an infected joint, especially hip or shoulder, is important because fluid in that space can cut off the blood supply

Pyogenic arthritis should be treated with antibiotic therapy to cover Staphylococcus aureus based on local susceptibility patterns. This is generally intravenous therapy with an anti-staphylococcal

penicillin such as nafcillin if methicillin resistance is low, or IV clindamycin or vancomycin if resistance is prevalent.

Arthralgia is the presence of joint pain, whereas arthritis has signs of inflammation, such as warmth, erythema, swelling, and pain which is worse after inactivity. Arthritis may also be characterized by limited range of motion and limited muscle strength.

In acute rheumatic fever, the arthritis involves multiple joints (usually large joints such as knees, ankles, wrists and elbows) and is migratory in nature. There is a dramatic response to low doses of salicylates.

Viral causes of acute arthritis include parvovirus B19, rubella, hepatitis B, hepatitis C and arthropod borne viruses of the alphavirus genus (such as Chikungunya virus.)

Synovitis

Transient synovitis (or toxic synovitis) is a diagnosis of exclusion

The differential diagnosis for painful hips is different based on the age and gender of the patient

Trauma
Dislocations (see also XXXLD.5.)

Subluxation of the patella presents as acute severe pain, often occurring after a pivoting or twisting motion and associated with a popping sound. On physical exam the knee is held in 20-30 degrees flexion with the patella lateral to the femur.

To prevent recurrence of patellar dislocation, rigorous quadriceps strengthening exercise should be a key part of post-injury rehabilitation

Shoulder dislocation tends to occur either between 18 and 30 years of age (predominantly in males) and in the elderly.

Ligamentous (sprains, strains)

The severity and type of pain may be helpful in determining the severity and type of ligamentous injury sustained

Sprains should be managed initially with rest (generally for 48-72 hours), ice (20 minutes every 2 to 3 hours for the first 48 hours post-injury), compression (with the joint in a neutral position) and elevation (15-25cm above the heart).

Care should be taken to prevent re-injury while the sprain is still healing to prevent more severe injury

In soft tissue injury, ice packs should be used with a soft cloth as a barrier between the ice and skin. The injured area should be iced for 20 minutes at a time every 2 to 3 hours.

Grade one sprains may cause mild discomfort with little to no loss of function. Grade two sprains have tenderness and swelling, with or without ecchymosis, and functional loss is mild to moderate. Grade three sprains have significant, diffuse tenderness, ecchymosis and swelling.

When the ankle is injured in a pre-pubertal adolescent, a growth plate fracture may occur instead of a sprain because the ligament is stronger than the growth plate

Bone (see also XXXI.D.5.)

Compartment syndrome occurs when there is an increase in the pressure within a semi-rigid structure, which can exceed capillary perfusion, leading to ischemic damage and considerable morbidity and mortality.

Abnormal gait in young children may be a sign of an occult fracture.

Some growth plate fractures and injuries lead to the formation of a physeal bar, which leads to growth arrest of the bone.

Miscellaneous
Scoliosis

Scoliosis is either idiopathic or related to neurologic disorders, musculoskeletal disorders or connective tissue disorders. The most common type, idiopathic scoliosis in adolescence, involves rotation of the spine which may continue until linear growth is complete. Extreme curvature greater than 50-75 degrees may lead to impaired respiratory function

The back should be examined at health supervision visits, and girls should be screened for scoliosis twice (at 10 and 12 years of age), while boys should be screened once at 13 or 14 years of age, according to several professional organizations, including the American Academy of Pediatrics. This screening includes inspecting the back upright, then having the patient bend forward with feet together and knees straight, with the examiner looking for asymmetry of the scapulae, ribs or paraspinous muscles and using a scoliometer to detect at least 6 to 7 degree curve prompting scoliosis radiographs.

For scoliosis with Cobb angle 20 to 40 degrees, bracing can be considered. If curves are greater than 40 degrees, spinal fusion is often the optimal treatment

Kyphosis

Kyphosis may cause back pain and give an undesirable cosmetic appearance and

should be treated with the goal of achieving pain relief and acceptable appearance,

as well as preventing neurologic deficit. Good follow-up is also important.

Avascular necrosis (Legg-Calve-Perthes disease)

The typical age group for Legg-Calve-Perthes disease is between 3 and 10 years of age

Legg-Calve-Perthes is more common in boys than girls

Legg-Calve-Perthes disease is an important consideration in the differential diagnosis of a child with a limp

Apophysitis

Osgood-Schlatter disease is caused by overuse of the extensor mechanism at the knee, leading to a traction apophysitis where the patellar tendon inserts into the tibial tuberosity

Osgood-Schlatter disease presents as pain with activity (especially running, jumping or kneeling) over the tibia tubercle and distal patellar tendon. It is generally a self-limited condition, occurring in growing individuals and resolving with skeletal maturity.

Sever disease (calcaneal apophysitis) occurs due to overuse injury, from activities such as running, gymnastics or basketball. It should be managed with supportive care, including stretching and heat, in addition to heel cups or heel lifts ranging from ¼ to 5/8 inch.

Slipped capital femoral epiphysis

Slipped capital femoral epiphysis can present with groin pain, hip pain, knee pain, and/or limp.

Myositis

A patient with suspected myositis should have complete history and physical examination to attempt to differentiate viral, bacterial and rheumatologic causes. Serum creatinine kinase can confirm the diagnosis of myositis.

Suspected viral: symptomatic treatment with fluids and NSAIDs until resolution of symptoms, generally within one month; corticosteroid therapy may be helpful in HIV related myositis.

Suspected bacterial: MRI helps to define nature of infection. Surgical debridement with culture of the purulent drainage and antibiotic therapy are keys to treatment. Hyperbaric oxygen may play an adjunctive role in treating Clostridium perfringens infection. IVIG may play a supportive role in severe group A streptococcal infection.

Suspected rheumatologic: MRI and flow cytometry may be useful to replace EMG and muscle biopsy in making a diagnosis; corticosteroids and other immunosuppressants are often used in treatment regimens

Suspected medication side effect: discontinue medication suspected of causing the myositis (such as statins) and manage any consequential rhabdomyolysis with fluids and alkalinization.

Etiologies of myositis: viral, trauma, adverse effects of medications such as statins and zidovudine, inflammatory diseases such as dermatomyositis, bacterial infection, fungal infection or parasitic infection

Back pain

Differential diagnosis for back pain in children and adolescents includes: referred pain (from retrocecal appendicitis, psoas abscess or pyelonephritis), muscle strain, spondylysis, spondylolisthesis, herniated disk, vertebral osteomyelitis, neoplasms (osteosarcoma, osteoid osteoma, aneurysmal bone cyst), and Scheuermann disease (idiopathic kyphosis).

In the evaluation of a child with back pain, the following evaluations may be necessary in certain circumstances:

Spine radiographs (standing anterior posterior, lateral and oblique): for injury from frequent hyperextension or physical examination with midline pain increased with hyperextension of lumbar spine while standing.

Laboratory test (WBC with differential, peripheral smear, CRP, ESR, LDH, blood culture) if systemic symptoms such as fever or weight loss are present MRI if neurologic symptoms, such as weakness, gait abnormalities or fecal or urinary incontinence.

Spondylolisthesis should be managed by avoiding vigorous activity, wearing a brace, and possibly spinal fusion in serious cases.

Developmental dysplasia, subluxation of the hips

Ultrasonography is the most precise way to confirm developmental dysplasia of the hip(s) in young infants with imaging.

One sign of hip subluxation is asymmetry of the gluteal and thigh folds.

It is possible to initially have no abnormal signs of hip subluxation in developmental dysplasia of the hip(s).

Females and infants who had breech presentation have higher rates of developmental dysplasia of the hips.

Isolated hip clicks are not usually due to dysplasia.

i. Bone cysts

Unicameral bone cysts are benign bone lesions usually located in the proximal humerus and femur in children which occur during active skeletal growth and resolve spontaneously with maturity. Aneurysmal bone cysts are expansile and generally managed with curettage or arterial embolization, with recurrence rate of 10 to 20%. Differential diagnosis includes eosinophilic granuloma, osteofibrous dysplasia, fibrous dysplasia, fibromas, osteoblastoma, osteoid osteoma, chondromyxoid fibroma, chondroblastoma, enchondroma, multiple hereditary exostoses, and osteochondroma.

Skin Disorders

Newborn skin
Pigmentary and vascular lesions

A port wine stain in the distribution of the ophthalmic division of the trigeminal nerve is more likely associated with a leptomeningeal angiomatosis (Sturge-Weber syndrome).

A tunable dye laser offers effective cosmetic palliation of port wine stains.

Pustular lesions
Erythema toxicum

The lesions are firm, yellow-white, 1- to 2-mm papules or pustules with a surrounding erythematous flare, usually appearing by second dayof life. Wright- stain of lesions will show eosinophils

Transient neonatal pustular melanosis

Lesions consist of evanescent superficial pustules, ruptured pustules with a collarette of fine scale, at times with a central hyperpigmented macule, and/or hyperpigmented macules. They are present at birth. Wright-stain of lesions will show polymorphonuclear leukocytes. Gram stain will be sterile. This distinguishes it from staphylococcal pustules.

Neonatal bullous impetigo

Flaccid, transparent bullae usually begin in the diaper area without surrounding erythema; caused by Staphylococcus aureus.

Atopic dermatitis (eczema)

The appropriate treatment of eczema consists of emollients, corticosteroids, antibiotics, and allergen elimination when appropriate.

Factors that worsen eczema are drying, chemical irritants, heat, and physical trauma. Rash is characterized by pruritus and erythematous papules acutely, erythematous, excoriated, scaling papules subacutely, and lichenification chronically.

It is generally more acute in infancy and involves the face, scalp, and extensor surfaces of the extremities. The diaper area is usually spared.

Older children may have a chronic appearance and localization of the rash to the flexural folds of the extremities.

Children with atopic dermatitis are prone to recurrent infections, particularly with

taphylociccus. aureus and herpes simplex.

Infectious rashes and infestations
Bacterial infections
General

Staphylococcus aureus is the most common cause of skin infections followed by

Streptococcus pyogenes (Group A streptococcus).

Staphylococcus. aureus is colonized in neonates within the first week of life, however, Streptococcus pyogenes is uncommon in neonates due to maternal antibodies.

Defects in the mucocutaneous barriers produced by trauma, surgery, foreign surfaces (sutures, shunts, intravascular catheters), and burns increase the risk for infection.

Staphylococcus. aureus may cause impetigo, cellulitis, ecthyma, or abscess. It may cause toxin-mediated disease- scalded skin syndrome or toxic shock syndrome. It rarely causes necrotizing fasciitis.

Streptococcus. pyogenes may cause impetigo or pyoderma leading to streptococcal toxic shock syndrome or necrotizing fasciitis.

Streptococcus. pyogenes or methicillin-sensitive S. aureus may be treated with topical mupirocin or orally with penicillin, amoxicillin, a first-generation cephalosporin, or a macrolide.

If methicillin-resistant Staphylococcus. aureus, clindamycin or trimethoprim/sulfamethoxazole may be used and vancomycin may be necessary for severe infections

For Staphylococcus aureus, abscessdrainage is usually necessary for treatment.

Impetigo

S. aureus is the primary cause of both bullous and pustular impetigo.

Staphylococcal scalded skin syndrome (SSSS)

A scarlatiniform erythema develops diffusely and may appear wrinkled with sterile, flaccid blisters and erosions.

Areas of epidermis may separate in response to gentle shear force (Nikolsky sign).

The desquamative phase begins after 2-5 days of cutaneous erythema.

Patients may have pharyngitis, conjunctivitis, and superficial erosions of the lips, but intraoral mucosal surfaces are spared.

Staphylococcal scalded skin syndrome (SSSS) is mediated by an exotoxin- exfoliatin A or B-released by certain strains of staphylococci.

Methicillin-resistant Staphylococcus aureus infection

It manifests as pustules or boils with accompanying erythema, swelling, or edema.

It is often fluctuant and very painful.

It often affects the lower extremities and buttock and is recurrent, often difficult to treat.

Papular urticaria

Papular urticaria represents a hypersensitivity reaction to insect bites.

It consists of transitional lesions in various stages of evolution between delayed- onset papules and immediate-onset wheals.

The most characteristic lesion is an edematous, red-brown papule.

Scabies

The first sign often consists of 1- to 2-mm red papules, some of which are excoriated, crusted, or scaling.

Threadlike burrows are the classic lesion of scabies but infants may present with bullae and pustules.

The palms, soles, and scalp are often affected in infants.

In older children and adolescents, the interdigital spaces, wrist flexors, anterior axillary folds, ankles, and buttocks are affected.

All contacts and family members of a child with scabies will require treatment even if they have no rash because a latent period of about 1 month follows an initial infestation.

The treatment of choice for scabies is permethrin 5% cream (Elimite) applied to the entire body from the neck down. Treatment of the scalp should also be included for infants. The medication is left on the skin for 8-12 hours prior to bathing.

Fungal infections

Granuloma annulare lesions are smooth while tinea corporis lesions are dry and scaly. They are both annular lesions with usual central clearing.

Tinea pedis and atopic dermatitis are distinguished by history and presence of fungi of tinea pedis on KOH preparation.

Tinea versicolor lesions are typically reddish brown macules in those with lighter skin and may be either hypopigmented or hyperpigmented in those with darker skin.

Tinea versicolor begins near follicles and enlarges to form patches. Commonly on the neck, upper chest, and back; Areas do not tan after sun exposure.

Molluscum contagiosum

Consists of discrete, pearly, skin-colored, smooth, dome-shaped, papules; they typically have a central umbilication. A self-limited disease but may be treated.

Curettage is the treatment of choice when old enough to tolerate the pain.

For younger children, cantharidin may be applied to the lesions (not on the face) and covered with adhesive bandages. A blister forms at the site of application, and the molluscum is removed with the blister.

Warts-condyloma acuminatum (see XVII.D.3)

Pediculosis

The ova or nits are glued to hairs or fibers of clothing, hatch in 1 to 2 weeks, and require another week to mature. Once the eggs hatch, the nits remain attached to the hair as empty sacs of chitin. Freshly hatched larvae die unless a meal is obtained within 24 hours.

Malathion 0.5% in isopropanol is the treatment of choice for head lice.

It should be applied to dry hair until hair and scalp are wet, and left on for 12 hours. A second application 7-9 days after initial treatment may be necessary.

All household members should be treated at the same time. Clothing and bed linens should be laundered in very hot water.

Cellulitis (see also XXII.A.3)

Management of cellulitis in a neonate should include empiric therapy with methicillin or vancomycin and gentamicin or cefotaxime.

In a child older than 2 months without fever, lymphadenopathy, and normal white blood cell count, treatment of cellulitis on an extremity may be done as an outpatient with dicloxacillin or cloxacillin or cephalexin.

If MRSA is suspected, clindamycin should be used.

If fever, lymphadenopathy, or constitutional signs are present, parenteral therapy should be initiated with nafcillin, clindamycin, or vancomycin.

Necrotizing fasciitis

Common predisposing conditions in neonates are omphalitis and balanitis after circumcision. Surgical debridement and parenteral antibiotics are necessary.

Hair loss

Alopecia areata

Rapid and complete loss of hair in round or oval patches on the scalp, immune- mediated, non-scarring;

Spontaneous resolution in 6 to 12 months with recurrence being common

May be associated with autoimmune diseases; increased incidence in those with trisomy-21 (Down syndrome.)

Trichotillomania

Characterized by the presence of hair shafts of different lengths in the area of alopecia

Tinea capitis

Clinical appearance is variable. May be "black-dot ringworm," characterized initially by many small circular patches of alopecia in which hairs are broken off close to the hair follicle; or may be diffuse scaling with minimal hair loss

A kerion is an elevated, boggy granulomatous mass often with pustules. It is a severe inflammatory reaction in response to tinea capitits.

Systemic therapy (eg, griseofulvin, itraconazole) is necessary to eradicate tinea capitis.

Griseofulvin therapy is safe and does not require routine laboratory studies to monitor its toxicity.

Telogen effluvium

A sudden loss of large amounts of hair, often with brushing, combing, and washing of hair. Normal hair growth will return within approximately 3-6 months. Precipitating cause may include childbirth, a febrile episode, surgery, or acute blood loss.

Also accounts for the loss of hair by infants in the first few months of life.

Neurocutaneous syndromes
General

Heterogeneous group of disorders characterized by abnormalities of both the integument and central nervous system

Most disorders are familial and believed to arise from a defect in differentiation of the primitive ectoderm.

Neurofibromatosis

Six or more cafe au lait spots > 0.5 cm in diameter suggests the diagnosis. Autosomal dominant inheritance

Neurofibromas usually do not appear until after puberty.

Tuberous sclerosis

The earliest sign of tuberous sclerosis may be hypopigmented macules.

Sturge-Weber syndrome

A sporadic vascular disorder

Facial capillary malformation (port-wine stain), abnormal blood vessels of the brain (leptomeningeal angioma), and abnormal blood vessels of the eye leading to glaucoma.

Patients present with seizures, hemiparesis, stroke-like episodes, headaches, and developmental delay.

Pigmented lesions
Hyperpigmentation

A helpful diagnostic sign in urticaria pigmentosa is pigmented lesions that flare after being rubbed.

The early warning signs of malignant melanoma are:

Asymmetry, Border irregularity or bleeding, Color variation, and Diameter enlargement; However, many of the melanomas diagnosed in children have none of these features.

Hypopigmentation
Acne

Angiofibroma may be mistaken for acne.

First-line treatment of acne vulgaris is with topical medications: retinoic acid and benzoyl peroxide.

In teenagers receiving corticosteroids, a characteristic form of acne may develop- with acne all in the same stage of development.

Systemic antibiotics should be prescribed when there is papulopustular or nodular acne.

May use tetracycline, doxycycline, erythromycin, or trimethoprim-sulfamethoxazole.

Other
Hemangiomas
Natural history

“Strawberry” hemangiomas may be present at birth or may become apparent in the first 2 months of life.

They enlarge and spontaneously involute by 5 to 9 years of age.

Therapeutic options

Indications for therapy for hemangiomas include severe ulceration/maceration, bleeding, ophthalmic sequelae, hepatic hemangioma.

Therapy includes pulsed dye laser, oral corticosteriods or propranolol.

Erythema multiforme, Stevens-Johnson syndrome

The spectrum of severity of erythema multiforme ranges from targetoid lesions to Stevens-Johnson syndrome.

Contact dermatitis

Contact dermatitis is characterized by linear vesicles and papules.

Rhus dermatitis (eg, poison ivy) is not spread by fluid contained in the vesicular or bullous lesions.

Short- and long-term effects of sun exposure

Sun damage to the skin is additive and leads to aging of the skin as well as an increased incidence of skin cancers.

Ectodermal dysplasia

A heterogeneous group of disorders characterized by a constellation of findings involving defects of 2 or more of the following: teeth, skin, hair, nails, and eccrine and sebaceous glands.

Sebaceous nevus

A relatively small, sharply demarcated, oval or linear, elevated yellow-orange plaque that is usually devoid of hair.

Occurs on the head and neck of infants

Lesions become verrucous and studded with large rubbery nodules as patient matures.

Dermoids

A dermoid usually appears as a well-circumscribed rounded or oval, gray or pinkish-yellow mass with a dry surface from which short hairs may protrude.

Epibulbar dermoids may cause visual disturbance by encroaching on the visual axis or by contributing to the development of astigmatism, which may lead to amblyopia.

Ichthyosis

A disorder of keratinization in which keratosis pilaris and hyperkeratosis are common

Onset generally occurs in the first year of life over extensor surfaces of legs. Ichthyosis commonly occurs in children with atopic dermatitis.

Keratolytic agents (eg, lactic acid, citric acid) are effective therapies in the management of ichthyosis vulgaris.

Psoriasis

Erythematous papules that coalesce to form plaques with sharply demarcated, irregular borders

Without treatment, a thick silvery or yellow-white scale develop

Pityriasis rosea

A herald patch, a solitary, round or oval lesion that may occur anywhere on the body usually precedes the generalized eruption.

Approximately 5 to 10 days after the appearance of the herald patch, a widespread, symmetric eruption involving mainly the trunk and proximal limbs becomes evident.

Typical lesions are oval or round, <1 cm in diameter, slightly raised, and pink to brown.

The developed lesion is covered by a fine scale, which gives the skin a crinkly appearance.

The long axis of each lesion is usually aligned with the cutaneous cleavage lines, a feature that creates the so-called Christmas tree pattern on the back.

The lesions may be asymptomatic or mildly to severely pruritic. Duration of the eruption varies from 2 to 12 weeks.

Secondary lues (syphillis) may mimic pityriasis rosea.

Seborrheic dermatitis

Greasy, scaly, erythematous papular dermatitis.

Diffuse or focal scaling and crusting of the scalp, sometimes called cradle cap in infants.

Cutaneous manifestations - endocrine, metabolic, nutritional disorders Acrodermatitis enteropathica is a manifestation of zinc deficiency.

The rash has a distribution over perioral, acral, and perineal areas and on the cheeks, knees, and elbows.

Acanthosis nigricans is associated with insulin resistance.

It is characterized by hyperpigmented velvety, hyperkeratotic, plaques that are most often localized to the neck, axillae, inframammary areas, groin, inner thighs, and anogenital region.

Collagen Vascular and Other Multisystem Disorders

Systemic lupus erythematosus (SLE)
Clinical manifestations
Usual

The most common presenting complaints of children with SLE include fever, fatigue, hematologic abnormalities, arthralgia, and arthritis.

SLE is often characterized by periods of flare and disease quiescence or may follow a more smoldering disease course.

Rheumatic fever is distinguished by carditits, migratory polyarthritis, erythema marginatum, subcutaneous nodules, and/or chorea, usually 2 to 4 weeks after acute group A streptococcus pharyngeal infection.

SLE may have similar symptoms but usually has a positive ANA titer.

Unusual

Some other possible manifestations include: malar rash, discoid rash, photosensitivity, oral ulcers, serositis, arthritis, renal manifestations, seizures, psychosis, hemolytic anemia, thrombocytopenia, lymphopenia, carditis, hepatosplenomegaly, or scleritis.

Laboratory evaluation
Antinuclear antibody testing

A positive ANA test is the usual finding in systemic lupus erythematosus.

A low-titer ANA may be seen in unaffected individuals and family members. A positive ANA occurs in many conditions other than SLE.

The ANA is specific for SLE but not sensitive. It does not correlate with disease activity.

Anti-double stranded DNA determination)anti-DNA):

Anti-double-stranded DNA is important in establishing a diagnosis of systemic lupus erythematosus.

It is sensitive for SLE and correlates with disease activity in some individuals. Helpful in monitoring patient's respond to treatment.

Complement concentrations

CH50, C3, and C4 complement concentrations are usually decreased in active SLE but increase with control of disease.

Helpful in monitoring patient's response to treatment.

Hematologic evaluation

May include immune-mediated cytopenias (hemolytic anemia, thrombocytopenia or leukopenia), anemia of chronic inflammation, hypercoaguability, thrombocytopenic or thrombotic microangiopathy

Course and complications

Infection is a major cause of mortality in systemic lupus erythematosus.

Renal disease is a common SLE complication. Blood pressure and urinalysis monitoring is critical.

Central nervous system lupus may include: optic neuritis, seizures, psychosis, cerebritis, stroke, transverse myelitis, depression, cognitive impairment, headaches, pseudotumor cerebri, peripheral neuropathy, chorea, or cranial nerve palsies.

A serious infection in a patient with SLE receiving immunosuppressive therapy may present with: shaking chills, leukocytosis and/or neutrophilia (especially in the absence of steroid therapy), increased numbers of band forms or metamyelocytes on peripheral blood smear.

Therapy

The major complications of corticosteroid therapy in systemic lupus erythematosus include: growth disturbance, weight gain, striae, acne, hyperglycemia, hypertension, cataracts, avascular necrosis, and osteoporosis.

Drugs useful in the treatment of systemic lupus erythematosus include: hydroxychloroquine, methotrexate, leflunomide, azathioprine, mycophenolate mofetil, and cyclophosphamide.

Anti-DNA and serum complement determinations are helpful in guiding treatment in a patient with systemic lupus erythematosus.

Neonatal lupus syndrome

The clinical picture of neonatal lupus includes: characteristic annular or macular rash typically affecting the face (especially the periorbital area), trunk, and scalp. The rash typically appears within the first 6 weeks of life after exposure to ultraviolet light and lasts 3 to 4 months; however, it can be present at birth.

Infants may also have cytopenias and hepatitis.

Conduction system abnormalities range from prolongation of the PR interval to complete heart block, rarely resulting in progressive cardiomyopathy.

The noncardiac manifestations of neonatal lupus are usually reversible, but congenital heart block is permanent.

Conduction system abnormalities can be detected in utero beginning at 16 weeks of gestational age.

Because maternal autoantibodies gain access to the fetus via the placenta at the 16th week of gestation, all pregnant women with circulating anti-Ro or anti-La antibody (or those with a history of offspring with neonatal lupus or congenital heart block) are monitored by a pediatric cardiologist with regular fetal electrocardiography from 16 weeks of gestation until delivery.

If fetal bradycardia is found unexpectedly during in utero monitoring, screening for maternal anti-Ro and anti-La antibodies is warranted.

Vasculitis syndromes
Henoch-Schonlein purpura (HSP)
Clinical manifestations
Many cases follow a documented upper respiratory infection.
The hallmark rash of HSP is present before other symptoms 75% of the time: palpable purpura starting as pink macules or wheals and developing into petechiae, raised purpura, or larger ecchymoses.
The skin lesions are usually symmetric and occur in gravity-dependent areas (lower extremities) or on pressure points (buttocks).
The skin lesions often evolve in groups, typically lasting 3 to 10 days, and may recur up to 4 months after initial presentation.
Subcutaneous edema localized to the dorsa of hands and feet, periorbital area, lips, scrotum, or scalp is also common.
Musculoskeletal involvement, including arthritis and arthralgias, is common, occurring in up to 75% of children with HSP. The arthritis tends to be self-limited and oligoarticular, with a predilection for the lower extremities.
Gastrointestinal manifestations of HSP occur in up to 80% of children with HSP. They include abdominal pain, vomiting, diarrhea, paralytic ileus, melena, intussusception, and mesenteric ischemia or perforation.
Renal involvement occurs in up to 50% of children with HSP, manifesting as hematuria, proteinuria, hypertension, frank nephritis, nephrotic syndrome, and acute or chronic renal failure.
Neurologic manifestations of HSP, due to hypertension or central nervous system (CNS) vasculitis, may also occur. They include intracerebral hemorrhage, seizures, headaches, and behavior changes.
Other less common potential manifestations of HSP are orchitis, carditis, inflammatory eye disease, testicular torsion, and pulmonary hemorrhage.
May present with initial abdominal pain or joint complaints.
Diagnosis

The diagnosis of HSP is made clinically but common laboratory findings include leukocytosis, thrombocytosis, mild anemia, and elevations of ESR and CRP.

Occult blood is frequently found in stool specimens.

Serum IgA values are often elevated but are not routinely measured.

Kawasaki disease (see also XIV.D.5.)
Clinical manifestations
Fever is characteristically high (≥101°F), unremitting, and unresponsive to antibiotics.
Bilateral nonexudative bulbar conjunctival injection with limbal sparing
Erythema of the oral and pharyngeal mucosa with strawberry tongue and dry, cracked lips
Edema and erythema of the hands and feet
Rash of various forms (maculopapular, erythema multiforme, or scarlatiniform) with accentuation in the groin area
Nonsuppurative cervical lymphadenopathy, usually unilateral, with node size >1.5cm
Perineal desquamation is common in the acute phase.
Periungual desquamation of the fingers and toes begins 1 to 3 weeks after the onset of illness and may progress to involve the entire hand and foot.
The differential diagnosis of Kawasaki disease includes: adenovirus, measles, scarlet fever, EBV, enterovirus, toxic shock syndrome, drug hypersensitivity reactions.
Laboratory abnormalities

May include: leukocytosis with neutrophilia and immature band forms, anemia, thrombocytosis after 1 week.

Hyponatremia, elevated ESR and CRP, elevated serum transaminases, hypoalbuminemia, abnormal plasma lipids

Sterile pyuria, Elevated serum gamma glutamyl transpeptidase; Pleocytosis of CSF, leukocytosis of synovial fluid

Management

High-dose intravenous immune globulin and aspirin therapy should be started within 10 days of initiation of symptoms.

IVIG results in rapid resolution of symptoms. IVIG and aspirin together reduce the likelihood of coronary disease by 2 to 4%.

Juvenile rheumatoid (idiopathic) arthritis (JIA)
Clinical presentations
Arthritis may initially be subtle or acute and often include morning stiffness with a limp or gelling after inactivity.
Involved joints are often swollen, warm to touch, and painful on movement or palpation with reduced range of motion but usually are not erythematous.
Arthritis in large joints, especially knees, initially accelerates linear growth, causing the affected limb to be longer and resulting in a discrepancy in limb lengths.
Continued inflammation stimulates rapid and premature closure of the growth plate, resulting in shortened bones.
Systemic-onset disease (SoJIA) is characterized by arthritis, fever, and prominent visceral involvement, including hepatosplenomegaly, lymphadenopathy, and serositis (pericarditis).

The characteristic fever, defined as spiking temperatures to ≥39°C, occurs on a daily or twice-daily basis for at least 2 weeks.
The fever is often present in the evening and is frequently accompanied by a characteristic faint, erythematous, macular rash.
The evanescent salmon-colored lesions, classic for systemic-onset disease, are linear or circular and are most commonly distributed over the trunk and proximal extremities.
Macrophage activation syndrome (MAS) is a rare but potentially fatal complication of SoJIA that can occur at any time during the disease course. It is also referred to as secondary hemophagocytic syndrome or hemophagocytic lymphohistiocytosis (HLH).
MAS classically manifests as acute onset of profound anemia associated with thrombocytopenia or leukopenia with high, spiking fevers, lymphadenopathy, and hepatosplenomegaly.
Patients may have purpura and mucosal bleeding, as well as elevated fibrin split product values and prolonged prothrombin and partial prothromboplastin times.
The erythrocyte sedimentation rate (ESR) falls because of hypofibrinogenemia and hepatic dysfunction, a feature useful in distinguishing MAS from a flare of systemic disease.
The typical patient with pauciarticular (oligoarthritis) juvenile rheumatoid arthritis has ≤4 joints affected in 1st 6 months after presentation.
Rheumatoid factor is usually negative in juvenile rheumatoid (idiopathic) arthritis.
Clinical course and prognosis
Eye disease

The ocular complications of juvenile rheumatoid (idiopathic) arthritis: uveitis- more likely in oligoarthritis, less likely in systemic arthritis.

If a patient is ANA positive, he or she is more likely to present with eye disease.

Cardiac disease

Cardiac complications may include: pericarditis or pericardial effusion.

Differential diagnosis

Juvenile rheumatoid (idiopathic) arthritis is often a disease of exclusion.

The differential diagnosis includes but is not limited to: systemic lupus erythematosus, juvenile dermatomyositis, sarcoidosis, and the vasculitic syndromes, scleroderma, seronegative spondyloarthropathies, bacterial arthritis, arthritis due to a viral illness, acute rheumatic fever, toxic synovitis, immunodeficiencies, inflammatory bowel disease, pain syndromes, and malignancies.

Examination of joint aspirate will distinguish between juvenile rheumatoid (idiopathic) arthritis and septic arthritis-which will likely be positive for an organism and have a cell count of > 50,000-100,000 cells/mm3.

Lyme disease as a mimic of pauciarticular (oligo) juvenile rheumatoid arthritis Lyme disease should be considered in children with oligoarthritis living in or visiting endemic areas.

Although a history of tick exposure, preceding flu-like illness, and subsequent rash should be sought, they are not always present.

Acute rheumatic fever is characterized by exquisite joint pain and tenderness, a remittent fever, and a migratory polyarthritis.

Therapy
Pharmacologic agents

The goals of treatment are to achieve disease remission, prevent or halt joint damage, and foster normal growth and development.

Children with oligoarthritis often show at least partial response to nonsteroidal anti-inflammatory drugs, with improvement in inflammation and pain.

Those who have no response after 4-6 weeks of treatment with NSAIDS or who have functional limitations, such as joint contracture or leg length discrepancy, benefit from injection of intra-articular corticosteroids, such as triamcinolone hexacetonide.

A minority of patients with oligoarthritis show no response to NSAIDs and injections, so require treatment with disease-modifying antirheumatic drugs (DMARDs), like patients with polyarticular disease or systemic JIA.

These drugs would include methotrexate, sulfasalazine, anti-tumor necrosis factor-alpha drugs,-etanercept or infliximab, and anti-CD20 drugs.

The complications of and nonsteroidal anti-inflammatory drug therapy in juvenile rheumatoid (idiopathic) arthritis: bleeding, gastritis, abnormal liver function test results, and encephalopathy.

Other therapies

There is a need for a comprehensive program (including physical therapy) for the management of juvenile rheumatoid (idiopathic) arthritis.

Dietary evaluation and counseling are needed to ensure appropriate calcium, vitamin D, protein, and caloric intake.

A social worker and nurse clinician can be important resources for families, to recognize stresses imposed by a chronic illness, to identify appropriate community resources, and to aid compliance with the treatment protocol.

Other rheumatic disorders
Dermatomyositis
Clinical presentation

The classic picture of dermatomyositis includes: proximal muscle weakness and/or rash at presentation with muscle tenderness, fevers, fatigue, dysphonia, or dysphagia.

The rash is photosensitive and may produce an erythematous “shawl sign” over the shoulders or heliotrope (blue-violet) rash over the eyelids.

There may be erythema over the nasolabial folds.

Gottron papules may be present as red plaques over certain joints.

Diagnosis

The appropriate laboratory evaluation of dermatomyositis includes: serum levels of muscle-derived enzymes such as creatine kinase, aldolase, AST, ALT, and LDH.

ALT is often elevated on presentation while creatine kinase may be normal. Patients usually have a normal ESR and negative RF.

Most patients are positive for ANA. MRI detects areas of disease.

Scleroderma

The clinical manifestations of localized scleroderma-morphea and linear scleroderma-can extend from the dermis to the bone.

The initial skin manifestations include erythema or a bluish hue seen around an area of waxy induration.

Early edema and erythema are followed by indurated, hypopigmented or hyperpigmented, atrophic lesions linear scleroderma varies in size from a few centimeters to the entire length of the extremity, with varying depth.

Patients sometimes present with arthralgias, synovitis, or flexion contractures. Children with en coup de sabre may have symptoms unique to central nervous system (CNS) involvement, such as seizures, hemifacial atrophy, ipsilateral uveitis, and learning/behavioral changes.

Localized scleroderma is much more common than systemic sclerosis and has a better outcome.

Sarcoidosis

Sarcoidosis in childhood can have granulomatous lesions in any part of the body, with accompanying fever, weight loss, general malaise.

In order children pulmonary involvement causing bilateral hilar adenopathy and parenchymal infiltrates may only present as a persistent, dry cough and restrictive changes on PFTs.

May have isolated hepatosplenomegaly

May have a maculopapular rash, ocular involvement, or arthritis

Ankylosing spondylitis

Presents as oligoarthritis in lower extremities particularly the hips, positive HLA- B27, with eye involvement

There is often sacroiliac joint tenderness.

Axial arthritis is usually absent until later in the disease course There is absence of RF and ANA.

Sclerosis of sacroiliac joints is seen on radiographs.

Other arthritis and arthralgia syndromes
Arthritis of inflammatory bowel disease

Arthritis may occur in patients with inflammatory bowel disease. It may involve polyarthritis of large and small joints.

Postinfectious arthritis

A patient with postinfectious arthritis will have sterile joint inflammation after a recent infection such as group A streptcoccus or viral infection. Presents as oligoarthritis affecting the lower extremities

It is usually transient, lasting less than 6 weeks. It includes toxic synovitis. Common particularly after rubella, hepatitis B, mumps, measles, varicella, EBV, CMV

The management of a patient with postinfectious arthritis includes NSAIDs, physical activity, physical therapy, and penicillin prophylaxis for arthritis caused by streptococcus.

Reactive arthritis

Occurs after an enteropathic or urogenital infection

Symptoms present 2-4 weeks after the infection, usually oligoarticular and in the lower extremities.

Is variable and may remit or progress to a spondylarthritis. May have cutaneous symptoms, fever, malaise, fatigue

Hypermobility syndrome

The treatment of hypermobility syndrome is by explanation, counseling the patient to avoid excessive movement.

May presents as intermittent pain in a child, especially a girl aged 3 to 10 years that is increased with activity.

The physical findings in hypermobility syndrome include generalized joint laxity, musculoskeletal complaints, and normal skin.

Functional joint complaints

A patient with functional joint complaints has symptoms that are relieved by rest and worse with activity, usually at the end of the days and nights. There is no objective joint swelling.

May have hypermobile or normal joints; There are normal radiograph and lab findings.

The management of a patient with growing pains (benign nocturnal pains of childhood) includes education, reassurance, and healthy sleep hygiene.

Inherited disorders of connective tissues (see also VII.D.8)
Marfan syndrome

Marfan syndrome consists of disproportionate overgrowth of the long bones, anterior chest deformity, arm span greater than height, reduced upper to lower segment ratio, arachnodactyly, wrist sign, thumb sign, scoliosis, and pes planus.

Also increased aortic diameter, mitral valve prolapse, ectopia lentis, myopia, normal serum levels of homocysteine and methionine, and may have a fibrillin-1 protein mutation.

Ehlers-Danlos syndrome

May present with easy bruising and poor wound healing; Soft, velvety skin that is hyperelastic.

Lax joints that are easily subluxed, unusual scarring, with fragility of skin and blood vessels, mitral valve prolapse, or aortic aneurysm.

Disorders of the Eye

External disorders
Alignment and movement disorders
Strabismus

1.) Early detection and treatment of strabismus is important to prevent amblyopia and promote binocular vision.

2.) Pseudostrabismus (appearance of crossed eyes) can be ruled out by holding a flash light in front of a child's eyes and looking to see whether the reflection of this light is properly centered in each eye. If the eyes are aligned with one another then the reflection from the light will be in the same spot of each eye. If strabismus is present then the reflection from the light will not be in the same spot of each eye.

Nystagmus

1.) Nystagmus, a condition of voluntary or involuntary eye movement could signify important eye or central nervous system pathology.

Conjunctivitis
Allergic and infectious conjunctivitis differ in the following aspects

1.) Allergic conjunctivitis is the eye's reaction to an allergen while infectious conjunctivitis is caused by bacteria or a virus.

2.) Allergic conjunctivitis is not contagious while infectious conjunctivitis is contagious.

3.) Allergic conjunctivitis is managed by avoiding the offending allergen and applying topical antihistamines, mast cell stabilizers, or nonsteroidal anti- inflammatory agents. For bacterial conjunctivitis, use antibiotic drops or ointment and viral conjunctivitis resolves spontaneously with time.

Differential diagnosis of conjunctivitis includes systemic conditions like uveitis.
Neisseria gonorrhea, Staphylococcus, Streptococcus, Pseudomonas, Chlamydia tachomatis, HSV are prevalent causes for conjunctivitis in neonates while H. influenza (usually nontypable), Streptococcus pneumonia, Moraxella catarrhalis, Adenovirus; Enteroviruses and HSV are prevalent in older children.
Clinical manifestations of conjunctivitis (influence of age) include 1.) Redness, itchiness in one or both eyes

2.) Pain and tenderness in the eyeball

3.) Conjunctival discharge: purulent, mucoid or mucopurulent depending on the cause

4.) Conjunctiva shows hyperemia and chemosis. Eyelids are usually swollen.

Treatment of conjunctivitis depends on its cause:

1.) Viral conjunctivitis generally resolves with time.

2.) For bacterial conjunctivitis, use antibiotic drops or ointment.

3.) For allergic conjunctivitis, the easiest way to manage the reaction is to avoid

the allergen. Applying topical antihistamines,mast cell stabilizers, or nonsteroidal anti-inflammatory agents helps.

Conjunctivitis can be prevented by neonatal prophylaxis, frequent handwashing using clean towels and avoiding allergens' exposure.

Orbital and periorbital (preseptal) cellulitis

Differences between preseptal and orbital cellulitis include:

1.) Antecedent trauma or bacteraemia (Preseptal) vs. Antecedent sinusitis (Orbital).

2.) Periorbital erythema, warmth, tenderness (Preseptal) vs. Proptosis, chemosis, ophthalmoplegia, decreased visual acuity (Orbital).

3.) Mean age 21 months (Preseptal) vs. mean age 12 years (Orbital).

About 75 percent of orbital cellulitis cases are related to sinusitis, especially ethmoiditis. Staphylococcus and Streptococcus are the most common pathogens of orbital cellulitis followed by fungi, Mucorales and Aspergillus spp.

Orbital cellulitis results from microbial infection with subsequent inflammation of the post-septal aspect of the eyelids. The most common routes of infection are from adjacent sinuses or teeth and direct inoculation through penetrating lid injury. In addition to bacterial and fungal Rhinosinusitis, other causes for orbital cellulitis include ophthalmic surgery, peribulbar anesthesia and dacryocystitis.

Clinical manifestations of orbital cellulitis

1.) Eyelid swelling with or without erythema. 2.) Eye pain/tenderness.

3.) Pain with eye movements. 4.) Subtle Proptosis.

5.) Ophthalmoplegia with or without diplopia. 6.) Fever and occasional leukocytosis.

Laboratory tests include CBC with differentials, CMP, blood cultures and smear for Gram stain and culture of nasal discharge.

Treatment for orbital cellulitis is antibiotics or antifungal, diuretics for increased intra-ocular pressure, nasal decongestant if cellulitis is secondary to sinusitis and in some cases, surgical intervention.

The complications of orbital cellulitis could be severe and include blindness, hearing loss, cavernous sinus thrombosis, intracranial brain abscess, septicemia and meningitis.

The most common causes of preseptal cellulitis are Staphylococcus aureus, Streptococcus pneumoniae, other streptococci, and anaerobes.

Preseptal cellulitis arises from infection due to local facial or eyelid injuries, insect or animal bites, acute dacryocystitis, or sinusitis.

Preseptal cellulitis typically presents with ocular pain, eyelid swelling, and erythema.

Diagnosis of preseptal cellulitis is based on clinical evaluation looking for insect bite, local face or eyelid trauma. If orbital cellulitis is suspected, CT or MRI is used to confirm.

Treatment with antibiotics is recommended with surgery reserved for complicated orbital cellulitis.

Contrast-enhanced CT scanning of the orbits and sinuses is helpful for distinguishing between preseptal and orbital cellulitis. Imaging studies are indicated if any of the clinical signs or symptoms point to orbital cellulitis rather than preseptal cellulitis.

Stye, chalazion

A chalazion and a stye are both lumps in or along the edge of an eyelid but chalazia are usually larger, less painful and tend to develop farther from the edge of the eyelid than styes.

Warm compresses, antibiotic ointments, steroid (cortisone) injections and/or surgical draining is used to manage stye and chalazion.

Nasolacrimal duct obstruction

Nasolacrimal duct (NLD) is presented with increased tear meniscus, reflux of tears/mucoid discharge through puncta with palpation of lacrimal sac, and occasionally redness of the conjunctiva.

NLD is treated by lacrimal sac massage, lacrimal duct probing, balloon catheter dilation, Stenting or surgery, if needed.

Ptosis

Congenital ptosis is one that is present at birth and is important to be differentiated from acquired as it can lead to serious complications with visual system development and maturation.

Internal disorders

Congenital glaucoma

Congenital glaucoma presents with epiphora, photophobia and/or blepharospasm.

Congenital glaucoma may be associated with port wine stains involving the upper and lower eyelids.

The white pupil

Cataracts

1.) Congenital infections, inherited conditions, radiation, and corticosteroid use can promote congenital cataracts.

2.) Gray or white cloudiness of the pupil may signify congenital cataract.

3.) Eye injury, excessive sun or radiation exposure, family history, obesity, high blood pressure, diabetes, family history, alcohol abuse and smoking increase the risk of cataracts.

Retinoblastoma

1.) Leukocoria, strabismus and to a lesser extent decreased vision, ocular inflammation and family history signify the presence of retinoblastoma.

2.) Retinoblastoma could be hereditary and is passed in an autosomal dominant pattern.

Papilledema, papillitis

Advanced papilledema may cause decreased vision and brief recurring visual blackouts.

Papilledema requires immediate diagnostic evaluation and effective management as it could manifest a neurologic emergency. It is often accompanied by headache, pulsatile tinnitus, nausea, vomiting, and diplopia. Blindness can occur without treatmentas a severe complication of papilledema.

Retinopathy of prematurity

Preterm babies who received oxygen therapy require retinal examinations at 4 to 6 weeks of age as they are at high risk for Retinopathy of prematurity (ROP).
ROP screening should be performed for infants with

1.) Birthweight ≤1500g or Gestational Age (GA) < 30 weeks.

2.) Birthweight between 1500g and 2000g or GA > 30 weeks but at high risk clinical course.

Miscellaneous
General
Red eye is evaluated primarily by the measurement of visual acuity and findings on penlight examination in deciding to manage or refer.
Amblyopia
Strabismus, more nearsighted, farsighted, or astigmatic in one eye than the other eye, and rarely certain eye conditions such as cataract may lead to amblyopia.
Foreign bodies, corneal abrasions
Excruciating eye pain, foreign body sensation and irritability in the eye, eye pain disrupting the ability to work, drive or sleep, teary eyes and blurred vision are common presentations of corneal abrasion.
Administration of topical or oral analgesics, topical antibiotics, and cycloplegics along with daily follow-up until healed is advised for corneal abrasions.
Suspected ocular foreign bodies can be examined by a slit lamp examination, ophthalmoscope or imaging modalities (radiograph, CT, Ultrasound

biomicroscopy).

Trauma to the eye
Initial emergency assessment and treatment followed by ophthalmology consult. Imaging studies with additional emergency treatment as needed are necessary to evaluate ocular trauma.
Bruising, tenderness and swelling around the eye, redness of the eye, diplopia, numbness of the cheek, nose or teeth; epistaxis are clinical signs of a blow-out fracture of the orbit.
Tumor or hemangioma affecting vision
Proptosis, impairment of eye movement, ptosis, optic atrophy, and loss of vision are the consequences associated with tumors in the periorbital area.
Ear, Nose, and Throat Disorders
Ears
Congenital malformations
External and middle ears that are malformed could be a manifestation of renal anomalies, ear malformations and craniofacial malformations.
External ear
The pathogenesis of otitis externa involves the breakdown of the skin-cerumen barrier. Following this is skin inflammation as well as edema, which leads to pruritus, ultimately prompting scratching. This whole process impairs epithelial migration and increases ear canal pH. The result is an ear canal that is dark, warm, moist, and alkaline which is ideal for bacterial infections. Bacteria include: Staphylococcus epidermidis, Staphylococcus

auricularis, Pseudomonas aeruginosa, streptococci, and diphtheroids. Also known to infect the external ear are fungi such as Aspergillus and Candida.
Otitis externa differential diagnosis include the following: chronic suppurative otitis media, ear canal carcinoma, otomycosis, contact dermatitis and psoriasis.
Otitis externa management: topical agents such as antibiotics, antiseptics, glucocorticoids and acidifying solutions, as well as wick placement as required. For severe pain, NSAIDs are used.
For children with recurring otitis externa: during swim season use acidifying or alcohol drops as a daily prophylaxis; dry the ears gently with or without the head titled post swimming; and avoid placing cotton swabs in the ears.
Presentation of aural foreign bodies: pain, discomfort, bleeding, decreased hearing or discharge (otorrhea). In cases that are delayed, swelling of the ear canal and erythema may be observed.
Hematoma of the external ear can be manifested as accumulation of blood between the perichondrium and cartilage. Immediate needle aspiration as well as incision and drainage procedures are used as treatment and to prevent further complications.
Middle ear
Acute otitis media

1.) Etiology, epidemiology

i.) Otitis media is frequently found in infants who are fed with propped bottles

ii.) AOM pathogens include the following: bacteria – primarly Streptococcus pneumoniae, Moraxella catarrhalis, beta-lactamase producing Haemophilus influenae; viruses – RSV, coronaviruses, pcornaviruses, adenoviruses, influenza viruses; and other pathogens – Chlamydia trachomatis and Chlamydia pneumoniae.

iii.) Pathogenesis – upper respiratory tract infections (viral) are usually involved as well as colonization with an otopathogen. The respiratory mucosa of the nose, nasopharynx, and eustachian tube become edematous (inflammatory edema). As a resut, the isthmus is obstructed and middle ear mucosa secretions follow. Through aspiration, reflux, or insufflations, bacteria and viruses that colonize the respiratory tract enter the middle ear. This results in discharge (suppuration) accompanied by clinical signs of AOM.

iv.) 60 – 80 % of infants have an episode of AOM before they turn one, and 80 – 90% by age 3. Incidences increase in the fall and winter months. Additional risk factors include: day care exposure, lack of breast milk, tobacco smoke exposure, family history and pacifier use.

v.) Those at greater risk for recurrent or chronic middle ear disease later on in life are infants who have AOM.

vi.) An important cause of AOM in older children is nontypeable H. influenza (NTHi).

vii.) Common pathogents in young infants include: S. aureus, group B streptococcus and gram-negative organisms. These organisms are similar to those found in older children with AOM.

viii.) Bacteriology of AOM and bullous myringitis is identical.

2.) Diagnosis

i.) If AOM is present in the first 6 weeks of life, careful management and follow-up are required in order to avoid potential complications which include: hearing loss, disseminated infections, bactermia and/or meningitis.

ii.) Pneumatic otoscopy can best diagnosis middle ear effusions. Indication of middle ear effusion is diminished tympanic membrane mobility.

iii.) A major complaint of AOM is otalgia, along with other symptoms which include otorrhea, hearing loss and tympanic membrane bulge. Other non-specific symptoms present including fever, headache, irritability, vomitting, diarrhea, apathy and anorexia. Complications that are severe include tinnitus, vertigo, nystagmus, facial paralysis and aural swelling. Prevalent clinical syndromes are otitis-conjunctivitis and bullous myringitis.

3.) Therapeutic options

i.) Treatment in normal cases of AOM: high-dose amoxicillin or a one-time dose of IM ceftriaxone. Treatment for complicated of AOM cases: high-dose amoxicillin- clavulanate. Alternative treatments for children with non-anaphylactic penicillin allergy: cefdinir, cefuroxime, cefpodoxime. Anaphylactic penicillin allergies: azithromycin or clarithromycin. In cases of H. influenzae or M. catarrhalis, clindamycin is ineffective. Due to its inadequacy in AOM, trimethoprim-sulfamethoxazole is not recommended.

ii.) Decongestants and antihistamines are not recommended for treatment of AOM as their efficacy has been discredited via studies.

iii.) If there is no improvement of symptoms (persistent symptoms such as fever and/or pain, bulging tympanic membrane and other suppurative complications) 48 – 72 hours after the initation of antibiotics, immediate treatment should be implemented with amoxicillin-clavulanate or ceftriaxone, and if needed, tympanocentesis following antibiotics.

iv.) Patients with AOM that are symptom free may have persistent middle ear effusion which could last up to 2 – 3 months.

v.) For patients with mild symptoms and signs of unilateral AOM, watchful waiting may be appropriate for the first 48 hours with follow-up before initiation of antibiotics.

vi.) For patients with AOM who are in extreme pain and are immunocompromised, unresponsive to therapy, seriously ill and have extremely high fever (hyperpyrexia), surgical management such as tympanocentesis and/or myringotomy (drainage procedures) should be considered as a third-line therapy. Other complications that could prompt drainage include meningitis, mastoiditis, facial paralysis, labyrinthitis and suppurative complications.

vii.) Drainage procedures can be complicated by persistent otorrhea and/or perforation, ossicular chain injury, otitis externa, ear canal skin injury, ear canal stenosis, implantation cholesteatoma and tube migration to the inner ear, which is rare.

Otitis media with effusion (secretory otitis media)

1.) Diagnosis

i.) Predisposing factors: suspected or diagnosed language and speech delay or disorder, hearing loss independent of OME, developmental delay, autism spectrum disorders, uncorrectable visual impairment and craniofacial disordres. Additional risk factors: day care exposure, family history, viral infections and allergies.

ii.) The primary cause of diminished tympanic membrane mobility is middle ear effusion. Other etiologies: retraction due to scar

tissue or tympanosclerosis, perforation of tympanic membrane, and ensuing treatment of AOM. In rare cases, eustachian tube dysfunction and barotraumas may be a cause.

iii.) Chronic sinusitis and craniofacial abnormalities are normally linked with secretory otitis media.

iv.) For OME that persists for ≥3 months, children should undergo a hearing evaluation in order to decide management therapy.

a.) Watchful waiting is appropriate for hearing loss ≤20 dB ad without speech, language or developmental problems.

b.) Watchful waitingis appropriate, while a surgical evaluation is recommended, for hearing loss between 21 – 29 dB.

c.) Surgical referal for hearing loss ≥ 40 dB.

2.) Therapeutic options

i.) Management in asymptomatic patients: 3 months of watchful waiting followed by evaluation and hearing test repeated every 3 – 6 months until symptoms abate / effusion has resolved or there is an implication for surgical referral. An option for some cases is antibiotic treatment as a single 10 – 14 day course For chidren with

severe and recurrent OME, surgical managmeent after consultation is an option.

ii.) Due to failure in efficacy in trials and increase risk of side effects, decongestants and antihistamines are not reccomended therapies for OME.

iii.) Management options: watchful waiting, pharmacotherapy and surgical intervention. Pharmacotherapy – if failure of initial course of amoxicillin, antimicrobials effective against beta-lactamse producing bacteria as a second course is recommended. Surgery – dependent upon risk for langauge, speech or learning problems and severity of hearing loss.

iv.) Symptoms and risk factors determine the proper management of a child with OME.

a.) Watchful waiting is the optimal therapeutic option (see i. above)

b.) Treatment with antimicrobials is an option but not required.

While antimicrobial therapy has a modest short-term benefit for 2 – 8 weeks per some studies, there are no known long-term benefits. Within two weeks of discontinuing the antimicrobial, the initial benefits become insignificant.

c.) Surgical referral recommended:

Within three months, if suspected or diagnosed language or speech disorder, developmental delay, hearing loss independent of OME and uncorrectable visual impairment.
Hearing loss ≥40 dB, structural damage to the middle ear (i.e. tympanosclerosis, or retraction pocket cholesteatoma) or tympanic membrane
Bilateral OME ≥3 months, unilateral OME ≥6 months, cumulative duration of OME ≥12 months or hearing loss ≥21 dB, without language, visual or developmental problems.

3.) Complications

i.) Retraction of tympanic membrane or complications with tympanostomy tube placement could result in cholesteatoma.

ii.) OME is generally asymptomatic but could possibly present with discomfort, hearing loss, sleep disturbance and behavior changes. iii.) In young children with OME, the development of speech and language may be impacted by hearing impairment.

Chronic suppurative otitis media

1.) Hearing amplication (available for children of all ages) 2.) Bacteriology differs from AOM.

i.) Common bacterial pathogens include: P. aeruginsoa, S. aureus, Klebsiella pneumoniae, Proteus species and diptheroids.

ii.) The most common bacterial pathogens in AOM: S. pneumonia, nontypeable H. influenzae, and M. catarrhalis.

3.) Management

i.) Ear drainage culture and sensitivities to determine pathogen. Treatment with topical and/or systemic antibiotics along with granulation tissue control and appropriate aural toilet follow. Topical aminoglycosides or cortisporin are other otpions, as well as surgery if symptoms do not improve.

Recurrent otitis media

1.) Effective management: second-line antimicrobials effective against beta-lactamase producing strains of M. catarrhalis and H. influenzae, as well as susceptible and most nonsusceptible strains of S. pneumoniae. Highly effective antimicrobials: amoxicillin- clavulanate, cefuroxime axetil, and IM ceftriaxone.

2.) Recurrent otitis media can be caused by cleft palata, chronic sinusitis, dysmotile cilia syndrome, primary ciliary dyskinesia, and immunodeficiency.

3.) Otitis media follow-up evaluation is critical to determine if recurrent and/or chronic middle ear effusion is evolving.

4.) Subtherapeutic doses of antimicrobial prophylaxis (i.e. an aminopenicillin or sulfonamide) may help control. It is possible that bacterial immunity to antimicrobials could develop. Following initial antibiotic treatment failure, second-line antimicrobials (i.e. amoxicillin-clavulanate, cefuroxime axetil, cefdinir, cefpodoxime, and IM ceftriaxone) should be used as treatment.

Other (not otitis media)

1.) Referred otalgia causes: dentition eruption, dental pain, TMJ dysfunction, otitis media, otitis externa, cerumen impaction, mastoiditis, tonsillitis and pharyngitis.

2.) AOM is most common cause of otorrhea from tympanostomy tube. Another cause of otorrhea is reinfection of the middle ear either from nasopharyngeal reflux through the eustachian tube or an ascending infection from the nasopharynx.

3.) Intratemporal complications of middle ear include: perforation of tympanic membrane, tympanomastoiditis, petrositis tympanosclerosis and acquired cholesteatoma.

4.) After placement of a ventilation tube, infection and otorrhea may result due to an

ascending infection from nasopharyngeal reflux, tube lumen obstruction or premature dislodgement of tube.

5.) Intracranial complications of middle ear include: meningitis, extradural abscess, subdural abscess, brain abscess, sigmoid sinus thrombosis and otitic hydrocephalus.

Otorrhea

1.) Watery otorrhea that is persistent should be promptly managed as it may be indicative of a cerebrospinal fluid leak.

2.) Purulent otorrhea causes: AOM or chronic otitis media with tympanic rupture; subsequentially following an infection or trauma due to otitis externa.

3.) Bloody otorrhea: trauma from usage of cotton swabs, ear thermometers, cerumen removing devices, scratching due to eczema as well as H. influenza, M. pneumoniae, and some viral infections.

Inner ear
Benign paroxysmal vertigo presentation: intermittent episodes (most prevalent within three years of birth) of dizziness, loss of balance, unsteadiness, lightheadedness, horizontal nystagmus, nausea and vomitting.
Most common differential diagnoses in children with balance disturbance: otitis media, cholesteatoma, labyrinthitis, menier disease and cerebellar dysfunction.
Inner ear complications may also be caused by bacterial and viral infections such as cytoegalovirus, mumps and rubella.
Hearing loss
Etiologies

Conductive hearing loss in children

Congenital:

Microta/atresia, tympanic membrane abnormalities, ossicular malformations

Acquired:

infection (AOM, OE, ossicular eriosion), foreign body (cerumen), cholesteatoma, trauma (ossicular disruption, TM perforation)

Moderate to severe conductive hearing loss after trauma may represent ossicular disruption

Blows to the head will cause disruption in the ossicular chain, temporal bone fractures, transverse fractures, penetrating trauma into the inner ear may damage cochlea.

Commonly used pediatric drugs might produce sensorineural hearing loss

Aminoglycosides, loop diuretics; Chemotherapeutic agents,(cisplatin) thalidomide, retinoid, chemicals like quinine, lead and arsenic.

Acoustic trauma produces high- frequency hearing loss

Acoustic trauma produces high frequency hearing loss ranges > 4000 Hz. Exposure to continuous noise greater than 85dB or sudden noise greater than 140 dB.

A mild conductive hearing loss may be subtle and may present as ignoring behavior, increasing the television volume, etc.

Can present as ignoring behavior, increasing TV or video game volume, asking to repeat questions or commands, not hearing instructions, behavior problems, problems with ability to understand and regulate emotions.

Neonatal risk factors for a sensorineural hearing impairment:

Hyperbilirubinemia, infection, craniofacial deformities, family history, neonatal depression, low birth weight, prolonged ventilation, ototoxic drugs, certain syndromes (eg, Waardenburg), infections (CMV, measles, mumps, rubella, varicella, syphilis), craniofacial deformities, family history, neonatal depression, low birth weight, prolonged ventilation, ototoxic drugs used during pregnancy, alcohol, isotretinonin, cisplatinum, Syndromes Waardenburg- hypertelorisim pigmentary abnormalities hearing loss to varying degrees Pendred Syndrome: goiter and enlarged vestibular aqueducts , Alport syndrome progressive sensorineural hearing loss, as well as progressive nephritis.
Diagnostic techniques

Tympanometry provides a graph of the middle ears ability to transmit sound energy (admittance or compliance) or impede sound energy as a function of air pressure in the external canal. Measured functions include air pressure, sound pressure, and measure TM function.

Age dependent hearing techniques include:

Newborn- 1 month: otoacustic emission testing, auditory brainstem response (ABR)

All Ages: OAE, Tympanometry

Birth to 9 months- ABR; Birth to 6 months- Behavioral Observation Audiometry (BOA)

9 months to 2.5 years:: Visual reinforcement audiometry (VRA)

2.5 ro 4 years- Play Audiometry

4 years to Adult: Conventional Audiometry The auditory system can be evaluated at any age.

Abnormalities of the TM can change the shape of the curve or the tympanometric gradient width. Rounded peak is consistent with effusion; estimation of ear canal volume can be helpful if large, suspect opening in TM via tube or perforation.

Only 50% of children who had hearing loss were correctly identified by targeted screening. Reliability is questionable therefore ongoing assessments are imperative. High false positive rates.

Findings of tympanomety can be normal in sensorineural hearing loss, auditory neuropathy.

Therapeutic options

Hearing loss 30 % of the patients can be unilateral or bilateral and it is more common with meningioccal meningitis. Every child with meningitis should have a hearing test; it appears early and worsens with time.

Alport syndrome is a genetic syndrome associated with sensorineural hearing loss, progressive nephritis, and eye abnormalities.

Mastoiditis

Mastoiditis can be a complication of supprative otitis media; mastoid antrum provides a conduit for infection spread.

Bugs of mastoiditis include:

Streptoccus pneumonia, S. pyogenes, Staphylococcus aureus, Haemophulus influenza, can be polymicrobial wth anaerobes, peptostreptococcus, actinomyces, bacteroides, gram negative bacilli, mycobacterium tuberculosis

Clinical manifestations of mastoiditis include:

postauricular pain, erythema and tenderness of tissue over mastoid, outwardly displaced pinna, history of AOM, fever, otalgia, headache, rhinorrhea, hearing loss, neurological complications and possible subperiosteal abscess.

Appropriate therapy for mastoiditis includes:

Culture specific intravenous antibiotics, myringotomy, tympanostomy tube placement, and mastoidectomy.

Laboratory evaluation of mastoiditis includes:

culture of middle ear fluid, CT scan which may show haziness or destruction of mastoid outline, plain radiography can should clouding of the mastoid or coalescence of the air cells. CT with enhancement or MRI is the most recommended test especially if there is worry for vascular thrombosis.

Nose and nasopharynx

Choanal atresia

Signs and symptoms of choanal atresia include:

respiratory distress soon after birth that is relieved by crying, inability to feed normally, in cases of unilateral CA: chronic mucoid discharge on affected side, chronic mouth breathing

Evaluation of child with choanal atresia includes inability to pass a firm catheter into oropharynx through the nares. Radiographic view of the choanae are helpful

50 to 70% of affected infants have other congential anomalies Associated congential anomalies occur more often in the bilateral cases of choana atresia. For example, in CHARGE.

Epistaxis

Evaluation of severe recurrent epistaxis includes:

hematologic evaluation (for coagulopathy and anemia) nasal endoscoy, and diagnostic imaging for AVM malformation, Family history is important.

Differential diagnosis of epistaxis includes:

local trauma, allergic disease, chronic sinusitis, nasal foreign body, medication use, intranasal neoplasm, intranasal polyps, septal perforation, vascular malformations, leukemia, Wegner granulomatosis, coagulopathy due to liver disease, bleeding disorders Von Wildebrands, hemophilia, platelet dysfunction.

Rhinitis a.General

Nasal congestion can be associated with cocaine use in adolescents

Complications of topical decongestants include safety between therapeutic and toxic doses is unknown, prolonged use can cause rhinitis medicamentosa, rebound nasal congestion.

Neonates are obligate nose breathers.

Differential diagnosis for chronic rhinitis includes:

choanal atresia, cocaine use, sinusitis, nasal polyps, cystic fibrosis, foreign body, allergy

Allergic rhinitis (see also VIII.B.)

Allergic rhinitis history:

Nasal congestions identified as mouth breathing, snoring, nasal voice, nasal irritation, paroxysmal sneezing, nasal and palatal puritis, nose blowing, sniffling, snorting occasional cough, allergic salute, itchy eyes, postnasal drip.

Physical examination for Allergy: endematous nasal turbinates edematous, pale to bluish hue, cobblestoning from lymphyoid hyperplasia in posterior oropharynx, dark discoloration underneath eyes, Dennie lines, transverse nasal crease across bride of nose, open mouth, receding chin, elongated face, arched hard palate.

Nonallergic rhinitis, history to focus on symptoms: duration, exposures, reaction patterns, chronicity, triggers, seasonal variation, environmental influences, allergies medical history headache, malaise, fatigue

Physical exam: eczema, wheezing, occasional fevers, lymphadenopathy.

Understand the treatment of allergic rhinitis.

Treatment of allergic rhinitis includes:allergen avoidance, indoors from 5 am to 10 am, air conditioning to remain on during spring and fall, dehumidifier, dust mite covers on bed and pillows, hypoallergenic pillows and comforters, remove pets from bedrooms, wash linens in hot water to denature dust mite dander, intanasal corticosteroid.

Infectious rhinitis

Know that group A streptococcal infection can present as protracted nasopharyngitis in infants and younger children.

Infants and young children with GAS infection present as protracted nasopharyngitis.

Recognize the systemic effects of oral decongestants and antihistamines in infants and young children.

Systemic effects of oral decongestants and antihistamines in infants and young children include cardiac related sequale with palpitations and tachycardia and stimulatory effects like insomnia, nervousness and irritability.

Polyps

Recognize that nasal polyps occur in children.

Conditions associated with nasal polyps include: Cystic fibrosis, chronic allergic rhinitis, chronic sinusitis and asthma.

The presence of nasal polyps is an indication for a sweat test, even in the absence of failure to thrive.

Common cold

Epidemiology: cold season begins in Septemberand remains until spring. Results from a number of viruses. Early fall: rhinovirus, parainfluenza. Late fall: RSV, coronavirus. Increase during winter months with final surge of rhinovirus, infections in spring. Most common in children less than 6 years, who get 6 to 8 colds a year, who have poor hygiene, and attend child care centers. In the teenage

years frequency decreases to four to five colds a year; Adults with young children get more colds than those adults who do not have children.

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Microbiology of common cold includes: Rhinovirus, parainfluenza, RSV and coronavirus.
Colds can trigger asthma in children.
Trauma

Hematoma of the nasal septum is a nidus of stagnant blood which is good medium for bacterial growth or abscess formation; it can cause septal perforationor destruction, with saddle nose deformity.

Physical findings of a hematoma of the nasal septum include:

Swelling pain and fluctuance, septal asymmetry with bluish or reddish hue to mucosa palpation feeling for swelling fluctuance or widening of septal space.

Management includes:

Evacuation of the clot with reapproximation of the perichondrum to cartilage, broad spectrum antibiotics, evacuate reoccurrence pack the nostril.

Foreign body

Nasal foreign bodies present with epistaxis, malodorous copious drainage, blockage of affected nostril.

Adenoids (see XXIII.G.1.)
Sinuses
Acute sinusitis

Diagnostic imaging using coronal CT of the sinuses is best. Plain radiographs do not image the sinuses correctly.

The common clinic presentation of acute sinusitis in children includes: swelling, headache, fever, nasal discharge and foul breath, very rarely there is facial pain.

The microbiology of acute maxillary sinusitis includes: pneumococci, H. influenza, and M. Catarrhalis.

Acute sinusitis unresponsive to conventional therapy may require surgical referral.

Surgical procedures: opens the osteomeatal unit improving sinus drainage.

The treatment of acute sinusitis includes amoxicillin for 14 days, if there is worry for b-lactamase organisms amoxicillin-clavulanic acid is preferred. If there are allergies, azithromycin, TMP-SMX, and clarithromycin may be used. Ineffective drugs include first generation penicillins, erythromycin, cephalexin.

Complications that occur with ethmoid sinusitis include: Ptosis, abscess, facial infection, and intraorbital extension of infection.

Intraorbital and intracranial complications may occur as a result of frontal sinusitis as the Veins of Breschet communicate freely to the dura.

Development of the maxillary and ethmoid sinuses occurs at 2 growth spurts 1 to 3 years and 7 to 12 years. Sphenoid pneumatized at 3years of age, frontal sinuses at 6 years.

Sinusitis differential should include- toothache, sore throat, persistent cough, and poorly controlled asthma.

Nasal swabs and throat cultures may not correlate well with cultures of the sinuses as the cultured area is usually polymicrobial and is usually a contaminated specimen.

There are no proven benefits with the administration of decongestants and antihistamines in the treatment of acute sinusitis.

Chronic sinusitis

Predisposing factors for chronic sinusitis in children include: Allergy, immune deficiency, primary cilliary dyskinesis, cystic fibrosis.

Trauma

Frontal sinus fracture requires surgical repair because there are freely communicating veins of Beschet which can spread infection.

Throat
Infections
Viral (see also IX.H.)

Viral infections of the throat are the most common infections. Viruses like adenovirus, coronavirus, enterovirus, rhinovirus, RSV, EBV, HSV, metapneumovirus.

Bacterial
Peritonsillar abscess

Signs and symptoms of peritonsillar abscess in children include: sore throat, fever, trismus, dysphagia, ipsilateral otalgia.

Needle aspiration usually showing mixed oropharyngeal bacteria, GAS CT of neck is most helpful for imaging. Laboratory studies including CBC and inflammatory markers are useful, especially for monitoring response to therapy.

Treatment for peritonsillar abscess includes: amoxicillin + clavulanic acid, penicillin + metronidazole, ampicillin sulbactam, ticarcillin+ calvulate, cefoxitin, clindamycin. Ineffective drugs include those which do not provide anaerobic or b-lactamase coverage. Surgical drainage with needle/incision or tonsillectomy; to send aspirate for gram stain and culture to better direct antibiotic therapy; Adjuvant corticosteroid therapy is controversial.

Microbiology for pertonsillar abscess includes: Streptococcus pyogenes, staphylococcus aureus, H, influenza, anaerobic bacteria, such as prevotella, bacteroides and peptrostreptococcus.

Tonsillitis, pharyngitis

Antibiotic therapy is indicated in cases of uvulitisor pharyngitis with GAS for the prevention of of rheumatic fever as a complication.

Throat culture in pharyngitis is useful for group A strep infection and N. gonorrhoeae. Throat and nasopharyngeal cultures are of no value for predicting etiology of otitis, pneumonia, meningitis, or sinusitis.

The differential diagnosis of exudative tonsilopharyngitis includes: Adenovirus, EBV, GAS, enterovirus, Neisseria gonorrhea.

Persistence of positivity of GAS after a course of treatment in some children can be because of a carrier state and it can confound later throat swabs. This must therefore be eradication of the carrier state by giving clindamycin 20mg/kg divided q 8 for 10 days. In such cases, eradication of carrier state with Clindamycin 20 mg/kg/day q8 hours for 8 to 10 days can be helpful.

Retropharyngeal abscess

Treatment of retropharyngeal abscess includes: third generation cephalosporin combined with ampicillin-sulbactam or clindamycin for anaerobic coverage. Surgical drainage is necessary to increase penetrance of antibiotic if antibiotic therapy is not adequate.

Microbiology of retropharyngeal abscess includes Polymicrobial GAS, anarerobic bacteria, Stapylococcus aureus, H influenza, Klebsillea, mycobacterium avium intracellularae.

Clinical signs of retropharyngeal abscess include Fever, irritability, decreased oral intake, drooling, neck stiffness, torticollis, and refusal to move neck. Also, can have muffled voice, stridor, respiratory distress, obstructive sleep apnea, and cervical lymphadenopathy.

Imaging studies that aid in the diagnosis of retropharyngeal abscess include CT neck which can show deep infections, increased width of retropharyngeal space, and air fluid levels. Lateral inspiratory neck radiographs are also useful.

Mouth and oropharynx
Tongue, oral cavity, uvula, salivary glands

Herpetic Gingivostomatitis: HSV type 1 occurs 3 to 50 days after exposure, contact with infected person, fever, irritability, malaise, small vesicles on ginigiva which rupture and become painful ulcerations, gingiva appear “fire engine red”, involves buccal mucosa and lips; spares posterior pharynx, check with PCR testing, spontaneous resolution 10 to 14 days, symptomatic support.

Herpangina: Coxsacie virus, epidemics during fall and winter, malaise, sore throat, low grade fever for several days before vesicles in mouth, 4-5 vesicles under the soft palate of tonsillar pillars, red macules progess to large vesicles which then ulcerate and become 2-4 mm. Pharyngeal vesicles and unlcerations. Resolve in 10 days symptomatic support.

Hand Foot Mouth Disease: Coxsackie, occurs during fall and winter, malaise sore throat, low grade fever, intraoral vesicles and vesicles in locations like the palms and soles which have erythematous margins.

Apthous Ulcers: minor apthous ulcer - 5mm in size, appear unattached to ginigiva heal in 7 to 10 days, Major apthous ulcers are on kertinazied mucosa

- attached to ginigiva or palatal tissue, larger, last longer, result in scaring. Yellow pseudomemnranous slough with erythematous boarder, thought to be

caused by vitamin deficiency, treatment includes topical analgesic, vitamin B and C supplementation.

Children with short lingual frenulum require no intervention unless there are troubles with feeding as a baby or speech or articulation issues.

The causes of parotitis: viral: mumps bacterial: staph aureus, recurrent idiopathic: poor oral hygiene, S. pneumonia, sialadenitis: Staph Aureus, alpha hemolytic strep, H. influenza, Stretococcus pyogenes, Stretptococcus. pneumonia, Moraxella Catharralis, Escherichia Coli, anaerobic bacteria.

The differential diagnosis of preauricular swelling includes: parotitis, lymphadenitis, tumor, and lymphosarcoma.

The clinical presentation of cold panniculitis includes inflammation of the subcutaneous fat. Presents will ill-defined erythematous to bluish indurated plaques or nodules that arise within hours to days on exposed surfaces, they persist for 2 to 3 weeks and heal without residual issues.

Bifid uvula is associated with submucous cleft palate and middle ear effusion.

Cleft lip, cleft palate

Feeding a newborn infant with a cleft palate consists of small frequent feeds. Infants have difficulty with suction in mouth. Children need to be positioned upright to so that milk may not escape through the nostrils. They also needed to be burped 2 to 3 times during the feed because they swallow more air. Bottles should be positioned upright or an angled bottle should be used, with a cross cut nipple to increase size of aperture. Child should feed for 30 minutes at a time. Use of a squeezable bottle is recommended to provide less forces from the newborn.

Clinical problems in children with cleft palate include: inability to clear middle ear therefore constant effusion which can lead to hearing impairment and possible speech and learning impairment, velopharyngeal insuffiency where the soft palate cannot seal, therefore leading to high pitched nasal voice, speech problems also related to dental malocclusions as a result of clefts or surgical repair with fistula formation. Dental: missing lateral incisors or poorly formed teeth or lack of enamel because of osseous defect in maxillary alveolar bone.

Children with cleft palate have difficulty properly opening and ventilating their Eustachian tube because of trouble with the tensor and levator veli palatni muscle.

Submucous cleft and recurrent chronic otitis media occur as a result of inability to clear the Eustachian tube which leads to chronic middle ear serous fluid buildup. Most of these children receive myringotomy tubes.

Management of mandibular hypoplasia is necessary because failure of the mandible to grow leads to tongue obstruction in the back of pharynx which blocks the ability of the palatal shelves to fuse. Tongue then falls backwards and causes obstruction and respiratory distress. In some cases children need a jaw expansion procedure.

Cleft palate deformities can be associated with Trisomy 13, velocardiofacial syndrome, teratogen use, skeletal, craniofacial and eye deformities.

Teeth
Normal development

Delayed dental eruption is defined as exposure of a tooth or multiple teeth through the oral mucosa which is overdue, according to the population based on chronological age. Causes of delayed dental eruption include: hypothyroidisim, hypopituitarism, ectodermal dysplasia, rickets.

Counseling parents regarding their child's dental care includes:

Oral hygiene with daily brushing with fluoridated toothpaste to prevent dental caries. Children less than 8 years of age are not able to coordinate brushing properly, therefore parent needs to assist the child. Diet should not include frequent fruit juice or sweetened beverages from a bottle or sippy cup.

Trauma

Therapeutic options for an avulsed tooth if it is a permanent tooth should be reimplanted 20 minutes after injury. Success may be achieved if delay is less than 2 hours, failure is common because of the inability of the root to be reabsorbed or ankylosis. The likelihood of normal reattachment depends on the viability of the periodontal ligament.

Cavities

Topical fluoride application with fluoridated toothpaste for children older than 6 years of age should be a "pea sized" ¼ gram on a toothbrush. For children less than 2 years who are at risk for cavities a "smear" of toothpaste should be used.

Nursing cavities are found in those children with frequent ingestion of sugar via bottle/ sippy cup especially at nap or bedtimes, and there is a visible layer of tartar noted on the teeth.

Infection

Dental infections can present as:

Swelling below the jaw with mandibular dental abscess, periorbital swelling with maxillary dental abscess

Anaerobic infections such as streptococcus mutans are associated with dental infection, they adhere to enamel, produce abundant acid, and survive at low pH. Once the enamel surface cavitates other oral bacteria like lactobacilli can colonize the tooth, produce acids, and foster further demineralization and further injury with periodontal disease.

Adolescent Medicine and Gynecology

Adolescent development
Physiologic
Pubertal staging (sexual maturity rating)
General:
The stages of sexual development::
Thelarche – in girls, onset of breast development
Pubarche – in girls, onset of sexual hair growth
Menarche – the start of menses
Spermarche – appearance of sperm in seminal fluid
Gonadarche – onset of pubertal function of gonads (make sex hormones)
Andrenarche – production of adrenal androgen that starts pubarche
Between 10-13 years of age, puberty begins in most boys, with testicle enlargement being the first change, pubic hair and growth of penis following, and sequential growth at peak height velocity.
Between 9-12 years of age, puberty begins in most girls, with breast enlargement being the first visible sign, with growth at peak heigh velocity and menarche (12.5 years is average age) following.
Stage 2-5 of sexual maturity progression can take up to 2.5 years to complete.
Delay of bone age is usually up to two years and can be considered normal.
When compared to adults, teens' upper body segment to lower body segment ratio is lower during growth spurts.

In cases of delayed puberty (commonly thought to be autosomal dominant with or without incomplete penetrance), some studies have shown that

youth with delayed puberty have either a parent or sibling with delayed puberty.

Gonadotropin-releasing hormone (GnRH) – stimulates release of follicle stimulating hormone (FSH) and luteinizing hormone (LH), in a pulsatile manner.
Male:
Development of secondary sexual characteristics sequence: testicular growth, pubarche, penile growth, peak height velocity.
Peak height velocity is usually reached after genital sexual maturity in most boys.
Enlargement of testicles > 4mL in volume is the earliest sign of puberty
60% of boys may have gyenocomastia, which is a normal pubertal occurrence. Two years after onset it can resolve. If there is asymmetry of breast development, pathology is not implied.
Female:
Development of secondary sexual characteristis sequence: breast budding (estrogen), pubarche (androgen effect), peak height velocity, menarche.
Menarche onset: 9-14 years of age; bone age of 12.5 years.
Girls heights will increase 6-8 cm post menarche.
At stage 4 of breast development, bleeding from menarche occurs. If bleeding occurs at stage 2 for breast development, menarche is not likely. In this case, other etiologies need to be considered.
A white discharge due to the effects of estrogen, called leukorrhea, may occur three to six months before menses.

Growth spurt – increase in height and weight

Peak height velocity in adolescents is more parallel with sexual maturity than chronological age.
5-6 cm/year = prepubertal height velocity average
9-10 cm/year = accelerated rate during peak growth spurt in adolescence
2-4 years = pubertal growth spurt completion
An x-ray of the left hand and wrist is usually used to measure bone age. This involves the least radiation exposure. Gruelich and Pyle is the published standard for bone age reference.
Changes in bones are caused by sex hormones (i.e. widening is caused by androgen, epiphyseal fusion is caused by estrogen).
A two year delay in bone age can be a result of constitutional delayed puberty.
Formula for adolescent stature estimate (midparental height, in inches)
Boys = [(maternal height + 5) + paternal height] / 2
Girls = [maternal height + (paternal height – 5)] / 2
*If using metric units in centimeters, replace 5 with 13

Changing laboratory parameters

Hematocrit increases to about 39% in males who are in sexual maturity stage 1 and 43% in stage 5.
Hemoglobin concentrations in males is about 14-18 g/dL

Increased alkaline phosphatase could be due to a growth spurt
Cholestrol concentrations increase
Total cholesterol = <170 mg/dL
Borderline = 170-199 mg/dL
High = >200 mg/dL
LDL cholesterol = <110 mg/dL is acceptable
Borderline = 110-129 mg/dL
High = >130 mg/dL
HDL cholesterol = > 40 mg/dL
Adolescent blood pressure is correlated with height and age.
Hypertension in adolescence = BP in the upper 95th percentile

Failures and variations in adult growth and development

Growth velocity is helpful for an adequate evaluation of short adolescents, not one single point value.
Constitutional delay = during childhood, a lower percentile height; delayed pubertal growth; and one parent with a history of short stature as a child, delayed puberty, and ultimately normal stature.
Hormone level checks with further investigation is required for girls after 13 years of age with no signs of puberty (thelarche, pubic hair) and boys after 14 years of age (no enlargement of testes or lengthening of penis).
Constitutional delay = most common cause of delayed puberty in boys
Delay of bone age by 2 or more years. Correlates better with height rather than chronological age.
Pubertal delay in females who are very short may be the first sign of Turner syndrome.
Pubertal delay in females with Turner syndrome is related to primary ovarian failure.
Estrogen, and other hormones that accelerate growth rate, also accelerate epiphyseal closure and may possibly limit growth potential.
A sudden halt in rate to normal growth in an adolescent requires an evaluation with lab tests which include: LH, FSH, and bone age.
Precocious puberty in boys:
Volume of testicles ≥4 mL before age 9.
MRI scans of the brain can show central nervous system abnormality (up to 75% of scans)
Risks: teasing, low self-esteem, school avoidance and decline in academic performance.
Premature development can cause emotional trauma to boys. In order to help the patient and his parent(s) deal with the stressors involved, psychological support is needed.
Psychological support is needed for both patients and parents

Psychologic growth and development

Development of a self-identity
Peer relationships and group belonging is critical to an adolescent's development of self-identity. Those who do not have these ("a loner") may have psychological difficulties.
Self-esteem and the emotional life of an adolescent are greatly impacted by physical development. Adolescents respond differently to rapid body changes, i.e. boys who develop early can have more self-confidence and are likely to be more successful in academics,

athletics and social groups than their peers, whereas girls who develop early can have concerns over body image and a lower self-esteem.
An adolescent's search for personal identity can be found in their experimenting in behaviors or various styles of dress.
Rebelling against the family's image is a normal process in an adolescent's search for identity. Through periods of rebellion and rejection, adolescets will usually assent to and repeat parental values.
Adolescent physical maturation rate does not parallel cognitive development rate.

Psychologic separation from the family

Enables adolescent to later return to the family. Through separation an adolescent may be able to find other peers and adults in order to diversify their psychosocial development.
The peer group is an important channel for separation from the family in young adolescents.

Relationship to the peer group

Homogenous peer groups in young adolescents = similar behavioral, dress and grooming standards.
Adolescent health is greatly impacted by peer groups, which give off peer pressure thus engagement in unhealthly behaviors (i.e. sex, alcohol, soking, drugs, and negative schoool attitudes) is likely.

Consequences of the emergence of a poor self-image

Many problems (familial and peer relationships, school underachievement, depression, suicide, risk taking behavior, etc.) are associated with poor self image. A positive self-image can be fostered through examples set in the lives of the parents and in other authority figures. Acceptance of the adolescent, as well as notice of admirable positive qualities and praising them for it, is very important in developing a postitive self image.

Family influence

Turmoil in the family can be expressed through behavioral problems. If the behavior of an adolescent appers to exceed limits of normal experimentation, information about social and family background (where behaviors occur) should be sought.
Sources of stress within families is very important to identify, as well as figuring out coping mechanisms for stress. Hoping to return to a more simpler time, adolescents may go into regression. This can be considered normal unless the adolescent becomes increasingly absorbed in these regressing activities, in which further evaluation may be required.
Family plays a crucial role in modeling behaviors of drinking alcohol, cigarette smoking, violence, and conflict resolution.

Media influence

Media can have a very strong impact on adolescents.
A risk factor for adolescent aggressive behavior, is being frequently exposed to violence on television at an early age. Screening for activities such as fire setting and cruely to animals is also very important.

Pro-tobacco media and marketing exposure endorse attitudes about tobacco that are positive.

Sexuality

Risk taking behaviors, such as sexual experimentation, is very common in adolescence (teens). In early adolescence, it is common to experiment with both homosexual and heterosexual relationships.
Peers and the media are the most common sources of information about sexuality, not parents, doctors or teachers.
Counseling parents about emerging sexuality: experimentation is normal and is expected; sexual orientation is set/developed during late adolescence.

ealth issues of adolescence

Preventive healthcare:
When evaluating adolescents, the following risk factors should routinely be reviewed: school performance, extracurricular activities, proper nutrition and exercise, sleep, sexuality issues and substance abuse.

Gynecology:

General:
In order to rule out sexually transmitted infections (STI's), pelvic examinations are necessary. Also useful in the diagnosis of STIs are swabs for culture, cervical motion tenderness and palpation for masses.

Vaginal discharge

Leukorrhea occurance before the onset of menses is normal.

o Management: reassurance, good hygiene eduation (front to back wiping, cotton underwear), sitz bath use (room temperature water, air drying), avoidance of douching and bubble bahs, no antibiotic or vaginal cream use.

The majority of vaginal discharges that are pathologic are related to sexual activity.
Uncomplicated pathologic vaginal discharge evaluation: speculum exam, bimanual exam, wet prep and culture.

Dysfunctional uterine bleeding

Differential diagnoses: tubal pregnancy, threatened abortion, pelvic inflammatory disease, coagulopathies and endocrine and hormone disorders such as hypothyroidism and polycystic ovary syndrome.
A workup for iron deficiency must be included in evaluation
Menometrorrhagia common causes: hypothalamic-pituitary-ovarian axis immaturity, platelet dysfunction, von wildebrand disease, and endometritis.
Dysfunctional uterine bleeding reveals anovulation.
10 days or more of menstrual bleeding is not physiologic.
Osteoporosis risk (due to ovarian failure)

Preferred treatment: reassurance, supplementation with iron, NSAIDS, and combination oral contraceptives.

Amenorrhea
Primary amenorrhea is defined as lack of menses by age 16 years or by two years after sexual maturation
The differential diagnosis of primary amenorrhea include:
Anatomic variants of the genital tract
Pregnancy
Ovarian pathology
Adrenal disease
Hypothalamic/pituitary disorders
Secondary amenorrhea is the cessasation of menses after menarche
The etiologies of secondary amenorrhea includes:
Hypothalamic pituitary ovarian axis abnormality with the disrupting agent ranging from neoplasm such as craniopharyngioma or pituitary adenoma, to physical or emotional stress
Exercise induced amenorrhea occurs due to reduced adipose tissue, which causes lower levels of leptin, which is a natural trigger for Gonadotropin Releasing Hormone(GnRH). Reduced levels of GnRH produce levels of Leutenizing Hormone (LH) and Follicle Stimulating Hormone (FSH) that are too low to stimulate ovulation
The most common cause of secondary amenorrhea in adolescents is polyscystic ovarian syndrome (PCOS).
Adolescents with PCOS often present with hirsutism or hirsutism equivalents, menstrual irregularities, obesity, and/or acanthosis nigricans.
Recognize that anorexia nervosa associated weight loss may be preceded by amenorrhea. Typically, menses ceases when body weight falls to 85% of ideal for age and height.
Recognize that menstrual irregularity is part of the criteria for diagnosing amenorrhea but not for diagnosing bulimia.
The management of exercise-induced amenorrhea involves restoring and maintaing a normal body weight, which will allow the natural return of menses.
Dysmenorrhea
The differential diagnosis of dysmenorrhea, including primary dysmenorrhea (without organic disease) and secondary dysmenorrhea.
There is a role of prostaglandin inhibitors in the treatment of primary dysmenorrhea.
The cost and benefit of various treatment options for primary dysmenorrhea.
Recognize that primary dysmenorrhea is a common cause of episodic school absence in adolescent girls.
The pathophysiology of primary dysmenorrhea includes dysfunction at the hypothalamus, pituitary, or ovary. There is decreased hormones (LH and FSH) in Primary dysmenorrhea. These hormones are required to initiate ovulation.
Pregnancy:

Medical considerations:

In order to improve physiologic outcomes for young adolescent mothers; good prenatal care and nutrition should be employed.
The younger the mother the greater the risk of complications for both mother and fetus.

Emotional, social, and economic issues are associated with teenage pregnancy.

emotional- dealing with the overwhelming knowledge of having a baby while still attempting to understand your own situation in life

social- support for mother and baby from family members as well as from the babies father

economic- majority of teenagers will not have jobs that will support themselves or a new baby and therefore other sources of financial support are sought (e.g. welfare, family members).

Prevention:

Forms of contraception available to adolescents include: oral contraceptive pill (OCP), transdermal, injectable, vaginal rings, subdermal implant, and emergency contraception (list does not include non-medicinal methods)

Complications associated with various forms of contraceptives:

OCP (listed below), transdermal- skin irritation and rash at the site of application, dysmenorrhea, along with higher incidence of breast discomfort/pain. Injectable - weight gain, loss of bone density. Vaginal ring- vaginitis, headache, leukorrhea, nausea, vaginal discomfort, subdermal implant- irregular bleeding, Emergency contraception- headache, fatigue, nausea, and dizziness (contraindication: allergy, pregnancy, undiagnosed genital bleeding).
Effectiveness of the different forms of contraception vary, with implants being the best at preventing pregnancy (0.05%) and female condoms being the worst (5% chance of pregnancy with perfect use).
Relative contraindications to using OCPs include: smoking.
Absolute contraindications to using OCPs include: history of deep vein thrombosis, pulmonary embolism, cerbrovascular accident, Factor V

Leiden mutation (or other thromboembolic conditions), and migraine with aura (or other neurological changes).

Note that adolescents that are in abstinence programs are just as likely to engage in sexual activity as all other adolescents.
A multitude of obstacles may prevent adolescents from effective use of contraception. These include fear of disapproval by parents and practitioners, language and cultural barriers, fear of pelvic examination, cost, side-effects (e.g. weight gain).
Know that prior to discussing birth control with a physician many adolescents or sexually active teens and may be using inadequate methods of birth control.
The most common reasons that adolescent girls and boys do not use contraceptives include lack of access to confidential services. Specific reasons female adolescents may avoid contraception include fear of undergoing pelvic exams, weight gain, loss of bone density, thromboemolism, or pelvic inflammatory disease.
Recognize that often times adolescents are more susceptible to not using contraception due to peer pressure and lack of cognitive maturity (i.e. more impulsivity).
Many adolescents have poor compliance with OCPs.

Compliance with a contraceptive method is directly related to the lack of perceived side-effects, older age of user, desire to avoid pregnancy, and comfort with the selection of the contraception method.
Pediatricians will see many adolescents who have not engaged in sexual activity and in those situations it is the responsibly of the practitioner to attempt to prevent consequences of unprotected sexual activity through education and counseling.
Consequences of sexual behavior (except pregnancy): Sexually transmitted infections: Gradnerella vaginalis is normal vaginal flora but is more common in individuals that are sexually active.
Neisseria gonorrhoeae and Chlamydia trachomatis are the two most common bacteria causing pelvic inflammatory disease (PID), however, other causes include anaerobes and gram-negative bacilli.
Clinical manifestations of PID include lower abdominal pain, abnormal uterine bleeding, and pain during or immediately after menses. Other non- specific findings include vaginal discharge, fever, urethritis, and chills. Finally, on physical examination common findings implicating PID include cervical motion tenderness along with adenexal tenderness on bimanual examination.
Lab tests for PID include a pregnancy test, microscopic examination of vaginal discharge, CBC, urinalysis, and tests for Chlamydia, gonorrhea, syphilis, hepatitis B, and HIV.
The only contraceptive method that will reduce the risk of sexually transmitted disease, including HIV/AIDS is condoms.
Trichomoniasis and genital warts are often times present in asymptomatic boys.
A cause of menometrorrhagia is gonorrhea and chlamydial infections.
Chlamydia and gonorrhea will not produce vaginitis in adolescents but will produce cervicitis.
Clinical characteristics of a patient that would implicate bacterial vaginosis include vaginal discharge typically with an unpleasant odor described as a “fishy smell.” Of note 50 to 75% of women with bacterial vaginosis will remain asymptomatic.
The current treatment for PID involves treatment for both Chlamydia and gonorrhea. Therefore patients diagnosed with PID should adequate coverage for Neisseria gonorrhea and Chlamydia, Azithromycin to treat Chlamydia and ceftriaxone for gonorrhea.
Treatment for genital warts includes three possible approaches: either chemical or physical destruction, surgical removal, or immunologic therapy. The preferred approach depends on the number and extent of lesions that are present.
Differential diagnosis of uerthritis in boys includes dysfunctional voiding, urethral stricutures.
Urethritis in adolescent boys should be treated as if both gonorrhea and Chlamydia are present due to the high rate of coinfection. Therefore, standard urethritis treatment in boys includes Azythromycin 1G x 1 dose or Ceftriaxone 125mg IM injection x 1 dose.
Oral Acyclovir is indicated in the treatment of genital herpes to either reduce the duration of outbreaks or as a daily prophylaxis in order to reduce the number of recurrent outbreaks. One should note that Acyclovir does not cure genital herpes, but will only reduce the duration or occurrence of outbreaks.
Indication for hospitalizations of an adolescent with PID include lack of reliable follow-up, pregnancy, lack of tolerance to oral medications, nonadherence to therapy, severe clinical

illness, complicated PID with abscess, or the need to rule out other etiologies of symptoms (e.g. appendicitis).
Monitor patients with PID and right upper quadrant pain for Fitz-Hugh- Curtis (perihepatitis) as there is an approximately 10% association.
PID is a risk factor for a future ectopic pregnancy and infertility.
The most common agents causing sexually transmitted infections in adolescence include: trichomonas, gonorrhea, herpes, chlamydia, syphilis, HIV, and human papillomavirus.
It is important to educate and counsel adolescents to use condoms during anal and vaginal intercourse.
Bacterial causes of vaginitis include peptostreptococcus, bacteroides, gardnerella.
There is a much higher risk of mortality associated with pregnancy and delivery than with any contraceptive method currently available.
Indications for a Papanicolaou smear in adolescence includes: Initial Pap smear should be started at age 21 and if normal should occur every 3 years until the age of 65.
All sexually active adolescents should be screened yearly for Chlamydia trachomatis and Neisseria gonorrhoeae.
Higher-risk adolescents, including those with previous history of sexually transmitted infections or multiple sexual partners, should be screen every 6 months for Chlamydia trachomatis and Neisseria gonorrhoeae.
Chronic Illness
There is an increased incidence of psychopathology (e.g. depression, anxiety, substance abuse) in adolescents with chronic illness or disability.
When medical regimens are discussed rather than dictated improved compliance can be seen in chronically ill youths.
Barriers to adherence in chronically ill patients include time, financial costs, pain, inconvenience, embarrassment, and/or the acknowledgment of personal vulnerability.
The most difficult part of achieving adherence for new treatment plans involves changing established behaviors.
Psychologic issues affecting an adolescent with a chronic illness or disability may include depression associated with loss of previously routine activities. Further, issues may include anxiety and depression associated with peer reactions or realization that some life goals may need to be altered.
Youths with chronic illness will need to have a plan in place to transition from pediatric to adult health care in order to avoid major disruptions in service.
Parents of chronically ill adolescents may need extra support and guidance in allowing children to have more autonomy in managing their own illness.
Eating disorders
An early warning sign of anorexia nervosa is amenorrhea
Anorexia nerovosa- key diagnostic features include: weight loss of more than 15% of ideal body weight, disturbed body image (fear of becoming fat or gaining weight), and amenorrhea in postmenarchal females of at least 3 cycles.
Bulimia- key diagnostic criteria include:
Recurrent episodes of binge eating occurring at least twice weekly for at least 3 months,
Recurrent, inappropriate compensatory behaviors to prevent weight gain.
Self-evaluation not accurately influenced by body shape or weight.

Differential diagnosis for anorexia nervosa and bulimia include:
Inflammatory bowel or celiac disease, Addison disease, thyroid dysfunction, hypopituitarism, diabetes mellitus, brain tumor, occult malignancy, other psychiatric disorders (e.g. schizophrenia, substance abuse, obsessive compulsive disorder).
Treatment for anorexia nervosa and bulimia both focus on a multiple disciplinary, team approach. This includes the involvement of a nutritionist, psychiatrist, as well as support from family members and pediatrician.
Inflammatory bowel disease (e.g. ulcerative colitis) and achalasia can mimic eating disorders.
Individuals that are obese or have a preoccupation with thinness may be at risk of developing anorexia nervosa or bulimia.
Hospitalization for both anorexia and bulimia include:
hypothermia (<35 degrees Celsius), bradycardia (heart rate < 40 beats per minute), weight loss less then 70% of ideal body weight, refeeding syndrome (phosphorous < 2mg/dL, marked edema), cardiac dysrhythmia (e.g. QTc > 460 msec), acute refusal of food
Factors affecting the prognosis for both anorexia and bulimia include severity of malnutrition, along with degree of disease affected change (e.g. osteoporosis). Further, patient's family and friend support during treatment along with individual characteristics of mental well being all have an impact on the success of treatment.
Behavioral health issues
Factors that predispose adolescents towards delinquent behavior includes:
Parental psychiatric illness, ADHD, severe behavioral problems before 5 years of age (e.g. fire setting, cruelty to animals), severe head trauma.
Health problems common among delinquent youth:
Injury, sexually transmitted infection, dental problems, inability to read, cigarette, alcohol, and drug use and abuse.
Parental involvement with school and extracurricular activities along with knowledge about a child's friends are protective factors for delinquency.
The association between parental involvement in children's health care and adherence to treatment.
Features that tend to worsen a patient's adherence to treatment include:
Lack of symptoms (e.g. hypertension), more than one treatment, side effects, multiple daily doses, lack of perceived seriousness of illness, inability to keep a medication calendar and dosing in synchrony with daily activities (e.g. brushing teeth) may enhance adherence.
The five leading causes of death among teenagers are:
Accidents (unintentional injury) accounts for approximately 50% of deaths, homicide,suicide, cancer, heart disease.
The options for postcoital contraception for female rape victims should be available to be used when unprotected sexual assault has occurred: levonorgesterol, estrogen plus progesterone, mifepristone, copper intrauterine device, Ulipristal.
Sexual assault is the most frequent type of trauma associated with post-traumatic stress disorder (PTSD.) Signs of PTSD in rape victims includes:
Reexperiencing the event (e.g. nightmares), avoidance of certain behaviors or activities, inappropriate arousal (e.g. easily startled), social or occupational impairment. Nonviolent

strategies for conflict resolution with adolescents includes: negotiation, mediation, empathy for the other person, listening to others before acting, and forgiveness or reconciliation.
Firearms are a leading cause of death among adolescents.
Teen's involvement in school violence may include:
Perpetrator, victim, encouraging bystander, nonviolent conflict resolver.
Generalized somatic complaints may be a sign of stress in adolescents.
Emanicipation of a minor is a legal mechanism by which a minor is freed from control by his or her parents or guardians. The minor takes responsibility for decisions on parenthood, independent living and financial decisions.
Certain situations that require parents be advised of a child's intent include: suicidal or homicidal ideation.
Special considerations for intellectually challenged patients: State and federal statutes that govern the care of intellectually challenged persons.
At an adolescent visit it is important to not only do a thorough physical examination, but to also do a functional assessment of the patient which should include discussions regarding peer and family relationships, school, and work.
There are a number of strategies that a pediatrician may use to facilitate the parent- child exchange of information about sexuality. Some strategies that may be used include educational videos and open discussions about sexual maturation. The pediatrician should facilitate and encourage the conversation between patient and parent. Regardless of the strategy employed it is important for parents to have a conversation regarding sex with their children.
Anticipatory guidance with regards to preventing adolescent "accidents" should include: drinking and driving, use of seat belts and bicycle helmets, and firearm safety.
Teenagers are more likely to divulge confidential information with regards to sexual activity or illegal substance use when interviewed without their parents. This information is important to gather in order to complete a comprehensive evaluation, along with dictating treatment options , treatment for sexually transmitted disease and substance abuse counseling.

Sports Medicine and Physical Fitness

Injury prevention
Trauma sustained by juvenile athletes is frequently due to incomplete healing of a previous injury.
Factors that influence participation in contact sports by healthy children and adolescents:

Sports participation can play an important role in the social development of children and adolescents by teaching leadership and team-building skills and encouraging physical fitness. Most children can be cleared easily for such participation, but not all sports are safe for children and adolescents, and some medical conditions warrant special consideration with regard to participation in some sports. In all cases, the clinician should perform a complete preparticipation sports physical examination and discuss the implications of the sports the patient plans to pursue.

Sports can be classified by contact level. Children who have splenomegaly, acute hepatomegaly, and contagious skin lesions should avoid contact sports but can participate in noncontact sports.

Children who have a single kidney should be advised to avoid high-contact sports, but those who have other single organs, such as ovaries and testes, can be cleared because the risk of injury is low or protective gear can be worn. Protective eyewear can reduce the risk of injury in the case of a single functional eye. It is prudent for the clinician to advise such patients of the risk to the remaining organ during sports participation, and some advocate having the parents and the athlete sign a document that indicates their understanding and acceptance of such risk.

Sports also may be classified by intensity, dynamic and static demand. This classification is especially important for children and adolescents who have cardiovascular diseases. For example, children who have ventricular dysfunction should avoid high dynamic sports; those who have significant essential hypertension, left heart obstructive disease, or an increased risk of aortic dissection should avoid high static sports. Accordingly, a girl who has Marfan syndrome should avoid gymnastics, a sport that has potentially high static intensity. Several cardiac diseases, such as acute pericarditis or myocarditis, cardiomyopathy, severe pulmonary hypertension, and right- to-left shunting, are absolute contraindications to participation in sports. A full listing of these conditions is outlined in the 36th Bethesda Conference guidelines.

Patients who have well-controlled epilepsy can be cleared to participate in most sports, but those whose seizures are poorly controlled should be advised to avoid sports in which they could sustain significant injury should a seizure occur during participation, such as swimming, diving, and riflery. Patients who have diabetes should be

encouraged to participate in sports, but they should pay attention to hydration and insulin therapy, especially with sports that last longer than 30 minutes.

> The risks in a conditioning program for junior high school athletes at the beginning of the sports season are:

Preventive conditioning is an important part of sports programs for preadolescents and adolescents because participation can help to improve strength and overall performance. Programs should include both aerobic conditioning and strength training. The most significant risks to the athlete involve strength, or resistance, training, but such risks can be minimized by adhering to a few guidelines.

Weight training programs that consist of high-repetition activities with low resistance are safe for both preadolescents and adolescents. Training should begin by teaching proper technique with no weight, adding small increments of weight as strength increases. Sessions lasting 20 to 30 minutes undertaken 2 to 3 days a week are sufficient to improve strength. Power lifting programs, which involve low-repetition, high- resistance exercises, should not be undertaken by preadolescents because of the risk of injury.

In general, such programs should be avoided until skeletal maturity is reached. Injuries reported in studies evaluating strength training programs include muscle strain, epiphyseal injuries in the wrist, and apophyseal injuries, but most of these can be avoided by using proper technique. Participation in these programs does not decrease linear growth.

Although preventive conditioning has been shown to improve strength and performance, there is no evidence that participation in such programs can prevent catastrophic injury. However, athletes who have particular recurrent injuries, such as to the shoulder or ankle, can reduce their risk of further injury by participating in specific strengthening exercises focusing on those areas.

Evaluation for sports participation

Guidelines for children with Down syndrome concerning participation in sports: Routine screening for atlanto-axial instability (AAI) in Down syndrome by lateral cervical radiograph in neutral/flexed/extended position between 3 to 5 years of age. Pre-participation cardiac evaluation by cardiologist if patient has congenital heart disease.
Febrile illness can results in decrease heat tolerance, increase heat illness and dehydration. In addition, fever may be heralding sign of an underlying condition that may put athlete at additional risk during exercise, such as myocarditis, pulmonary infection or infectious mononucleosis with splenomegaly. Athlete should be advised to avoid sport until febrile illness has resolved.
Conditions that should be evaluated by a cardiologist prior to sports participation:

Red Flags in the History or Physical Examination:

Syncope or near-syncope on exertion
Chest pain or discomfort on exertion
Palpitations at rest
Excessive shortness of breath or fatigue with activities
Family history of Marfan syndrome, cardiomyopathy, long QT syndrome, or clinically significant arrhythmias
Family history of premature, sudden death
Irregular heart rhythm
Weak or delayed femoral pulses
Fixed, split second heart sound
Any systolic murmur graded 3/6 or greater
Any diastolic murmur
Stigmata of Marfan syndrome
Chest pain in Turner syndrome

Cardiovascular conditions for disqualification from sports: Absolute Contraindications to Sports Participation
Pulmonary vascular disease with cyanosis and large right-to-left shunt
Severe pulmonary hypertension
Severe aortic stenosis or regurgitation
Severe mitral stenosis or regurgitation
Cardiomyopathies
Vascular form of Ehlers-Danlos syndrome
Coronary anomalies of abnormal sinus origin
Catecholaminergic polymorphic ventricular tachycardia(CPVT)
Acute phase of pericarditis
Acute phase of myocarditis (at least 6 months)
Acute phase of Kawasaki disease (at least 8 weeks)

Metabolic control, self-esteem, and body image may be better in the physically fit child. Therefore, physical fitness and regular exercise should be encouraged in all children who have type 1 diabetes.

Exercise in a child who has diabetes does require specific attention, however. During periods of exercise, extra calories or lower insulin doses may be needed to prevent hypoglycemia.

Some patients have a delayed hypoglycemic response to exercise, sometimes hours later. Blood glucose monitoring to assess the effects of exercise and the response to interventions allows determination of an effective regimen for the individual child. The stress of exercise when metabolic control is poor (eg, during hyperglycemia, especially with ketosis) may worsen metabolic control, so it may be necessary to delay exercise at these times.

> It is important to assess and document neurocognitive function in a preparticipation sports examination for comparative purposes in sideline evaluation of an athlete with a head injury. The exam tests the athlete's decision making abilities, as well as his/her reaction time, memory and attention span.

Although sports participation can have many physical and social benefits for children and adolescents, there are risks, including athletic concussion, which is described as mild traumatic brain injury that is caused by an impact to the head. Numerous guidelines and literature reviews have examined athletic concussion, but few have focused specifically on the pediatric athlete. Medical examination frequently is mandated by schools and organizations before children can participate in sports, and preparticipation assessment of an athlete is an opportune time to focus on how to prevent head trauma as well recognize concussions. Loss of consciousness is not the only indication that an athlete has suffered a concussion; in fact, most sports participants who sustain concussions do not lose consciousness. Documenting thehistory of the number of prior concussions, timing and severity of each, and the description of resulting symptoms can help establish a baseline for medical care.

Although conventional neuropsychological assessment is a well-established and recommended methodology for the evaluation of children and adults who have sustained neurologic injury or who have neurodevelopmental problems, the use of comprehensive neuropsychological evaluations at a preparticipation visit is impractical and expensive because of the extensive time needed to complete an evaluation.

Abbreviated "baseline" neuropsychological testing for athletic purposes has been introduced, but sufficient data regarding the reliability, validity, and clinical utility of neuropsychological instruments used in this model are lacking for pediatric athletes. Similarly, the value of such baseline testing in making individual decisions about return to play after a head injury has not been demonstrated. However, neuropsychological testing still can play a valuable role in evaluating and managing certain athletes after injury, especially those who experience poor recovery and multiple injuries, and in developing appropriate posttraumatic educational management plans if cognitive difficulties exist.

Experts agree that the earliest an athlete should return to play is when no signs or symptoms of any type are apparent at rest or exertion; neurologic examination results are normal; and neuroimaging, when performed, provides unremarkable results. Some advocate for even more conservative standards with pediatric athletes, including always removing them from the day's competition after a concussion and not allowing return to play unless cleared medically. Seasonal and lifetime sports exclusion guidelines are not

well-established or evidence-based for pediatric athletes, but a conservative approach is inarguably appropriate.

If a physician disqualifies an athlete from one sport, he or she should attempt to direct them to another sport.

Sports injury and treatment

General
Bruises, hematomas
Considerable amount of blood can be lost with a deep hematoma of the thigh.
Bruises and hematomas are treated with ice packs, compression.

Hematomas and bruises cause pain, swelling and tenderness over the area of skin discoloration or deep hematoma may last weeks to months before it heals, it will change color and slowly shrink in size. Treating a superficial hematoma is similar to the treatment used for other soft tissue injuries. Using the R.I.C.E method is recommended (Rest, Ice, Compression and Elevation). Apply ice to the area for 15 minutes, several times per day. Mild hematomas and contusions typically heal within about five days. A more serious hematoma is an epidural hematoma, in which a blood vessel in the brain ruptures and a blood clot forms between the skull and the brain's protective covering (the dura). This clot grows slowly and puts pressure on the brain that, if not treated promptly, can result in death. If you suspect a head injury but don't see any initial signs or symptoms, you should follow the head injury first aid treatment steps.

Heat illness

Infants and children < 4 years of age are more susceptible because they adjust to heat more slowly

Symptoms of heat illness / Heat Exhaustion:
Mild dehydration
Core temperature 100.4° to 104°F (38° to 40°C)*
Profuse sweating
Thirst, nausea, vomiting, confusion, headache
Feels faint or has collapsed
Symptoms of Heat Stroke:
Usually severe dehydration
Core temperature may be >104°F (40°C)*
Flushed with hot, dry skin
Dizziness, vertigo, syncope, confusion, delirium
May be unconscious
Shock
The complications of heat stroke

Patients who present with the signs of heat stroke have been injured by core temperatures greater than 104°F to 105.8°F (40°C to 41°C). The severity of injury is cumulative, so exposure to a very high temperature (109.4°F [43°C]) for a relatively brief time may produce an injury that is similar to one produced by exposure to a lower temperature (106.2°F [41.2°C]) for a longer period of time. The onset of severe neurologic dysfunction (delirium, coma, seizures) often is identified as one of the features that distinguishes heat stroke from heat exhaustion. In general, severe neurologic

dysfunction is not observed until the rectal temperature is greater than 105.8°F (41°C). Confusion or delirium is the first sign of a deteriorating neurologic status. Coma and seizures can follow rapidly and often are associated with a poor outcome.

Shock

Hypotension is a common accompaniment of heat stroke and is frequent when the temperature exceeds107.6°F (42°C). In the early stages of heat stroke, vasodilation leads to a low blood pressure, even though the cardiac index is increased and the central venous pressure is normal. More severe heat stroke produces irreversible myocardial impairment. Electrocardiographic findings that are indistinguishable from coronary ischemia may be observed. In the later stages of heat stroke, patients often become hypovolemic from sweat losses. Hypotension in heat stroke frequently is associated with diminished cerebral perfusion, which causes cerebral ischemia. These effects compound the injurious effects of heat on the CNS and may produce severe and potentially permanent neurologic dysfunction. Serum lactate concentrations frequently are elevated in heat stroke. Even after the blood pressure has returned to normal and vasopressors no longer are necessary, blood lactate values may remain elevated and only gradually return to normal. This delay is not unique to heat stroke; it is seen following circulatory shock and in patients who have hepatic dysfunction, which commonly accompanies heat stroke. In addition, vasomotor tone may remain abnormally low, even after normal temperature and intravascular volume have been restored. This outcome may be caused by the direct effects of heat on the heart and vascular system, or it may be mediated by the effects of endotoxin released from the heat-injured gastrointestinal (GI) tract or other inflammatory mediators.

GI Abnormalities and Hepatic Injury

Heat stroke causes a number of GI abnormalities. Although mild heat stroke may cause little more than diarrhea, more severe heat stroke produces significant injury to the GI tract, including mucosal swelling, petechiae, and hemorrhages. Even after resuscitation and return to normal temperature, GI

tract injury can continue to evolve. As a consequence of these injuries, circulating endotoxin rises and potentially toxic free radicals may be generated. Injury to the GI tract probably contributes significantly to the hypotension and multisystem failure observed in heat stroke. The liver may be injured severely in heat stroke. As a metabolically active solid tissue, the liver normally is a major site of heat production in the body. During periods of hyperthermia, the temperature of the liver is among the highest of any site in the body, placing hepatic tissue at high risk of injury. In addition, portal circulation perfuses the liver with a variety of toxic substances generated by the GI tract during heat stroke, including endotoxin and free radicals. Patients who survive heat stroke demonstrate rapidly rising alanine aminotransferase and aspartate aminotransferase concentrations that peak at 48 to 72 hours after injury and gradually return to normal after 10 to 14 days. Bilirubin can be elevated, but severe hyperbilirubinemia is unusual. Prolongation of the prothrombin time is common.

Biopsy of the liver after heat stroke shows few changes in mild heat stroke, although apoptosis can be an early finding. After severe heat stroke, the liver shows widespread abnormalities, including areas of cholestasis and necrosis.

Renal Failure

Renal insufficiency is a common finding in heat stroke and can affecti at least 50% of the patients who have nonexertional heat stroke and an even higher proportion of patients who suffer from exertional heat stroke. The usual clinical findings are those of prerenal azotemia, with elevation of blood urea nitrogen to a greater extent than creatinine. These abnormalities respond well to rehydration and usually correct within the first few days. Dialysis or other forms of renal support are required infrequently in patients who have no pre- existing renal disease.

Hematologic Abnormalities

Abnormalities in hematologic and coagulation values commonly follow heat stroke. The hematocrit declines rapidly in the first 24 hours following heat stroke. Although a portion of the decrease can be explained by rehydration, the reduction in hematocrit often is much greater than would be caused by rehydration. The cause of this anemia may be multifactorial. Red blood cell half-life is shortened after heat stroke. Further, red blood cells that have been heated in vitro have greater membrane rigidity and increased osmotic fragility, which may contribute to red blood cell damage and ultimately lead to early removal from the circulation. Individuals suffering from heat stroke tend to have a much greater number of spherocytes, which may have a shortened life span. Thrombocytopenia is common, although the platelet count may remain normal in mild heat stroke. The cause of thrombocytopenia has not been defined. The platelet count often is normal at the time of presentation, but declines steadily for the first 24 hours. In a recent series, 45% of patients had evidence of disseminated intravascular coagulation. Coagulation abnormalities include prolongation of the prothrombin time and partial thromboplastin time and elevation of the d-dimer. Abnormalities of plasminogen activator inhibitor 1 also have been described. Although bleeding diatheses are reported most commonly, some patients may experience a period of hypercoagulability shortly after the onset of heat stroke. Within 24 hours, these findings are replaced by a picture of disseminated intravascular coagulation. Such abnormalities generally resolve within a few days. In spite of the coagulation abnormalities, clinically significant hemorrhage is uncommon.

Hypersegmented neutrophils frequently are observed in the peripheral circulation for the first few hours but are cleared rapidly from the blood. These neutrophils, which have five or more nuclear lobes (Fig. 1), are termed “botryoid” nuclei because the morphology resembles a cluster of grapes. Although the cause of this abnormality is not known, probably these cells are undergoing the morphologic changes of apoptosis.

Postmortem Abnormalities

Death from heat stroke does not produce any distinguishing postmortem abnormalities. Nonspecific findings include hepatocellular necrosis, disseminated intravascularcoagulation, and in children, intrathoracic petechiae. Such histologic abnormalities are not immediately apparent at the onset of heat stroke; the patient must survive for at least 6 hours for the abnormalities to evolve. If the patient dies rapidly from heat stroke, the histologic abnormalities associated with heat stroke are not observed. In the absence of external evidence that overheating may have been the cause of death, the observed postmortem abnormalities are sufficiently ambiguous that heat stroke may not be considered in the differential diagnosis. For this reason, it is important to search for environmental clues that heat may have been the cause of an otherwise unexplained death.

It is important to be attentive to adequacy of hydration and maintenance of proper electrolyte balance. If sweat losses are not replaced, patients may become hypovolemic, which can lead to decreased sweating and, in extreme cases, hypotension. Significant amounts of sodium are lost in sweat, so electrolyte replacement is essential to avoid hyponatremia. Individuals who will be exercising in a hot environment should be encouraged to consume liquids containing electrolyte solutions (ie, "sports drinks") to avoid the risk of hyponatremia. Consumption of large quantities of hypotonic fluids should not be encouraged because of the risk of electrolyte imbalance.It is important for supervisory officials to encourage appropriate intake of fluids during activities. Research has shown that if fluid intake is left up to personal preference, most individuals do not drink sufficient quantities to maintain proper hydration. Although figures do not exist for pediatric patients, current recommendations for adults are to consume 500 mL of fluid within 2 hours prior to exercise (assuming the patient is euvolemic). During exercise, approximately 250 mL

of fluid every 20 minutes is recommended to offset sweat losses. Studies have shown that acclimated individuals are more likely to consume liquids to maintain hydration than are those who are not acclimated. Therefore, special attention should be paid to individuals who are not yet acclimated to hot weather.

Although it is important to emphasize adequate hydration, hydration alone does not prevent heat illness. It is possible for well-hydrated individuals to suffer heat illness or heat stroke if they continue to exercise at a rate that generates heat more rapidly than it can be transferred to the environment.

Evaluation and Management of an athlete with heat illness:

Treatment of Heat Exhaustion:

To prevent injury from heat-related illnesses, it is important to maintain a high index of suspicion. Heat exhaustion should be suspected if an individual is exercising in a warm environment and feels faint or nauseated or is confused or vomiting. Effective treatment requires immediate removal from the heat source, cessation of exercise, and hydration. If the patient has significant CNS symptoms (ataxia, confusion, seizures, coma) or symptoms of heat illness do not resolve within 20 to 30 minutes, the diagnosis of heat stroke should be considered strongly and the patient treated accordingly.

6. Treatment of Heat Stroke

Cool the patient until core temperature is <102.2°F (39°C)
Rehydrate with intravenous fluids
Administer vasopressor therapy if shock persists after rehydration
Monitor for anemia and thrombocytopenia
Treat coagulopathy if clinical hemorrhage is present
Treat seizures
Monitor for cerebral edema
Institute mechanical ventilation for respiratory failure
Liver transplantation not indicated for severe liver failure
Children have less tolerance to exercise than adults when air temperature is greater than body temperature Exertional heat stroke is reported more commonly among adolescents and adults than among young children. The reasons for this discrepancy are unknown, but it

may be that the discomfort that precedes heat illness usually causes younger children to decrease their activity level before the onset of injury. Unfortunately, adolescents and adults may be sufficiently motivated to ignore the discomfort until they collapse from heat stroke. The mechanisms of heat-related illness:

Exertional heat stroke is caused by increased heat production over a period of sufficient duration to raise core temperature to injurious levels. Vigorous

exercise is not a problem if the duration of the exercise is brief or if the environment is cool. The combination of prolonged exertion in a warm, humid environment, however, is dangerous. Dehydration can contribute significantly to the risk of heat stroke while exercising. Nonexertional heat stroke occurs in the absence of excessive physical activity among individuals who are exposed to excessively warm, humid environments and are unable to disperse heat produced by basal metabolic processes. Sleeping infants may be at risk if they are covered with excessive bedding. Because of their limited motor skills, infants may be unable to remove the blankets in response to overheating; they depend on their caretakers to provide a safe thermal environment. Both infants and young children are at risk for nonexertional heat during the summer if they are left unattended in an automobile exposed directly to the sun or in hot environments. Measurements of automobile temperatures in the summer show that the temperature can reach 145°F (62.8°C) in as few as 40 minutes, even in a light-colored vehicle that has the windows partly open. Heat stroke occurs rapidly in such an inhospitable environment. Disabled children and adolescents as well as the elderly may have limited mobility and be unable to leave their dwellings during hot weather. Patients who present with the signs of heat stroke have been injured by core temperatures greater than 104°F to 105.8°F (40°C to 41°C). The severity of injury is cumulative, so exposure to a very high temperature (109.4°F [43°C]) for a relatively brief time may produce an injury that is similar to one produced by exposure to a lower temperature (106.2°F [41.2°C]) for a longer period of time.

Return to play criteria

Criteria for return to play in sports after a head injury:

Current concepts of managing concussions related to sports dictate that an athlete manifesting any signs or symptoms of concussion be removed from the practice or game immediately, not be allowed to return to play that day, be monitored carefully, and receive medical attention. These same principles should be relevant to nonathletic activity. Return to play is allowed only when the player's signs and symptoms have resolved and when he or she has demonstrated the ability to progress stepwise through several levels of activity without any recurring symptoms. Activity should move through the following steps, with advancement only if there are no symptoms. If symptoms recur, the athlete should rest for 24 to 48 hours and try to progress again, dropping back if symptoms recur.

The recommended steps for a logical and safe progression are:
Complete rest, although bed rest is not indicated; the patient may participate in activities of daily living.
Light (low-intensity) aerobic exercise, such as walking, without a component of resistance. Weightlifting is prohibited.
Activity specific to the sport, such as running or skating. At this

step and the next, resistance training may be added.

Training drills without contact, followed by mental status testing.
Full-contact training after clearance by medical personnel.
Participation in a game.

Such a regimen must be individualized to each athlete, and progress should be monitored by clinicians who have the appropriate level of training. The primary care physician and the experienced athletic trainer make a good team in determining the appropriate course of action in most cases, and the trainer can serve as a liaison with the coach to make sure the plan is executed properly.

The criteria for return to play following an eye injury includes ophthalmologic evaluation
The criteria for return to play following various injuries: shoulder, knee, ankle, stress fracture, shin splints: athlete should be free of pain and swelling during and after exercise; have full range of motion. Flexibility and stability of the injured extremity; and have return of 95% of normal strength. In addition, they highly recommend stepwise return to competition. Finally, it is critical that the athlete regain the ability to perform sport-specific maneuvers.
The criteria for return to play after a neck injury includes an appropriate medical evaluation.
The appropriate management of a player with an acute neck injury are:

Appropriate on-field management of a suspected cervical spine injury follows a similar protocol to basic life support, which begins with a focus on airway management, breathing, and circulation (ABCs). In the United States, American football poses the greatest risk for cervical spine injury and accounts for approximately 50% of such injuries in youth sports. Cervical spine injuries also can occur during water sports, particularly diving, when an athlete hits the water at a high velocity with the neck flexed, similar to the spear tackle in football. When approaching the athlete who is suspected of having a cervical spine injury, an initial desire to assess and treat the neck directly is common. Appropriate management of the injured cervical spine, however, begins with assessment of the ABCs while attempting to keep the head and neck in a stable position. If the injury occurs in football, an appropriate initial step includes listening for breathing or putting a hand beneath the jersey and shoulder pads to check for thoracic expansion. If the athlete is unconscious, it is imperative to

establish the airway first, which includes assuring that the tongue is not obstructing the airway. The football helmet never is removed on the field. If the airway is obstructed, the face mask should be removed, but the helmet and shoulder pads should remain in place to ensure alignment of the cervical spine.

If the athlete is found in the prone position, establishing airway control can be difficult. Treating the prone athlete involves initial assessment of the airway. If the athlete is unconscious and not breathing, the airway is established in the supine position. In this scenario, the log roll technique is used to move the patient to the supine position. This technique, which should be practiced before any injury occurs, involves rolling the injured player by two assistants controlling the body and the team leader controlling the head. It is essential to keep the head turning at the same speed as the body to lessen any chance of additional injury to a potentially unstable cervical spine.

Five radiographic views should be obtained in all patients in whom cervical spine injury is suspected. Flexion and extension views never should be obtained when cervical instability is suspected.

Overuse syndromes

The management of an athlete with an overuse injury:

Overuse injuries, by definition, result from repetitive strain at cartilaginous junctions. The recommended therapeutic approach is rest, ice, and anti- inflammatory medications. Exercise that does not aggravate the injury but preserves conditioning may be continued during recovery, especially for the competitive athlete. This exercise program can be prescribed for the athlete by the coach and trainer, often in consultation with a sports medicine specialist.

The most common type of overuse injuries in athletes:

Common overuse injuries are Osgood-Schlatter disease of the tibial tuberosity, often seen in athletes who engage in repetitive running and jumping motions, and Sever disease of the calcaneus.

Head

The sport most commonly associated with a head injury are: Closed-head injury is a common medical problem in young athletes who play in high and medium-contact sports

Common Sports Characterized by Level of Contact

High-contact Sports

- Basketball
- Football
- Soccer
- Martial Arts
- Rugby

Medium-contact Sports

- Baseball
- Fencing
- Cheerleading
- Skiing
- Volleyball

Noncontact Sports

- Running
- Swimming
- Tennis
- Weight Training

Eye and ear

A.The indications for the use of goggles for eye protection in sports activities are:

Any sports in which balls, racquets or flying objects are present pose a potential for eye injury. Sports such as racquetball, tennis and badminton may seem relatively harmless, but they involve objects moving at 60 miles per hour or faster. During a typical game, a racquetball can travel between 60 and 200 miles per hour. Another potential danger is that the racquets themselves move at high speed in a confined space and could strike a player. Flying objects aren't the only hazard. Many eye injuries come from pokes and jabs by fingers and elbows, particularly in games where players are in close contact with each other. Basketball, for example, has an extremely high rate of eye injury. So does swimming, where no flying objects are involved. These are great reasons to wear protective eyewear. Enhancing performance is another important aspect of eye protection. It used to be common for people with mild to moderate prescriptions to simply participate in sports without wearing their eyeglasses or contacts.

The acute management of an eye injury are:

If a globe rupture is suspected, the examination should stop and a simple protective eye shield should be placed. Immediate referral to an ophthalmologist, potentially in a tertiary care center, is critical because surgical closure often is required. It is important to avoid any manipulation (such as patching) that could increase intraocular pressure and possibly cause extrusion of intraocular contents. Broad-spectrum parenteral or oral antibiotics should be administered early in the course to reduce the risk of endophthalmitis, which

can have devastating effects.

Simple corneal abrasions (10 mm) can be treated with topical antibiotics. Occasionally, an ophthalmoplegic agent may be used to relieve pain associated with ciliary spasm. A suspected corneal abrasion in a contact lens wearer warrants immediate referral to ophthalmology to rule out a corneal ulcer.

Retained foreign bodies in the scleral or palpebral conjunctiva often can be removed in the primary care setting. After instilling the topical anesthetic, the object may be brushed away by using a cotton-tipped swab. This removal should be followed by copious irrigation and treatment of any underlying corneal abrasion.

Lessons for the Clinician

Corneal abrasions are common, but when the history or examination results are not consistent with the expected course, globe rupture should be considered.
Visual acuity testing is an important part of the physical examination that must not be overlooked when patients have perceived visual impairment or other eye complaints.
Early closure and prevention of endophthalmitis are paramount in preserving vision.
Hyphema is a common presentation to a children’s hospital that has potentially permanent sequelae of blindness or amblyopia. Typically, hyphema is associated with trauma and often includes periocular signs of trauma, such as bruising, skin abrasion, subconjunctival hemorrhage, corneal abrasion, or even an orbital fracture. Appropriate evaluation includes a complete blood count, ultrasonography of the eye (B scan), and coagulation studies if a bleeding diathesis is suspected. If there is suspicion of nonaccidental trauma, a child abuse team should be notified and the appropriate evaluation initiated, including a skeletal survey and head CT scan. A dilated fundus examination should be performed by an ophthalmologist and remains an important part of the evaluation for nonaccidental trauma.

If the patient is African American, a thorough history regarding sickle cell anemia should be obtained, and hemoglobin electrophoresis is recommended if sickle cell disease is considered. Both sickle cell anemia and sickle cell trait can have devastating consequences on an eye that suffers a hyphema. The presence of either condition dramatically changes the treatment plan from noninvasive to surgical on an emergent basis. Patients who have sickle cell anemia have a significantly worse visual prognosis from a hyphema, and the use of acetazolamide is contraindicated.

Mouth

The indications for the use of mouth guards in athletics are:

Mouth guard use in high-contact sports, in addition to reducing oral and dental injuries, is theorized by some to reduce the risk of concussion and should be strongly encouraged in all athletes. These high contact sports are: Basketball, Football, Soccer, Martial Arts, Rugby.

Neck

Water sports are an important cause of cervical injuries.
Improper handling of a neck injury will increase the neurologic deficiency.

Shoulder

The appropriate initial management of a shoulder dislocation are:

Shoulder Dislocation Treatment Self-Care at Home

If a sling is not available, rig one by tying a long piece of cloth in a circle (a bed sheet or towel may do nicely).

A pillow placed between the arm and body may also help support the injured shoulder.

Medical Treatment

Treatment may include medications to lessen pain. After a dislocation is confirmed by radiograph, many people require medicine to lessen pain and help relax the surrounding muscles during the reduction procedure (relocating the joint to its healthy alignment). The patient may require a mild sedative as well to allow the body to relax. Most people can have their dislocated shoulder relocated in the emergency department, but a few difficult cases require a general anaesthetic in an operating room.
Many successful techniques for relocating a dislocated shoulder are available. A doctor may need to try more than one technique on the patient before a method is found that suits the particular dislocation. The technique may also vary if the dislocation is not the usual inferior type.
After the patient's shoulder is back in place, they will be sent home in a sling or shoulder immobilizer (a sling-like device that attaches to the body to lessen movement at the shoulder joint). A prescription pain reliever is often needed to control pain.

Follow-up

The patient shouldsee an orthopedic doctor for a follow-up examination within a few days. Pain relieving medications may be modified and the joint examined to see that relocation has been maintained. The doctor may

reexamine for injury to structures damaged by the original trauma.

After a period of immobilization (usually a few weeks), slowly and gradually begin to increase the range of motion at the shoulder joint. This helps to preserve natural movement and lessen the risk of recurrent dislocation. When good progress is made with range of motion, strengthening exercises may be added to help the patient return to full function.

Elbow

Posterior elbow dislocations comprise over 90% of elbow injuries. Early recognition of this injury is required due to the need for early reduction, given a higher

likelihood

for poor function and possible neurovascular compromise with delays in reduction.

In

posterior elbow dislocations, the patient often describes falling on an outstretched hand.

Workup

Before reduction of the injury, obtain anteroposterior and lateral radiographs of the elbow. If clinicians feel comfortable trying a reduction in the field, they may do so before obtaining radiographs; however, postreduction films should be obtained and the affected limb's neurovascular status should be documented pre-and

postreduction.

Postreduction films are also necessary to ensure adequate reduction and to evaluate for fractures. The recreation of a normal radiocapitellar line (the line drawn through the shaft of the radius through the center of the capitellum) should be evident on all radiographic views. Ultrasound is also used to diagnose elbow pathology by presenting real-time, high-resolution imaging of tendons, ligaments, and nerves. CT scanning may be useful in the evaluation for the full extent and location of fracturesthat may occur with a complex elbow dislocation. MRI has little use in an acute elbow dislocation

Physical Therapy

Early range of motion (ROM) exercises in stable, reduced elbow dislocations have been shown to be associated with an improved outcome. However, immobilization of the affected elbow for longer than 3 weeks in patients following an elbow dislocation has been associated with loss of ROM compared with patients who start early ROM exercises.

Medical Issues/Complications

Brachial artery disruption may be seen in any elbow dislocation and special attention should be made in open dislocations.

Ulnar nerve injury may occur in up to 15% of elbow dislocations. Perform an initial neurovascular assessment and frequent reassessments.

Median nerve injury is frequently seen in conjunction with brachial artery injuries because both of these structures are in close anatomic proximity. The median nerve function should always be assessed with a higher suspicion of injury if a brachial artery injury is known to exist. The median nerve may also be injured or entrapped during a reduction.

Associated fractures for elbow dislocation include those of the radial head or neck (5-10%), medial/lateral epicondyle avulsions (10%), those of the coronoid process (10%), and fractures of the distal radius, ulna, and proximal humerus (10%).

Compartment syndrome may develop in the forearm fascia or biceps tendon due to massive swelling, which may occur in an acute elbow dislocation. Compartment syndrome must be considered in the differential diagnosis in the presence of persistent patient pain, particularly when exacerbations of pain occur with passive finger and wrist extension of the dislocated arm.

Ectopic calcification, primarily around the collateral ligaments, is common after an elbow dislocation, provides no limitations and requires no intervention.

Myositis ossificans may also be seen if significant hemarthrosis developed with the elbow dislocation.

Surgical Intervention:seek surgical intervention by an orthopedist if any signs of neurovascular compromise, associated fractures, or nonreducible dislocations are present.

Consultations:Obtain orthopedic consultation if any signs of neurovascular compromise, associated fractures, or nonreducible dislocations are present.

Return to Play

A follow-up examination before the patient returns to play is necessary to reassess motion of the formerly dislocated elbow following the immobilization and early range of motion period. Most individuals can return to play 3 to 6 weeks following an elbow dislocation.

In sports in which an elbow dislocation occurs in a player's nondominant arm, return to play may occur at the earlier end of the rehabilitation spectrum — as long as motion is back to a level that is acceptable to the physician and suitable for the athlete. For elbow dislocations that occur in a player's dominant arm, return to play may take a longer time period. Throwing sports, such as baseball, may require the patient to undergo periods of rest up to 3 months following a dislocation, followed by a strengthening rehabilitative program once full motion is achieved.

> Evaluation of the young athlete who has elbow pain begins with a history that includes questions about hand dominance, position played, number of pitches per week, types of pitches (ie, does he or she throw breaking pitches), number of teams played on, and amount of rest between outings. The athlete may complain

of decreased throwing velocity, distance, or accuracy concurrent with the elbow pain. Physical examination may reveal swelling and tenderness over the distal upper arm, medial epicondyle, and proximal forearm. Radiographs should include anteroposterior, oblique, and lateral views, often with comparison views of the other elbow. Such films appear normal in about 85% of cases but can show hypertrophy or fragmentation of the medial epicondyle or subtle apophyseal widening.

Rarely, an avulsion fracture of the medial epicondyle or intra-articular loose body may be documented, suggesting a worse injury. Magnetic resonance imaging is reserved for severe injuries, those not responding to conservative treatment, when there is concern for intra-articular bodies, or for more skeletally mature patients. In these patients, the clinician should evaluate the patient for UCL injury, excessive physeal widening, or intra-articular bodies that may necessitate surgical intervention.

The primary treatment for medial apophysitis is rest for 4 to 6 weeks or until the patient is asymptomatic; application of ice for swelling and oral analgesics for pain are adjuvant treatments. If there is a flexion contracture, the patient may wear an elbow extension brace. During the period of rest, the athlete can continue with general conditioning, with special emphasis on core and shoulder strengthening. After the pain is relieved, the child can increase throwing activities incrementally under supervision. Most athletes can return to pitching by 12 weeks after treatment begins.

The primary approach to these injuries should be prevention. The USA Baseball Medical and Safety Advisory Committee specify throwing limits that vary according to age. In general, they recommend limits of 75 to 125 pitches/week or 50 to 75 pitches/outing, depending on age; They also recommend a season limit of 600 pitches and that athletes compete no more than 9 months per year, with 3 months of rest from overhead activity.

Although less common, other elbow injuries are possible in the young athlete. Fractures, most commonly medial epicondyle avulsion, occur, but the onset of pain tends to be much more acute, with a specific inciting event. UCL injury occurs in adult athletes but is rare in children. Avascular necrosis or osteochondrosis of the capitellum (Panner disease) is analogous to Legg-Calvé- Perthes disease. It appears as a focal lesion of capitellar subchondral bone and the overlying cartilage and generally affects children younger than 10 years of age.

Pain is located laterally. Olecranon apophyseal injury occurs with forceful triceps contractions during overhead throwing, similar to what occurs with Osgood- Schlatter disease. Affected children often have acute pain and swelling over the posterior elbow and decreased range of motion.

Wrist

Distal radial epiphyseal injury is common in young gymnasts.Scaphoid fracture is associated with a poor prognosis

Knee

Patellofemoral instability in children may be due to inherent malalignment, overuse, trauma, or a combination of these factors. Patients generally fall into one of two categories: those who experience one episode of dislocation or those who have recurrent instability. Common presenting complaints are anterior knee pain and a sense of giving way. These are ubiquitous complaints among patients seen in general practice and orthopedic clinics. History, physical examination, and radiographic imaging can help differentiate patients who have patellofemoral instability from those who have patellofemoral pain syndrome or other causes of anterior knee pain. Factors contributing to anterior knee pain include weak quadriceps or trunk muscles; tight hamstring, calf, or hip muscles; improper hip or ankle rotation; osteochondral lesions; patellar tendinosis; quadriceps tendonitis; arthritis; and other traumatic and overuse injuries.

Clinical Evaluation

The goal of clinical examination is to evaluate anatomic structures via observation and direct palpation. When findings on the history suggest acute instability, it is important to rule out associated patella fractures or tears of the anterior cruciate or medial collateral ligaments. In cases of recurrent instability, patients should be examined while bearing weight to assess alignment and rotation in the back and lower extremities. Gait analysis can reveal a tendency toward internal hip rotation, tibial torsion, or foot pronation. Relative to the opposite leg can provide a better indication of anatomic alignment. A positive patellar apprehension test is an indication of patellar malalignment or medial patellofemoral ligament disruption. While the patient lies supine, signs of apprehension or discomfort are monitored when an attempt is made to displace the patella. The degree of medial and lateral translation, normally between 25% and 50% of the width of the patella, provides an indication of stability. The Q angle is calculated by measuring the relative positions of the anterior superior iliac spine, midpoint of the patella, and the tibial tubercle. A normal Q angle is between 5 and 15 degrees for men and 10 and 20 degrees for women. A Q angle that exceeds normal values is indicative of malalignment.

Patellofemoral dysfunction

Patients who have increased hip internal rotation, lower leg external rotation, knock knee alignment, flat foot, tight hamstrings, tight heel cords, and poor quadriceps muscle tone are at increased risk for anterior knee pain. This condition is referred to as patellofemoral or anterior knee pain syndrome. Patellofemoral Syndrome (PFS) is a common cause of anterior knee pain that typically affects adolescent athletes who engage in running, jumping, and squatting sports. Females tend to have wider pelvises than do males and, therefore, have higher Q-angles that make them more susceptible to developing PFS.

Internal derangement of the knee (IDK) is the term used to cover a group of disorders involving disruption of the normal functioning of the ligaments or cartilages (menisci) of the knee joint.

The normal knee joint has two collateral ligaments, two cruciate ligaments and two semilunar cartilages (menisci). Any of these may be involved in a derangement, some being more damaged than others. In some instances more than one structure is disrupted.

Assessment of the ACL on physical examination:

Employs the anterior drawer and Lachman tests; the anterior drawer test involves grasping the proximal tibia and pulling the leg forward with the knee flexed to 90 degrees and foot stabilized. The Lachman test is performed by flexing the knee 15 to 30 degrees, followed by attempting to pull the tibia forward with one hand while holding the femur stationary (Fig. 4). Excessive anterior tibial translation (forward motion) with either test signifies an ACL injury; although the Lachman test is more sensitive, (1) both tests have decreased sensitivity immediately after an injury due to significant pain and swelling.

Physical exam findings of MCL Rupture:

Valgus stress testing in full extension and at 30 degrees of flexion is performed to test the integrity of the MCL. In full extension, the MCL serves as a secondary stabilizer, and the knee remains stable as long as the cruciate ligaments and posterior capsule are intact. In 30 degrees of flexion, cruciate

ligaments and the posterior capsule are loose and do not resist valgus forces. The MCL is the primary stabilizer to valgus force at 30 degrees of flexion, and instability is present if the MCL is damaged.

Physical exam to assess for meniscal pathology:

The McMurray test often elicits positive results in patients who have meniscal abnormalities. To test for medial meniscal pathology, the patient is positioned supine, with the ipsilateral hip flexed to 90 degrees and affected knee maximally flexed. The knee is extended gradually while applying a valgus force and external rotation of thetibia (Fig. 12). The maneuver provides an axial load androtational force to the meniscus and elicits pain.

> Treatment of PFS focuses on pain relief and improving patellar tracking. Knee bracing, patellar taping, and antiinflammatories usually provide significant relief. Extensive physical therapy consisting of iliotibial band stretching and medial quadriceps strengthening helps to improve patellar tracking. Core strengthening to improve pelvic control and minimize medial knee deviation also may be beneficial.

If 4 to 6 months of aggressive therapy and modalities fail to provide relief, patients should be referred to an orthopedic surgeon. Surgical treatment options include vastus medialis tightening, tibial tubercle realignment, and lateral retinacular release.

> The criteria for orthopedic consultation for a knee injury
> All tibial tubercle avulsion fractures should be referred to an orthopedic surgeon.
> If a patient develops significant leg or calf pain that is unresponsive to narcotic therapy, compartment syndrome should be considered. Although rare, it has been reported and warrants an emergent orthopedic consultation if suspected.
> Patients who have septic bursitis or those who fail conservative management should be referred to an orthopedic surgeon.
> Patients suspected of having an ACL tear should be placed into a knee brace or immobilizer and given crutches for protective weightbearing. An elective outpatient referral to an orthopedic surgeon should be made within 7 to 14 days.
> Failure of nonoperative treatment of MCL injuries warrants referral to an orthopedic surgeon for consideration of operative repair.
> Surgical treatment and orthopedic referral are warranted for patients who have unstable lesions and those who fail nonoperative treatment for OCD (osteochondritis dissecans) lesions.
> Another common disorder that can cause anterior knee pain is prepatellar bursitis. A bursa is a fluid-filled sac between a bony prominence and the overlying skin that reduces friction and skin irritation. The prepatellar bursa primarily reduces pressure exerted on the skin overlying the patella when a person is kneeling on hard surfaces. The bursa can become irritated and inflamed in young athletes who spend a lot of time on their knees, such as wrestlers and baseball catchers. The bursa also can fill with blood, which may result in significant swelling. Affected patients present with tenderness, redness, and pain with knee flexion. Swelling generally is located superficial to the patella. Swelling at the medial and lateral joint lines is more consistent with an effusion or swelling in the knee joint itself. Imaging studies generally are not useful in diagnosis. Symptoms usually resolve with rest, ice, and compressive dressings. Large fluid collections may be aspirated for comfort. The

aspirate should be sent for crystal analysis and Gram stain to rule out infection or gout. Patients who have septic bursitis or those who fail conservative management should be referred to an orthopedic surgeon.

Ankle

The treatment of uncomplicated ankle injuries is as follows:

If there is no bone-specific tenderness and the athlete can walk without much of a limp, the pediatrician can suggest exercises as a precursor or possible alternative to physical therapy. The premise is that the initial regimen of RICE (Rest, Ice, Compression, and Elevation) is helpful for the first 48 hours after injury. The process of ankle mobilization can proceed after that time. The initial step is to start theankle moving (tracing the alphabet with the first toe is a good start), after which progressive strengthening can proceed. The following exercises can enhance strength and can be learned easily in the pediatric office. An elastic band is used to strengthen the muscles gradually as they flex against resistance. These exercises are done in three sets of 15 repetitions daily for a 6-week period. After 6 weeks, the exercises usually can be discontinued. If the ankle injury persists for more than a few weeks and if radiographs are negative and there is no bone specific tenderness, referral to a physical therapist is indicated. The prevention of ankle injuries is especially important in any young athlete who has suffered a previous ankle injury. Multiple studies have demonstrated that the most common cause of an ankle injury is the existence of a previous ankle injury. The prevention of an ankle injury involves the implementation of programs designed to strengthen the muscles around the ankle, including the peroneus muscle groups on the lateral aspect of the lower leg and the tibialis posterior muscle on the medial aspect of the leg. The same exercises used to strengthen the injured ankle are useful in preventive strengthening.

Epiphyseal injury is possible in an apparent ankle sprain in a child whose growth plates have not closed.
If there is no bone-specific tenderness and the athlete can walk without much of a limp, the pediatrician can suggest exercises as a precursor or possible alternative to physical therapy. After 6 weeks, the exercises usually can be discontinued. If the ankle injury persists for more than a few weeks and if radiographs are negative and there is no bone-specific tenderness, referral to a physical therapist is indicated.

D.Nutritional requirements 1.Hydration and rehydration

For the juvenile athlete, fluids should not be restricted during an athletic event or practice.

Weight gain and loss

The appropriate amount of weight loss per week for athletes who participate in sports with weight categories are as follows:

Sports in which a lean body mass is desirable or competing at the lowest possible weight is seen as an advantage include distance running, cross country skiing, swimming and diving, dance, figure skating, gymnastics, and weight-class football and wrestling. Other sports, such as football, rugby, basketball, and power lifting, emphasize weight gain in the form of lean body mass. Whether attempting to lose weight and body fat or gain weight and muscle mass, some youth may resort to unhealthy dieting and exercise practices, supplement use, and drug ingestion.

Some practices, including rapid weight loss with voluntary dehydration, the use of anabolic steroids, and inappropriate use of stimulants or insulin, may be fatal. Inappropriate weight loss also may result in medical complications such as delayed physical maturation and potential growth impairment; oligomenorrhea and amenorrhea; development of eating disorders; depression; an increased incidence of infectious diseases; and renal, endocrine, gastrointestinal, and thermoregulatory disturbances. Disordered eating behaviors are prevalent in both female and male athletes. It is reported that 10% to 15% of high school boys in "weight sensitive sports" practice unhealthy weight loss behaviors, with numerous studies reporting such behaviors in wrestlers.

Voluntary dehydration frequently is used to lose weight, and techniques can include fluid restriction, spitting, use of laxatives and diuretics, wearing of rubber suits during activities to cause sweating, and excessive sweating in saunas and steam baths. Dehydration results in greater body heat storage, causing an excessive increase in the core body temperature and a decreased blood volume, and decreased exercise tolerance. It also increases the risk of heatrelated illness.

No optimal values for body composition have been established for any sport. Rather than a specific percentage of body fat for an individual athlete, a range that is realistic and appropriate should be recommended. If it is appropriate that an adolescent loses weight, healthy weight loss should not exceed 1.5% of the total body weight per week or 1 to 2 lb/wk. Weight loss beyond these guidelines results in the breakdown and metabolism of muscle, causing muscular weakness. An appropriate diet for most athletes consists of a minimum of 2,000 kcal/day. The ideal method of losing weight is to consume 1,750 fewer kcal per week and expend 1,750 kcal more per week by exercising. A well-balanced diet that contains approximately 55% to 65% of calories from carbohydrates, 15% to 20% of calories from protein, and 20% to 30% of calories from fat is recommended.

Many athletes who participate in sports with weight categories practice weight control that may be pathogenic and pathologic.

erformance-enhancing drugs

Signs and symptoms of the use of performance-enhancing drug or steroid use:
Acne
Hirsuitism
Hypertension
Liver tumors
Psychosis and aggression
Emotional lability
Increases in low-density lipoprotein (LDL)-cholesterol
Decrease in high-density lipoprotein (HDL)-cholesterol
Virilization (females)
Premature closure of epiphyseal growth plates
Dysplasia of collagen, resulting in tendon rupture
Decreased sexual function and desire
Testicular failure
Gynecomastia Growth Hormone
Behavioral changes
Coarsening of facial features
Growth of facial bones

Enlargement in thickness of fingers and toes
Increase in skull circumference
Broadening of the nose
Enlargement of the tongue
Growth of the mandible
Increased separation of the teeth
Cardiovascular disease
Diabetes
Hypertension
Peripheral neuropathy

We must be able to approach our patients with well directed questions to elicit the information that we need without being judgmental. Simply asking patients if they or their friends are using anything to improve their appearance or performance is a good starting point.

Questions to Ask Regarding Substance Use

Do you (or your friends/teammates) use anything to help you in sports or competition?
Do you take any herbs, vitamins, or other natural medicines?
Do you use anything to help improve your workouts?
Do you take any supplements to help your appearance?

Physical examination findings of acne, hirsuitism, gypertension, virilization (females), and gynecomastia may suggest use of anbolic steroids. Behavioral changes, coarsening of facial features, growth of facial bone, enlargement in thickness of fingers and toes, increase in skull circumference, broadening of the nose, enlargement of the tongue, growth of the mandible, increased separation of the teeth may suggest growth hormone

use.

The use of performance enhancing drugs can be confirmed by laboratory evaluation and checking levels of these drugs in blood and other enzyme and hormone levels affected by it.

Know the side effects of anabolic steroids and other performance- enhancing drugs (eg, creatine, androstenedione).

Side effects of anabolic steroids:

Men may develop:

Prominent breasts
Baldness
Shrunken testicles
Infertility
Impotence Women may develop:
A deeper voice
An enlarged clitoris
Increased body hair
Baldness
Infrequent or absent periods

Both men and women might experience:

Severe acne
Increased risk of tendinitis and tendon rupture
Liver abnormalities and tumors
Increased low-density lipoprotein (LDL) cholesterol (the "bad" cholesterol)
Decreased high-density lipoprotein (HDL) cholesterol (the "good" cholesterol)
High blood pressure (hypertension)
Heart and circulatory problems
Prostate gland enlargement
Aggressive behaviors, rage or violence
Psychiatric disorders, such as depression
Drug dependence
Infections or diseases such as HIV or hepatitis if drugs are injected
Inhibited growth and development, and risk of future health problems in teenagers

Side effects of androstenedione in men include:

Acne
Diminished sperm production
Shrinking of the testicles
Enlargement of the breasts In women, side effects include:
Acne
Masculinization, such as deepening of the voice and male-pattern baldness

In both men and women, androstenedione can decrease HDL cholesterol (the "good" cholesterol), which puts the user at greater risk of heart attack and stroke.

Adverse effects related to human growth hormone range in severity and may include:

Joint pain
Muscle weakness
Fluid retention
Carpal tunnel syndrome
Impaired glucose regulation
Cardiomyopathy
High cholesterol (hyperlipidemia)
Diabetes mellitus
High blood pressure (hypertension)

Diuretics taken at any dose, even medically recommended doses, predispose athletes to adverse effects such as:

Dehydration
Muscle cramps
Exhaustion
Dizziness
Fainting

Potassium deficiency
Heart arrhythmias
Drop in blood pressure
Loss of coordination and balance
Heatstroke
Death

Possible side effects of creatine that can decrease athletic performance include:

Stomach cramps
Muscle cramps
Nausea
Diarrhea
Weight gain

High-dose creatine use may potentially damage kidneys and liver.

Although stimulants can boost physical performance and promote aggressiveness on the field, they have side effects that can impair athletic performance.

Nervousness and irritability, which make it hard to concentrate on the game.
Insomnia, which can prevent an athlete from getting needed sleep.
Dehydration.
Heatstroke.
Addiction or tolerance, meaning that athletes need greater amounts to achieve the desired effect, so users take doses that are much higher than the intended medical dose.

Other side effects include:

Heart palpitations
Heart rhythm abnormalities
Weight loss
Tremors
Mild high blood pressure (hypertension)
Hallucinations
Convulsions
Stroke
Heart attack and other circulatory problems

Physical fitness

Regular exercise is important to promote good general health.
Skeletal maturity is important in dictating the appropriate type of physical training.

Substance Abuse

Epidemiology
Current data
Use or abuse of multiple drugs is more common than the use or abuse of any single drug

Alcohol and tobacco use often begins in adolescence or preadolescence. Studies show that 46% of adolescents have tried alcohol by eighth grade, and by senior year in high school 77% of adolescents have started drinking. Currently smoking rate among teenagers is 8.1% and 12.4% of teenagersbecomes lifetime users. Teenagers start smoking at age 12 years and convert to regular smokers by age 14 years. Marijuana use among high school seniors was reported to be 32.4% last year.

evelopmental patterns

The initial age for experimentation with the major drugs of use or abuse less than 18 years. Earlier exposure to drugs of use/abuse is associated with an increased probability of dependence.

Risk factors

Genetic

There is genetic predisposition to alcoholism.

Familial

Adolescent substance abuse is associated social issues in the family such as parental drug use/abuse, child abuse, family disruption, and family tolerance of alcohol use.

Peer group

An excellent predictor of drug use and abuse in an adolescent is extent of drug use among close friends.

School

Early academic failure is predisposing factor to adolescent behavioral dysfunction, including substance use/abuse.

Protective factors against substance use and abuse in youths are feelings of connectedness to school, family, and community.

Laboratory evaluation

Ethical and practical

Urine screening in caring for a patient with a known drug use problem is reasonable to collect urine to confirm a state of abstinence or to confirm a state of suspected recent use.

Doctor-patient relationship might be adversely affected from urine screening for drugs without the knowledge and/or consent of the teenager.

To prevent accidental or purposeful contamination, dilution or substitution and ensure a reliable urine sample for drug screening collection should be under observation.

Applicability and limitations

The approximate duration of positivity of urine screening varies depending on drug, dose, and frequency of use.

Urine drug screen may yield false-positive and false-negative results due to varying retention time between use and identification in collected specimen.

Role of primary care practitioner

Evaluation by interview
Recognition of drug-related dysfunction
Adolescents should be evaluated for drug use and abuse as a possible etiology when dysfunctionssuch as , delinquency, school failure, promiscuity, running away from home, family conflict, depression or suicide attempts exists.
Adolescents with substance abuse disorders often display psychiatric symptoms

Obtaining a history

It is important to know not only type of drugs which are being used/abused by youth, but also the frequency of use, circumstances, and the possible risks associated with the drugs.
It is extremely important to consider privacy and confidentiality in eliciting a drug history from an adolescent since providing .confidentially to the adolescent yields greater disclosure of sensitive information.
It is necessary to obtain information from parents about their own drug use and abuse and any concerns about their child's drug use and abuse.

Collateral contacts

Information collected from school or police authorities in evaluating drug

use and abuse is extremely valuable.

Coordinating role

Primary care physicians hold a position to coordinate and manage drug abuse treatment in youths across service systems,school, mental health facilities, drug and alcohol treatment centers.

Anticipatory guidance

Pediatricians are at a unique position to offer appropriate anticipatory guidancee to adolescent patients and their parents about the dangers of drug and alcohol use or abuse and methods to adopt protective behaviors and prevent danger related to risk behavior, avoidance of drinking and driving, surreptitious drug use, peer group behaviors, and appropriate parental monitoring).
Primary care providers need to discuss parental role in preventing substance abuse by teenagers, parents' drug andalcohol use/abuse patterns, and their perception. Parents should be reminded about their potential influence on adolescent behavior.

Office counseling

The stages of drug and alcohol use are experimentation, non-problematic use, problem use, abuse, dependence and secondary abstinence. It is essential to counsel and provide anticipatory guidance to teenagers on drug andalcohol experimentation and abuse during yearly well care visits.

Evaluating and preparing for referral

Primary care physicians should screen adolescents routinely at least on yearly basis, initiate appropriate intervention and be familiar with community resources and refer patients with a substance abuse disorder for treatment.

Opportunities for community initiatives

Primary care physicians can serve as a resource for schools and other community based drug and alcohol prevention programs.

Safe Rides are effective in preventing accidents from impaired driving and works with communities to ensure safe and sober driving. Students Against Driving Drunk (SADD) works with schools and community to provide prevention tools in order to handle issues of underage drinking, other drug use, driving with impaired or altered mental status, and other destructive decisions that teenagers might take.

Referring for treatment

Primary care practitioner is obligated to periodically reassess the progress of a patient referred for substance use/abuse treatment.

Those who use or abuse drugs and alcohol have the potential to relapse for the rest of their lives.

Specific substances

Overview

Street drugs are often adulterated and an overdose may be secondary to either a combination of drugs or a drug other than the one alleged to have been taken by the overdose victim.

Alcohol

Physiologic consequences of alcohol consumption are increased triglycerides resulting in fatty liver. Fatty liver may cause necrosis in liver and lead to alcoholic hepatitis which later turns into fibrotic liver or cirrhosis. Alcohol impairs the metabolism of many drugs, and causes retention and toxicity of drugs by utilizing the hepatic microsomal enzyme system that is shared by other drugs.

The major behavioral consequences of alcohol use orabuse are euphoria, grogginess, talkativeness, impaired short-term memory and increase in pain threshold.

The signs and symptoms of an acute alcohol overdose are disorientation, lethargy, obtundation or coma leading to respiratory depression in an adolescent. In case of large ingestion a person may have epigastric pain, anorexia, emesis, mid abdominal pain and heme positive stool.

The management of alcohol overdose in an acute care includes providing ventilator support for respiratory depression until elimination of alcohol is done by the liver. If blood alcohol level is >400 mg/dl dialysis needs to be considered.

Marijuana

The major physiologic consequences (somatic consequences) attributable to marijuana abuse involves pulmonary, cardiovascular , reproductive and immunological system.

Respiratory- initial bronchodilation followed by bronchoconstriction and neoplastic change with regular use. Cardiovascular- tachycardia and elevated blood pressure. Reproductive- decrease levels of pituitary gonadotropins, testosterone, and spermatogenesis, impaired sperm motility, irregular ovulation. Immunological- affects antitumor activities and immunosuppression. Infants born to mothers who smoked marijuana during pregnancy are short, have small heads and are also weigh less.

Major behavioral consequences of marijuana use or abuse in an acute setting are euphoria, relaxation and disinhibition. Adverse consequences are impaired problem solving skills, difficulty in conversing and organizing thoughts, interference with co-ordination, impaired judgment of time, speed and distance, slowing of reaction time. Regular use of marijuana leads to inability to learn,

short attention span, poor memory and dysphoric reactions. Marijuana has the potential for physiologic addiction and dependency. Its use or abuse often precedes use of other drugs.

Tobacco

Physiologic consequences due to smoking tobacco are: respiratory- increased chronic cough, sputum production and wheezing; gastro hepatic: interference with drug metabolism (phenacetin, theophylline, imipramine); reproductive: decrease fetal weight and increased perinatal mortality and morbidity. Neuroendocrine: increased epinephrine level leading to elevated blood pressure, respiration and heart rate.

The behavioral consequences of repetitive use of tobacco are increased tolerance, physical withdrawal symptoms, increased difficulty controlling desire and use of tobacco despite experiencing harmful effects and tendency to give higher priority to tobacco use than other activities.

Major physiologic consequences attributable to chewing tobacco are oral problems (e.g. receding gums, gingivitis) and precancerous leukoplakia along with addiction.

Using pharmacotherapy along with behavioral therapy throughformal smoking cessation programs provides higher cessation and lower relapse rates. Nicotine in the form of patch, gum, inhaler, nasal spray, lozenge and micro tab are available and should be initiated after going through the 5 A's (e.g. Ask, Advice, Assess and Arrange). Bupropion and Varenicline may be used when patients are older than 18 years.

Opiates

Major physiologic consequences (somatic consequences) attributable to the use of opiates are euphoria, decrease in pain sensation, cutaneous flushing and pin point pupils. User may have fat necrosis, lipodystrophy and atrophy of the extremities. User also has loss of libido and constipation. The method of opiate administration are injecting intravenously or subcutaneously, snorting or sniffing or smoking.

Major behavioral consequences of opiate use and abuse are apathy, sedation, impairment of judgment, disinhibition, psychomotor retardation, and short attention span. There is a known potential for physiologic addiction.

Signs and symptoms of an acute opiate overdose are drowsiness, slurred speech, decreased level of consciousness, seizure, miosis and in case of severe overdose midriasis, respiratory depression, cyanosis and pulmonary edema.

Amphetamines

The major physiologic consequences (somatic consequences) attributable to low dose amphetamine are increased physical activity, tachycardia, irregular heart rate, elevated blood pressure and decrease in appetite. High dose amphetamines users may have slow cardiac conduction, hypertension, hyperpyrexia, seizure. Long term users are at risk for psychosis, cerebrovascular damage, tooth decay with receding gums and infections like HIV, hepatitis B and C.

Methods of amphetamine administration are oral ingestion, intravenous, smoking or absorption across mucous membranes.

Major behavioral consequences of amphetamine use and abuse are euphoria, sensation of increased energy, aggression, hypervigilance, grandiose or labile mood, hallucinations with paranoia, interference with personal functioning and argumentativeness. There is a known potential for physiologic addiction

Signs and symptoms of an acute amphetamine overdose include dilated pupils, tachycardia,bradycardia, arrhythmias, hypertension, nausea, vomiting, sweating, chills, chest pain and convulsions.

Management of an acute amphetamine overdose starts with treating agitation and delusional behaviors with haloperidol or droperidol. Cooling blanket may be used for hyperthermia, lorazepam or diazepam is used for sedation and managing hypertension and arrhythmia.

Hallucinogens

The major physiologic consequences of hallucinogens are variable depending on the substance abused. LSD (Lysergic acid diethylamide) causes dizziness, dilated pupils, nausea, flushing, elevated temperature and tachycardia. MDMA (Methylenedioxymethamphetamine) related somatic symptoms are nausea, jaw clenching, teeth grinding, blurred vision and impairment of memory and ability to perform cognitive task . The somatic consequences of Phencyclidine are shallow breathing, flushing, hyper salivation, nystagmus, generalized numbness of limbs, los of motor co-ordination and ataxia.

Major behavioral consequences of hallucinogen use and abuse are euphoria, emotional lability, and altered perception, distortion of body image, panic attacks and even toxic psychosis. LSD can cause synesthesia which means seeing smells and hearing colors. LSD is not addictive like PCP and MDMA.

Signs and symptoms of acute hallucinogen intoxication vary depending on the drug used. PCP in high dose can present with hypotension, generalized seizure and cardiac arrhythmia.

General management of acute hallucinogen intoxication entails ensuring a dark quiet room safe from injury. Recent oral ingestion of PCP warrants induction of emesis or gastric lavage due to its slow absorbability in the GI tract along with providing hydration and increasing drug excretion by acidification of urine. A noncomatose patient with agitation may be given diazepam. Seizure with LSD is also treated with benzodiazepines.

Cocaine

The major physiologic consequences attributable to cocaine are due to sympathomimetic affects; dilated pupils, tachycardia, hypertension and hyperthermia. Chronic rhinorrhea,

nosebleed and loss of smell can result from snorting cocaine. Inhalation of cocaine can cause burn injury. Cocaine causes low birth weight, premature birth and developmental disorder in babies born to mothers who used cocaine during pregnancy. Cocaine may be used by snorting, inhaling and in IV form.

Behavioral consequences of cocaine use and abuse include euphoria, increased motor activity, decrease in fatigability and mental alertness. Unwanted effects from chronic use are anxiety, irritability and paranoid psychosis. There is a known potential for physiologic addiction to cocaine use.

The signs and symptoms of an acute cocaine overdose are dilated pupils, delusion, paranoia, tachycardia, hypertension, hyperthermia, diaphoresis, piloerection and hyperreflexia. Uncommon symptoms are seizure, hypotension and dysrhythmia.

Inhalants

Physiologic consequences of the use/abuse of inhalants are euphoria, drowsiness, a lingering headache, sleep, slurred speech, dizziness, diplopia, ataxic gait, and disorientation. Rhinorrhea, epistaxis, sneezing, coughing, excess salivation, and conjunctival injection are manifestations of mucous membrane irritation. Gastrointestinal symptoms are nausea, vomiting, diarrhea, and abdominal cramps. Some experience dyspnea, or wheezing. Nitrites can cause headache, syncope, lightheadedness, hypotension, cutaneous flushing, tachycardia, methhemoglobinemia and transient changes in EKG.

Variety of agents are used as inhalants, e.g., organic solvents, fuels, toluene, paint thinner, glues, spray paint, hair spray, gasoline, Freon, propane, lighter fluid, nitrites, shoe polish.

The signs and symptoms of an acute inhalant overdose are restlessness, general muscle weakness, dysarthria, nystagmus, disruptive behavior and occasional hallucinations

Death from overdose of inhalants occurs due to cardiac dysrhythmias.

The management of an acute inhalant overdose is mostly supportive and focused on controlling arrhythmia and stabilizing respiration and circulation.

Anabolic steroids

Anabolic steroids cause physiologic changes to multiple organ systems. Musculoskeletal- increase muscle mass and strength but shorter adult height, endocrine- decrease production of testosterone, decrease spermatogenesis and testicular atrophy by suppressing LH and FSH, gynecomastia from peripheral conversion of androgen into estradiol and estrone; in females steroids causes hirsutism, acne, deepening of the voice, clitoral hypertrophy, and male-pattern baldness. Gastrointestinal effects-elevated hepatic enzymes, increased cholesterol and decrease in HDL. Thrombotic consequences include strokes, myocardial infarctions, and limb loss.

Anabolic steroid users/abusers may have mental status and behavioral changes such as irritability, aggressiveness, euphoria, depression, mood swings, altered libido, and psychosis.

Over-the-counter medicines

Over-the-counter cough and cold preparations (e.g., pseudoephedrine, dextromethorphan) are abused/ used inappropriately by teenagers.

Disorders of Cognition, Language, and Learning

Developmental delay, intellectual disabilities:

Clinical features, presentation:

Language development in infancy and early childhood is a better predictor of cognitive function than motor development. Phonemic awareness is the ability to attend to phonemes (speech sounds) that are used in syllables and words and is a critical language skill in the development of reading.

Intellectual disabilities (ID) is defined as a disability characterized by significant limitations both in intellectual function and in adaptive behavior as expressed in conceptual, social, and practical adaptive skill. This disability originates before the age of 18.

The 3 most common causes of ID are Fragile X syndrome, Fetal alcohol syndrome, Down syndrome.

Children who have mild intellectual disability (ID) (IQ: 50-55 to 70) and no comorbid disorders are self-sufficient in activities of daily living (ADL) and communication. They can be expected to learn at one half to two thirds normal velocity. They can reach a third- to sixth-grade reading level by late adolescence and can be expected to be employed in competitive unskilled, semiskilled, or in some cases, skilled jobs. The prevalence rate of mild ID is 20 to 30 per 1,000, and affected children often are identified in kindergarten and early elementary school.

Individuals who have moderate ID (IQ: 35-40 to 50-55) are able to do ADL and express basic needs. They learn at one third to one half velocity. They can be expected to achieve a first- to third-grade reading level. They may be able to function in a supportive employment setting, but more often they work in sheltered workshops that provide constant supervision. In adulthood, they often live in group homes. The prevalence rate of moderate ID is 5 per 1,000, and the condition most often is identified in preschool.

Children who have severe ID (IQ: 20-25 to 35-40) have limited language skills and need support with ADL. They might be able to learn to read simple signs such as stop and exit. They will continue to need help with ADL from caregivers as adults. The prevalence rate of severe ID is 3 per 1,000, and affected children are identified before age 3 years. These individuals often have identified genetic, medical, and neurologic causes.

Children who have profound ID (IQ: <20-25) require assistance for ADL. The prevalence rate of profound ID is 1 to 2 per 1,000, and affected children are identified prior to age 2 years. They have the highest rates of identified genetic, medical, and neurologic causes.

Mild intellectual disabilities and borderline intelligence may not be recognized until the child enters school ie, in kindergarten, or first or second grade.

Females who have fragile X syndrome have highly variable presentations. Approximately 50% of those who have a full fragile X mutation are intellectually disabled, with features and behaviors similar to those of affected boys, although they typically are less severely affected than males who have equivalent mutations. The other 50% are intellectually normal. Males who have Fragile X syndrome have both motor and speech delays and display behaviors such as hand-flapping and wrist-biting. They are almost always intellectually disabled, with abilities falling within the moderate range of intellectual deficit.

Fetal Alcohol Syndrome (FAS) is the most common preventable cause of intellectual disability (ID) Children who have FAS exhibit deficits in measures of attention, learning,

executive functioning, and visuospatial processing. Young adults who have FAS and normal IQs demonstrate deficits in the areas of attention, verbal learning, and executive function that are more severe than suggested by IQ alone. Their social abilities often plateau at the 6-year-old level, and their interpersonal relationship skills also are delayed.

A helpful rule regarding the development of articulation is the Rule of Fourths. The average 2-year-old child should be understood by strangers half the time (2/4), the average 3-year-old child should be understood ¾ of the time, and the average 4-year-old should be understood all the time (4/4). Children may have difficulty with the pronunciation of certain sounds until they are 7 years of age, but 100% of speech should be understood by age 4 years. In normal speech development, the first eight consonant sounds that develop are m, b, y, n, w, d, p, and h. The last eight consonants are sh, th (as in thirty), s, z, th (as in the), zh sound typically spelled as s (as in pleasure), l, and r, which develop by age 7.

Etiologies:

A family history is essential when evaluating a child who has ID. Such a history includes a three-generation family history, with attention to other family members who have ID, developmental delays, psychiatric disorders, congenital malformations, miscarriages, stillbirths, and early childhood deaths. The family history may help guide the clinician to a diagnosis, particularly when other family members are affected.
Prenatal causes of ID are a number of chromosomal anomalies and genetic, metabolic and neurologic disorders, congenital infections, prenatal toxin and drug exposure.
Perinatal causes of intellectual disabilities are complications related to prematurity, CNS bleeding, periventricular leukomalacia, breech or high forceps delivery, multiple births, placenta previa, preeclampsia, and perinatal asphyxia.
Postnatal (acquired) causes of intellectual disabilities are undernutrition and

environmental deprivation (lack of physical, emotional, and cognitive support required for growth, development, and social adaptation) during infancy and early childhood may be the most common causes of ID worldwide. Viral and bacterial encephalitis (including AIDS-associated neuroencephalopathy) and meningitides, poisoning (eg, lead, mercury), and accidents that cause severe head injuries or asphyxia may result in ID.

Physical stigmata of the most common genetic syndromes are Fragile X syndrome:

The clinical presentation of fragile X syndrome in males varies according to age. Prepubertal boys may be large for age (in both height and weight) and have relative macrocephaly. They often exhibit hypotonia and may have gastroesophageal reflux and recurrent otitis media. They have motor and speech delays and display behaviors such as hand-flapping and wrist-biting. They are almost always intellectually disabled, with abilities falling within the moderate range of intellectual deficit. As they grow, their faces elongate, and the prominence of the forehead, ears, and jaw becomes more noticeable. After puberty, they have striking macroorchidism.

Fetal alcohol Syndrome: presents with distinctive facial features, including small eyes, an exceptionally thin upper lip, a short, upturned nose, and a smooth skin surface between the nose and upper lip (short palpebral fissure), flat philtrum, deformities of joints, limbs and fingers, slow physical growth before and after birth, vision difficulties or hearing problems, short stature, small head circumference and brain size (microcephaly), poor coordination, heart defects etc.

Trisomy 21 (Down syndrome):

This is usually characterized by hypotonia, hyperflexibility of joints, flat facial profile, slanted palpebral fissures, poor Moro reflex, excess skin on the back of the neck, abnormal ears, dysplasia of the midphalanx of the fifth finger, and single palmar crease.

Williams syndrome:

This is characterized by distinctive facial features: short upturned nose, flat nasal bridge, long philtrum, flat malar area, wide mouth, full lips, dental malocclusion and widely spaced teeth, micrognathia, and periorbital fullness; developmental delay coupled with strong language skills, and cardiovascular problems, such as supravalvular aortic stenosis and transient hypercalcemia.

Common metabolic causes of intellectual disabilities are inborn errors of metabolism such as phenylketonuria, homocystinemia, glycogen storage

disorders, and disorders of amino acid metabolism.

Common chromosomal causes of intellectual disabilities are Cri du chat syndrome, Fragile X syndrome , Klinefelter's syndrome, Mosaicisms, Trisomy 13 (Patau's syndrome), Trisomy 18 (Edwards' syndrome), Turner syndrome and Down syndrome.

Common inheritance patterns of intellectual disabilities are X linked recessive, autosomal recessive.Common infectious causes of intellectual disabilities are congenital infections caused by rubella, cytomegalovirus, Toxoplasma gondii, Treponema pallidum, or HIV.

Common teratogenic causes of intellectual disabilities are anticonvulsants such as phenytoin, and valproate; chemotherapy drugs, lead, methylmercury etc.

Screening and diagnostic evaluation:

Indications for specific genetic tests in children with intellectual disabilities are

1) to focus the primary care physician's anticipatory guidance regarding common symptoms and complications, 2) to predict and manage associated conditions, 3) to determine long-range outcomes and prognosis, and 4) to provide appropriate counseling to the family regarding recurrence risk in future pregnancies. Identification of a specific cause also may eliminate the need for additional unnecessary testing.

Although neuroimaging has been recommended, a recent consensus statement suggests that the absence of physical findings, such as microcephaly, macrocephaly, and focal motor abnormalities, in children who have MR decreases the likelihood of a positive result. In cases in which neuroimaging is performed, magnetic resonance imaging generally is the study of choice. Imaging studies also may provide information about the timing of an insult and help rule out specific disorders. Routine electroencephalography for children who have MR is not indicated unless clinical evidence suggests a seizure disorder.
Metabolic disorders cause MR in a small percentage of patients. However, an increasing number of disorders are amenable to treatment if identified at an early age. The newborn screening helps to identify some of the disorder very early and treat appropriately.
Additional metabolic testing should be considered when history (parental consanguinity, family history of similar problems, developmental regression, and episodic

decompensation) or physical findings (hypotonia, declining growth, hepatosplenomegaly) suggest a specific cause.

Theraputic Options:

Children with intellectual disabilities and/or autism spectrum disorder who have symptoms of hyperactivity and short attention span may respond to medication like stimulants.

Autism spectrum disorder:

Clinical features, presentation:

The hallmark of autism is abnormal social interactions. Children lack the ability to share interests with others (joint attention skills) using verbal or nonverbal communication. They commonly show weakness in eye contact. Their interaction may range from aloofness and an unawareness of other people to having varied or odd interaction with others. Language development commonly is delayed, and children may have immediate or delayed echolalia, unusual intonation, and repetitive speech. Children who have autism may engage in repetitive play and show little imaginative play. They may focus on sensory aspects of objects or develop obsessions about unusual objects (stop signs, elevators, etc.). They often have difficulty handling transitions and may engage in repetitive hand or body movements. Many affected children have cognitive impairment too.

A child with isolated speech and language delay is more common than ASD. The first step is to determine whether the speech delay is isolated or whether there are global delays. Receptive language as reported by the parents is very helpful. If receptive language is on time, such as two-step or three-step commands, then speech delay may be more isolated. Motor delays can contribute to speech delays, especially to articulation, but receptive speech should be normal. Hearing testing should be done as part of the work-up. Also, an autism screening tool is helpful. This will help to differentiate between ASD and isolated speech and language delays.

The main difference between ASD and ID is that, most people with an intellectual disability show relatively even skill development. Individuals with autism typically show uneven skill development. They have strengths in some areas, especially visual tasks and difficulties in other areas such as problem solving. Regardless of their level of intellectual functioning, problems with social awareness and understanding persist in people with Autism.

During the 1960s and 1970s, the vast majority of those diagnosed with autism had an intellectual disability (up to 75%) but today, only about 40% of patients with ASD have ID. These patients are recognized as higher functional population. Although commonly associated with general intellectual disabilities, pproximately 75% of people with autism have a non-verbal Intelligence Quotient (IQ) below 70. Autism can also occur in individuals of normal, or even superior intelligence

A child with profound hearing loss will have higher IQ, better relationships, better non-verbal communication when compared to a child with ASD. This helps to differentiate between the two and also an audiology testing helps to confirm the profound hearing loss.

Atypical developmental patterns that may be indicators of autistic spectrum disorder are 1. Children who have autism may demonstrate simultaneous hyposensitivities and hypersensitivities for different stimuli within the same sensory modality. For example, the

slightest sound of water dripping may irritate the child, yet the child may seem oblivious to his mother calling his name loudly. 2. Affected children may explore toys visually in unusual ways,

holding objects very close to their eyes, looking at objects out of the corners of the eyes, or demonstrating an unusual head tilt. 3. Others may have oral aversions and intolerance to certain food textures that contribute to self- imposed restricted diets. 4. The motor skills of children who have autism may seem advanced, especially running, climbing, and jumping skills. However, some children may have subtle deficits in fine motor skills, coordination, and motor planning and sequencing of movements.

Etiologies

Autism's theory of causation is still incomplete. It has long been presumed that there is a common cause at the genetic, cognitive, and neural levels for autism's characteristic triad of symptoms. However, there is increasing suspicion among researchers that autism does not have a single cause, but is instead a complex disorder with a set of core aspects that have distinct causes. In other words, completely different underlying brain dysfunctions have been hypothesized to result in the common symptoms of autism, just as completely different brain problems result in mental retardation. Various genetic, prenatal, perinatal, postnatal causation has been tried to explain the cause for autism but none of them has been able to describe the exact cause of Autism

Screening and diagnostic evaluation:

Diagnostic criteria for the autism spectrum disorders are: 1)Lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (eg, by lack of showing, bringing, or pointing out objects of interest) 2)Lack of social and emotional reciprocity 3)Marked impairment in the use of multiple nonverbal behaviors, such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction 4)Delay in or total lack of the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime). Researchers propose that all four criteria be present to make the “provisional diagnosis” of autism in the very young child(less than 3 years of age).; However after 3rd birthday full re-evaluation using the full DSM-IV-TR criteria, combined with a standardized tool, is recommended to diagnose Autism.

Screening for the autism spectrum disorders in the medical home should be completed at 18, 24, and 30 months of age and whenever a concern is raised. The ASDs surveillance and screening algorithm published in the AAP ASD clinical report states that if a child has two or more risk factors (eg, older sibling having an ASD, parent concern, other caregiver concern, or physician concern), the primary care physician should make three simultaneous referrals: to an early intervention or school program (depending on the child's age), to an ASD specialist or team of specialists for a comprehensive evaluation, and to audiology (if this has not already been done).

Therapeutic options:

Management of autism generally is the province of early intervention specialists, educators, therapists, and behavior management specialists. Early intervention programs and schools

should provide services that are individualized, appropriate, and intense and should address development, social skills, behavior, and academic issues. Medications should be considered when behaviors (eg, self-injurious behaviors, aggression, anxieties, obsessions, stereotypies, sleep disorders) negatively affect or prevent the progress of educational, therapeutic, and behavioral intervention. There is no medical "cure" for autism, and only one medication (risperidone) has been approved by the United States Food and Drug Administration for use in children who have ASDs and aggressive or self-injurious behaviors

Speech and language disorders: Definition of:

Presentation (normal variations):
Normal disfluency of childhood occurs in preschool children during the attainment of language skills (age 2 to 3 years) and that stuttering represents a clinical disorder after 5 years of age
Articulation of all consonant sounds is not complete until 6 years of age. Speech development progresses in tandem with language. The first eight consonant sounds that develop are: m, b, y, n, w, d, p, and h. The last eight consonants typically acquired are sh, th as in the word "think," s, z, th as in the word "the," l, r, and the zh sound usually spelled as s, as in the word "treasure." Consonant blends such as sp, tr, and bl also may not emerge until early school age. At age 2 years, a child who is developing typically can create speech that is intelligible to an unfamiliar adult at least 50% of the time and at age 3 years, at least 75% of the time. Children may continue to have difficulty with pronouncing some sounds until about 7 years of age. Nonetheless, 100% of speech should be intelligible by about age 4 years.

Etiologies:

Language and speech disorders are features of many genetic and chromosomal disorders. For example, children who have Down syndrome typically show verbal skills below those expected on the basis of cognitive functioning. Children who have fragile X and Klinefelter syndromes may present with delayed language skills with or without characteristic physical features. Toddlers and preschoolers who have William syndrome are delayed at early stages of language development, although they ultimately exhibit communication and social skills in advance of other aspects of their cognitive functioning. Language and speech disorders also are features of neurologic

diseases such as seizure disorder and cerebral palsy. In school-age children with no obvious genetic or neurologic condition, the prevalence of language disorders is approximately 2% to 3%; the prevalence of speech disorders is between 3% and 6%.

Speech and language disorders are more common in boys than girls and more common among children who have a family history of language, speech, or reading disorders in a first-degree relative than among children who have a negative family history. So it is very essential to obtain a family history in these patients.

Screening and diagnostic evaluation:

Children presenting with language delay should receive a full audiologic assessment, using the assessment techniques that are appropriate for the child's age. Children who have normal hearing and indications of impairments of cognitive or social skills should receive a comprehensive developmental assessment. Early intervention services for children from birth to 3 years of age can provide an assessment of the child's level of functioning in each developmental domain. All children with speech or language delays should be referred to speech-language pathologist. He/She will verify the degree and nature of language or speech delay, as well as make recommendations for the frequency and type of treatment. In general, therapy is effective for improving specific skills and communication. For toddlers who have delayed development, some treatments focus on the nature of adult input to the child, capitalizing on the importance of environmental input in stimulating language development. Other approaches treat children in group settings or classrooms. These children generally benefit from participation with children who are developing typically and who provide good models for communication.

> Mild-to-moderate delays in language development, such as not saying single words at 15 to 16 months of age and not speaking in phrases after 24 months of age, among children who have a positive family history for Reading Disability should give a pediatrician cause for concern. As children age, subtle language problems, such as difficulties with word finding or use of nonspecific or vague words (eg, "thingies" or "stuff"), may reflect language problems that warrant attention. During an office visit, ask parents of 3- and 4-year-old children about their child's interest in and ability to recite nursery rhymes or play rhyming games. Not reciting nursery rhymes at 4 years of age and the inability to recognize words that rhyme by 5 years of age may reflect early problems.
>
> For children already in school, difficulty decoding single words or unfamiliar words, slow reading, and problems with spelling are highly suggestive of a reading disability. At the beginning of first grade, children should begin attempting to sound out words and regularly identify the first sound in a word. As the year progresses, they should recognize the ending sound and medial sounds. Although fluency is not expected at this point, children should be

reading by the end of first grade. Regularly sounding out words is expected during second grade. It is usual for children to "read" a word inappropriately on impulse, but if the child does so regularly and is unable to return to the word and attempt to decode its component sounds, there may be cause for concern. A lack of desire and resistance to complete any reading task combined with difficulty with reading grade level books is a red flag, especially in the presence of other risk factors such as family history or language delays.

> Developmental screening involves the use of a brief standardized tool to identify children at risk of developmental delay. A formal developmental screening tool should be administered at the 9-, 18-, and either 24- or 30-month health supervision visits as well as when there are concerns regarding development. In addition, an autism-specific screen is recommended when the child is 18 and either 24 or 30 months old. If development screening tool results raise concerns, additional evaluation is needed to establish a diagnosis and direct treatment.

General developmental screening tools, such as the Ages and Stages Questionnaire, are effective for identifying children at risk for language and other developmental delays. An autism-specific screening tool, such as the Modified Checklist for Autism in Toddlers (M-CHAT), can identify a child at risk for autism. Other questionnaires, such as the Child Behavior Checklist, may identify the presence of attentional issues, hyperactivity, or oppositional behaviors.

Therapeutic options:

When a child is diagnosed with speech or language delays, the therapist should use evidence-based practice in developing an intervention plan. The interventions can range from clinician-directed to child-centered or a hybrid of the two approaches. Clinician-directed therapy is a highly structured approach. The clinician controls the situation and provides reinforcement and feedback on the performance. In child-directed interventions, such as indirect language stimulation or developmental/pragmatic approaches, the clinician sets up a situation offering opportunities for the child to provide responses that are natural in the situation. This approach is very useful for a child who is either noncompliant or not a very assertive communicator. In the hybrid approach, the clinician focuses on a few specific language goals, maintaining control but facilitating spontaneous responses using the language goals that are being targeted.

Learning disorders:

Clinical features, presentation:
Children who have LD perform overall within the normal range of intelligence and adaptive functioning but show isolated specific academic skills deficits.
Cognitive abilities and academic skills develop at different rates for individual children.
Reversals (b and d) in writing can be normal for children through 7 years of age.
Earlier signs of LD may assist in earlier identification. Through the use of developmental surveillance at routine health supervision visits, the pediatrician may recognize significant risk factors, such as a prior medical history of prematurity or family history of LD and educational difficulties. A history of preschool developmental problems can also indicate that the child is at increased risk for LD. The child who has a preschool speech and language disorder may later experience educational difficulties in areas such as comprehension of language-based instruction or the phonemic processes used in the development of early word reading or decoding. Difficulty with recognition and drawing of shapes in the preschool period may portend problems in letter recognition or writing. Such language and visual motor difficulties may also be associated with problems with the sound/symbol associations needed for reading. Performance of formal developmental screening at the 30-month visit may identify these related preschool problems; performance at the 48-month visit may identify specific problems in early decoding, writing, and sound/symbol association.
Close to 3 million children from the ages of 6 to 11 years are affected by learning disabilities (LDs). The category of specific LD includes disabilities in mathematics and written expression, but the most common form of specific LD is reading disability (RD) or dyslexia. RD accounts for 80% of cases of LD and occurs in an estimated 5% to 10% of the general pediatric population, with the most recent population surveys placing the figure at 6.5%.

Males who have RD outnumber similarly affected females. Learning disability is defined as a disorder in one or more of the basic psychological processes involved in understanding or in using language, spoken or written, that may manifest itself in an imperfect ability to listen, think, speak, read, write, spell, or do mathematical calculations" and "includes such conditions as perceptual disabilities, brain injury, minimal brain dysfunction, dyslexia, and developmental aphasia. Dyslexia is defined as "difficulties with accurate and/or fluent word recognition and by poor spelling and decoding abilities" that "typically result from a deficit in the phonological component of language."

The reported prevalence of dyslexia (specific reading disability) ranges from 5% to 17% in the general population. The rate of dyscalculia (specific learning disability in mathematics) is 4% to 6%. Individuals may have learning difficulties that occur independent of intelligence. Frequently, individuals who have high intelligence are not identified as having a learning issue or are considered to lack motivation. Children who have above-average intelligence may have uneven profiles when evaluating their cognitive, social, and emotional development.

The prognosis for children who have LD can range from academic and social success to ongoing vocational and personal-social problems in adulthood. Affected children are at increased risk for poor academic performance, not completing high school, and exhibiting behaviors that, in some cases, may escalate and lead to expulsion. For a clinician presented with a child in the midst of school failure, the long-term effects on the student's self-esteem and behavior may overwhelm concerns about academic failure. Longitudinal tracking of children who have Reading disability reveals persistent reading problems into young adulthood. However, with proper parent, school, and community support, children who have LD can reach high levels of achievement.

Problems encountered in content classes (eg, social studies, history) may reflect reduced reading comprehension, impaired short-term memory, or a slow reading rate.

Signs and Symptoms lf Learning Disability and School Failure

Differential diagnosis in a child who presents with academic difficulties is attention-deficit/hyperactivity disorder, intellectual disability, depression, Obsessive-compulsive disorder etc. A child should undergo a comprehensive physical examination to rule out visual and auditory problems, disordered sleep, nutritional deficiencies, thyroid disease, history of an acquired neurologic insult, or epilepsy. An older child should be evaluated for use of illicit drugs or alcohol.

Children who have attention problems generally exhibit distractibility and disorganization. And this may be confused with learning disability. So detailed evaluation is essential in order to differentiate between the two.

Etiologies:

The causes of school-related difficulties are usually multifactorial eg, learning difficulties, attention deficit hyperactivity disorder, depression, social issues, environmental factors etc. Significant evidence points toward both genetic and neuroanatomic bases for both LD and RD. Family and twin studies suggest that 50% of the problems in performance can be accounted for by heritable factors, with environmental influences greater in children who have lower IQ scores. Genetic linkage analyses suggest loci on chromosomes 2, 3, 6, 15, and 18, with four candidate susceptibility genes (DYX1C1, KIAA0319, DCDC2, ROBO1)

identified. These genes are involved in neuronal migration and axonal growth. Identified anatomic abnormalities include ectopias in cortical layer 1, focal microgyria, and defects in the thalamus and cerebellum. Differences have also been described in the temporal, parietal, and occipital brain regions. Abnormal circuitry may account for the sensory, motor, perceptual, and cognitive problems seen in LD, including the phonologic deficits, motor deficits, and problems in auditory discrimination, borne out in recent functional magnetic resonance imaging studies.

Patients who have CNS-based chronic conditions (eg, epilepsy, myelomeningocele) have an increased incidence of learning disabilities.

Patients with post-concussive syndrome have high incidence of temporary learning problems.

Screening and diagnostic evaluation:

School distress and increased effort of learning are the two most common indications for psychological, educational, and medical evaluation of a child with poor school performance. In 2006, 7% of children (1 in 14) had been suspended from school at least once during the year. In some students, signs of school distress can present as internalizing behaviors resulting in frequent absences or social disengagement; other students may present with externalizing symptoms such as "acting out" (ie, negative attention seeking at school) leading to frequent detentions or aggressive behavior that may eventually result in suspension or expulsion. Aggressive behavior toward vulnerable students (bullying) is particularly in the spotlight due to increased awareness of recent acts of homicide and suicide occurring in schools. In the 2005 to 2006 school year, 78% of public schools experienced one or more violent incidents; 17% of these were reported as serious violent incidents.

The presence of strong family support has been shown to have a positive effect on the prognosis of children who have learning disabilities. Support at home via interactive learning activities (use of readers, audiobooks) is especially important for children who have learning disabilities because they often are overwhelmed, disorganized, and frustrated in learning situations. Specific academic support such as one-on-one tutoring in the home frequently is sought to help focus on homework. Family support can augment specialized educational intervention. Educational strategies such as remediation work targeting the underlying cognitive function that is impaired and phonologic programs (those that improve word decoding) have shown efficacy up to the sixth grade. If the area of weakness is unlikely to be corrected, circumvention strategies are useful in improving the outcome. Such strategies include using a keyboard or verbal exams for a child who has motor dysgraphia, using audiobooks for a child who has dyslexia, and using a calculator for the child who has dyscalculia (math disorder).

There are some limitations of psychoeducational and/or neuropsychologic tests for learning disabilities. In general, this type of psychoeducational testing is beyond the scope of a general pediatric practice; although some clinicians who have additional experience in this area (eg, pediatricians who may be board certified in neurodevelopmental disabilities or developmental and behavioral pediatrics) may feel comfortable administering and interpreting some measures in the office. In either case, it is helpful for primary care

physicians to become familiar with local resources in the schools and in the community where this testing can be obtained. When LD is suspected, the clinician should guide the family to these resources and assist in the referral process

Blood, urine, and imaging studies generally are not indicated or diagnostically useful in the evaluation of school distress and LDs. Notable exceptions include neurologic findings suggesting a focal lesion, skin findings suggesting a neurocutaneous syndrome, and physical findings or past medical history suggestive of disorders such as syndromes or nutritional disorders that have a genetic or metabolic cause. In such cases various genetic testing and neuroimaging (eg, SPECT, PET, fMRI)) can be done to determine the exact cause of LD.

Commonly Used Tests for Evaluation of Suspected Learning Disabilities are:

Therapeutic options:

Educational criteria for placement in special classes are different in different communities

Children showing academic difficulties require psychoeducational evaluation to determine their cause and eligibility for special education services. The results of the evaluation may be used to establish an Individualized Education Plan (IEP) that includes evidence-based educational remediation, accommodations, and modifications. Special education in each local school district is mandated and regulated under the federal Education for All Handicapped Children Act (PL 94-142). This act was reauthorized and revised as PL 101-476 under the title Individuals with Disabilities Education Act (IDEA). The law provides that every child identified via screening or observation as meeting the definition for developmental delay be referred for a comprehensive multidisciplinary team assessment. The team works in collaboration with the family to establish an IEP that contains defined education and therapy strategies and objectives.

The 504 plan is part of a United States federal law passed in 1973 (the Rehabilitation Act) that extended civil rights to individuals who have disabilities. The Act allows for reasonable accommodations such as a special study area, as necessary, for students who have disabilities. The IEP process is mandated by the recently reauthorized (2004) IDEA, which is enacted to provide every United States child with a free and appropriate education that meets the unique needs across the lifespan (birth to 21 years) of the individual who has a qualifying disability and prepares that person for future educational, employment, and independence opportunities.

Other factors influencing learning and performance:

General:

Multiple factors other than intellectual and learning disabilities that can cause academic underachievement are chronic lung disease, neurological disorders, socioeconomic status, as well as psychological disorders

Family factors:

Children need a stable emotional base to attend to learning. Family conflict may shift a student's attention away from school. Poverty, substandard living conditions, or homelessness may create conditions that make school success a secondary concern. Parents' negative attitude toward their own schooling also may hinder a child's focus on education. Maternal depression, especially during preschool years, can have a deleterious

effect on a child's cognitive and emotional development. Abuse and neglect often affect school

performance adversely. Children in foster care experience high rates of school failure and drop out.

Physical impairments and chronic illness:

Children with complex partial or absence seizures, Tourette syndrome, visual problems, mild conductive hearing loss may present as complaints about school performance.
Any acute or chronic medical condition may increase school absences. About 75% of all school absences are attributed to health problems. School absences may be more likely to affect school performance when the absence is prolonged (≥6 d) or cumulatively excessive (≥10 d per quarter, ≥30 d/y). Some common conditions are associated with cognitive issues (eg, iron deficiency anemia associated with a small, yet nonreversible decrement in IQ), with increases in school absences and school problems (eg, asthma), and with early school failure (eg, frequent ear infections). Other chronic illnesses i.e., inflammatory bowel disease, malignancy, epilepsy, diabetes, arthritis, hemophilia, human immunodeficiency virus infection, cystic fibrosis and their treatments affect school performance.

Medications:

Some common medications may affect a student's school performance, such as antihistamines and decongestants. Medications used for seizures and asthma also may affect classroom performance.

Emotional disturbances (eg., anxiety, depression):

Emotional disorders such as anxiety, depression, can manifest as academic or behavioral problems. Mood disorders may mimic ADHD and result in school problems. Mood disorders often present with greater intensity and pervasiveness than ADHD. Oppositional defiant disorder may present in school-age children as negative, hostile, and defiant behaviors and cause significant school problems.
Chronic school failure can lead to depression, anxiety, substance use, other risky behaviors, reduced motivation for school work, juvenile delinquency, and school drop-out. Studies show that adults who fail to complete high school are more likely to experience poorer health outcomes. Smoking rates are higher among adults who finished less than a high school education (in 1999, 32% compared with 23% of all persons). Low educational attainment is associated with higher age-adjusted death rates (in 1998, 562 deaths/100,000 who had less than a high school education, 466/100,000 for high school graduates, and 224/100,000 for those who had more than a high school education). Mothers who have completed less than a high school education are less likely to breastfeed (in 1993 to 1994, 43% versus 58.1%), and among breastfeeding mothers, they are less likely to continue to breastfeed for 3 or

more months (in 1993 to 1994, 44% versus 56%).

Students with learning disabilities are at increased risk for behavioral/mental health disorders.

Special sensory deficits

Hearing impairment:

Clinical features, presentation:

Hearing impairment that begins after the age of 5 years has less of an impact on language development than an earlier onset of hearing impairment.

The relationship between the various levels of hearing impairment (mild, moderate, severe, profound) and language development are as follows:

Untreated chronic hearing loss can affect language development, leading to speech delay and difficulties with articulation. Such loss also can manifest as a child asking people to repeat themselves, not hearing instructions, ignoring a speaker, or listening to loud television or music. In extreme cases, children can present with behavioral problems or other issues that are manifestations of the effect that hearing loss has on their ability to communicate. Because language serves as the foundation for children to learn other cognitive skills, the effects of hearing loss also can be seen in areas that require interactions for learning, such as a child's ability to understand and regulate emotions and accomplish complex motor skills. The earlier the underlying hearing loss in affected children can be identified by parents and practitioners, the earlier the children receive appropriate interventions and accommodations to minimize the impact on their cognitive, social, and emotional development.

Infants with a congenital hearing impairment achieve the normal prelinguistic language milestones like smiling, cooing, babbling, gesture games at the normal time. However, early hearing loss can be highly detrimental to the linguistic and cognitive development of an afflicted child. Yet, early intervention in the hearing- challenged child may largely offset such developmental loses.

Newborn hearing screening may not be conducted if the baby was born at home or in birthing centers without facilities for hearing screening. In such cases hearing test must be conducted within the first postnatal month.

Deafness is associated with an increased risk of learning disabilities and a resultant low reading level.

Etiologies:

The common causes of congenital deafness are in utero infections, severe hyperbilirubinemia, respiratory distress, prolonged mechanical ventilation, exposure to ototoxic drugs (gentamicin), history of head trauma, meningitis, genetic disorders such as Usher syndrome, Waardenburg syndrome, Pendred syndrome and mitochondrial disorders. Certain infections can cause hearing loss at any age and one of the most common culprits is bacterial meningitis. It is thought that over 30% of bacterial meningitis cases result in some degree of hearing loss-from mild impairment to profound deafness. In a 2006 study, the Gallaudet Research Institute reported that 3.2% of American youth with hearing loss had suffered meningitis.

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Screening and diagnostic evaluation:

Newborn hearing Screening is conducted by using evoked otoacoustic emission (OAE) testing, auditory brainstem response (ABR), or a combination of the two. OAE detects the

evoked sound from the cochlea in response to clicks or tones; ABR measures the electroencephalographic waveform response from the vestibulocochlear nerve. ABR has the potential to detect hearing loss due to auditory neuropathy, a condition to which the neonatal intensive care unit population is particularly prone, in addition to hearing loss resulting from middle and inner ear disorders. Furthermore, ABR is minimally affected by external or middle ear debris. A two-stage screening, in which ABR is used to confirm abnormal OAE results, yields the lowest number of false-positive results, thereby reducing referral rates and decreasing parental anxiety.

Any infant who fails the initial screen should be referred to an audiologist for a full evaluation no later than by 3 months of age. This evaluation should include a wide range of diagnostic tests to confirm and determine the nature of the hearing loss. Such tests include physical examination of the outer ear, OAE, diagnostic ABR, tympanometry, behavioral observation audiometry, visual reinforcement audiometry; play audiometry, conventional audiometry etc. If any audiologic evaluation confirms hearing loss, other investigations are necessary. Because children who have hearing impairment rely heavily on their other senses, a vision screen and developmental screen should be performed on all children showing any degree of hearing impairment. If a syndrome is suspected, referral to a geneticist should be considered.

Causes of an acquired hearing impairment are otitis media with effusion, otitis externa, cerumen impaction, foreign body in the external ear, trauma, Cholesteatoma, Lyme disease, bacterial meningitis, fifth disease, syphilis etc.

Therapeutic options:

Major approaches to the education of deaf children are: The goal of early hearing detection and intervention (EHDI) programs is for all infants to receive targeted interventions by 6 months of age to permit them to be exposed maximally to spoken language and have the greatest likelihood of developing at the same rate as their peers who hear normally. Early intervention services can help children to make the most of their hearing devices and to develop visual communications skills to augment their hearing. Such services maximize the ability of children who have hearing loss to develop well and succeed both academically and socially. Depending on the degree and type of hearing loss, a broad range of hearing devices exists to circumvent the defective part of the hearing pathway. Cochlear implants are used for children with sensorineural hearing loss. Early implantation allows exposure to sound during the critical period in development and leads to improved hearing, language, and speech development. For children with acquired hearing loss, the cause has to be treated in order to improve hearing in these children.

Visual impairment:

Most common causes of visual impairment are refractive errors, strabismus, amblyopia. Some of the congenital causes of visual impairment are cataracts, glaucoma, retinoblastoma, retinitis pigmentosa. Some of the other rare causes are macular degeneration, diabetic retinopathy, trauma etc.

Screening and diagnostic evaluation:

Age-appropriate acuity testing uses instruments that require increasing cognitive ability, ranging from recognizable cartoon pictures (Allen test) or symbols of readily identifiable shapes (LH test) for ages up to 3 years, to choosing which of four letter shapes are presented (HOTV test) or which direction a letter E is pointing (tumbling E test) for ages up to 5 years, to Snellen letters or numbers for ages 5 years and older. The best test for checking acuity is the highest level that the child can complete. Testing is performed at the appropriate distance for the chart used (usually 10 ft), with a line of figures presented at one time. One eye is tested, with the examiner making all efforts to occlude by a patch or wide eye cover the opposite eye at all times, thus avoiding inadvertent "peeks." Differences of two lines of visual acuity between the eyes or vision less than 20/40 acuity in either eye require referral for additional evaluation.

Congenital infections which are associated with development of vision impairment are cytomegalovirus, rubella, herpes, and toxoplasmosis.

Etimologies:

Ophthalmologic problems such as strabismus, amblyopia, nystagmus, visual acuity defects, and blindness can lead to reading or academic problems.

Therapeutic options:

Guidelines endorsed by the AAP recommend visual assessment at birth and at all subsequent routine health supervision visits. Anatomy and gross visual assessments should be checked from birth to 3 years of age, about the age when visual acuity can be measured reliably and ophthalmoscopy can be attempted. Vision should be assessed whenever there is a complaint about vision:

Children who have visual impairment need to learn in specialized environments to motivate them to move and improve their abilities to interpret sensory information accurately. Young infants should be encouraged to be mobile and orient themselves to the environment using touch, smell, and auditory input. Beginning in preschool, the goal is to teach skills that promote independence in the performance of daily activities. Depending on the degree of any associated impairments, the child may attend a regular school program with input from a teacher of the visually handicapped. The educational program should emphasize sensory experiences and auditory programs. The school-age child should have an Individualized Educational Plan. Again, depending on any associated impairments, the child may participate in regular community-based classroom activities, focusing on reading, writing, travel needs, and eventually vocational training and independent living. Braille is used for nonvisual communication and audio recordings supplement reading.

Diagnostic evaluation and community-based treatment:

Diagnostic assessment instruments:

Cognition, intelligence, and adaptive behavior:

In the past, academic testing was compared with the student's estimated potential, as measured by an IQ test, with LD diagnosed when significant discrepancy was demonstrated between IQ and reading scores. This approach was problematic because children whose IQ scores were below average would not meet such

discrepancy criteria and children who had extraordinarily high IQ scores could be considered to have LD in spite of reading scores solidly within the range of age expected norms.

> Both achievement tests and a test of intelligence are valuable in the evaluation of children with school learning problems. Domain-specific academic and cognitive testing is the formal diagnostic process used to determine the presence and extent of LD. In some situations, it is appropriate for this testing to take place through the school district as part of the IEP process. In other situations, parents may prefer to seek private psychological services for this testing. Such an evaluation tests a child's cognitive abilities, including the areas of language processing, memory, attention, and nonverbal reasoning, as well as specific academic achievement in core areas, including reading, mathematics, and written expression
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> An IQ score reflects a child's performance on an intelligence test relative to that of children of the same age. In short, a child's IQ score reveals the extent to which his or her performance on the test departs from average. The IQ score represents a construct of "intelligence" that includes a combination of verbal and nonverbal processing skills, such as vocabulary, information about the world, reasoning, short-term memory, and speed of information processing; these skills, together, are represented by the IQ score.

Classification ratings for IQ ranges as they are distributed along the normal curve.

> To evaluate specific learning disabilities, such as a reading disorder, disorder of written expression, or math disability, IQ tests typically are used in conjunction with achievement tests. Achievement tests are designed to measure what a child has actually learned, including mathematical problem-solving, reading, spelling, writing, or an understanding of science concepts. Most achievement tests focus on a particular subject and measure a child's learning with questions of varying difficulty. The child's score then either is compared with that of a child of the same age or grade or measured against an objective standard. When used to diagnose a specific learning disability, a child's academic achievement in one or more areas is compared with his or her intellectual abilities. If a child's ability in one or more areas of achievement, as measured on standardized tests, is significantly lower than expectations based on age, education, and intelligence, the probability is high that a learning disability exists.
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> Overall, the general rule of thumb is that the older the child, the more stable the IQ. By age 4 years, the correlation with IQ 12 years later is relatively high (r=0.77). Although many older children show little fluctuation in their IQ scores, research has indicated that a subset of younger children show wide fluctuation in IQ scores. Finally, even older children may show some fluctuations in scores in

response to major stressors such as a loss of a parent, divorce, or change in schools. With these possible exceptions, by around age 10 years, IQ scores generally are relatively stable.

> IQ is influenced by genetic factors (eg, the child's genetic makeup), familial factors (eg, parents' IQs and education and quality of the home environment), educational factors (eg, quality of educational opportunities and teaching), and other factors, such as the community in which the child lives. Environmental influences on the development of intelligence include access to stimulating or enriching experiences, caregivers who help the child learn problem-solving skills, access to books and sources of knowledge, good

nutrition, a high level of social support, parental involvement in the child's learning and education, an enriched language environment, good school attendance, good schools, and stable neighborhoods.

The Wechsler Intelligence Scale for Children (WISC-IV) is used to assess a child's mental ability in comparison with the abilities of other children of the same age via a numerical score referred to as the full-scale intelligence quotient (IQ). The scores on a cognitive test are used to predict how a person will function academically. The WISC-IV consists of 15 subtests: 10 core subtests and 5 additional optional subtests. The subtests are grouped into four composite scales known as factor scores. The individual factor scores provide more detailed information regarding the child's mental ability than does the full-scale IQ score. The verbal comprehension factor assesses skills such as verbal knowledge and how a person uses verbal skills in novel situations. The perceptual reasoning factor evaluates the ability to reason and organize material that is seen without the use of words. The working memory factor is based on the ability to remember information and either to manipulate it or use it to perform calculations. The processing speed factor assesses the speed of processing information. So full subtest profile of scores on standardized IQ tests is more useful than the full-scale IQ score alone.

Most intelligence tests assess a range of verbal, visual-spatial, and problem- solving skills. Because they target multiple cognitive skills, IQ tests are composed of subtests that measure specific areas of functioning. Scores on these subtests are combined to yield measures of verbal and nonverbal problem-solving abilities, as well as a full-scale IQ score.

Academic achievement:

Standardized scores measure performance for a group of students, such as all the students in one school, at a certain grade, or in a single school district.

Standardized scores employ a procedure in which scores are reported in terms of standard deviation units from the mean. When scores are standardized to the normal bell curve, two thirds of the students in the sample fall between -1 and +1 standard deviations from the mean. Scores also may be expressed as percentiles or, in some cases, grade equivalents. Standardized scores are less useful for evaluating change in an individual student. A single student may be learning more every day and growing in ability or skill, but still be at the same relative rank compared with his or her peers. Achievement tests measure what a child has learned in specific subject areas such as reading or mathematics. Criterion-based testing evaluates how much a student knows or can do using a specific set of standards, not by comparison with other students, which probably makes it more useful for identifying whether a student is improving or meeting objectives. A child who has a significant discrepancy (determined by each state) between an expected achievement score based on an intelligence quotient test and the actual score may have a specific learning disability and be eligible for specialized educational services.

Educational and developmental programs:

Extracurricular activities are very essential to promote the self-esteem of learning- disabled children.

As students who have learning difficulties progress in their education, the emphasis changes from remediation to accommodation. Accommodation encompasses modifications

in how tasks are given to students so that affected children may complete the same school work as other students. Such accommodations allow students who have learning issues to present their knowledge without being affected adversely by their disabilities Audio texts are useful for students who read slowly. Other potential accommodations allow a change in timing, formatting, setting, scheduling, response, or presentation of the assignment or test. Such accommodations do not alter what the test or assignment measures in any significant manner. Examples of such accommodations include a Braille version of a test for a student who is blind or taking a test alone in a quiet room for one that has attention-deficit/hyperactivity disorder.

Federal legislation for the provision of services to infants, toddlers, and preschool children who have disabilities has evolved since 1986. From its inception as part of PL99-457 (the Education of the Handicapped Amendments) through the most recent changes defined in PL 105-17 (Individuals withDisabilities Education Act, Part C), legislative efforts uphold the rights of students and parents to the key components of a free and appropriate public education. Federal regulations define early intervention services as services that "are designed to meet the

developmental needs of each child eligible under this part and the needs of the family related to enhancing the child's development." The goal of early intervention therapy is to enhance the development of infants and toddlers who have disabilities and minimize their potential for developmental delays. The services are designed to meet the needs of the child and family and promote the child's development in natural environments. Participation in the Part C early intervention system for infants and toddlers is voluntary for the family; they have the right to accept or decline specific early intervention services. Thus, declining such services does not constitute child neglect. Early intervention services were established to allow children who have developmental problems to reach their potential. Research studies have identified specific times in which a child's brain is especially efficient at learning specific information. Coordinated, community- based multidisciplinary programs for early intervention represent effective public policy because they not only help to improve some children's cognitive outcome, but they also aid in family functioning.

The Individuals with Disabilities Education Act (IDEA) of 1990 (PL 101-476) defines the guidelines for education of children in the United States who have specific learning disabilities. According to the "least restrictive clause" in IDEA, children who have learning disorders should be integrated into the mainstream classroom as much as possible. Children who have more significant learning issues (eg, autism spectrum disorder, cognitive impairment) may require a self- contained classroom that provides more individualized and intensive educational support.

Public Law 94-142, the Education of the Handicapped Act, passed in 1975, gave children between the ages of 5 and 18 years access to a free, appropriate public education. Services to children between the ages of 3 and 5 years of age were optional. In 1986, the law was amended as 99-457, which established early intervention programs for children from birth to 3 years who had developmental delay. The law also mandated that preschool children receive services. The law was amended again in 1990 with 101-476, the "Individuals with Disabilities Education Act" (IDEA). The portion entitled Part H, the Program for Infants and Toddlers with Disabilities, required states to develop and implement community- based

systems of care that are coordinated, family-based, and culturally competent, involving greater interagency collaboration. It mandated early identification and provision of services to infants and toddlers who have developmental delays and established conditions. In 1997, the IDEA amendment, PL 105-17 (Part C, formerly Part H), encouraged the states that did not serve the at-risk population to track and monitor such children so that they could be referred when needed. Eligibility for such programs is dependent on whether a child has any developmental disability,

In a recent survey in United States public schools, school failure, defined as grade retention (repeat of grade), occurred in 10% of kindergarten through eighth-grade students at least once. Such children's can present as internalizing behaviors resulting in frequent absences or social disengagement; other students may present with externalizing symptoms such as "acting out" (ie, negative attention seeking at school) leading to frequent detentions or aggressive behavior that may eventually result in suspension or expulsion. Aggressive behavior toward vulnerable students (bullying) is particularly in the spotlight due to increased awareness of recent acts of homicide and suicide occurring in schools.

Behavioral interventions:

The use of rewards is more effective in promoting behavioral change than is punishment. Children with learning, developmental, and behavioral problems should respond best to token economy, which consists of providing rewards or privileges for the child's positive behavior and losing these for negative behaviors. Extinction is the denial of all attention after a child engages in negative behavior, which can be an effective approach but often causes a transient increase in negative behavior that can be discouraging for a parent. Although spanking may immediately decrease a child's negative behavior, it also teaches a child that hitting is acceptable and results in other discipline approaches losing their efficacy.

Counseling:

Education of families is also critically important to help children's with LD to them access appropriate treatment. At a minimum, families should leave the physician's office understanding that RD/LD is not due to a primary visual deficit and that letter reversal, a common finding in typically developing 7 year olds, is not diagnostic of RD. Common misconceptions include regarding the child as lazy or as not trying hard enough. Another misconception is perpetuated when a family is told that repeating a grade could help the student "catch up." So it is very essential that family understands the diagnosis of LD and help the child adequately.

Use of community resource:

Community resources available for parents are information's available in American Academy of Pediatrics pamphlet "Learning Disabilities: What Parents Need to Know." Further information and support can be offered through referral

to parent and LD advocacy organizations, such as the National Center for Learning Disabilities (www.ncld.org), the Learning Disabilities Association of America (www.ldanatl.org), LDOnline (www.ldonline.org), the International Dyslexia Association (www.interdys.org), the National

Dissemination Center for Children with Disabilities (www.nichcy.org), and the Parent Advocacy Coalition for Educational Rights Center (www.pacer.org).

Complementary and alternative therapies:

When the effects of the Feingold diet, sugar restriction, megavitamins, and fish oil were studied, no beneficial role in the treatment of learning and behavioral conditions was found. However, zinc treatment was found to improve certain skills (eg, hyperactivity, impulsivity, and socialization) for children who have ADHD. Limited research has been conducted in immunotherapeutic fad therapies for the treatment of allergic conditions in children. Conventional specific allergen immunotherapy is the only proven therapy for the treatment of allergic rhinitis and venom hypersensitivity. Other forms of immunotherapy, such as Candida immunotherapy and homeopathic desensitization, have not been shown to be beneficial in the treatment of allergic conditions. However, enzyme- potentiated desensitization is promising in the treatment of allergic rhinitis. Additional research is needed to examine immunotherapeutic therapies and to delineate further the role of antioxidants for children who have allergic conditions.

Behavioral and Mental Health Issues

Developmental stages
Pregnancy, birth, first days after birth
Prenatal care
It is important to advise a mother about the environmental effects on her fetus of alcohol, cigarette smoking, drug use, and nutrition
The prenatal visit serves several purposes that benefit both the family and the pediatrician. The objectives of the visit are to

establish a “positive pediatrician-family relationship” gather information from the family

Pprovide anticipatory guidance and enhance parenting skills identify and discuss high-risk issues.

prenatal anticipatory guidance can be useful in supporting immunization compliance

additional topics that could be discussed include feeding choices or breastfeeding support, car safety seat use, circumcision, and safe sleep practices.

it is important to identify resources for maternal/ familial support during the perinatal period and early infancy

High-risk pregnancy

There is a spectrum of reactions of parents to a malformed infant when diagnosed at birth or in utero.
Learning that there are serious problems in their fetus or newborn creates a crisis for the parent that is likened to experiencing the death of a loved one. Parents experience shock, followed by the grieving stages that are associated with a death in the family: denial, bargaining, confusion, anger, and depression.

Parents typically work through this process differently from each other, which may cause further pain and suffering.
In addition, they may be faced with extraordinarily difficult decisions such as whether to terminate the pregnancy or whether to withdraw life support.
It is important for the pediatrician to be a good listener and to offer support.
It also has been shown repeatedly that providing information about the fetal/newborn diagnosis and potential outcomes allows parents to regain some control of the situation.
They may choose to pursue further research, seek another opinion, and

consider options and form opinions.

Parent-infant attachment

When compared with adolescents who are not fathers, adolescent fathers are more likely to come from low-income families, reside with a single parent, and underperform academically.
The father may initially desire a close relationship with his child, but this often becomes difficult over time as parenting responsibilities increase.
It is most likely that adolescent fathers will separate from the mother and form a new family rather than increase their involvement with their children with time.
The increased financial burden of a child increases the probability of an adolescent father dropping out of school to find work and reduces the likelihood of the father continuing with his education and pursuing a career.
The multiple stressors he encounters may make it difficult for him to plan ahead, and this includes lack of consistent condom use.
His immaturity, inexperience, low self-esteem, lack of knowledge of development and discipline, and lack of good role models increase the likelihood of poor parenting skills and the likelihood of physically abusing both his child and partner.
Strained marital relationships can lead to increased marginalization of the father in the family and distancing from his children. Over time, this leads to decreased involvement with his children.
Seeking proximity to a parent during times of stress to re-establish a sense of well being is consistent with healthy parent-infant attachment.

Infant

Colic
Colic generally develops in the first two to four weeks of life and persists through the third to fourth months of life. Peak at 2 to 3 months of age.
Stages of crying: Studies in the United States in middle class infants show crying occupies about 2 hours per day at 2 weeks of age, about 3 hours per day at 6 weeks, and gradually decreases to about 1 hour per day by 3 months. Crying is most common between the hours of 3 PM and 11 PM.
Various psychosocial and dietary factors have been implicated in the cause of infantile colic.
Some studies suggest it is due to a disturbance in an infant's sleep-wake cycle or an infant state regulation disorder.
History is important in the diagnosis of colic; physical examination and laboratory tests usually are not helpful .

Colic is an ill-defined syndrome of paroxysmal fussiness characterized by inconsolable agonized crying, drawing up of the legs, abdominal distention, and excessive gas.
A colicky infant is one who is healthy and well fed but cries for more than 3 hours a day, for more than 3 days a week and for more than 3 weeks - rule of threes.
Always rule out other causes of crying in an infant before diagnosing infantile colic.
Undetected corneal abrasion, urinary tract infection, child abuse, unrecognized traumatic injuries should be ruled out first.

Management of colic:

Parents need to be reassured and parental anxiety must be relieved.
One should encourage a quite environment, rhythmic stimulation like gentle swinging or rocking maybe helpful.
Change the feeding habits so that the infant is not rushed, has ample time to burp and can be fed more frequently so as to decrease gastric distention if that is a contributing problem.
Medications like phenobarbital elixir and dicyclomine should be discouraged because of the risk of adverse reactions. Simethicone has not been shown to ameliorate colic.
A trial of ranitidine or other proton pump inhibitors may be helpful if gastroesophageal reflux is contributing to the symptoms.
For colic that is refractory to behavioral management, a trial of changing the feedings, eliminating cow's milk from the formula or mothers diet can be considered.
Diagnosis of food-induced colic is established by the implementation of several brief trials of hypoallergenic formula.
In infants with food allergen–induced colic, symptoms are generally short- lived, so prolonged restricted diets are generally unnecessary.
Periodic re-challenges should be done every three to four months to determine when eliminated foods can be returned to the infant's diet.

Feeding

There are normal variations in feeding and it is important to distinguish these from practices that reflect poor parenting like feeding to quiet the infant and propping the bottle
Infants should be fed when hungry, warm, and dry, not just when they are fussy in an attempt to quiet them.
In addition, the bottle should be held, not propped, regardless of the infant's age.
Propping a bottle increases the risk of choking and the development of otitis media.
Parents also should be advised that their infant should not be put to bed with a bottle of formula or juice, a practice that could lead to dental decay.
A parent should not force a baby to eat. If the child stops feeding, the parent should try to burp the baby. If the infant still does not want to feed after burping, he or she has had enough. If the baby prefers the formula to be warmed, the parent should not use the microwave, which might create hot spots that can burn a baby's mouth.
Children can have feeding problems for various reasons including oral motor dysfunction, cardiopulmonary disorders leading to fatigue, gastrointestinal issues causing pain, social or emotional issues and problems with regulation.

Rocking movements

Normal individuals may demonstrate repetitive movements .

It is important to distinguish normal from abnormal repetitive movements in children.
Movement disorders or dyskinesias are a diverse group of entities associated with abnormal excessive, exaggerated, chaotic, or explosive movements of voluntary muscles. They are generally the result of abnormalities of the extrapyramidal system or the basal ganglia.
Movement disorders in children are typically hyperkinetic patterns. The abnormal movements are activated by stress and fatigue and often disappear in sleep. They are typically diffuse and migratory (chorea), but may be isolated to specific muscle groups (segmental myoclonus, palatal myoclonus) and may not disappear in sleep.
Repetitive movements may represent self-stimulation in a neglectful environment

Temperament

Types of temperament in infants:

The difficult child who withdraws from new stimuli and is easily frustrated.
The easy, highly adaptable child who has regular biologic cycles.
The slow-to-warm-up child who needs extra time to adapt to new circumstances.
About 10% of children were generally less adaptable, had increased activity levels, and tended to be emotionally negative. These children were considered "difficult."
"Easy" children, about 40% of the group, had regular eating and

sleeping schedules, adapted well to new situations, and tended to have positive moods.

A third group, comprising about 15% of the sample was characterized as "slow-to-warm-up." These children tended to be quiet and took longer to adapt to new situations.
Parental counseling should address the temperamental characteristics of infants

Toddler and preschool

Toilet training
The appropriate age for toilet training is related to cultural influences, neurophysiologic readiness, and the child's motivation
Signs of a neurophysiologic readiness for toilet training includes the ability to follow directions, be aware of their urges, have dry periods (of approximately 2 hours or more), have the desire to remain dry and emulate older family members, and have the necessary motor skills to sit still on the toilet and pull clothes and underwear up and down.
Sex differences have been noted in regards to toilet training, with girls tending to achieve toilet training milestones approximately 2 to 3 months ahead of boys.
On average, girls achieve urine control at approximately 32.5 months of age versus 35 months for boys; the corresponding values for stool control are 31.5 months and 34.7 months of age.

Some of the approaches to toilet training include:

Passive/child-oriented method: Children are introduced to a potty-chair and gradually progress from simply sitting on it to using it appropriately with the encouragement of positive reinforcement
"Train-in-a-day" method: involves an intense period of instruction designed to achieve continence in 24 to 48 hours. The training is conducted in one room and generally begins

with a doll that can wet in the potty to demonstrate toileting actions. Then the child is taken through the same motions and subsequently provided with large amounts of liquids to induce frequent urination. The child also is given frequent reminders and checked for dryness every 3 to 5 minutes with positive reinforcement (eg, praise, toys, or food) for all successful toileting behaviors.

If successful toilet training is delayed, physicians should perform a history and physical examination to screen for signs of developmental delay, genitourinary abnormalities, or constipation.

If the evaluation does not reveal any abnormalities, parents should be reassured and encouraged to de-emphasize the process in an effort to transfer more responsibility to the child

Parents should be reminded to provide incentives for desired toileting behaviors and to avoid criticism. All caregivers for the child should follow

a consistent toilet training plan.

Habits

Thumb sucking

The natural history of thumb sucking: onset in utero or in the first few months after birth, peak at 18 to 21 months of age, resolution by 4 years of age

Thumb and finger sucking is a benign habit that usually has no consequences or concern.

Children who suck their thumbs chronically have a higher incidence of paronychia, herpetic whitlow, irritant eczema, and accidental ingestion and can develop calluses and, rarely, digital deformities that require surgery.

Dental changes in primary, mixed, and secondary dentition include malocclusions, such as posterior cross bite, anterior open bite, and excessive over bite

Older children who suck their thumbs are subject to social stigma. Peers as well as their parents may ridicule them, and they may be treated as immature and less socially acceptable.

Treatment for thumb sucking should not be pursued before the child is 4 years of age. Even in older children, if the behavior is infrequent or does not interfere with dentition, cause social stigma, or harm self- esteem, therapy is not necessary.

A variety of techniques can be used to stop thumb or digit sucking:

Positive reinforcement techniques, negative or aversive therapies, competing response therapy, and dental appliances all have been used to treat nonnutritive sucking habits.

Positive reinforcement should be given when the child is not sucking the thumb or finger.

Negative or aversive therapies include a bad-tasting substance put on the nails to deter or remind the child not to put the thumb or fingers in the mouth. A variety of over-the-counter nontoxic substances are sold for this purpose. A sock, adhesive strip, splint, or glove can be used to remind the child not to put the thumb or fingers in the mouth.

Once the child and parent are aware of when the sucking habit occurs, a competing response can be used as an alternative to the thumb or finger entering the mouth.

If all else fails either a palatal crib or rake, can be a reminder not to suck the thumb or finger. Palatal appliances ideally should be placed during the spring or summer, when other activities can distract the child. Usually 3 months with the appliance is sufficient to change

or eliminate the thumb sucking. The earliest a palatal appliance should be considered is during the mixed or permanent dentition stage.

Masturbation

Children follow a progression of appropriate sexual behavior based on age and developmental level.

Common early sexual behaviors include touching one's genitals in private or in public, attempting to touch others' genitals, showing one's genitals to others, or attempting to view others while they are naked.

Typically, these early sexual behaviors diminish by 5 years of age, once the child has a better understanding of cultural norms and has received explanation from his or her parents about social norms and exhibiting these behaviors in private.

When a child reaches puberty, sexual behaviors become more intentional with the goal of producing sexual arousal or orgasm. This behavior typically occurs in private.

Masturbation in public suggests poor awareness of social reality.

Masturbation seldom produces self-induced injury in childhood.

Differential diagnosis of excessive masturbation: sexual overstimulation, environmental deprivation and genital disease (itching).

Excessive masturbation that is not easily redirected may be related to family dysfunction or stress, underlying developmental disability, mental health issues, under stimulation in the home, or possible neglect, physical or sexual abuse.

Medical conditions that result in genital itching such as vaginitis, vaginal discharge, lichen sclerosus, diaper rash, pinworms, or eczema should be ruled out.

The Child Sexual Behavior Inventory is a good tool to help distinguish developmentally appropriate sexual behavior from pathologic behavior in children ages 2 to 12 years.

If behavior is assessed to be within normal limits, the clinician should proceed with parental reassurance and instructions on redirection of behavior and establishment of clear boundaries.

Anticipatory guidance

Knowledge of milestones allows the physician to reassure parents regarding normal development and prevents overanxious behavior.

There is a relation of accident proneness to family stress, disorganization, and hazardous environment.

It is important to counsel parents on setting limits for their toddlers.

Pediatricians can provide parents with age-appropriate anticipatory guidance. During the 12-month visit, for example, the pediatrician might explain that the child soon will begin to experience struggles over autonomy and independence.

More frequent temper tantrums can be expected in the second year of life, as the toddler inevitably encounters frustration while seeking

autonomy, despite remaining heavily dependent on caregivers.

Temper tantrums, breath-holding spells

It is the involuntary stopping of breathing due to a painful or frustrating event.

They occur in almost 5% of children, typically between the ages of 6 and 18 months.

A BHS of sufficient duration may lead to loss of consciousness or a seizure.

There are two major types of BHSs.
The cyanotic spell occurs when the child is upset. The child may give a cry, followed by forced expiration, apnea, cyanosis, and loss of consciousness.
Pallid spells are less common and occur following a painful experience or when the child is startled. The child stops breathing, becomes pale and hypotonic, and may have a tonic seizure. These seizures do not imply a diagnosis of epilepsy because they are provoked by the BHS.
Electroencephalography may show slowing but not epileptiform activity and, therefore, is not indicated.
The parents should be advised that while frightening to witness, a BHS is benign and will not lead to epilepsy, brain damage, or death.

Management of breath holding spells:

Picking up or comforting a child who has become angry or frustrated in an attempt to avert a BHS should be discouraged because it may reinforce an unwanted behavior.
Parents should be consistent and not reinforce the behavior after the child recovers.
Because BHSs are involuntary, behavioral therapy for the child is not indicated.
Parents and caregivers need to be instructed in appropriate behavioral modification approaches.
Hematologic conditions such as iron deficiency and transient erythroblastopenia have been reported in conjunction with a BHS. Because anemia can exacerbate BHS, obtaining a complete blood count is indicated in severe cases.

Temper tantrums are common events experienced by more than three-quarters of children between the ages of 18 and 48 months.

Boys and girls are affected equally, and the median duration of a tantrum is 3 minutes (75% lasting 1.5 to 5 minutes).
Tantrums are composed of 2 temporally related and overlapping components: anger and distress.
Anger tends to occur early in the tantrum and is manifested by behaviors such as screaming, kicking, hitting, stomping, throwing, and stiffening when

held. Whining, crying, and seeking comfort from others characterize distress

The anger component tends to peak and wane at the beginning of a tantrum, regardless of the total tantrum duration. Prolonged tantrums generally contain a longer distress component than do shorter episodes.

Management plan for temper tantrums:

One of the most important components of care is to “catch the child doing well” and provide positive reinforcement for desirable behaviors when the child is not having a tantrum.
These rewards focus largely on time and attention from the caregiver; material rewards are occasionally appropriate.
Planned ignoring has also been used successfully, although there may be an initial increase in the problem behavior when this approach is instituted.

A star chart or token economy is an effective behavior modification approach but is more appropriate for an older child.
Time-out and other negative reinforcements may increase tantrum frequency. Time-outs should be limited to approximately 1 minute per year of age
Referring the parents to a counselor would be appropriate if they have exhausted simpler approaches, if they have difficulty developing a consistent approach at home, or if the child's symptoms exceed the usual duration, frequency, or pattern for temper tantrums.

Managing biting:

Distraction and redirection through a change in activity or environment are the best means of discipline for discouraging inappropriate behavior.
Any verbal directives or comments made in response to biting should be expressed in a neutral tone and kept simple (eg, "No biting").
A brief response also limits the potential for the child to receive additional adult attention that he or she could perceive as rewarding for the behavior.
A child should never be bitten back.
Adults caring for the child should praise appropriate behaviors and monitor for any cues before biting to prevent and interrupt behavior whenever possible.

Head banging

Head banging does not indicate a sensory deficit.
Head banging may occur in about 25% of infants and toddlers, tends to be more common in boys, and peaks by about 18 to 24 months of age.
Most normally developing children outgrow the habit by age 3 years.
Head banging may relax or comfort the child and, therefore, may be performed as a toddler falls asleep or wakes during the night.
Children who are teething may also bang their heads, possibly to distract themselves from the pain.
Head banging also may occur when a child is frustrated or during a tantrum.
Some children may bang their heads to gain attention, particularly if the parents have a strong reaction to the behavior.

Middle childhood

Fears and phobias
Phobias are a subcategory of anxiety disorders.
The definition of phobias from the Diagnostic and Statistical Manual of Mental Disorders, edition IV criteria is excessive anxiety and worry occurring more days than not for at least 6 months and associated with at least one of the following symptoms: restlessness, feeling tired easily, difficulty concentrating, irritability, muscle tension, and sleep disturbance.
Fears can be described as the unpleasant feelings that arise in response to realistic danger.
Certain fears are more likely to occur at specific developmental stages and at certain ages.
Infants often are afraid of loud noises and strangers; toddlers are of storms, the dark, and separation from their parents; preschool-age children fear monsters and ghosts; and school-age children are frightened by bodily injury, punishment, and failure.

Specific phobias are fears that are excessive, not based in reality, last for at least 6 months, and affect normal daily function. Specific phobias are divided into five subtypes: animal (eg, excessive fear of snakes, spiders, insects, or dogs), natural environment (eg, fear of heights), blood injection-injury (eg, fear of the dentist, needles), situational (eg, flying phobia), and other (eg, fear of loud noises such as thunder).

Because treatments of fears and phobias vary, pediatricians need to understand the different therapeutic interventions. The following list describes ways a parent can help a child manage his or her fears.

Identify the problem.

Help the child to clear his or her mind of past fears. Relax. Encourage the child to take a break. Suggest activities that reduce stress such as walking, reading a book, or taking deep breaths.

Provide physical comfort. Have the parent give a sense of safety and security.

Limit exposure to the media.

Help the child think of positive thoughts to clear his or her mind of fears.

Talk to the child in a calm and understanding way and encourage the child to express his or her thoughts.

Do not allow the child to avoid the feared stimulus. Avoidance breeds more anxiety.

Phobias that affect normal functioning at home, school, or in social situations require the expertise of mental health practitioners.

Three primary approaches can be undertaken to manage phobias: traditional behavioral strategy, cognitive behavioral therapy, and medications.

Traditional behavioral strategy involves desensitization or exposure therapy that focuses on changing the response to the feared object or situation. Gradual repeated exposure to the cause of a phobia might help the child learn to conquer it.

Cognitive behavioral therapy encompasses a more comprehensive form of treatment. The therapist helps the child learn new ways to view and cope with the feared object or situation.

A variety of pharmacologic agents, such as anxiolytics, antidepressants, sedatives, and beta-blockers, have been used in the treatment of childhood phobic disorders. However, the efficacy of pharmacologic treatment for phobic disorders in children has not been well studied.

Childhood fears are common, follow an expected developmental progression, and often can be handled with parental guidance and counseling from the pediatrician.

Lying and stealing

When repetitive and persistent patterns of behavior in regards to lying and stealing are present always rule out a severe psychiatric disturbance .

Anticipatory guidance

Parental monitoring during middle childhood (ages 5 to 10 years) can have alifelong effect on children's health.

There are many health-related problems that can result from poor parental monitoring, including injuries, antisocial behavior, substance abuse, and sexually transmitted infections. Evidence suggests that parental monitoring can prevent many of these outcomes.

Multiple studies have demonstrated that lower levels of parental monitoring are associated with higher levels of aggression in boys.
Preteens and adolescents who perceive less parental monitoring are more likely to drink alcohol and smoke. Exposure to media intended for adult audiences may play a role in this link.
Poor parental monitoring during the teen years is associated with an increased likelihood of risky sexual behavior.
For parental monitoring to be effective, a trusting and loving relationship between a parent and a child is necessary.
Parents and children need to be able to communicate without excessive conflict to prevent a "flight to peer" response, a distancing strategy in which children spend more time with peers, resulting in a greater amount of unmonitored activities.
A poor parent-child relationship can decrease a parent's desire to

monitor because the parent may develop an attitude that behaviors not seen do not need to be corrected. Parents can be encouraged to spend time talking with their children about family values, attending their children's activities, listening to their children's concerns, and becoming acquainted with their children's friends.

For misbehavior, monitoring must be coupled with discipline. Parents should be encouraged to use nonphysical responses such as discussing why the behavior is wrong, setting limits, redirecting toward a positive behavior, promoting empathy, and withdrawing privileges. Parents should offerfrequent
It is important to counsel parents about the involvement of their children in middle school in extracurricular activities such as music and sports: under involvement, over-competitiveness and socialization

Adolescence

Anticipatory guidance
Aggressive negative behavior may be adolescent rebellion: contrast frequency, severity, duration of symptoms to differentiate this from behavioral issues.
Behavioral changes common with the onset of early adolescence include: fatigue, increased sleeping, irritability, secretiveness and easy embarrassment.

Externalizing behaviors and conditions

Aggressive behaviors (e.g., aggression, ODD, CD, antisocial behaviors)
Clinical features and presentation
Aggressive behavior can be symptomatic of a normal developmental stage like temper tantrums in toddler years, rebellion in adolescence.
It is important to distinguish normal from abnormal behaviors.
The disruptive behaviors associated with normal developmental stages usually are short-lived, and normal aggressive behavior should not cause significant and persistent dysfunction.
Accurate assessment of a child who exhibits aggressive or disruptive behavior requires determination of the frequency, intensity, and chronicity of the behavior within the context of the developmental stage.

The impact of the behavior on the child's functioning and those around him or her also should be examined.

Etiologies

Environmental and biological contributors to the development and maintenance of aggressive behaviors:
Neglect
Psychological maltreatment
Parental discipline
Media
Lead exposure
Exposure to violence in the media has been shown to contribute to aggressive behaviors in youth, and media violence adds to the cumulative risk for antisocial behavior.
Aggressive behaviors in childhood may result from psychological maltreatment that destroys a child's sense of self and personal safety.
Psychological maltreatment, a repeated pattern of damaging interactions between parent and child, includes interactions such as belittling, rejecting, denying emotional attachment, and modeling of inappropriate behaviors.
In addition, witnessing intimate partner violence is another form of psychological maltreatment that may lead to aggressive behavior in children and adolescents.
There is an association between corporal punishment and aggression in children, adolescents, and adults.

Screening and diagnostic evaluation

The steps involved in the evaluation of a child with aggressive behavior include:

Parent Interview

The parent interview should include gathering information regarding the child's temperament as an infant and young child because disruptive behaviors often begin early in life.
An assessment of the child's developmental history and current status is important for identifying any delays in language, cognition, or other areas that could affect a child's behavior.
Parents should be asked to provide specific information about the child's disturbing behavior, such as when the behavior started and if there were any psychosocial stressors (eg, divorce, homelessness, death in the family, community violence, illness in the family) at that time that could be affecting the child's behavior.

Teacher Interview

Information sought during the teacher interview is similar to that gathered during the parent interview. Teachers should be asked about the onset of the abnormal behavior as well as its frequency, intensity, changes over time, situations in which it occurs, and potential exacerbating factors.

Child Interview

Interviewing younger children provides an opportunity to observe the child's behavior with someone other than a parent.
Older children and adolescents may be asked directly about their perceptions of the issue as well as about family and social functioning.

Standardized Rating Scales

Rating scales allow for a comparison of the child's behavior with that of other children the same age. “Broad” rating scales often are administered first that assess for symptoms associated with a variety of disorders, such as depression, anxiety, aggression, withdrawal, inattention, hyperactivity, and delinquent behavior.
The Youth Self Report, Child Behavior Checklist (Parent Report Form) and Teacher's Report Form, the Behavior Assessment System for Children, and Conners' Parent and Teacher Rating Scales – Revised are used commonly in primary care settings.
Rating scales are based on the opinions of others. They should not be used in isolation for diagnosis.
They should be used in combination with other sources of data like interview data, academic grades and interpreted in the context of the information obtained from other sources.

Therapeutic options

Parent Management Training

The basis of parent management training is teaching parents techniques to alter their children's behaviour. Strategies include positive reinforcement for desired behavior; differential attention attending to desired or positive behaviors and actively ignoring mild inappropriate behaviors; effective instruction giving, using appropriate consequences like mild punishment and consistency.
For older children, a token/point system (response/cost system) may be incorporated into the training.

Cognitive Problem-solving Skills Training

Children showing disruptive behavior focus more on aggressive stimuli, over attribute hostile intent, are deficient in social problem-solving skills, and often lack behavioral self-awareness.
Training programs that focus on cognitive problem-solving skills address these issues by teaching children a step-by-step approach to solving interpersonal problems by revealing the cognitive processes that underlie social behavior.
Appropriate behaviors also are developed through modeling, role-playing, and reinforcement (eg, verbal praise).

Medication

Although stimulants are a reasonable choice to address impulsive aggression in children believed to have underlying ADHD, Guanfacine or Clonidine may cause less irritability.
When the underlying issue is anxiety, selective serotonin reuptake inhibitors (SSRIs) can be helpful like Fluoxetine.

Mood stabilizers such as the antiepileptic drugs valproic acid and lamotrigine also can be helpful in treating aggressive children, as can atypical antipsychotics such as risperidone and aripiprazole.

Disruptive behaviors (Oppositionality, ODD, CD)

Clinical features, presentation

Starting fires and cruelty to animals may indicate an underlying psychiatric disorder.

During the course of normal development, families may experience periods when a child exhibits temper tantrums during toddler years or rebellion during adolescent years. These behaviors, when limited in time, are considered normal developmental occurrences.

Oppositional defiant disorder (ODD) is defined as including ongoing symptoms of "negativistic, defiant, disobedient, and hostile behavior toward authority figures."

Affected children tend to break the rules at school, lose their tempers easily, blame others for their errors, pick on other children, argue with authority figures and adults, and display excessive anger.

When children who have ODD become more persistently angry and heighten their behavioral outbursts, they are described as having conduct disorder.

This is defined in the DSM-IV as "a repetitive and persistent pattern of behavior in which the basic rights of others or major age appropriate social rules are violated." Behaviors include displays of significant aggression toward others, hurting animals, intentional destruction of property, school truancy, stealing, and running away from home.

Children who have CD are more likely to be suspended from school or have police involvement.

The young child who demonstrates ODD as a preschooler may develop CD as he or she enters middle school.

Some children exhibit signs of both disobedience and anger, components of both ODD and CD.

The overall prevalence of CD in the United States ranges from 6% to 16% for males and from 2% to 9% for females.

The young child who exhibits aggressive behavior across various settings is particularly at risk for the eventual development of CD and is more likely to have persistent aggressive behaviors into adulthood (as manifested by anti-social personality disorder (ASPD).

Children who manifest aggressive behaviors earlier in life are more likely to have symptoms of greater severity and participate in more severe criminal offenses during adolescence and adulthood.

Children who display symptoms of both ADHD and CD are at significant risk for poor outcomes. They display greater serious antisocial behaviors and are at greater risk for delinquent behavior and ASPD in adulthood than children who have either ADHD or CD alone.

Etiologies

The environmental (e.g., family systems, community) and biological (eg, genetics, co-existing conditions) contributors to oppositional and defiant behaviors:

Child neglect in the first 2 postnatal years may be a more important precursor of childhood aggression than later neglect or physical abuse at any age.

Psychological maltreatment, a repeated pattern of damaging interactions between parent and child, includes interactions such as belittling, rejecting, denying emotional attachment, and modeling of inappropriate behavior.

In addition, witnessing intimate partner violence is another form of psychological maltreatment that may lead to aggressive behavior in children and adolescents.

A family history of aggressive behavior, including a history of incarceration, places children at increased risk. Parental use of physical discipline provides children with a model for using aggressive behavior as a solution to conflict. This example can lead to future aggressive behavior in the child.

Exposure to violence in the media has been shown to contribute to aggressive behaviors in youth, and media violence adds to the cumulative risk for antisocial behavior.

Aggressive behavior reported by teachers has been linked to elevated blood lead levels. A higher number of youth living in juvenile detention centers have had exposure to lead compared with the general population.

Screening and diagnostic evaluation

Common conditions occurring in concert with oppositional defiant or conduct disorder include: ADHD, mood disturbances, decreased IQ.An estimated 65% to 90% of boys who have conduct problems also have ADHD.

Mood disturbances such as depression also may be associated with CD, and youth may exhibit rage or outbursts during depressive periods Increased risk for anxiety, although the association between CD and anxiety disorders is less consistent than that between CD and other disorders.

Decreased IQ, language deficits, and impaired executive functioning are associated with CD and ODD.

Evaluate a child with defiant, oppositional, or delinquent behavior (school-family information, developmental milestones, child/adolescent interview, rating scales).

Parent Interview

The parent interview should include gathering information

regarding the child's temperament as an infant and young child because disruptive behaviors often begin early in life.

An assessment of the child's developmental history and current status is important for identifying any delays in language, cognition, or other areas that could affect a child's behavior.

Teacher Interview

Teachers should be asked about the onset of the abnormal behavior as well as its frequency, intensity, changes over time, situations in which it occurs, and potential exacerbating factors.

Child Interview

With younger children, the interview provides an opportunity to observe the child's behavior with someone other than a parent. A diagnosis should not be based on a child's behavior in the evaluation; many children do not act out during a medical appointment. Because children younger than 10 years may not be reliable in their self-reports of behavior, diagnosis should not be based solely on their reports. However, their reports of internalizing symptoms, such as anxiety and depression, are more reliable and likely to be more valid than parent and teacher reports of these symptoms.

Standardized Rating Scales

Older children, the child's parents, and the teacher should be asked to complete standardized rating scales.

"Broad" rating scales often are administered first that assess for symptoms associated with a variety of disorders, such as depression, anxiety, aggression, withdrawal, inattention, hyperactivity, and delinquent behavior.

The Youth Self Report, Child Behavior Checklist (Parent Report Form) and Teacher's Report Form, the Behavior Assessment System for Children, and Conners' Parent and Teacher Rating Scales – Revised are used commonly in primary care settings.

In more severe cases, the pediatrician must decide whether to treat the behavior personally or refer the child and family to a mental health professional.

Therapeutic options

Evidence-based interventions are available for addressing disruptive

behavior problems in children. Research indicates that parent management training and cognitive behavior therapy, usually in the form of problem- solving skill training, is the most effective. In addition, research has shown that the two interventions used together are more effective across a wider range of variables at home and with peers than either technique alone.

Parent Management Training

The basis of parent management training is teaching parents techniques and strategies to alter their children's behavior and change the consequences of that behavior. Strategies include positive reinforcement (eg, social praise, tokens/points) for desired behavior; differential attention (ie, attending to desired or positive behaviors and actively ignoring mild inappropriate behaviors); effective instruction giving, using appropriate consequences (ie, mild punishment); and consistency across caregivers, situations, and settings.

For older children, a token/point system (response/cost system) may be incorporated into the training

Cognitive Problem-solving Skills Training

Aggressive behavior is not simply the reaction to environmental events.

Training programs that focus on cognitive problem-solving skills address these issues by teaching children a step-by-step approach to solving interpersonal problems by revealing the cognitive processes that underlie social behavior. Appropriate behaviors also are developed through modeling, role-playing, and reinforcement (eg, verbal praise).

Medication

Selection of medication should begin with insight into the biologic conditions that predispose the individual to aggression.
In addition to the child's history, family history can be very helpful in determining the causes of a child's disordered behavior. ADHD with impulsivity, anxiety, and affective instability, including bipolar disorder, are underlying psychiatric diagnoses that may be present in aggressive children.
Children believed to have underlying ADHD, guanfacine or clonidine may cause less irritability.
When the underlying issue is anxiety, selective serotonin reuptake inhibitors (SSRIs) can be helpful. Currently, the United States Food and Drug Administration approve only fluoxetine for children. Sertraline, citalopram, escitalopram, and others often are used off-label.

Due to concerns about increased suicidality in children and adolescents, most general pediatricians prefer to consult a psychiatrist before prescribing these medications.
Mood stabilizers such as the antiepileptic drugs valproic acid and lamotrigine also can be helpful in treating aggressive children, as can atypical antipsychotics such as risperidone and aripiprazole. A psychiatrist should prescribe these medications and follow children who are taking them.
The pediatrician's role in treating children taking any of these medications, particularly SSRIs, mood stabilizers, and antipsychotics, primarily involves familiarity with drug-drug interactions and possible adverse effects.

Antisocial behaviors, delinquency

Clinical features, presentation

Antisocial personality disorder (ASPD) is defined by DSM-IV as “a pervasive pattern of disregard for, and violation of, the rights of others that begins in childhood or early adolescence and continues into adulthood.” Symptoms include poor school performance, truancy, poor self-esteem, low frustration tolerance, persistent lying or stealing, frequent fighting, and an apparent lack of remorse or empathy. Young adults who have ASPD may have had a history of childhood CD.

Etiologies

Antisocial behavior may be indicative of other disorders like depression, anxiety and psychosis.

The environmental and biological contributions to the development and maintenance of antisocial behaviors include neglect, physical discipline, psychological maltreatment, media exposure and lead exposure

Screening and diagnostic evaluation

Older children, the child's parents, and the teacher should be asked to complete standardized rating scales in addition to providing information during the interviews.

Rating scales allow for a comparison of the child's behavior with that of other children the same age. “Broad” rating scales often are administered first that assess for symptoms associated with a

variety of disorders, such as depression, anxiety, aggression, withdrawal, inattention, hyperactivity, and delinquent behavior.

The Youth Self Report, Child Behavior Checklist (Parent Report Form) and Teacher's Report Form, the Behavior Assessment System for Children, and Conners' Parent and Teacher Rating Scales – Revised are used commonly in primary care settings.

Internalizing behaviors and conditions

Anxiety
Clinical features, presentation

Anxiety disorders are diagnosed when fears, worries, or anxiety occur outside the range of normal developmental responses or are extreme and cause significant distress or impairment in functioning (school, home, social settings).

In anxiety disorders, the fear response is no longer adaptive and is either out of proportion to a stressor or occurs when there is no threat.

Epidemiology of anxiety disorders:

Anxiety disorders are the most common psychiatric illnesses in children and adolescents, affecting 8% to 10% of this population.

The age of onset of anxiety disorders can be as early as the preschool years, but the condition generally does not cause substantial impairment until school age.

Anxiety disorders can occur throughout the lifespan, but most adult anxiety disorders begin by early adolescent years.

Girls have twice the likelihood of developing anxiety disorders as boys. Anxiety disorders can cause family distress and dysfunction, school refusal and avoidance, and social isolation. Untreated anxiety disorders increase the risk of developing further anxiety disorders, depression, and substance abuse.

The long-term social impact of untreated anxiety disorder includes decreased academic and occupational achievement and poorer social and relationship outcomes.

Anxiety disorders can be accompanied by comorbid conditions, such as medical illnesses, secondary anxiety disorders, attention- ficit/hyperactivity disorder, disruptive behavior disorders, learning disorders, and mood disorders.

Anxiety disorders tend to run in families, and this finding is believed to be related to a combination of genetic predisposition and environmental factors.

Some young children have temperament styles that cause them to be more anxious and avoidant in new or unfamiliar situations, also known as behavioral inhibition. This temperament style increases the risk of developing an anxiety disorder in childhood.

Environmental and psychosocial stressors, including school issues, social difficulties, family dysfunction, trauma, and loss can contribute to anxiety disorders.

A specific phobia is fear of an object or situation that is not developmentally expected and is out of proportion to the actual danger and manifests as an anxiety response that is extreme and unreasonable (eg., avoidance or intense anxiety and panic).

The specific fear must be present for at least 6 months and cause the child substantial distress or impairment in functioning to be considered a true phobia.

Social phobia (social anxiety disorder) is the most common anxiety disorder in teens, likely because of the increased importance of peers and social context in adolescent development.

Youth who suffer from social anxiety disorder have severe anxiety in social situations that is extremely distressing and can lead to avoidance and substantial deterioration in overall function.

Generalized anxiety disorder can have its onset in childhood and

adolescent years.

The anxiety centers around everyday events and responsibilities in the child's life, such as school, friends, health, future, and finances, but the distress and worry are excessive, unrealistic, or unhelpful and persist for at least 6 months.

Children may report feeling tense and irritable; have frequent muscle aches and pains; and experience poor sleep, tiredness, and difficulty concentrating. Affected individuals may have academic performance anxiety that interferes with starting and completing assignments and taking tests due to fear of failure.

A pattern of avoidance can develop to prevent a feared event or the youth may seek excessive reassurance from others that nothing bad will happen.

Screening and diagnostic evaluation

Several anxiety disorder screening tools have been well validated for use in children and adolescents.

The Multidimensional Anxiety Scale for Children (MASC) is a self-report screen for identifying anxiety disorders in children and adolescents.

The Screen for Child Anxiety Related Emotional Disorders (SCARED) and the Spence Children's Anxiety Scale (SCAS) both have parent and self-report versions to screen for anxiety disorders in youth.

Therapeutic options

> Treatment of childhood anxiety includes both specific and nonspecific treatment approaches.

Specific approaches are evidence-based treatments for anxiety disorders and include structured psychotherapies (cognitive behavioral therapy [CBT]) and pharmacotherapy (selective serotonin reuptake inhibitors [SSRIs]).

Nonspecific treatments include activities that decrease stress, improve mood, and support well-being, including sleep hygiene, healthy eating, regular exercise, predictable routine, and social supports.

Anxiety disorder treatment guidelines recommend the use of CBT as first-line treatment for children who have anxiety disorders when this therapy is available. CBT includes psycho education and cognitive and behavior skills training to help individuals understand and better manage anxiety.

The cognitive strategies include identifying anxiety symptoms and somatic symptoms related to anxiety, developing realistic thinking, and problem-solving. Behavioral strategies consist of relaxation and stress management techniques as well as gradual exposure to the feared situation to modify the anxiety response through systematic desensitization and habituation.

Many children experience improvement in their anxiety with CBT alone and do not require medication.

Medications help decrease overall anxiety symptoms and panic response and should be considered if the anxiety is moderate to severe.

SSRIs have been approved for treating children who have depression and OCD. SSRIs do not have United States Food and Drug Administration (FDA) approval for the treatment of anxiety disorders. However, considerable evidence supports their efficacy.

CBT in combination with an SSRI is recommended for moderate-to- severe anxiety disorders. Randomized, controlled trials of SSRI medications (sertraline, fluoxetine, fluvoxamine, and paroxetine) show effectiveness in treating children and adolescents who have anxiety disorders.

Initial doses should be minimal, with very gradual increases, as tolerated, according to symptom response and adverse effects, remembering that the medications take several weeks to start working and improvement continues for several months at a given dose

Suicidal ideation and suicide attempts are not as common in childhood anxiety disorders as in depressive disorders but they may occur.

Post Traumatic Stress Disorder

Posttraumatic stress disorder (PTSD) is an intense and disabling response to a traumatic event.

Acute PTSD occurs 1 to <3 months following the event, whereas chronic PTSD persists for 3 months or longer. The symptoms fall primarily into three clusters.

The first includes reexperiencing the event. This reaction manifests clinically as distressing memories, nightmares, or the more extreme “flashbacks” to the event itself. During reexposure (either from internal cues such as reexperiencing in the mind, or external, such as visiting the

scene of the accident), the person suffers from intense anxiety and distress.

The second cluster can, to some extent, be seen as a reaction to the first. This response is characterized by avoidance of reexposure to the trauma and numbing to the psychological responses the exposure creates, including a more generalized emotional numbing.

The third symptom cluster involves a constant hyperalert state. The person may have difficulty falling or staying asleep, may have an increased startle response, may have more trouble concentrating, and may be more generally irritable or angry.

Approximately 70% of children have had exposure to a traumatic event before age 16 years; however, despite the large amount of exposure, the 6- month prevalence of PTSD in these patients is ~3.7% for boys and 6.3% for girls.

Protective factors against PTSD include, in descending order of importance connections to positive caretakers, feelings of self-worth, feelings of hope, importance to others, talents valued by others, religious faith, and socioeconomic advantage.

Specific risk factors for developing PTSD also exist. These include: Lack of resilience or negative prior life experiences, which would include a history of other traumatic exposures, preexisting psychological problems, and substance abuse

Nearly 100% of children who see a parent being killed or sexually assaulted acquire PTSD. This observation is important because it appears that in children, threat to a parent is equivalent to threat to self.

PTSD develops in 90% of sexually abused children, 77% of children who see a school shooting, and 35% who witness violence around their home.

Dissociation, a psychological mechanism whereby a person attempts to protect him or her by separating from reality, was found to be strongly predictive of posttraumatic symptoms.

Resources available to a child after a traumatic exposure are critical. The most important of these is an attentive parent or guardian; thus, attending to parents' mental health can actually be the most effective intervention in aiding the child's recovery.

Treatment for PTSD is complicated and multifactorial.

There is some evidence that selective serotonin reuptake inhibitors (SSRIs) have some utility. Additionally, SSRIs can lead to activation

(irritability, poor attention, and sleep disturbance), which can mimic the hyperarousal cluster of PTSD symptoms.

Clonidine was found to decrease basal heart rate, anxiety, impulsivity, and PTSD hyperarousal.

Propranolol also was found to decrease reexperiencing and hyperarousal. Opiates may be beneficial, but should be used with caution. Even controlling for subjective pain, one study found that there was a significant linear association between morphine dosage and 6- month reduction in PTSD symptoms.

Neuroleptic agents, such as risperidone, have shown mixed results. Benzodiazepines have not been found to be beneficial in treating PTSD- specific symptoms, and may worsen some symptoms characterized primarily by disinhibition (aggression, acting out).

In children, the strongest empirical evidence to date supports Trauma- Focused Cognitive Behavior Therapy (TF-CBT).

CBT begins with the child directly discussing the traumatic event, and then proceeds to anxiety-management techniques, such as relaxation and correction of distorted trauma-related thoughts. Through this procedure, the children learn that they do not have to be afraid of their memories. CBT often is accompanied by psychological education and parental involvement.

Research shows that the better parents cope with the trauma and the more they support their children, the better their children will function. Therefore, it is important for parents to seek treatment for themselves so as to develop the necessary coping skills that will help their children.

Obsessive-compulsive disorder (OCD):

It is an anxiety disorder involving obsessions (distressing intrusive thoughts or images) and/or compulsions (repetitive behaviors or rituals performed to relieve distress and anxiety associated with the obsessions) that are unwanted, cause substantial anxiety, and interfere with functioning (consuming more than 1 hour per day).

The most common obsession themes are illness, contamination and danger-related.

The most common compulsions are cleaning, washing rituals and checking behaviors.

OCD does not always involve observed compulsions, and the individual can suffer from repetitive images or thoughts (e.g., of violent, religious, or sexual nature) that are extremely distressing.

Children who have OCD generally have poor insight into their illness and may not recognize the obsessions and compulsions as irrational. They may have trouble going to school, find they are unable to concentrate in class, have difficulty getting out of the house or getting dressed, and have decreased food intake related to obsessions and compulsions.

Etiologies Genetic

Taken together, family, twin, and adoption studies make a strong case for genetic influences operating in the transmission of major depression.
Adoption studies document up to an eight-fold increase in depression and a fifteen-fold increase in suicide among the biological relatives of adoptees who have affective illness.
Despite strong associations of heritability, one third to one half of children develop depression in the absence of any family history of depressive illness.
Genetic risk factors seem to play a greater role in adolescent depression than in prepubertal depression, with the most potent risk factor being parental depression.

Biochemical factors

Endocrine studies implicate several different neurotransmitter systems in depression, including the noradrenergic, serotonergic, cholinergic, and dopaminergic systems.
Abnormal levels of these neurotransmitters (and their metabolites) in blood, cerebrospinal fluid, and urine as well as measures of platelet receptor functioning have been detected in depressed patients.
Several other abnormalities have been associated. These include cortisol hypersecretion, dexamethasone nonsuppression, and hyposecretion of growth hormone in response to an insulin challenge, hypersecretion of growth hormone during sleep, functional and structural brain imaging changes, and sleep electroencephalographic abnormalities.

Environmental factors

Stressful events can precipitate depression in children who have no identifiable constitutional susceptibility.
Studies of depressed adults and adolescents show significantly more stressful life events or losses in the year prior to the onset of depression.

Low self-esteem and perceived worthlessness also play an important role in the development and perpetuation of depression.
Other factors include abuse or neglect, parental substance abuse, low socio-economic status and education level, stress related to adolescent development or issues of sexuality.

c.

Mood and affect disorders
Clinical features, presentation

Depressive disorders may present a variety of symptoms. They can be sad, irritable, or angry and may present with school or behavioral problems.

They can demonstrate somatic complaints (headache, stomachache, and muscle weakness), decreased or increased appetite, fatigue, insomnia, hypersomnia, or disturbed sleep-wake cycles. Some children can develop psychotic features, such as paranoid delusions or auditory hallucinations.

Some develop self-injurious behaviors or suicidal ideation, plan, and intent. Anxiety symptoms may be present and may predate the development of depressive symptoms. Behavioral problems ranging from oppositional defiance to frank conduct disorder may occur in conjunction with an underlying mood disorder.

Normal adolescents with depression can present with a variety of intense moodiness, impulsivity, and erratic behavior. The biologic correlates of depression include sleep issues and change in appetite .

Epidemiology:

Prevalence rates for depression range from 1% to 2% of prepubertal children to 3% to 8% of adolescents.

Depression in prepubertal children and bipolar disorder in any age group are equally common in both sexes. However, unipolar depressive disorders in adolescents are more common in girls than in boys (ratio of 3:1). Early onset of puberty in girls increases the risk for depression.

There is a wide variability in the presentation of childhood bipolar disorder.

Acting out and oppositional behaviors are more likely to be in youth with depression.

Etiologies

There is an association of depression with chronic illness. These can include illnesses like cystic fibrosis, juvenile diabetes, epilepsy, inflammatory bowel disease, sickle cell anemia, organ transplantation, and cancer.

There is an association between depression and substance use/abuse.

Anxiety disorders often coexist with depressive disorders. There is also an association between anger/hostility and anxiety and depression in adolescents' depression and substance abuse are more common in teens with sexual orientation issues (eg, gay, bisexual, transgender).

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Comorbid disorders in depressed children and adolescents include anxiety disorders, attention-deficit/hyperactivity disorder (ADHD), substance abuse, and conduct disorder.

Screening and diagnostic evaluation

It is important to distinguish between a major depressive disorder, dysthymia, brief grief reactions, and adjustment disorder with depressed mood.

The DSM IV diagnosis of major depression requires at least 2 weeks of predominantly depressed, bored, or irritable mood and an additional four or more depressive symptoms (eg, sleep disturbance, decreased interest in pleasure, guilt, hopelessness) that together cause significant distress or impaired functioning.

Dysthymic disorder is manifested by a depressed or irritable mood for most of the day, for more days than not, for at least 1 year.

Unlike major depression, dysthymia is milder, requiring only two of the following symptoms: appetite and sleep disturbance, poor energy, poor concentration, and hopelessness.

However, because of the chronic nature of the disorder, it can interfere with a child's peer relations, attachments to caretakers, and development of social skills. Children who have dysthymia are also at increased risk for the development of subsequent mood disorders.

Adjustment disorders are manifested by depressive symptoms (not enough to fulfill criteria for depression) and precipitated by maladaption to common forms of psychosocial stress (eg, loss of a loved one, family conflict, failure or rejection at school).

Adjustment disorders do not predict later dysfunction; they have a good short- term prognosis, with a median episode length of 7 months and a 97% recovery rate.

Bereavement resembles an adjustment disorder in that it is associated with a stressful event. "Normal" bereavement is the reaction to the loss of a loved one. For children, it can lead to a temporary impairment in school and social functioning.

The reaction to the loss can be so intense that it fulfills criteria for major depression, but bereavement is not considered a disorder unless these symptoms persist for more than 2 months and cause significant impairment or dysfunction.

Uncomplicated bereavement gradually diminishes with time, and previous levels of functioning are restored.

Supplementation of the clinical history with self-report questionnaires can be enormously helpful, particularly in the primary care setting. Examples of such instruments include the Children's Depression Inventory (CDI), which assesses the severity of depression in prepubertal school-age children.

Beck Depression Inventory (BDI), Reynolds Adolescent Depression Scale (RADS-2), and the Mood and Feelings Questionnaire (MFQ) (9) are screening tools for older adolescents.

The Guidelines for Adolescent Depression in Primary Care (GLAD-PC) Toolkit also has child and parent report measures and scoring instructions on their web site.

Therapeutic options

Empirically validated treatments for depression include psychopharmacologic medications (either alone or in combination with supportive psychotherapy), cognitive behavioral therapy (CBT), and interpersonal therapy (IPT).

Antidepressant medications have demonstrated efficacy in the treatment of depression in children and adolescents.

Fluoxetine should be the first choice for medication management of depressive symptoms in children and adolescents.

CBT has a common therapeutic format that includes:

Psychoeducation; behavioral modification involving active scheduling of enjoyable activities; cognitive restructuring to reframe distortions and allow for new, more rational thoughts; and learning self-relaxation techniques; Most studies of CBT have suggested that it is an extremely effective intervention for depressive disorders in children and adolescents.

IPT centers on the problems in the patient's interpersonal interactions, seeking to change conflicted, hostile relationships into supportive, meaningful, and satisfying ones. The focus of the therapy usually is on development of social skills and investigation of the effect of shifting roles within family or peer groups, including adjustment to such events as parental divorce or remarriage.

Suicidal behavior

Self-poisoning after 6 years of age is not likely to be accidental.

Asking a child or adolescent about suicidal thoughts or actions will not "put such ideas into his/her head."

The warnings signs of suicide can include isolation from friends, giving things away, self-inflicted harm, etc.

Publicity regarding suicide may prompt other adolescents to attempt suicide. Vulnerable adolescents, particularly those who are depressed, become suicidal in the face of intense loss. Cluster formation can pose a true public health dilemma because too much media coverage (television, internet) can contribute to the contagious or imitative risk.

Epidemiology of mortality due to suicide:

Suicide is the third leading cause of death in the United States among adolescents and young adults ages 10 to 24 years.

From 2003 to 2004 the rate increased to 7.32 per 100,000. This increase reflects upward trends in three groups: females ages 10 to 14 years and 15 to 19 years and males ages 15 to 19 years.

Hanging/suffocation was the most common method of suicide in females in all age groups and firearms the most common method used by males.

There are ethnic disparities within these age groups: Native American/Alaskan Native youth and Hispanic females have a higher percentage of suicide attempts than their peers.

Few prepubertal children successfully commit suicide (suicide rate 0.6 per 100,000 for ages 5 to 14 years), but these children are at high risk for suicide attempts in their adolescence and young adulthood.

Epidemiology of suicide attempts:

% of children/adolescents considered suicide and 8.4% had attempted suicide within the past year. Both mood and anxiety disorders increase the likelihood of suicidal ideation. In prepubertal children, suicidal ideation can be

associated with disruptive disorders.

In adolescents, substance abuse and separation anxiety may predispose to such ideation.

Suicidal "gestures," such as superficial cutting or ingestion of a few extra pills, must be taken seriously. Any episode of self-inflicted harm may be a sign of attempted suicide. Homosexual adolescents are at risk for suicide.

Risk factors for suicide in children and adolescents include: mood disorder, substance abuse, loss of a loved one, family discord, social isolation, family history of suicidal behavior, previous suicide attempt, and availability of firearms.

How to assess a child or adolescent with suicidal ideation:

The following questionnaire helps in assessment of suicidal ideation:

Do you have thoughts of death or dying?
Do you wish you were dead?
Do you believe that things would be better if you were dead?
Do you have any intent to kill yourself or any plan to do so?
If you do have a plan, what is it?
Do you have the means necessary to carry out your plan?
Have you ever tried to kill yourself or hurt yourself before?

The indications for hospitalization of a child or adolescent at risk of suicide include actively suicidal children.

Factors that can be protective against suicidal behavior include religion, school engagement, family connectedness, coping strategies and reduction of firearms in the home.

Psychotic behavior, thought disorders

According to the DSM-IV diagnostic criteria for schizophrenia, at least two of the following symptoms should be present for a significant period of time during a 1- month period:

Delusions
Hallucinations
Disorganized speech
Grossly disorganized or catatonic behavior
Negative symptoms such as flat affect and paucity of thought or speech

Only one symptom is needed if the delusions are bizarre, hallucinations include a voice providing running commentary on the person's behavior or thinking, or two or more voices are conversing with each other.

Another criterion is the deterioration of social, occupational, and self-care functioning below the level achieved before onset.

For children and adolescents, this includes the failure to achieve age-appropriate levels of interpersonal, academic, or occupational development. The disturbances also must be present for at least 6 months.

Disorders of attention and impulse control

Clinical features, presentation

The prevalence rate of ADHD is higher in boys than in girls.

The spectrum of symptoms that can occur with ADHD subtypes include inattention, impulsivity and hyperactivity.

ADHD is difficult to diagnose in the early years of life.

ADHD-combined type reaches its peak prevalence of identification in the early elementary school years, but ADHD-inattentive subtype may not be identified until later in the school career.

75% to 85% of affected children continue to have symptoms of impulsivity and low attention into teenage years and adulthood.

Individuals who have ADHD show decreased hyperactivity as they get older, but adolescents may have more immature interactions with their peers.

Longitudinal studies have found elevated anxiety or mood disorders among adults who have ADHD, and males have decreased ability to handle stress.

Studies have found higher rates of divorce among adults who have ADHD compared with the general population.

However, many individuals who have ADHD can compensate for their weaknesses and attain above-average achievement.

The differential diagnosis of a child presenting with behavior problems in school include:

Developmental disorders: language disorder, learning disability, autism spectrum disorders

Medical disorders: anemia, lead intoxication, seizure disorder, substance abuse, prematurity, fetal alcohol syndrome, Tourette syndrome, sleep apnea

Genetic disorders: Klinefelter syndrome, Fragile X, Turner syndrome, Williams syndrome, Inborn errors of metabolism

Psychiatric disorders: adjustment disorders, mood disorders, oppositional defiant disorders and posttraumatic stress disorder

The most common presentation of ADHD in preschool children is problems with hyperactivity and impulse control.

Etiologies

Coexisting conditions like oppositional defiant disorder, conduct disorder, anxiety, depression, learning disabilities are frequently seen in children with ADHD

The neurochemical basis of ADHD:

> Neuroimaging studies have shown structural and functional differences in areas of the brains in patients who have ADHD compared with patients who have no ADHD.
> Certain regions of the brain, rich in dopaminergic and noradrenergic pathways and associated with executive function, seem to be particularly affected, including the prefrontal cortex, striatum, and cerebellum.
> Functional magnetic resonance imaging (MRI) studies suggest that these particular regions of the brain are less activated in children who have ADHD compared with those of age-matched controls during activities that measure executive function, a cognitive process of the frontal lobe that is used to solve problems or achieve a goal.
> Other studies, using brain MRI, have found reductions in prefrontal, frontal, and hemispheric volumes in individuals who have ADHD compared with controls.

Screening and diagnostic evaluation

The diagnosis of ADHD cannot be made by use of a specific test. Observation of behavior

in a physician's office does not usually reflect the situation at school.

Diagnostic studies in the evaluation of disorders of attention include laboratory, neuroimaging, psychoeducational testing, and continuous performance tests.

Although rating scales are not diagnostic in isolation, they help obtain standardized information from both parents and teachers.

Behavioral rating forms are used to document core symptoms of specific behavioral disorders for an individual child compared with his or her peers.

The Vanderibilt is an attention-deficit/hyperactivity disorder (ADHD)-specific rating form for children in the elementary school years that also provide information about comorbid symptoms such as oppositional defiant disorder. However, there are no normative data for adolescents.

The Conners is another ADHD-specific rating scale that has normative data for ages 6 to 18 years; a separate version is valid for children 2 to 6 years of age.

It is important to request information on symptoms and impairment from two or more settings in a patient with ADHD.

Patients with CNS-based chronic conditions like epilepsy, myelomeningocele and lead poisoning are at increased risk of ADHD

Therapeutic options

> The first-line agents for treatment of ADHD are stimulant medications. Stimulant medications improve attention in normal individuals as well as in children with ADHD. More than 80% of children respond to this class of medications with improvements in their symptoms. The generic forms are methylphenidate and amphetamines.

Other medications that are used include nonstimulants such as atomoxetine and guanfacine.
Atomoxetine is considered a second-line drug because it is less effective in treating ADHD than stimulants.
Medication alone is not sufficient for the treatment of ADHD.
It is important to communicate with the teachers of a child with ADHD when medications are used.

The medications used in treating ADHD include:

Stimulants:

Methylphenidates (MPH): Ritalin(MPH), Concerta(MPH), Metadate(MPH), Daytrana (MPH) and Focalin(dex MPH) Amphetamines (AMP): Dexedrine(dextro AMP), Vyvanse(lisdex AMP dimesylate), Adderal(mixed AMP)

Nonstimulants:

Strattera (Atomexitine)

The beneficial effects of ADHD medications: Children treated with stimulants show improvement in attention to task and decrease in impulsivity and hyperactivity. Stimulants also may improve parent-child interactions, reduce aggressive behavior, and improve a child's academic productivity and accuracy. The effect on academic performance is less strong.

The most common adverse effects of stimulants are decreased appetite, abdominal pain, headaches, irritability, and sleep problems. Gastrointestinal effects and headaches may be lessened if the medication is taken with food.

Less common adverse effects include weight loss, “rebound” effects, tics, social withdrawal, and affective changes. Rebound refers to temporary worsening of symptoms (irritability, increased activity, or mood swings) when the medication wears off. Administering a low dose of an immediate-release (short-acting) stimulant at this time may be helpful.

Social withdrawal, lethargy, or restricted affect may be a result of overdosing. Rare adverse effects include psychotic behavior that may present as hallucinations or mania.

Special caution is recommended before using stimulant medications in children or adolescents who have pre-existing cardiovascular disease or symptoms suggesting cardiovascular disease.

Potential for drug dependency exists. Avoid abrupt discontinuation in patients who have been on medication for prolonged period.

Contraindications to the use of stimulants include hypersensitivity to any component, idiosyncratic reactions to sympathomimetic amines, marked anxiety or agitation, use within or during 14 days of treatment with a MAO inhibitor, famiy history or diagnosis of Tourette’s syndrome or tics.

Some of the management strategies for a child with ADHD and its coexisting conditions include:behavioral management strategies, special education

placement, tutoring, cognitive monitoring strategies, psychotherapy and hypnosis.

The medical indications for the use of stimulant medications outside of school hours are mainly to prevent rebound effect. Administering a low dose of an immediate-release (short-acting) stimulant at this time may be helpful.

Classroom accommodations for children with ADHD may by implemented under Section 504 of the Rehabilitation Act (504 Plan) or under the Individuals with Education Disabilities (IED) Act.

Psychosocial Issues and Problems

Family and environmental issues

General issues

Anticipatory guidance reduces subsequent problems in critical life events. Accurate and effective information, encouragement to express emotions and helping facilitate communication between patients, caregivers and family members is vital for support during such events.

Regressive behavior and somatic complaints are commonly observed in children who are stressed.

Parental-focused positive reinforcement is critical throughout childhood and adolescence.

While transitioning their patient to adult care, a pediatrician can help their patients' families by discussing and sending a summary of patient's medical history and the care being received to the adult physician before the first visit. Pediatrician should assure and indicate willingness to be available for further consultations during the transition period.

Critical life events

Divorce

1.) The developmental stage of a child will have an effect on how he/she responds to divorce and to be a blended family.

2.) Divorce could result in either joint or sole custody accompanied by visitation by either parent. This could affect the emotional development of child that in

turn influences their social interactions, schooling and development process.

3.) A child's emotional adjustment to divorce may play a role in how they perceive their subsequent intimate relationships.

Death

1.) The developmental stage of a child will have an effect on how he/she responds to a death in the family.

2.) Pediatrician should provide support for child and family who had experienced death of a loved one by encouraging open discussion of reactions, thoughts, and feelings in the family. Age-specific and culturally sensitive guidance should be provided accordingly.

3.) A child who had lost a loved one goes through a multitude of emotions including shock, anger, denial, disbelief and sadness.

4.) Life-threatening or terminal illness results in family members responding initially with shock and grief, followed by a denial stage, anger and grievance period that eventually results in fear of death.

Sudden infant death

1.) Use of home monitors are costly and come with a social stigmatism and takes an emotional toll on family members resulting in increased depression and hostility during the initial period.

2.) Parents who experienced unexpected death of an infant due to SIDS need to becounseled and educated about the back-to-sleep program avoiding soft bedding. Maternal cigarette smoking pre and post pregnancy is a risk factorand siblings of SIDS victims have a 5 to 6 fold increased risk of dying of SIDS.

Impact of mass media

Adverse effects of watching TV on children include promoting aggressive and violent behavior, obscuring distinction between fantasy and reality, trivializing sex, increasing obesity and risk for suicidal behavior.

Young children up to 6 years of age spend, on average, two hours every day watching TV or other screen media. Older children spend around 7 hours a day on entertainment media. Entertainment media including TV should be avoided for children under age 2 and children over age 2 should be limited to 1 to 2

hours per day of high quality content. "Screen-free" zones with no TVs, computers, etc., need to be established in kids' bedrooms.

Children are vulnerable to commercial advertising on TV as they are unable to critically evaluate the content. This greatly influences their purchases

of toys, cereals, etc.

Socioeconomic factors

Adoption

Pediatrician should play a vital role in adoption by

verifying medical records to determine any pre-disposed or underlying genetic conditions a child may have. Immunization records and medical problems need to be addressed accordingly.

Foster care

Children in foster care have special health care needs as they are more prone to physical, mental, dental, developmental and psychosocial problems

Foster care children evaluation criteria should include formal mental health, developmental and educational evaluations, initial screening within 3 days and comprehensive assessment within 30 days of placement.

Pediatricians dealing with children in foster care should educate and engage child agencies, foster and biological parents and youth in developing an integral health plan addressing special health care needs. This plan should address behavioral and emotional issues related to trauma and confused loyalties, treat significant sleep disorders, support children through transitions and attachment issues, educate positive parenting strategies and reduce conflict or coercion by birth parents.

Foster care children need a medical home that may involve birth and/or foster parents, case workers and caregivers providing care coordination and case management.

Child welfare and foster care systems have the ultimate goal of ensuring the well- being, safety and permanency of children with their families. States have the primary responsibility of child welfare services. With the federal support,

the states fund these systems and oversee legislative actions. Suspected child abuse is brought to the attention of Child Protection Services whose caseworkers investigate the situation ensuing filing of a court petition if abuse or neglect has been substantiated. Child could then be placed in foster care, reunited with family, given custody to a relative, or given for permanent legal adoption with termination of biological parents' rights, as deemed appropriate.

Placement instability of children in foster care due to behavioral issues can worsen and the behavior can cause extended developmental and behavioral problems. Addressing theseproblems can improve the placement stability and lead to child's permanent placement.

Youth aging out of foster care would be in need of garnering social support and developing independent living skills including arranging for their own accommodation, employment, food, insurance, etc.These young adults are at a greater risk for welfare dependency, homelessness, crime, incarceration, teenage pregnancy and substance abuse.

Discipline

Effective discipline involves a positive and supportive parent-child relationship and includes positive reinforcements to increase desired behavior and removal of reinforcement or punishment to reduce undesired behaviors. Common strategies include time out, time in, "catch 'em when they're good," and verbal reprimands.

Cultural issues in medical care

Specific problems, conditions

Enuresis

Primary enuresis is urinary incontinence in a child who has never achieved dryness while secondary enuresis is incontinence in a child who has been dry for at least 6 months.

Urinary incontinence that exists during day time is diurnal and one that exists nighttime is nocturnal.

Etiological factors for nocturnal enuresis include developmental delays, organic illness, psychological distress, genetic and gender factors. The prevalence is greater in boys than girls.

Nocturnal enuresis is prevalent among school-age children.

Events such as the birth of sibling, moves and/or stress could lead to regressive bed-wetting.

Secondary diurnal enuresis

necessitates prompt referral to a pediatric urologist.

Nocturnal enuresis is frequently associated with a family history.

Laboratory studies for enuresis are unlikely to be positive unless additional clinical findings are present.

Nocturnal enuresis can be treated by enuresis alarm, pharmacotherapy with imipramine, DDAVP therapy, hypnotherapy and counseling.

Parents need to be counseled about coping with enuresis without causing psychological problems.

Incontinence secondary to environmental stress, a "resistant" child, urgency incontinence are additional reasons for diurnal enuresis.

Diurnal enuresis can be treated by hypnotherapy, bladder-training exercises and counseling.
Pharmacologic treatment of enuresis has limited efficacy and can lead to adverse effects. DDAVP administration cannot achieve dryness unless combined with alarm therapy and is associated with seizures due to water intoxication. Imipramine therapy has a high relapse rate when discontinued and an adverse effect profile including significant risk of death with overdose.
Encopresis
Encopresis is the voluntary or involuntary passage of stools due to stool retention while Hirschsprung's disease (HD) is a blockage of the large intestine due to improper muscle movement in the bowel. Encopresis is observed in children older than 4 years of agewhile HD exists since birth. Encopresis is associated with stool in the rectum while HD has no stool in the rectum.
Encopresis is a symptom rather than a developmental variation after the age of 4 to 5 years. Bowel movement restriction by a child causes stool retention in the colon and rectum. When the colon gets filled with impacted stool, liquid stool can leak out of the anus resulting in the soiling of clothes.
Constipation with dry, hard stool, large stool passage that clogs the toilet, delay and avoidance of bowel movement, stool leakage in child's underwear, UTIs and abdominal pain are symptoms of stool retention
Encopresis can be associated with urinary tract infections resulting from incomplete emptying of the bladder and may also lead to enuresis due to pressure the bladder.
Common causes of encopresis include constipation leading to intentional withholding of stools, emotional stress, and in some cases organic diseases like Hirschsprung disease, neurological and metabolic disorders.
Treatment options for encopresis include colon clearance of the retained stool with stool softeners, colon lubricants, rectal suppositories, enemas or oral fluids followed by positive reinforcement encouraging healthy bowel movements. Psychotherapy may be useful in some cases.
Encouraging a high fiber diet with increased fluid intake, stool softeners, positive reinforcement encouraging healthy bowel movements are ideal for managing stool withholding during toilet training.
Intentional withholding of stools can be treated by cleaning stool with bowel irrigation or stool softeners. Emotional stress related encopresis can be managed by psychotherapy and positive reinforcement.
Medical therapy, behavioral modification, and counseling when combined results in greater efficacy of encopresis management.
Delayed bowel training is observed during toilet training years with bowel dysfunction.
Psychosomatic disorders
Conversion reaction is a condition normally observed when a psychological emotional problem translates to a physical one. They could result from physical/sexual abuse or any stress-related incidents. Seizures, motor or gait dysfunctions, sensory complications are accompanied. Management should focus on identifying the stressors and model of the symptoms, primary gain of underlying conflict, secondary gain and hysterical personality.

This may also produce a condition in which the person is unconcerned about the disability arising from their symptoms, called la belle indifference.
Psychosomatic disorders are always a result of underlying stress related factors.
Psychosomatic complaints are stress or emotion related and the resolution of the emotion that underlies the physical symptom is the child's primary gain, and the change in the child's social or occupational stance resulting from the symptom is secondary gain.
Chronic pain syndrome is present with severe joint pain, muscle spasms and motor dysfunctions and may be accompanied with chest pain and/or palpitations, respiratory distress and urinary frequency and/or retention. Seizures, syncope, gait and sensory issues, abdominal pain may be observed as well.
Psychosomatic disorders need to be investigated with a cost effective strategy with focused medical and psychological evaluation so as to determine the underlying stressors.
Treatment options for psychosomatic disorders starts off with reassurance that there is no underlying organic disease; this may be followed by psychotherapy and/or physical therapy, hypnosis and anti-anxiety or antidepressant treatment on a need basis.
Differential diagnosis of conversion symptoms includes psychophysiology, hypochondriasis, malingering and somatic delusions.
Academic activities relating to school attendance and absenteeism are important to assess with every complaint of recurrent pain.
Psychosomatic disorders may be manifestations of parental anxiety and/orparental pressure for a child to perform.
Sibling rivalry
Sibling rivalry is common and parents who are apprehensive about that may be counseled appropriately.
Therapeutic plan for an aggressive sibling rivalry needs to focus on recognizing the differences between each child and creating an environment that helps them get along with each other. Arrange for fun family activities promoting teamwork, teach positive ways to gain attention, allocate time for individual attention, avoid comparisons and let them have their own space when appropriate.
Separation anxiety and school refusal
Separation anxiety is often a precursor to school phobia or refusal.
It is developmentally normal in the preschool and during the initial school months of kindergarten or first grade.
It is more prevalent in children from single parent homes and

may be triggered when in cases of a new sibling, physical move or family stress.

School absence related to separation anxiety is accompanied by a severe emotional distress to attend school, child preferring to stay with parents and parents being aware of the situation while that related to truancy involves child concealing absenteeism from parents, frequent antisocial behavior and not conforming to academic and behavior expectations.
Plan for abnormal separation anxiety disorder should include psychoeducation, positive reinforcement, individual and family behavioral therapy, including cognitive behavioral methods.
Maternal depression has a negative impact on behavioral, social and cognitive behavior of a child.

Sleep disorders

Sleep is of two types; non-rapid eye-movement (NREM) sleep and rapid eye- movement (REM) sleep and in a sleep episode, NREM and REM sleep alternate cyclically. Newborns do not have a regular sleep pattern and sleep around 16 – 18 hrs daily in discontinuous episodes, the longest lasting 2.5 - 4 hours. Over first year, their sleep matures into 9 - 13 hour blocks with single naps. Sleep reduces by 2 hours from age 2 through 5 (from 13 to 11 hours). Children usually discontinue napping between 3 - 5 years.

Children, in the first 12 months have sleeping/waking patterns influenced by feed timings.

Nightmares occur during REM sleep followed by waking up by 3 - 5 year olds where children respond to consoling. Sleep walking or talking is also seen in age groups 4 through 8 that can be managed by reassurance and timed awakenings.

Night waking starts around 9 months of age when children start experiencing separation anxiety.

It is important to alleviate parental concern towards infant sleep disorder. The child's sleep pattern and parent's reaction to those need to be understood to educate the parents about infant sleep patterns, reinforce night awakenings and identify appropriate parent responses and expectations.

Sleep disorders are common in childhood and are observed in about 1/5ths to 1/3rds of children.

Nightmares are scary dreams occurring during REM sleep followed by awakening where children are responsive to consoling and have recollection of the dream. Night terrors occur within 2 hours of sleep and are associated with a screaming and rapidly breathing child being non responsive to consoling and having no recollection in the mornings after.

A plan to manage an infant with inappropriate sleep patterns should include establishing sleep routines, avoiding stimulating activity during sleep times, not

removing the baby from crib during night awakenings and soothing them back to sleep.

A plan for managing a child with bedtime refusal or frequent awakening should include setting sleep timings and enforcing them and not yielding to the child for their tantrums or crying to avoid sleep times.

Management of night terrors involves educating the family about the issue and reassurance. It is important to make the child's room safe to protect from injury during thrashing behaviors, eliminating sources of sleep disturbance and maintaining a consistent sleep schedule.

Vulnerable child syndrome

Factors that predispose a child to the vulnerable child syndrome (VCS) include serious illness, hospitalization, or survival of a life-threatening event by a child and history of miscarriages, infertility, or maternal illness during pregnancy.

Health condition, real or perceived can cause parental anxiety about the child's well-being. This results in parents treating child as vulnerable, distorting the parental perception of child's health and affecting the normal parent-child relationship.

Anticipatory guidance to avoid VCS involves identifying and educating the families at risk for predisposed factors. Parents of sick children should have clear communication regarding the issue and treatment without undermining or exaggerating the severity.

Families need to be counseled about normal child behavior and development and unwarranted parental anxiety needs to be addressed.

Rumination and cyclical vomiting

Non organic causes of vomiting include rumination (associated with passive regurgitation of gastric contents into the mouth) and cyclic vomiting (typical of recurrent nausea episodes and vomiting without any medical causes). These non- organic causes are difficult to diagnose and differ from organic ones that are related to a specific medical issue.

Rumination can be managed by behavioral therapy, negative reinforcement or stimulating methods such as bitter taste on tongue.

Pain

Pain can cause sensory, psychological and behavioral suffering and the child's response and tolerance to pain could be a function of their developmental age.

Pain in children is treated based on the severity and the developmental stage. It can be managed by non-drug methods like heat or cold use, massage, rest. NSAIDs, antidepressants, epidural analgesia, patient-controlled analgesia may be helpful when appropriate.

Gifted child

Gifted child often attracts attention and requires more resources that could cause extra pressure for siblings, parents and other family members. Parents can play a vital role in identification and development of their gifted children but could also

feel unprepared to understand the nature of giftedness or raise the "exceptional" child.

Gifted children should be raised appropriately. Activities and education above age level can be encouraged when apt but focus should also be placed on developing normal social, behavioral routines without stressing only on the gifted child’s extraordinary talents and excusing them from ordinary behavioral expectations.

Chronic illness and handicapping conditions

Grief reaction in parents with children with handicapping condition is common. Different stages of grief are Denial, Anger, Bargainging, Depression and Acceptance [mneumonic: DABDA].

These stages can occur in any order.

Methods of treatment for handicapping conditions includes visual training, megavitamins, patterning [is a series of exercises designed to improve

the “neurologic organization” of a child's neurologic impairments] are INAFFECTIVE.

Effect of a chronic illness on siblings include:

guilt, anger, embarrassment, anxiety

Siblings may feel that their own needs are not being acknowledged or fulfilled, feelings of neglect which may trigger guilt towards their own good health and resentment towards their parents and ill sibling. Younger siblings may wish that the affected sibling would die so that they can get their parents back.

There is an increased child abuse among handicapped children (about 11 times higher than normal children).

Psychosocial factors are associated with but do not cause chronic illness (eg, asthma, seizures, inflammatory bowel disease).

Chronic illnesses are usually associated with high financial costs which affect the marriage and family economics. Frequent medical visits and hospitalizations can interfere with parental employment and undermine job performance and security. Areas of marriage that may be impacted: finances, self-esteem, sexuality, spirituality, social life, future planning, parenting style, recreation.

Chronically ill or handicapped child may progress to normal adult behavior including separation from their parents and can have emerging sexuality inspite of

their chronic illness. The role of pediatrician in making parents understand this is very important.

It is important to understand the psychosocial effects associated with the use of home medical equipment (eg, oxygen monitors, physical therapy, transportation, hygiene). Families experience an impact on the quality of life and mental health. Depression, anxiety, insecurity about child's future are common. Parents may feel inadequate to the task of caring, they can experience social isolation, fatigue, and emotional exhaustion.

It is very important to be supportive and nonthreatening while talking with parents whose children have chronic diseases .Appropriate ethical decisions relating to children with chronic and handicapping diseases depends on the best interest of the children.

Monitor growth and development in patients following all types of transplantation.

Psychosocial and family issues associated with all types of transplantation include depression, anxiety, high financial costs, nutrition issues, non compliance, and emotional functioning.

It is important to provide medical home for children with chronic conditions (eg, registry, medical summary, care plan, transition plan).

Violence

Family violence

Circumstances that may indicate intimate-partner violence include depression, substance use/abuse, and physical injuries.

Possible effects of intimate-partner violence on children arephysical abuse, injury while protecting mother, injury from assault directed at mother, learned aggression, post- traumatic stress disorder, and hypervigilance.

Common characteristics of violent children and adolescents are exposure to corporal punishment and intimate-partner violence, perception that the world is hostile, little awareness of options for conflict resolution, poor peer relations, and impulsiveness.

The important precipitants of violence by batterers are pregnancy, efforts by women to leave the home, initiation of separation or divorce, etc.

Societal violence

Abuse and neglect

Child abuse

Epidemiology

Consider the possibility of abuse or neglect in a child with failure to thrive.

A related caregiver is the abuser of a child in 90% of child abuse cases.

The siblings of abused children are at increased risk of abuse.

Intimate-partner violence frequently accompanies child abuse.

Neglect is the most common form of child abuse.

Etiology

The origins of child abuse are:

Child stresses (eg, handicap, hyperactivity), social/situational stresses (eg, poverty, isolation, family discord, multiple births, parent-child conflicts), and parent stress (eg, abused as a child, depression, substance abuse). Abusive and neglectful parents often have severely unrealistic expectations for their

children's behavior.

Signs and symptoms

Distribution of injuries

Fractures of ribs, scapulae, and sternum are rarely accidental. Difference

between cutaneous signs of physical abuse and accidental injury are human bites, usually >2.5 cm in diameter, have central or ovoid pattern and central

echymosis.

In contrast animal bites leave puncture marks or torn flesh.

Bruise over chest and abdomen mainly in a non-ambulatory child is highly suspicious of abuse.

Delay in seeking care, false histories of no trauma or of minor illness given by caretakers at the time of presentation should raise suspicion for physical abuse.

Distinguish between the physical findings of inflicted and accidental burns.

The pattern of inflicted burns frequently is that of immersion and involves the perineum or the extremities with well-demarcated borders, "stocking" and "glove" pattern of hands and feet.

Patterns that indicate accidental burns like "pouring" or "flowing" of a hot medium over the skin or splash marks.

Chemical burns from contact or ingestion occurring when an unattended child gains access to caustic household solutions should be evaluated for neglect.

Contact burns can have varied patterns (eg, curling iron, clothes iron, and cigarette) and may be accidental, although a careful history must evaluate for risk factors for abuse or supervision neglect.

Skin conditions such as staphylococcal or bullous impetigo, staphylococcal scalded skin syndrome, herpes, contact dermatitis, and toxic epidermal necrolysis can mimic burns

The most common fracture locations and types in physically abused children are:

Bruises

Staging of bruises: Development of bruises depends on skin color, depth of injury, location. Generally bruises are considered older if they are yellow, green, and brown.

Distinguish between cutaneous signs of physical abuse and of nonabusive skin conditions (eg, mongolian spot, coining, cupping, urticarial pigmentosa). Cutaneous signs of abuse are like that mentioned above however in abusive conditions need to be documented properly to avoid misidentification.

Injuries in children that are infrequently indicative of physical abuse are dislocated elbow, clavicular fracture, toddler, fracture of the tibia.

Ingestions

Ingestions may be manifestations of child abuse.

Fractures

It is very important to distinguish between inflicted fractures and conditions such as osteogenesis imperfecta, hypophosphatasia, infantile cortical hyperostosis, and osteoid osteoma .

Normal variants of bone mistaken for abuse include nutrient canals, cortical irregularities, metaphyseal beaks and spurs, distal ulnar cupping, normal symmetric peritoneal changes seen in infants, and ossification defects of the ribs. The role of a bone survey for fractures in suspected child abuse is very important as it can provide the age of the suspected fracture. It consists of radiographs of the skull, lateral cervical spine, ribs, pelvis, thoracolumbar spine, arms, hands, legs, and feet. Anteroposterior and lateral views should be obtained to examine all aspects of the bones fully. A repeat survey 2 weeks later can be helpful to identify healing fractures that exhibit new bone formation.

Fractures are present in a minority of physical abuse.

The chip fracture of metaphysis is commonly due to wrenching or pulling injuries.

A radionuclide bone scan can reveal subtle areas of skeletal trauma that may not be seen on plain-film radiograph studies of bones

Shaken infant syndrome

A skeletal survey is very important in a child with a subdural hematoma as intracranial hemorrhage in an infant or young child is highly correlated with physical abuse.

Abuse is the most common cause of serious intracranial injuries during the first year after birth.

In the absence of signs of cutaneous trauma it is important to recognize shaking as a possible cause of coma in infants.

Retinal examination must be done to identify retinal hemorrhage in suspected head trauma due to shaking.

Treatment, legal issues

Sexual abuse should be reported to law enforcement and must be reported to a state child protection agency by law.

Physicians are legally obligated to report suspected abuse under the state laws.

An investigation of unsubstantiated cases of child abuse produces stress in a family.

An unsubstantiated report/finding by a child protection agency does not necessarily mean that abuse or neglect did not occur.

The standard of proof in a civil court is the preponderance of evidence (ie, a lesser standard than in criminal proceedings re child abuse]

The problems associated with foster home placement includes : continued risk of child abuse , behavioral issues including risk taking behaviors, antisocial behavior, health care needs, mental health issues.

There is a need for a team approach in the management of child abuse which includes: Pediatrician knowledgeable and skilled in child maltreatment, hospital social worker, pediatric nurse, psychologist or psychiatrist, data coordinator.

Be aware of intervention options for families involved in child abuse are Respite care or "crisis nurseries," therapeutic preschools, temporary receiving or foster homes, and "parenting classes".

The role of a child advocate (guardian ad litem) in legal proceedings appointed by the court is very important and represents the child's legal best interest.

Circumstances that can lead to failure to substantiate child abuse: failure to locate child; failure to locate parents; parents' refusal to speak to investigators; duplicate reports; child's refusal to repeat history; non-English speaking family.

Recognize that many abused and neglected children are not removed from their parents or placed in foster care.

Neglect

Neglect of safety and neglect of medical care are forms of abuse.

Excessive quietness, compliance, or apathy in an infant or toddler may be a symptom of neglect or abuse.

Factitious disorder (Munchausen syndrome) by proxy [MSBP]

The signs of factitious disorder (Munchausen syndrome) by proxy:

recurrent sepsis from injecting fluids, chronic diarrhea from laxatives, false renal stones from pebbles, fever from heating thermometer, rashes from trauma, sugar or blood in the urine, etc.

Munchausen syndrome by proxy refers to the production of signs or symptoms of factitious illness in the child by the caregiver.

The morbidity and mortality rate is high for this type of child abuse.

The features of the parent of a child with factitious disorder (Munchausen syndrome) by proxy are: usually mothers, the perpetrator usually has a distant or uninvolved partner ,usually have at least some training in a health profession. They usually are very pleasant, cooperative, and appreciative of medical staff. Approximately 72% of women who perpetrate MSBP display abnormal illness behaviors themselves, including somatoform disorder and factitious disorder. Adults diagnosed with factitious disorder are more likely to abuse their children through MSBP than are controls.

Children with factitious disorder (Munchausen syndrome) by proxy may exhibit significant ongoing psychologic problems including severe withdrawal, paranoia, preoccupation with bodily integrity and vulnerability, and chronic invalidism.

The components of a management plan for a patient with factitious disorder (Munchausen syndrome) by proxy: Child protection authorities and the legal system should be engaged immediately. Arrange for foster placement to assure safety if child victim needs it. Both perpetrators and victims require psychiatric evaluation and follow-up. The primary concern is the safety of the patient.

First step: observe the relationship between the occurrence of symptoms or signs and the presence or absence of the caregiver. In some cases, such as suspected airway compromise, acute life-threatening events, repeated poisoning, or suspected failure to give medication, the use of covert video surveillance has been proven to be useful, although its use is controversial.

When MSBP is proven or strongly suspected:

The infant should be removed to a safe environment and child welfare authorities, local law enforcement, and hospital security should be notified as applicable. It is neither safe nor effective to confront the parent in the child's presence without ensuring the child's safety and the safety of the medical team. Psychiatry consultation and social services referral both are indicated and helpful, but should be deferred until the child's safety is assured.

Sexual abuse

Epidemiology

Age and gender distribution of sexual abuse:

About 30% of sexual abuse victims are under 6 years old, 30% are 6 to 12 years old, and the rest are 12 to 18 years old.

75% of reported victims are female and male victims are less likely to report.

97% of reported offenders are male. Females are more often offenders in child care settings, including babysitting.

Sexual abuse by stepfathers is nearly 5 times higher than by natural fathers. The absence of a protective parent from the home increases the risk of sexual abuse.

Most perpetrators of sexual abuse are known by the child before the abuse occurs.

History

Sexual abuse should be considered as the cause of physical symptoms such as vaginal, penile or rectal pain; discharge, bruising, erythema, or bleeding; chronic dysuria, enuresis, constipation or encopresis.

The techniques for obtaining information from a child suspected of being sexually abused.

The child, in another room, should complete a drawing series that can be used during the interview. This will put the child at ease and provide indirect entry to the topic of sexual abuse.

Find out how the child refers to his or her private part by showing the nude figure drawings appropriately matched to the child's skin color, race, and age to determine what words are used to identify body parts and whether the child recognizes "private parts" and what to do or say if something inappropriate happens to any body part. In young children, anatomically correct dolls may be substituted. They are best used in the hands of a trained interviewer. An open- ended question about private parts is, "What would you do or say if something happened to a private part or if someone else asked you to do something to their private part?" If the child says, "I would say no," the next question should be "Have you ever had to say no?" The interview proceeds to determine what happened, where things happened, the identity of the perpetrator(s), frequency, and bribes and threats used. The types of abuse and their frequency are used by law enforcement to determine the charges and number of counts. Questions and answers in quotes should be recorded in the interview report. Some jurisdictions allow or require videotaped recordings of the interview.

It is important to know which physical complaints can lead to a diagnosis of sexual abuse in young children, e.g.: genital discharge, genital pain.

In a suspected sexual abuse, the first detailed interview of a child is diagnostically critical. An explicit description and imitation of adult sexual behavior by children may indicate either victimization or observation of sexual acts (not fantasy).

Repetitive interviewing of an allegedly sexually abused child should be avoided as it creates rote quality to responses, increases likelihood of leading questions, increases chance of "learned" responses, is unnecessarily stressful, increases

chances of inconsistency/retraction.

The advantages of anatomically correct dolls for interviewing are the child who is nonverbal can point. The disadvantage is risk of overinterpretation .The drawing of genitalia by a child may indicate sexual abuse. When sexual abuse is suspected, the child should be interviewed alone.

Verbatim statements by a child may qualify as evidence in a criminal court.

Physical examination

Recognize physical findings consistent with sexual abuse

Female:

Hymen with new or healed lacerations or transactions, absence, or remnants; U or V shaped indentations in posterior or lateral portions of the hymen, Posterior fourchette lacerations, vaginal wall tears, perianal lacerations.

Acute findings: lacerations, abrasions, ecchymoses, edema.

Presence of sperm, semen, and pregnancy (in child prior to the age of consent). Suggestive findings: bite marks on the genitals or inner thigh or scarring or tears of the labia minora.

Straddle injuries result in trauma of labia and clitoris and rarely involve hymen.

Male:

Bite mark must be differentiated from laceration from zipper, forceful retraction of the foreskin may result in a dehiscence of the tissues ,non-specific transitory redness of penis.

Anal exam:

Abnormal anal findings are unsual.

Acute: lacerations, edema, abrasions, ecchymoses, fissures that come out past the anal verge.

Chronic: scars, dilation 20mm or more without rectal feces, altered anal contour.

Labial adhesions, vulvar erythema, and anal tags are not signs of abuse. Physical examination for suspected sexual abuse:

A chaperone should be present during the exam to rule out acute and chronic illness and evidence of medical or dental neglect and physical abuse.

The anal and female genital examinations are accomplished best with good lighting, magnification, and the ability to photograph findings. On rare occasions, sedation of the child with monitoring is necessary. A colposcope is used in referral centers. A digital camera with macro capabilities should record normal and abnormal findings adequately.

Girls generally are examined in the frog leg position. The labia are grasped by an assistant's gloved hands and pulled forward and laterally to reveal the hymen. The fourchette, urethra, and hymen walls should be observed. Findings should be recorded on an anatomic form. In the older female, it may be necessary to examine the hymen edge with a moistened cotton swab or with a Foley catheter inserted through the hymen opening that is inflated and pulled back against the hymen wall to detect abnormalities. Bimanual and speculum examinations are performed on adolescent females. A speculum is not used with preadolescent females. The speculum size should be as small as possible. The anal examination is external. Particular attention is paid to the opening size, presence of stool, symmetry of folds, and signs of old or new tissue injury. Midline skin tags are normal. The anus is considered to be dilated if the opening is more than 2 cm and no stool is present in the ampulla. The male genitalia rarely are injured in sexual abuse and may be injured nonintentionally by zippers and toilet seats. Accidental injury of the hymen is unusual.

Laboratory findings

The presence of a sexually transmitted disease may indicate sexual abuse

A chlamydial infection may be acquired from the mother at birth and persist Microbiologic tests are helpful in documenting suspected sexual abuse:

They include tests for HIV ,hep B, syphilis, Chlamydia and gonorrhea [culture or nucleic amplification test].

Forensic evidence

Sexually transmitted disease in a prepubertal child is presumptive evidence of sexual abuse only "gold standard" tests to diagnose sexually transmitted diseases in children are used because of the legal issues involved . Only

standard culture systems for the isolation of N. gonorrhoeae and C. trachomatis should be used

Evidence of seminal fluid is infrequently found in sexually abused children Seminal fluid is unlikely to be found/persist beyond 72 hours in a sexually abused child

Management

Sexual abuse can recur even when families are receiving treatment.

The current recommendation for antibiotic prophylaxis for prepubertal and adolescent sexual assault victims:

Neisseria gonorrhoeae

Children and adolescents: Ceftriaxone 125mg IM in a single dose
ORChildren >45kg and adolescents: Ceftriaxone 250mg IM x1 for adolescents and adults

Chlamydia trachomatis

Children <45kg: Azithromycin 20mg/kg (max 1g) PO in a single dose
ORErythromycin base/Ethylsuccinate 50mg/kg per day divided into 4 doses for 14 days
Children >45kg and adolescents: Azithromycin 1 g PO in a single dose
Doxycycline 100 mg PO bid for 7 days (if at least 8 yr and not pregnant)

Trichomoniasis and bacterial vaginosis:

Children <45kg: Metronidazole 15mg/kg per day (max 2g) PO divided into 3 doses for 7 days.
Children >45kg and adolescents: Metronidazole 2 g PO in a single dose

It is important not to assign blame to the victim in helping families cope with sexual abuse. All victims need psychological support.

Critical Care

Recognition of impending systemic failure
General

Hypotension is a late finding in shock and represents a loss of compensatory mechanisms.

Prolonged capillary filling time in a sick patient may indicate decreased cardiac output and compensatory peripheral vasoconstriction - both signs of shock.

Increase in respiratory rate is a sign of impending coma.

Invasive bacterial infections such as sepsis and bacterial meningitis are often associated with core temperatures greater than 41 degrees Celsius.

Malignant hyperthermia may occur during or after general anesthesia. It a hypermetabolic state caused by inhaled anesthetic agents and succinylcholine. There may be a family history of issues with anesthesia or a personal history of Duchenne muscular dystrophy, Noonan syndrome, or other myopathies.

Environmental temperatures may influence capillary refill time by causing peripheral vasoconstriction or vasodilation to maintain core body temperature.

Central nervous system

A unilateral dilated pupil is a sign of uncal herniation. The uncal portion of the temporal lobe is particularly sensitive to lateral compressive forces, which cause a CN III palsy.

Compensatory tachypnea, such as in respiratory disease, reflects an increase in respiratory rate. Hyperventilation may refer to increased respiratory rate, depth or both such that carbon dioxide levels are decreased.

Although brain death is a clinical diagnosis, confirmatory testing is recommended for all children less than 1 year. An EEG that shows 30 minutes of electrocerebral silence of a cerebral blood flow scan that shows absence of uptake are consistent with brain death.

Brain death is a primarily clinical diagnosis. In children below one year of age and those whose clinical exam is equivocal, an EEG with electrocerebral silence over 30 minutes and a cerebral perfusion scan without uptake are consistent with brain death.

Respiratory

Signs and symptoms of impending respiratory failure include increased work of breathing, dyspnea, grunting, flaring, retracting, decreased pH and increased CO2.

Signs of severe airway obstruction include stridor, wheezing, apnea, hypoxia, and hypercarbia.

Cardiac

Cardiogenic shock may present in the early phase (compensated) with increased heart rate and systemic vascular pressure. If allowed to progress, patients may deteriorate into late phase (decompensated) with poor perfusion in the periphery and viscera, decreased vascular pressure, respiratory failure, or altered mental status.

Renal

For patients with hypertensive emergency, intravenous therapy is recommended using sodium nitroprusside, labetalol or nicardipine. The goal is to reduce the blood pressure to the 95th percentile for age, sex, weight and height. A reasonable goal is to reduce the blood pressure 10% in the first hour of therapy, and 15% in the next 12 hours.

Prerenal azotemia is due to hypoperfusion of the kidney in conditions such as shock, hemorrhage or renal artery stenosis. The BUN:Cr ratio will be >20 and the fractional excretion of sodium will be >1%. Renal azotemia is typically caused by intrinsic renal disease, such as acute tubular necrosis or glomerulonephritis. The BUN:Cr ratio is <15 with serum uremia.

Hepatic

Signs and symptoms of impending hepatic failure include fever, nausea, vomiting, abdominal pain, jaundice, fetor hepaticus, ascites, bleeding diathesis, or irritability.

Electrolytes

Adrenal insufficiency is characterized by hyponatremia, hyperkalemia, hypoglycemia, relatively low cortisol and aldosterone, elevated ACTH, elevated renin, and ketosis and hyperpigmentation in older children. Patients may present with weakness, anorexia, vomiting or orthostatic hypotension.

Adrenal insufficiency and the syndrome of inappropriate antidiuretic hormone both feature hyponatremia. Adrenal insufficiency is also characterized by hyperkalemia, hypoglycemia, relatively low cortisol and aldosterone, elevated ACTH, elevated renin, and ketosis and hyperpigmentation in older children. Patients may present with weakness, anorexia, vomiting or orthostatic hypotension. SIADH features elevated urine osmolality, increase urine sodium, low BUN and uric acid and normal adrenal

function. Patients present with the symptoms of hypokalemia, such as nausea, vomiting, weakness and seizures.

Water intoxication in an infant typically occurs by feeding a baby free water. It presents primarily with symptoms of hyponatremia, such as lethargy, vomiting, seizures, Cheyne- respirations, or rhabdomyolysis.

Skin

A child with purpura and possible sepsis should be assumed to have meningococcemia caused by N. meningititis and started on intravenous penicillin or ceftriaxone until culture and sensitivities are available. Vancomycin may be added to the regimen due to concerns about resistance.

Emergency life support
General

Initial evaluation of an accident victim should assess the ABCDEs. Airway and cervical spine should be stabilized and secured immediately. Breathing must be determined to be effective. Circulation may be affected by primary cardiac causes or shock. Deficit refers to neurologic deficit, which is often present in children due to the initial trauma or resulting shock. Exposure is needed to complete a physical exam to search for causes and sequela of the trauma.

Airway and respiratory

A general guideline for the selection of endotracheal tube size is to add four to the age of the patient in years and divide the sum by four.

Positive End Expiratory Pressure (PEEP) is beneficial in a patient with pulmonary edema because the positive airway pressure prevents alveolar collapse, increases functional residual capacity, and decreases pulmonary vascular resistance.

Cardiac and circulatory (shock)

Cardiopulmonary resuscitation is ideally performed with at least two operators in a medical setting with bag and mask or endotracheal tube in place. In neonates, compressions to breaths should be a ratio of 3:1, with compressions at a depth of ½ the chest diameter with the hands encircling the chest and compressing with both thumbs in the center of the chest in the nipple line. In children and adolescents, compressions to breaths should be a ratio of 15:2, with compressions at a depth of at least 2 inches with the heel of one hand supported by the other in the center of the chest in the nipple line. The rate of compressions in all age groups should be at least 100 per minute. Breath volume should cause chest rise.

Initial management of septic shock should be with fluid resuscitation and broad spectrum antibiotics based on patient risk factors and local antibiograms. If these measures do not cause

significant clinical improvement, inotropes may be necessary. Cardiogenic shock however, may worsen with aggressive fluid resuscitation.

Initial therapy of hypovolemic or septic shock should include rapid administration of a normal saline bolus of 20cc/kg. Up to 80cc/kg may be given over the first hour with the goal of normalizing blood pressure and perfusion. If unstable after this volume, inotropic agents should be considered.

A bone marrow needle may occasionally be needed to administer fluid intraosseously to a patient in shock where peripheral access cannot be obtained quickly or several sites would be needed for the volume of fluid being administered.

Common conditions requiring emergency life support
Airway, chest
Upper airway obstruction

Foreign body aspiration and asthma may both present with coughing, increased work of breathing and the first episode of wheezing. In foreign body aspiration, a chest radiograph may show unilateral hyperinflation, as the right mainstem bronchus is the most common location; asthma will be bilateral. After initial episode of respiratory distress, a foreign body may have an asymptomatic phase, where reflexes fatigue, after which erosion or infection may develop, whereas in asthma the acute episode typically does not subside without treatment.

Pneumonia, bronchiolitis
Burns

Airway injury with an acute burn can present with blackened or swollen mucosa, decreased aeration, wheezing or crackles and abnormalities on blood gas.

Near-drowning

Hypothermia associated with a near drowning should be managed by removing the wet and using external heat sources, such as dry blankets and warming pads. Warmed oxygen may be given. In severe cases, warmed peritoneal dialysis, colonic flushes or bypass may be used. If CPR is needed, efforts should be continued until the core body temperature normalizes, even if efforts are unsuccessful prior to this.

The mechanism of cerebral edema in an asphyxiated patient is not well understood. Presentation is the same as other causes of cerebral edema and includes change in mental status, ataxia, seizures, dizziness, vomiting, and irregular respirations.

The best factor to predict the prognosis in submersion injury is the neurologic exam 24-72 hours after presentation. Children who are responsive on the scene with normal sinus rhythm and reactive pupils have tended to have good outcomes. Other factors include the duration of submersion and duration of resuscitation.

Hemothorax, flail chest

A flail chest is a segment of ribs that are broken in more than one place such that the broken rib segments are not connected with the remainder of the rib cage. Because of this, the motion is paradoxical to the rib cage during respiration. There is often lung contusion in the corresponding lung tissue and extreme pain with respiration.

Respiratory distress

Acute respiratory distress syndrome after shock is non-cardiogenic pulmonary edema due to permeable alveolar capillaries that causes fluid collection in the alveolar and interstitial spaces. The permeability is thought to be due to circulating inflammatory markers. Tachycardia, dyspnea and tachypnea are early signs. Exam may reveal crackles, which are typically in the peripheral, rather than central lung fields. Chest radiograph may reveal patchy atelectasis and air bronchograms.

Cardiac and circulatory

Cardiac failure occurs when the cardiac output cannot meet the needs of the tissues to maintain perfusion. Infants may present with decreased volume at feeds, sweating, fatigue, or dyspnea. Children may complain of fatigue, dyspnea, cough, weight gain, or exercise intolerance. Adolescents may have primarily abdominal complaints. Exam may reveal an S3 gallop, tachycardia, JVD, hepatomegaly, tachypnea, rales or wheezing, increased work of breathing, weak cry, edema, or poor weight gain. Chest radiograph will show cardiomegaly and bilateral perihilar fluffy infiltrates from pulmonary edema. An EKG will also demonstrate cardiomegaly, as well as decreased cardiac output. Serum BNP may also be elevated.

First line acute treatment of congestive heart failure is with intravenous diuretics. Afterload reduction with ACE inhibitors and Angiotension II Receptor blockers may be added as needed.

Complete heart block may present with syncope, sudden death, prominent pulses, elevated systolic blood pressure, or systolic murmur. Younger children may take frequent naps due to fatigue or have night terrors. EKG will show a complete dissociation between the P waves and QRS complexes.

Initial therapy for paroxysmal atrial tachycardia includes vagal maneuvers for older children. For infants, an ice bag may be placed on the face. Ocular pressure is not

recommended. If physical measures are not successful, adenosine may be used in stable patients.

Pericardial tamponade is caused by a rapid accumulation of fluid in the pericardium, such that the pericardial sac cannot stretch to accommodate the fluid. Signs and symptoms include distant heart sounds, pulsus paradoxus, tachycardia, dyspnea, JVD and hypotension. An EKG may show low voltage.

Long QT Syndrome may present as syncope. The prolonged QT interval is vulnerable for a re-entry tachycardia, frequently Torsades de Pointes. EKG may reveal a prolonged QT interval, T wave alternans, or notched T waves. Because it may be caused by a number of different gene mutations, patients may have no associated medical issues or be affected by autism, neurological deafness, syndactyly, micrognathia, or congenital heart defects.

Abdomen

An acute "surgical abdomen" may present in several different ways depending on the pathology. The patient may lie very still or writhe in pain. Fever can indicate an infectious etiology. Hypotension may be related to sepsis or hypovolemia, while hypertension may be reactive due to pain. Most etiologies involve abdominal pain, although the character and location depends on the affected organ. There may be abdominal distention, hypoactive bowel sounds, guarding and/or rebound tenderness.

A CT scan in acute abdominal trauma is a fast test that is sensitive and specific for spleen, liver, and kidney injuries in stable children. It is less useful for diaphragm, pancreas and intestinal pathology. Oral contrast is indicated if a bowel injury is suspected.

Initial evaluation of a patient with probable splenic rupture includes a thorough history of the trauma and physical exam. Exam may demonstrate abdominal pain or left shoulder pain. If there has been significant blood loss, the patient may present in hypovolemic shock. An abdominal CT scan with intravenous contrast in a stable patient can demonstrate the extent of injury.

Emergency Care

Fever

Fever without localizing signs have differential diagnoses that vary according to the age of the patient. Neonates are at risk for serious bacterial infections including sepsis and meningitis due to GBS, E.coli, L. monocytogenes, HSV, and enterovirus. Infants 1 to 36 months old are at risk of bacteremia and UTI.

The definition of fever is a rectal temperature greater than 38°C. Body temperature may be measured orally, axillary or rectally. Rectal temperatures should not be taken in neutropenic patients or those with diseases with rectal involvement.

A fever less than 39°C does not require treatment. Above this temperature, children generally become uncomfortable and may be treated. Acetaminophen may be given to children of all ages at a dose of 10-15mg/kg every 4 to 6 hours. Ibuprofen may be given to infants greater than six months at a dose of 10mg/kg every 6 hours.

Body temperature has a normal range of 36.6-37.9°C, with the lowest point at waking and highest in the early evening.

Seizures

Initial therapy for status epilepticus may include intravenous or rectal benzodiazepines, intravenous barbiturates or intravenous phenytoin.

The loading doses for anticonvulsants are: lorazepam IV: 0.05-0.1mg/kg, midazolam IV: 0.2mg/kg, diazepam IV: 0.2-0.5mg/kg, diazepam rectal: 0.2-0.5mg/kg, fosphenytoin IV: 15-20 PE, phenobarbital IV: 5-20mg/kg.

Wounds

General

A laceration through the vermilion border of the lip requires closure by a skilled physician, as a step off as little as 1mm can result in a poor cosmetic outcome.

Most wounds may be cleansed with irrigation using tap water or saline to decrease bacterial load and clear debris. Irrigation with antiseptic washes has not been shown to decrease bacterial counts any further than normal saline.

In a patient with a fever and swollen foot and fever after a puncture wound through a sneaker, pseudomonal infection should be suspected. Management should be done with surgical exploration and debridement under general anesthesia, followed by antibiotic therapy for 14 days.

Tetanus immune globulins should be given to patients who are not fully immunized against tetanus who have anything but an uncomplicated minor wound. This includes but is not limited to animal bites, human bites, dirty wounds, burns and crush wounds.

Puncture wounds, such as those due to animal bites, typically cause pain and little bleeding due to the small skin surface area that is disrupted. If the wound is through a nail, there may be a collection of subungual blood. If infected, the area will become erythematous and may have drainage or palpable swelling in the deeper tissues.

Due to the small surface area, irrigation of puncture wounds may be more difficult, leading to infection. Drainage of fluid may also be inhibited due to inflammation and small surface area. Surgical drainage may be necessary in these cases, as well as in nail injuries, where blood and pus may collect under the nail surface.

Depending on the cause of the puncture wound, parts of the causative object may break off under the skin and remain in the wound, such as with glass, pencils or animal teeth or claws. Because a puncture wound may extend further into the soft tissues than is suggested by the size of the wound on the skin surface, bacteria may be drawn into deep tissues and cause bacterial spread to deep tissues and bones. These infections may be relatively asymptomatic at first due to lack of erythema or drainage on the surface.

Lacerations should be cleaned with irrigation and explored for foreign bodies. Debridement may be necessary. Most wounds that present within 6 hours of injury can be closed primarily in the ED or office with single layer closure using simple interrupted sutures. Deeper lacerations may need a multiple layer closure. The wound should be bandaged and kept moist with a topical antibiotic ointment. Infection occurs in approximately 3-7% of uncomplicated lacerations. Other complications include poor cosmesis and loss of skin sensation due to severed cutaneous nerves.

Bites and stings (see also III.D.4.)
Rabies protocol

Rabies prophylaxis is not required for domestic animal bites where the animal can be observed for signs of disease for 10 days. Rodent bites rarely require prophylaxis, but local public health officials should be consulted.

Domestic animals should be quarantined and observed for 10 days for emerging signs and symptoms of rabies.

Dog, cat bite

Prophylactic antibiotics should be given to dog and cat bite victims if the wound is puncture, in the hand, close to a joint or in an immunocompromised host. Augmentin is the drug of choice.

Snake bite

Most snake bites in the United States are from non-venomous species and require only local wound care. Patients with venomous snake bites should have immediate assessment of respiratory and cardiovascular systems, with intravenous access established in a non-affected limb. Baseline labs, including coagulation studies, should be obtained. The affected area should be marked and frequently reassessed. If the patient meets criteria, antivenim should be initiated.

Spider bite

Management of necrotizing spider bites, such as the Brown Recluse, is with local wound care. Management of neurotoxic spider bites, such as the Black Widow, is with antivenim and supportive measures.

Human bite
Insect sting

Life threatening reactions to Hymenoptera stings include hypotension, wheezing, laryngeal edema, and delayed serum sickness.

Immunotherapy with insect venom is 98% effective in preventing subsequent reactions to Hymenoptera stings in patients with anaphylaxis.

Patients younger than 16 years who experience large local reactions or generalized urticaria to insect stings do not require skin testing or desensitization to Hymenoptera.

Trauma
Abdominal trauma
Multisystem trauma
Head injuries

Immediate life threatening complications of closed head trauma include subdural and epidural bleeding and acute phase swelling resulting in brainstem herniation and loss of regulatory mechanisms.

A patient with a closed-head injury and brief loss of consciousness may have an epidural hematoma, frequently caused by interruption of the middle meningeal artery. A head CT may demonstrate this; however, this modality may be of limited benefit as compared to MRI for lesions in the posterior fossa.

Severe brain injury may be present in a patient who has no external signs of trauma.

Ecchymoses in the orbital area, or “raccoon eyes,” and hemotympanum battle and sign (postauricular bleeding) are indicative of a basilar skull fracture.

The initial assessment of a patient with a head injury should include stabilization of the cardiopulmonary systems. Once stable, a full neurological exam should be undertaken to assess level of consciousness or neurologic deficits. If serial examinations are indicated, the Glasgow Coma scale provides an objective measure of a patient's responsiveness, with scores for ocular, verbal and motor responsiveness. If a patient is sedated, sedation should be lifted to peform exams.

Signs of a progressive increase in intracranial pressure include headache, vomiting, change in mental status, CN III palsy, hypertension, irregular respirations, and bradycardia.

Radiograph study of the skull may be negative in a patient with significant intracranial injury and may be positive in a patient without a significant intracranial injury.

The mainstay of outpatient management of minor head trauma is with serial neurologic exams and counseling to avoid strenuous mental activity, such as screen time or school work, until neurologic exam improves. Return to sports should be graded and based on exam and symptoms. Investigative imaging is only warranted in patients with mental status changes, focal neurologic findings, loss of consciousness greater than 5 minutes or age below 2 years.

Retinal examination in a patient with seizures or in a coma may point to the mechanism of injury such as abusive head trauma. A few other situations, such as motor vehicle accidents, cause retinal hemorrhages with this clinical picture.

Papilledema may not be present initially and may develop later in the course of intracranial hypertension.

Drug and alcohol use is associated with head injury due to associated risk taking behaviors, but use may also complicate the exam of a patient with head injury.

Temporal bone fractures are commonly manifested by bleeding from the external auditory canal or hemotympanum, hearing loss, facial paralysis, and CSF otorrhea.

Signs and symptoms of closed head trauma depend on the severity of the injury and range from mild headache and dizziness to nausea and vomiting, change in mental status, seizures or loss of consciousness.

Initial management of a patient with acute CNS trauma includes assessment of cardiorespiratory status. Once deemed stable, a full history and physical exam with special attention to mechanism of injury and full neurological exam should be performed, with imaging by head CT as necessary. Further management depends on severity of injury. This may be as simple as outpatient monitoring for patients with mild concussive injury. However, those with severe injuries may require emergent intubation, intracranial pressure monitoring, measures to decrease ICP, surgical intervention and monitoring in an intensive care setting.

Burns

First degree and small second degree burns may be treated on an outpatient basis with topical silvadene or Bacitracin and a sterile dressing that is changed daily. Blisters should be left intact. Pain relief may be achieved with over the counter analgesia.

Minor electrical burns caused by chewing on a household electrical cord affect only the visible surfaces of the mouth that came into contact with the cord and may be treated like any other burn. Electrical burns caused by high voltage sources and lightning show cutaneous entry and exit sites. The current follows the path of least resistance and may affect any organ system in that path, regardless of visible injuries. Cardiac conduction abnormalities are an immediate concern.

A first degree burn affects only the epidermal layers and will be dry, erythemetous, and painful. Second degree burns affect the epidermis and upper layers of the dermis and will be moist, edematous, mottled and painful. Third degree burns affect the dermis and underlying tissues and will be dry with white tissue or eschar and painless.

Airway and cardiovascular status must be assessed immediately in burn patients. Aggressive fluid resuscitation with lactated ringers solution by the Parkland formula (4 x wt in kilos x %BSA) should be instituted. Half of the IV fluids should be given in the first 8 hours and the rest over the next 18 hours. Any offending agent should be removed from the skin and the skin should be irrigated. The wounds should be draped in a sterile fashion until fully assessed. Analgesia should not be delayed. Patients with burns over 15% of BSA, associated with inhalation injury, resulting from suspected abuse, affecting the face, perineum, hands or feet, or electrical burns should be hospitalized.

Fractures, dislocations 6.
Neurovascular states

Several fractures are of particular concern for injury to underlying vasculature, such as fractures of the supracondylar humerus, distal femur, and proximal tibia.

Specific problems (eg, spiral fracture, "nursemaid's" elbow)

A greenstick fracture occurs in the long bones of children due to their flexible nature. Typically following a blow, one side of the cortex stretches while the

opposite side buckles. On radiograph, the bone may show bowing or an area of increased density in the buckled area.

The typical patient with a subluxation of the radial head (nursemaid's elbow) is a toddler that is pulled suddenly by the hand or distal arm. They will present with the arm held close to the body in flexion and pronation, reluctant to move the limb.

Treatment for subluxation of the radial head (nursemaid's elbow) is to hold the head of the radius in one hand while using the other hand to flex and supinate the wrist.

Clavicular fracture may present as palpable deformity or pain with movement of the upper extremity after a blow to the shoulder or fall on an outstretched hand. It may also present as a palpable deformity or fussiness in a neonate, often one who is large for gestational age (LGA).

Treatment is immobilization with a figure eight sling and pain relief. Surgical correction is needed for open and comminuted fractures.

Acromioclavicular separation in an athlete occurs after a direct blow to the superior aspect of the joint or blow to the joint on an outstretched arm. There are four subtypes based on which ligament is disrupted. Patients will complain of point tenderness of the AC joint and may note the affected shoulder is inferior with a step off defect. Radiographs will reveal a widened joint space.

Patients with possible spinal cord injuries should be immobilized on a back board and cervical collar. The initial evaluation should assess cardiorespiratory status. Once stable, a modified neurologic exam should be performed and plain films of the spine should be obtained. Any focal findings on neurological exam should be further investigated with other imaging modalities as indicated, such as MRI.

Pharmacology

Pharmacodynamics
Absorption

Some drugs should be taken with food while others should be taken on an empty stomach.

Factors that influence bioequivalence of drugs include differences in manufacturing, preparation, and even lot number. Crystal form, particle size, and drug characteristics which are not tightly controlled during manufacture can affect drug absorption

Hepatic drug metabolism

Drugs may alter the elimination and half-life of many other drugs.

Premature infants have slow elimination of caffeine and theophylline, and levels should be monitored.

Drugs such as phenobarbital, carbamazepine, phenytoin, and rifampin may stimulate hepatic drug metabolism.

Drugs such as erythromycin, ciprofloxacin, cimetidine, and omeprazole may inhibit hepatic drug metabolism.

Renal excretion

Drug elimination rates in sick newborn infants are highly unpredictable.

Dosage requirements of renally excreted drugs such asaminoglycosides are highly unpredictable in infants. Monitoring of drug concentrations is indicated.

Newborn infants require adjustment of antibiotic doses for renally excreated drugs. Renal function demonstrates an age-dependent increase in glomerular filtration rate: Examples include: aminoglycosides, penicillins, and digoxin.

Half-life

At average doses, a drug which has a much longer than average half-life can accumulate to toxic levels. Three half-lives yield 90% of steady state.

Interpretation of drug concentration

Nonadherence is the major factor when drug concentrations or drug actions are highly variable in an adolescent.

Drug concentration measurement must be carefully timed in order to yield accurate results.

Drug concentration is best obtained during steady state or during the terminal log- linear phase of the concentration-time curve. At this point, the drug concentration at the site of action is expected toproportional to plasma concentration.

Drug pharmacokinetics are influenced by drug metabolism, drug excretion, and route of administration.

Adverse drug reactions

Idiosyncratic drug effects are abnormal reactions to chemicals specific to an drug.

Drug allergy and rash may be idiosyncratic reactions

Dose related drug effects are proportionately related to the concentration of drug in the body and the duration of the exposure to the drug.

Information on drug toxicity and adverse reactions are limited to information from a relatively small population during drug trials. Post-marketing reporting of adverse drug reactions to the FDA is important for determining the actual toxicity and adverse reactions from a drug.

Drug interactions

Concomitant administration of certain drugs can alter concentrations of other drugs in the patient's regimen.

There is a potential for interactions between drugs and alternative therapies such as herbs.

Specific drug classes

Antibiotics

Serum sickness is a Type III hypersensitivity reaction. Symptoms include fever, malaise, rashes, edema, myalgia, lymphadenopathy, arthralgia, arthritis, and gastrointestinal complaints. It typically occurs between 7 and 21 days after exposure. Serum sickness–like reactions from drugs are characterized by fever, pruritus, urticaria, and arthralgias. Symptoms begin 1-3 weeks after drug exposure. As the reaction progresses, the urticarial skin eruption becomes erythematous and forms into round plaques with dusky centers.

There is a risk for hypersensitivity reaction to an antibiotic.

There is a risk of ototoxicity or nephrotoxicity when vancomycin is given with other nephrotoxic agents, and is age dependent. .Aminoglycosides also pose the aforementioned risks when administered.

Diuretics

Ototoxicity and nephrotoxicity are potential adverse dose-related effects of furosemide.

Long-term complications of furosemide therapy include: hypokalemia, contraction alkalosis, and hyponatremia.

Corticosteroids

Patients who are on long – term corticosteroid therapy are at risk for adrenal crisis due to suppression of the hypothalamic – pituitary – adrenal axis. They require stress steroid dosing prior to surgery to prevent adrenal crisis.

Children on long – term corticosteroid therapy should receive stress dosing for trauma, surgery, significant burns and moderate to severe illness. Children receiving chronic systemic corticosteroids should be carefully monitored for signs of infection including fever and skin rashes. Varicella in particular has been known to cause serious illness and even death. It is important to note that typical indications of inflammation may be masked by steroid treatment. Patients should be tested for tuberculosis prior to beginning chronic corticosteroid treatment. Growth should be monitored and eyes should be examined twice yearly. Children should be on

calcium and vitamin D supplementation and participate in weight bearing exercise. Bone densitometry may be used to monitor oeteopenia.

Children who are on chronic corticosteroid treatment are at risk for: adrenal insufficiency, growth supression, decreased bone mineralization, pathologic fractures, immunosuppression, cataracts, diabetes, weight gain, acne, skin atrophy, hypertension, peptic ulcer disease, and pancreatitis.

Immunosuppressants

Long-term risks of chronic immunosuppression include: infection, neoplasm, growth suppression, osteopenia, thyroid dysfunction, hirsutism, dysthymia, tremors, myopathy, neurocognitive deficits, cardiomyopathy, hypertension, lipid abnormalities, atherosclerosis, delayed tooth eruption, prolonged primary tooth retention and increased dental carries.

Antihistamines

Common side effects of H1 antihistamines may include sedation, drying of the mouth and eyes, urinary retention, constipation, excitation, nervousness, palpitations, and tachycardia.

Beta-blocking drugs

Side effects of beta-blocking drugs may includebradycardia.

Anti-inflammatory drugs

Risks associated with the use of aspirin and nonsteroidal anti- inflammatory drugs include anaphalaxis, urticaria, angioedema, and rarely asthma, rhinoconjunctivitis.

H2-blocking drugs

Common side effects of H2 antihistamines have been known to cause myelosuppression.

Beta-agonists

Medications include albuterol (short-acting), salmeterol (long-acting).

Pain management

Conscious sedation

Consious sedation is often achieved with anxiolytics such aspentobarbital, and benzodiazepines.

Moderate sedation

Light sedation – use of anxiolysis and/or analgesia that obtunds consciousness but spares normal protective reflexes such as: cough, gag, swallow, hemodynamic reflexes.
Deep sedation – Sedation obtunds consciousness and either blunts or posses a risk of blunting normal protective reflexes.
General Anesthesia – Use of hypnotics,, sedation and analgesia resulting in the loss of normal protective reflexes.

Understand what level of observation and monitoring is recommended for a patient undergoing procedural sedation

Thorough medical history, anticipating underlying medical problems that predispose the patient to sedation problems
Careful physical examination focused on the cardiorespiratory system and airway
Appropriate fasting
Informed consent
Pediatric drug dosing (mg/kg)
Appropriately sized equipment
A separate, dedicated observer to monitor sedated patients who may have airway compromise (induced or pre-existing)
Documentation of vital signs and condition on a time-based record
Emergency backup system, “code” team, and “crash cart”
Fully equipped and staffed recovery area
Discharge criteria documenting recovery from sedation

Overdosage of sedatives may cause prolonged sedation, nausea, vomiting, ataxia, desaturation, respiratorydepression. Patients should be carefully monitored for symptoms of overdose.

Questions that may help the clinician understand the indications and contraindications for moderate sedation include:

Will this experience be painful, just distressing, or both? Is the patient especially anxious? Has he or she had a previous negative experience? Are the parents anxious?
Is it necessary for the patient to be perfectly still or is a little movement acceptable?
What are the best options for providing the intervention? Are medications the best option, is there a role for behavioral interventions, or can both be used for optimal results?
How should the patient be monitored and who should do so? What plans must be made for recovery?
What preparations should be made for adverse events?
There should be an appropriate interval of fasting before moderate sedation.

Sedative analgesia

Medications used for sedative analgesia include: opioids, nitrous oxide and ketamine.

Non-pharmacologic techniques

Non pharmacologic techniques, such as biofeedback, hypnosis and distraction may be used for sedation.

Research and Statistics

Study design

The validity hierarchy for study design and type from weakest to strongest: observational studies, experimental studies, and meta-analyses.

Randomized clinical trials – are the gold standard, are best for justifying causality and have the fewest problems due to bias. This is the best type of study to establish efficacy of treatment of procedure. Disadvantage of great expense and long duration.
Controlled clinical trials - Tendency to show positive outcomes compared to randomized clinical trials, especially when using historical controls. Also has a disadvantage of great expense and long duration.
Cohort studies – used to study causes of a condition, disease course, and risk factors. Limitations include: great cost due to long duration of study, and loss of patient follow-up.
Case-control studies – used to study rare diseases or events, conditions which develop over long time periods, and for investigation of a preliminary hypothesis. This study design is generally inexpensive and conducted over a short period of time. The disadvantages are: largest number of errors and biases compared to other study designs, dependence on previously existing records, difficulty selecting an appropriate control group.
Cross-sectional and longitudinal studies – used to evaluate status of a condition or disease, and to evaluate diagnostic procedures. This design is inexpensive and short in duration. Disadvantages include: data represents a moment in time, and may not be representative of population.

Systematic review and meta-analysis – combines published data to draw a general conclusion; the disadvantage is that the data from multiple types of studies should be reported separately.
Descriptive epidemiologic studies – easily written, hypothesis generating. The disadvantages of this study design are susceptibility to biases.
Case reports/series and anecdotal evidence – these are easily written, and the observations provide information for investigators designing studies to examine causes of the observations. The disadvantages are that they are susceptible to many biases.
The power of a study is (1-β). It is the probability of finding a difference when one truly exists. Low power can occur due to small sample size or small difference.
Small sample size may limit the ability to detect adverse events.
Cross sectional studies are most likely to yield valid information about the accuracy of a diagnostic test.
Clinical trials are most likely to yield valid information about the benefits and/or harms of an intervention.
Cohort studies are likely to yield valid information about the prognosis of a condition.

Data analysis

Validity indicates whether a test measures what it is intended to measure.
Reliability indicates whether the results of a test are reproducible.
Bias is a measurement error related to the ways that the target and sample populations differ. It may distort the estimate of the association between exposure and outcome and affect the validity of the study.
Confounding refers to the presence of nuisance variables which are likely to be present in one group and are related to the outcome of interest. These variables can confuse the interpretation of results.
Generalizability is the ability to apply the results of a study to a particular population.
Intention-to treat analysis means that results from all patients who entered the trial were included in the analysis of the arm to which the patient was randomized regardless of subsequent events. This may help to maintain the power of a study by analyzing a larger sample size.
Number-needed-to-treat (1/Absolute Risk Reduction) refers to the number of people who would need to be treated to prevent one event. It is useful to help the clinician to determine relative risks and benefits of therapeutic interventions.
Type I errors (α) – conclusion that there is a difference when no difference exists.
Type II errors (β) – conclusion that there is no difference when a difference exists
Study results may be affected by the data source. Examples include: diaries, billing data, and discharge diagnostic codes.

Reading and interpreting results

Incidence – rate of the proportion of people in a population who develop a disease or condition in a specific period of time.
Prevalence – proportion of people in a population who have a disease or condition at a specific point in time.

Pre-test probability – prevalence of disease prior to diagnostic procedure; Also referred to as the index of suspicion.
Post-test probability – predictive value of a positive or negative test; (True positives/all positives) or (True negatives/all negatives).
Positive predictive value is the probability of disease given a positive test result.
Negative predictive value is the probability of no disease given a negative test result.
Sensitivity is the ability of a test to detect disease if disease is present; best used to rule out disease.
Specificity is the ability of a test to yield a negative result if no disease is present; best used to rule in disease.
Standard deviation describes the spread of individual observations around the mean.
Standard error describes the mean of the spread of observations.
The confidence interval defines an upper and lower limit for the associated probability. It indicates how much the estimate would vary in repeated samples.
Likelihood ratio is the ratio of true positives to false positives. Used with the prior odds of disease, it determines the odds after a test is completed.
Relative risk is the incidence of disease in people with the risk factor to the incidence of people without the risk factor. (Experimental event rate/Control event rate).
Odds ratio is a measure of risk used particularly in case-control studies. It is the odds that a person with an adverse outcome was at risk divided by the odds that a person without an adverse outcome was at risk.
Statistical significance is an event which would occur by chance with a p value of less than or equal to 0.05. It is a rejection of the null hypothesis. Statistical significance does not necessarily translate to clinical significance.
A specific, clear, structured, searchable clinical question can be developed usingPICO. (Patient/Problem, Intervention of interest, Comparison, Outcome).

Ethics for Primary Pediatricians

Autonomy, beneficence, and rights
Critical care, end of life, and limitations on medical intervention
Decisions to withdraw/withhold life-sustaining medical intervention

Decisions should be made after careful consideration of all factors recognized by both family and medical staff including: medical likelihood of particular outcomes, burdens on the patient and family, religious and culture decision- making frameworks, and input by the patient when possible

Decisions should not be based on fear of litigation but based on what is thought to be best for the patient

Recognize and apply ethical principles when involved in end-of-life care Effective communication with families is paramount.

Always keep in mind what is best for the patient

Recognize and apply ethical principles with regard to limitations on medical intervention

There is no moral distinction between withholding or withdrawing interventions that are not medically indicated

Continually evaluate results of treatments and the evolution of illness to recognize whether interventions continue to be the best medical and moral choices

Decisions to withdraw/withhold artificial hydration/nutrition

Case law supports it being withheld in the setting of adult vegetative or permanently unconscious patient who can be shown to have previously expressed a wish not to be maintained in such a state

For infants and children unable to make such decisions, decision is based on what families and caregivers decide best supports comfort and on the overall condition/outlook for the patient

Cardiopulmonary resuscitation (CPR) and "do not resuscitate" (DNR) orders Decisions should be supportive of agreed-on goals of care should be effectively communicated to all caregivers

Orders should clarify the plan regarding mechanical ventilation, the use of cardiac medications, chest compressions, and cardioversion

The order may change over time as goals of care change balance benefits and burdens to the patient

If orders do not exist, a full resuscitation may not be required depending on the desired endpoint for the patient

Futility

The concept of futility is used to support the forgoing of life-sustaining medical treatment against the wishes of patients and families by holding that clinicians should not provide futile interventions

If a desired physiologic outcome will not be achieved with an intervention, this approach could support a medical goal of doing no harm

Persistent vegetative state

The use of long-term mechanical ventilation is likely unjustified, as there will be no net benefit to the patient

Palliative care and pain management

Children with cancer should have adequate pain management to provide comfort at all stages of his or her illness Intensive symptom control Establish, revisit, and revise goals of care

Consider the holistic nature of symptoms

Control pain with scheduled and as needed treatments, both pharmacologic and non-pharmacologic

The doctrine of double effect: A patient requiring increasing pain or sedation medications is not to be thought of as having intentional increased respiratory depression or death due to medications, as long as the side effects are not the intended consequences but rather are instituted for the comfort of the patient.

This doctrine also applies to withdrawal of life-sustaining medical treatment

Physician-assisted suicide and euthanasia

These are distinct from palliative care and pain management

Physician-assisted suicide is legal in the U.S. only in the stage of Oregon Euthanasia is illegal in the U.S.

Maternal/fetal conflicts

A clinician should not oppose a pregnant woman's refusal of a recommended intervention unless the risk to the pregnant woman is minimal, the intervention is clearly effective, and the harm to the fetus without the intervention would be certain, substantial, and irrevocable

The U.S. Supreme Court has held that drug testing of pregnant women without consent was a violation of the Fourth Amendment, which provides protection from unreasonable searches

Patient-parent-pediatrician relationship

Obligations: veracity, fidelity, and confidentiality

Veracity is the principle of truth telling violated by lying or by omission of information related to patient care and informed consent.

It is important to keep patients and families informed about care, which can be difficult if a patient or family does not want to hear the truth.

Recognize and apply ethical principles involved in the patient-parent-pediatrician relationship regarding issues of fidelity.

Fidelity is legally enforceable.Is related to patient confidentiality involves: keeping our promises, doing what is expected of us, performing our duties, and being trustworthy, treating others with basic respect, being competent and capable of performing the duties required of your professional role, adhering to a professional code of ethics, following the policies and procedures of your organization and applicable laws,honoring agreements made with the patient Recognize and apply ethical principles involved in the patient- parent-pediatrician relationship regarding issues of confidentiality.

Very important in the care of adolescents: confidentiality regarding STI diagnosis and treatment, pregnancy and contraceptive services, drug or alcohol dependency counseling and services, crime-related injury, and mental health services between a patient and physician varies between states .

Informed consent/dissent/assent

For older children, informed assent affirms that the voice of the patient must be heard.

The age at which a competent patient may legally exercise voluntary and informed consent for medical care varies from state to state and may be limited to specific conditions (sexually transmitted infections, family planning, drug or alcohol abuse).

There may be strain on the three-tiered relationship if parental permission does not match with the best interest of the child.

For younger children, what is best for the child is to be done even if the parent(s) disagree.

The dissent (or disagreement) of a child is the opposite of assent and is also morally relevant.

Pediatric ethics requires clinicians and parents to override a child's dissent when a proposed intervention is essential to his or her welfare.

Otherwise, assent should be solicited and dissent should be honored. Understand the difference between informed consent and assent.

Informed consent is a patient's or parent's consent for medical care, which is legally enforceable.

Informed assent is the agreement to medical care by a patient, although it is not legally enforceable.

Minors as decision-makers

In seeking younger children's assent, a clinician should help them understand their condition, tell them what they can expect, assess their understanding and whether they feel pressured to assent, and solicit their willingness to participate.When this cannot occur, pediatric ethics requires that clinicians apologize to a child for acting to override dissent.

Older children or adolescents may have the cognitive and emotional capacity to fully participate in health care decisions. If so, the adolescent should be provided with the same information as would be given to an adult patient. In cases like this, the patient may be able to provide informed consent ethically but not legally.

The adolescent's parent(s) remain in a guiding and protective role. The process of communication and negotiation will be more complex should disagreement arise between the parent and adolescent.

Understand when it is appropriate to have a minor involved in making decisions about his or her medical care.

Concerning genetic testing for susceptibility to adult-onset diseases, it is appropriate to wait until the child or adolescent is mature enough to weigh the pros and cons and make his or her own decisions about genetic testing.

Adolescents who are competent may sometimes consent to be research subjects.

Children born with ambiguous genitalia should not undergo elective surgery until they are old enough to participate in informed consent for a procedure.

A minor seeking emergency medical care does not need parental consent.

A minor is exempt from needing parental permission if he/she is married, emancipated by court ruling, in the military, or financially dependent and living apart from parents. In some states runaways, pregnant minors, minor parents, and college students are included in this exception.

A minor is exempt for specific medical conditions: STI diagnosis and treatment, pregnancy and contraceptive services, drug or alcohol dependency counseling and services, crime-related injury, and mental health services.

The mature minor exception: Minor is capable of providing informed consent to the proposed medical or surgical treatment—generally a minor 14 yr or older who

is sufficiently mature and possesses the intelligence to understand and appreciate the benefits, risks, and alternatives of the proposed treatment and who is able to make a voluntary and rational choice. (In determining whether the mature minor exception applies, the physician must consider the nature and degree of risk of the proposed treatment and whether the proposed treatment is for the minor's benefit, is necessary or elective, and is complex.)

Advance care planning/directives

Discussions concerning resuscitation, symptom control, and end-of-life care planning should be initiated when the physician recognizes that a significant possibility of patient mortality exists.

A palliative care team or hospice program can help in such situations.

For nonverbal and preverbal children who are developmentally unable to make autonomous care decisions, children should be as fully involved in discussions and decisions about their care as appropriate for their developmental status. Utilizing communication experts, child life therapists, chaplains, social workers, psychologists, or psychiatrist to allow children to express themselves through art, play, music, talk, and writing will enhance the provider's knowledge of the child's understanding and hopes.

Understand the use of advance directives in pediatrics.

An advance directive is a mechanism that allows patients and/or appropriate surrogates to designate the desired medical interventions under applicable circumstances.

Discussion and clarification of resuscitation status should be included in advance care planning, and for children attending school in spite of advanced illness, may need to be addressed in that setting.

Decisions regarding resuscitation status in the out-of-hospital setting can be an important component of providing comprehensive care.

Concerning a prenatal diagnosis of a lethal anomaly, parents may decide to carry the fetus to term but not want the help of any resuscitative measures.

An adolescent with a chronic and/or life-threatening condition should be supported in developing an advance directive as part of a comprehensive plan for end-of-life care. This approach can involve parents, medical caregivers, and other professionals who are familiar with the child's condition and developmental abilities to understand death and to express wishes concerning the end of life.

Religious (philosophical) exemptions

Pediatricians need to remain sensitive to and maintain an attitude of respect for these differences, yet recognize that an obligation exists to provide effective medical treatment to the child.

An adult with decision-making capacity is recognized as having the right to refuse treatment on religious or cultural grounds, but children who have not yet developed this capacity are considered a vulnerable population that have a right to treatment.

In situations that threaten the life of the child or that may result in substantial harm, legal intervention should be sought if reasonable efforts toward collaborative decision-making are ineffective.

If a child's life is imminently threatened, medical intervention is ethically justified despite parental objections.

Pediatricians should try to open a dialogue with parents who refuse vaccinations for their child and try to understand the reasons for refusal. They should try to work with them to overcome their concerns over time during the course of visits.

Pediatricians should refer patients to reputable sources for vaccine information and discuss risks and benefits.

Physician concerns about liability should be addressed by appropriate documentation of discussions in the chart.

Physicians also might wish to consider having parents sign refusal waiver.

Ethics and the use of technology
New technology
Genetics

Molecular diagnostic tests are often used to diagnose malformation syndromes, mental retardation, or other disabilities wherein there is a clear benefit to the child.

In other cases, such as genetic testing for susceptibility to adult-onset diseases, it is appropriate to wait until the child or adolescent is mature enough to weigh the pros and cons and make his or her own decisions about genetic testing.

Cochlear implants

A serious complication of cochlear implants is an excessively high incidence of pneumococcal meningitis. All children receiving a cochlear implant must be vaccinated with the PCV-13 vaccine, Haemophilus influenzae type b conjugate vaccines, and appropriate annual influenza vaccinations.

Patients receive these implants by their first or second birthday, therefore, there is an ethical issue that the patient has no say in the surgery.

Decline of sign language and deaf culture.

Significant social divide between hearing and non-hearing parents of these deaf children.

The non-hearing parents view the implants as a means of discrimination related to their sociocultural identity.

Sex/gender assignment

A newborn is assigned a sex before (with ultrasound) or at the time of birth based on the external genitalia.

In case of a disorder of sex development, these genitalia may appear ambiguous, and additional components of sex (e.g., chromosomal, gonadal, hormonal sex) are assessed. In consultation with specialists, parents assign the child a sex that they believe is most likely to be consistent with gender identity, which cannot be assessed until later in life.

There is debate about surgical modification of patients with congenital adrenal hyperplasia and other disorders of sexual differentiation before the patient is able to be part of the decision.

Imperiled newborn infants
Delivery room resuscitation issues

All newborns born greater than 22 or 23 weeks of gestations should receive all resuscitation efforts available unless otherwise pre-determined by the parents.

In the neonatal intensive care unit

For the most immature infants at the limit of viability (22-25 wk gestation), decision-making about care is a complex process that involves the physician, other health professionals, and the family.

The challenge for all premature infants is not only to improve survival but also to reduce short-term complications and improve long-term neurodevelopmental outcome.

Adverse neurodevelopmental sequelae include cerebral palsy, seizures, hydrocephalus requiring a shunt, blindness, deafness, and cognitive impairment.

The risk of an adverse outcome increases with decreasing gestational age at birth.

Organ transplantation and donation

Based on either irreversible cessation of neurologic function of the brain and brainstem or a predetermined period of cardiac asystole.

In order to avoid a potential conflict of interest, the request for organ donation should be separated from the clinical discussion of either brain death, cardiac asystole, or withdrawal of life-sustaining medical treatment.

There should be a separate organ donation team.

Death by neurologic criteriashould follow strict criteria adhering to nationally accepted guidelines must be used to determine when irreversible cessation of brain and brainstem function has occurred, and to adequately document these findings.

Organ procurement from a non-heart beating donor can occur under either controlled (after planned withdrawal of life-sustaining medical treatment) or uncontrolled (after failed cardiopulmonary resuscitation) circumstances.

Asystole must be present for 5-10 minutes for cardiac death to be considered irreversible.

Enhancement therapies
General considerations

Concerning autonomy: if a product is available, and the patient wants it, why should he or she not haveaccess to it?

Concerning justice from the patient's perspective: "everybody is doing it."

Concerning nonmaleficence: Just because we have the ability and chance to do something, does it follow that we must do so?

Growth hormone

There is difficulty in identifying where "normal" height ends and that the demarcation of "idiopathic" short stature is arbitrary and subjective, as growth hormone is now FDA approved for idiopathic short stature.

Growth hormone therapy is very expensive and requires therapy over several years. Is the cost worth the benefit to an individual v. society as a whole in those patients other than those with isolated growth hormone deficiency (for example, Turner syndome).

Is insurance coverage for growth hormone a good use of resource allocation in terms of justice?

Performance enhancement

The use of substances such as anabolic steroids, creatine, and dietary supplements in hopes that these agents will improve athletic performance.

These performance enhancers have not been rigorously studied with regard to safety in children and adolescents.

Stimulants are used to improve cognition and late-night studying.

Are these substances ethically appropriate to achieve a competitive advantage?

Allocation of healthcare resources
Just allocation of health care

Justice should be upheld in regards to allocation of limited healthcare resources.

Unnecessary tests and therapies should not be ordered if not necessary, particularly in fee-for-service health systems.

Managed care issues

Managed care involves cost-sharing systems that may increase the incentive to order less testing and spend less on patient care as this may be encouraged monetarily for the provider.

However, it is not ethical to limit spending on patient care for only this reason.

Professionalism and institutional ethics
Cross-cultural issues

There is importance in increasing one's cultural competence, relating to professionalism. This involves self-awareness of one's own cultural values ("cultural awareness"), learning basic fundamentals of other cultures ("cultural knowledge"), developing an ability to apply cultural knowledge to patient encounters ("cultural skills"), seeking exposure to cross-cultural encounters ("cultural encounters"), and being motivated to achieve the previous ("cultural desire").

Use professional interpreters instead of family member or friend interpreters to prevent medical errors and avoid issues of confidentiality and disruption of social roles.

Institutional ethics committees

Committees help with policy development, education, and case consultation.

Members should include those with insight into the unique ethical issues arising in the case of children.

Should provide advice without mandating action or being determinative.

Ideally should adopt a collaborative approach that uncovers all the readily available and relevant facts, takes into account the values of those involved, and balances the relevant interests, while arriving at a recommendation based on a consistent ethical analysis.

Adopt attention to respect for persons, beneficence/nonmaleficence, and justice.

These committees have acquired considerable influence and are increasingly recognized by state courts as an important aid in decision-making.

The membership, policies, and procedures of a pediatric ethics committee should conform to accepted professional standards.

Professionalism

General issues

Practice according to the highest standards utilizing evidence-based practice, personal physician accountability, and quality improvement.

Put patients first-have a commitment to act in the best interest of the patient.

Competence and Integrity: board certification, hospital credentialing, peer review of practice.

Create and apply systems and processes to eliminate the opportunity for error, identify error before it impacts patients, mitigate the effects of error, and thus improve patient safety and outcomes.

Gifts

Acceptance of modest gifts does not "involve a serious conflict—in fact, refusal of a gift may constitute a social or cultural affront."

More expensive gifts do breach the appropriate boundaries in the professional relationship.

It is acknowledged that some families may wish to compensate physicians by services or with barter.

Nonmonetary payments, as with gifts, may become precursors to boundary violations and should be approached with caution.

Pediatricians should be aware that they are subject to influencefrom the pharmaceutical industry when they accept gifts or payment for CME credit.

Errors and malpractice

General approach to apology and disclosure of medical errors.

Focused Setting: Quiet, comfortable private room, seated, pagers and cell phones off.

Sincerity and Empathy: This is about the patient, not risk managers and lawyers.

Truthfulness and Honesty: What happened? How andwhy did it happen? A True Apology: Directly state, "I am sorry."

35 states have passed "apology laws" that protect apologies from being used against clinicians in court.

Fixing the Problem: Explain what will happen for this patient.

Preventing the Problem: Explain steps taken to prevent this error in the future. Listen: Be sure the family has time to be heard.

Mounting evidence suggests that prompt disclosure actuallymay decrease the likelihood of legal action.

Duty: patient-physician relationship.

Breach: duty was breached based on "standard of care", usually need expert testimony.

Causation: breach of duty was the proximate cause of the injury, usually need expert testimony.

Damages: proof of damages.

Previous four needed to be prove medical negligence.

Common conditions involved in malpractice suits: meningitis, appendicitis, pneumonia, brain-damaged infants, respiratory problems in newborns, asthma, specified nonteratogenic anomalies (eg. developmental dysplasia of the hip, spina bifida occulta, craniofacial anomalies).

Conflicts of interest

Avoid any conflicts of interest that will compromise keeping the patient's needs first.

Medical testimony and expert witness

Experts often are required to give an opinion on the relevant standards of care, whether those standards were breached in the case at hand, and whether the breach was the proximate cause of the injury.

The expert must demonstrate sufficient knowledge, experience, and education on the issue in question.

The AAP expects experts to be licensed, board-certified physicians who are competent in clinical practice.

Their testimony is expected to be "thorough, fair, objective, and impartial."

Their compensation is to be reasonable and not contingent on the outcome of the case.

Pediatrician expert witnesses must ensure that their work is "relevant, reliable, honest, unbiased, and based on sound scientific principles."

Physicians who may present a risk to patients

Physicians have a responsibility to remove themselves (or others) from the care of the patient if he/she is a risk to the patient in any way.

Research and children

Respect for persons, which ensures informed, voluntary consent; Beneficence, which maximizes benefits while minimizing risks; and

Justice, which assures that some individuals are not exploited to benefit others.

Healthy children may participate only in research that involves no more than minimal risk.

Children who have disorders or conditions may be eligible for studies involving greater risk.

Informed consent entails proxy consent from a parent and sometimes assent from the child.

Special medical circumstances
Brain death

Now called death by neurological criteria.

Strict criteria adhering to nationally accepted guidelines must be used to determine when irreversible cessation of brain and brainstem function has occurred, and to adequately document these findings.

Because the response to severe neurologic injury in the immature brain may differ from that of the mature brain, currently established guidelines require a longer period of observation of infants and younger children compared to older children or adolescents.

The period of observation may be shortened through the use of a confirmatory test such as a cerebral perfusion study (demonstrating absent blood flow to the whole brain).

Patients in comas or vegetative states have some brain functions, including motor function and, therefore, do not fulfill the neurologic criteria for death.

Care of patients with disabilities

The duty to promote beneficence and nonmaleficence requires that professionals share the moral burden of difficult decision-making with families.

Professionals must not simply tell children and families what can be done, but rather help them decide if what canbe done should be done.

Children with AIDS/HIV infection

Pediatricians are strongly encouraged to disclose human immunodeficiency virus status to school-age children who are infected but asymptomatic and to all symptomatic children and adolescents.

In contrast to adolescents and adults, disclosure of HIV status to children should be undertaken over time, providing sequential pieces of practical health information that match the developmental capacity of the child.

Planned disclosure to family members and friends can increase practical and emotional support for the HIV-positive person.

While providing a listening ear and emotional support, clinicians also can offer hope and reassurance about the availability of effective treatment that can result in improved quality of life and survival for people living with HIV infection in the United States.

Violence and child abuse
Intimate-partner violence (IPV)

A careful assessment of the child's well-being and safety is essential.

Pediatric clinicians are obligated to report to CPS any suspicion of child abuse or neglect or any concern that the child is in imminent danger.

Document IPV in a confidential, separate portion of the chart where a legal guardian who is accused may not access it.

Recognize and apply ethical principles regarding the issues of physical and mental abuse.

IPV and child maltreatment commonly co-occur, particularly in families that have additional psychosocial risk factors such as poverty, parental depression, and substance abuse.

IPV negatively affects children's social-emotional health, increasing the risk for internalizing and externalizing disorders and aggression with peers.

Violence in society

Adequate parental monitoring in middle childhood may prevent childhood aggression, and therefore, potential violence in society.

Child abuse

In the United States, physicians are required by law to report and document child abuse in the chart.

Report not only confirmed cases, but also cases where there is reasonable cause to suspect abuse.

Confidentiality should otherwise be upheld.

Complementary and alternative medicine

If parents are using alternative therapies for life-threatening illnesses for which effective conventional therapies are available, physicians must notify child protective services.

The four ethical principles of autonomy, nonmaleficence, beneficence, and justice should be used in decision-making when complementary and alternative medicine is to be discussed.

Pediatricians must educate themselves and their families about the risks and benefits of various treatments being considered.

All discussions and consents need to be as informed as possible and documented clearly, whether advice has been accepted or rejected by the family.

All responses of all patients to the chosen therapies of any type should be monitored.

If a family chooses a treatment outside the clinician's scope of practice, he or she still can evaluate responses, even if unable to refer or provide treatment.

Children in foster care

Child welfare should obtain appropriate medical consents and releases of information from the birth family, provide copies to the pediatrician, and educate health providers about the foster care agency's guidelines regarding consent and confidentiality.

Children and adolescents in foster care are children who have special health-care needs because they have a high prevalence of medical, mental health, developmental, educational, and dental health problems.

Expert consensus contends that child health, mental health, and developmental outcomes will be improved by child-centered medical home care while the child or teenager is in foster care.

Education, training, and evaluation

Curriculum resources

Identify curriculum resources for professional ethics in education, training, and evaluation.

Clinical ethics consultation.

Ethics for the Pediatrician series in Pediatrics in Review.

Evaluation tools

Identify evaluation tools for professional ethics with regard to the education, training, and evaluation of pediatric residents.

Evaluation questions at the end of Pediatrics in Review, Ethics for the Pediatrician series.

Patient Safety and Quality Improvement

Definitions used in discussions of patient safety

The failure to complete a planned action as intended or the use of a wrong plan to achieve an aim.

It is a mistake in action or judgment.

Most medical errors do not lead to adverse events.

A potential adverse drug event or "near-miss event" is a medication error that places a patient at significant risk of injury but does not actually result in harm.

An unexpected occurrence involving death or serious physical or psychological injury, or the risk thereof.

An ADE or a potential ADE may qualify as a sentinel event. Not all sentinel events are the result of a medical error.

An adverse event is an injury caused by medical management rather than by the underlying disease or condition of the patient.

It is preventable if based on the medical information known at the time, it could have been avoided.

Most common type is an adverse drug reaction.

An adverse event that could not have been foreseen based on the medical information known at the time, such as a patient with no known drug allergies who develops urticaria after taking a medication.

Epidemiology of medical error and harm

Approximately 70,000 hospitalized children experience an adverse event each year, of which 42,000 could be avoided.

The risk for an ADE is estimated to be three times higher in hospitalized children than in adults.

The estimated cost of medical errors in the United States is $37.6 billion annually.

Preventable adverse events comprise an estimated $17 to $29 billion, more than 50% of which represent direct costs to the health-care system.

An adverse drug event extends the length of hospital stay by 1.74 to 2.2 days and increases costs by $2,000 to $3,200.

A preventable ADE extends the hospital stay by 4.6 days and increases costs by $5,800

These figures represent dollar values from the 1990s; current estimates are likely even higher.

In a prospective study of medication orders in a children's hospital, 23.7% contained errors.

Medication dosing errors are common due to weight-based dosing.

Identify situations presenting high risk for adverse events in the management of pediatric patients.

Children who experience extended lengths of stay, complex medication regimens, and higher severity of illness are at increased risk of ADEs.

Neonates and young infants are at even greater risk due to their immature hepatic, renal, and immune systems.

Complex medication regimens increased the likelihood of preventable ADEs.

Parents whose English proficiency was limited were less likely to report an ADE, which may represent a subpopulation of children at even greater risk of harm.

Many over-the-counter children's medications are available in a variety of preparations and concentrations, which can contribute to confusion and subsequent dosing error.

Detecting and reporting adverse events

Relying on reporting by incident reports presumes that an individual recognizes a medical error or an adverse event when it occurs.

Staff may assume that someone else will report the event.

Incident report forms generally require a large amount of information, which makes them both time-consuming and cumbersome to complete.

Personnel who do take the time to report events often receive no feedback.

They may not see their efforts resulting in noticeable improvement, which can act as a disincentive for reporting subsequent events.

A natural hesitancy to point out one's own mistakes for fear of beinglabeled incompetent and a reluctance to point out others' mistakes for fear of being labeled a whistleblower.

Worry about discipline by an institution or licensing organization. Fear of litigation.

Chart reviews in search of medical errors and adverseevents are time- and labor- intensive, as well as cost-prohibitive.

Apply effective strategies to improve reporting of adverse events. Transitioning to a culture of blameless reporting.

Trigger methodology where the use of a drug such as flumazenil will trigger a location for a chart review.

Apply voluntary systems for reporting of adverse medical events. Obtain a thorough list of the patient's current medications.

Obtain an accurate list of the patient's allergies and adverse reactions Print legibly. Avoid the use of unsafe abbreviations. Write daily and units.

Obtain an accurate patient weight. Mind your decimals.

Include indication for therapy on orders and prescriptions. Educate patients and families.

Disclosure of medical errors

Use appropriate means to disclose medical errors to patients.

Health-care professionals should consult risk management experts or legal counsel for guidance with disclosure of adverse events and saying “I’m sorry.”

Apply appropriate methods of support for patients and their families after an error producing medical harm occurs.

Use appropriate methods of support for physicians and other health-care providers after an error producing medical harm occurs.

Health-care professionals must be supportive and nonjudgmental of their colleagues when medical errors occur.

Debriefing sessions can provide a therapeutic venue to discuss not only the medical aspects of the event, but also the emotional issues as they relate to theclinicians involved.

Methods to reduce medical adverse events

An individual’s actions are rarely responsible for an error or adverse event alone.

Root cause analysis (RCA) is a retrospective tool often used to determine the factors contributing to an error or adverse event so improvements can be made to prevent a recurrence.

It is system focused, so it helps to avoid blaming an individual whose actions are rarely responsible for an error or adverse event alone.

Begins with a detailed account and timeline of events leading up to the incident.

A multidisciplinary meeting is convened to review this summary and identify key contributory factors, or “root causes,” for the error or adverse event.

Once the root causes have been established, risk reduction strategies are developed with a time frame for implementation.

Anticipate system vulnerabilities by applying failure mode effects analysis (FMEA). Anticipates failures in human, process, and mechanical interactions.

The analysis then identifies areas most in need of improvement. Risks are evaluated in terms of severity, occurrence, and detection.

The risks are given a priority value assessing overall risk level; actions for risk reduction or elimination are then determined.

FMEA works well for processes that are relatively linear.

FMEA has been used to guide implementation of computerized order provider entry, reducing ordering errors in pediatric chemotherapy.

Similarly, the process could be used to help mitigate risk in the transition to a new electronic medical record in an office setting.

Evidence can serve as a powerful tool to support and drive quality improvement, as in the implementation of evidence-based guidelines

Implemented through the use of physician order sets, electronic templates, and alerts built into computerized provider order entry, as well as electronic documentation systems.

Use best-practice guidelines to reduce medical adverse events.

Best-practice guidelines reduce variation in care delivery, thereby reducing risk of errors.

The Muir Gray classification system uses a rating scale, with randomized controlled trials being ranked as the best or highest level of evidenced-based intervention.

Most clinical guidelines use evidence-based interventions and stress the application of interventions that have the strongest support from research studies.

Use effective methods of communication to reduce errors in the healthcare setting

Interventions can reduce error in situations (eg, stress, fatigue. distraction) at high risk for medical error.

The use of checklists.

Understand and apply methodologies to prevent medication errors.

Lean methodology focuses on reducing waste and improving work flow to increase productivity by eliminating steps that do not add value to a process.

Six Sigma strives to decrease variation through detailed data collection and analysis.

A commonly used Lean Six Sigma tool is a value stream map that outlines an entire process from start to finish.

Each step in the process is analyzed in three domains: personnel, information flow, and time.

Teams of health-care providers then address workflow and find ways to increase value at each step.

Some ways to increase value include reducing variation, streamlining processes, eliminating redundant information, and increasing employee engagement.

Lean Six Sigma can be used, for example, to improve efficiency in an outpatient office or an emergency department.

Clinical pharmacist participation on inpatient rounds has been shown to be an effective means of primary prevention in various settings.

Recommendations, including choice of drug, dosing regimen, and pharmacokinetic monitoring, help reduce errors and preventable ADEs.

Review of medication orders by pediatric pharmacists has been shown to be an effective means of preventing potential ADEs from reaching the bedside.

Pharmacists and nurses should be empowered and encouraged to double-check orders and contact the prescriber when an error is identified.

It can ascertain that required information is included in an order or prescription using forced format screens and essentially can eliminate the issue of illegibility.

Weight-based calculators with pediatric guardrails can ensure that dosing remains within safe parameters.

Many systems include checks for allergies and drug interactions, integration of available laboratory data, reminders to monitor serum drug concentrations, and promotion of standardized order sets.

Look alike product names or packaging can affect safety.

A poorly designed interface between the medical device and the user can contribute to errors.

Devices should be designed according to users' needs, abilities, limitations, and work environments.

Includes the design of the device's user interface, which includes controls, displays, software, labels, and instructions.

Severity of illness, intensity of treatment, and polypharmacy have been associated with ADEs.

ADEs are more likely to result in life-threatening consequences in intensive care unit (ICU) patients than in others.

Review the dosing regimen, indications for therapy, and potential adverse effects for every new prescription.

Provide written instructions and information whenever possible.

Encourage patients and families to contact you with any questions or concerns.

Principles of quality improvement applied to improving patient safety
Key principles of patient safety

Health-care leaders and organizations must drive a culture that improves quality and patient safety.

Apply knowledge of human factors in the design of systems and processes promoting patient safety.

Human factors is the science that focuses on understanding and supporting how people interact with technology.

Three major areas when evaluating medical-device-related adverse events from a human factors perspective:

User characteristics, including the person's abilities and training and her expectations of the device

Device design considerations, which focus on the device-user, interface, including instructions for use.

The environment in which the device is used, including the lighting, noise, distractions, and time constraints.

Promote effective team functioning in the prevention of medical error.

Team training can improve communication and situational awareness to create a safer health-care environment.

Understand the importance of assessment and redesign of healthcare process before error occurs.

A central concept in QI is implementation of incremental change, followed by measurement of the effects of these changes over time.

Understand the importance of creating and maintaining a learning environment (eg, morning report, meetings with partners) in improving patient safety.

Core principles of quality improvement

Analysis of variation in data helps to understand whether the variation is actual improvement.

A scientific method to improving human systems is quality improvement. Put evidence-based initiatives to use.

Apply the psychology of change (eg, motivating people to improve) to improve health-care systems.

Interdisciplinary team training programs employing simulation can be used to improve quality of care and develop strong patient safety environments.

Identify the components of the Langley Model for Improvement: Plan-Do-Study-Act

> Plan phase: the QI team states the project's objective and creates a detailed plan that specifies - who will be responsible for the changes; what the specifics of the changes will be; where the changes will be implemented; and when the changes will occur.
> Do phase: tests of change begin and the team documents any obstacles that are encountered, so they can be addressed
> Study phase: team begins to analyze the process and outcome data, and a record of lessons learned is kept.
> Act phase: the QI team considers adopting, adapting, or abandoning changes based on the results of the Study phase and integrates the findings into the next PDSA cycle.
> The number of PDSA cycles needed for a given QI project varies and depends on the individual nature of the initiative and the environment in which it is implemented.

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