

PNCB CPNP-PC® - Quiz Questions with Answers

Assessment and Diagnosis

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1.

You are assessing a 12-hour-old male in the newborn nursery. The baby was born full-term via vaginal delivery, and the pregnancy was uncomplicated. This is the mother's first child. Your assessment of the infant is unremarkable, except for a palpable abdominal mass in the left upper quadrant.

What is the MOST likely diagnosis?

Ureteropelvic junction obstruction

Wilms tumor

Renal vein thrombosis

Urolithiasis

Correct answer: Ureteropelvic junction obstruction

The most common cause of abdominal masses in the newborn period is hydronephrosis, a significant dilation of one or both kidneys, likely caused by an obstruction of the ureteropelvic junction. If spontaneous resolution does not occur by 6 to 12 months of age, or if symptoms arise or persist, surgical repair may be necessary. The longer the obstruction lasts, the less likely renal function will return to normal.

Wilms tumors and urolithiasis, although possible, are extremely rare in newborns. With renal vein thrombosis, sudden onset of gross hematuria may be noted, along with a firm, flank mass.

2.

The Salter-Harris classification of fractures describes growth plate fractures, ranging from types I to V.

The MOST common type seen in pediatrics, and described as a fracture that passes across most of the growth plate and up through the metaphysis, is:

Type II

Type I

Type III

Type V

Correct answer: Type II

Fractures of the long bones can produce permanent deformities in children if the fracture occurs through the growth plate. The outcomes depend on the location, type, age of the child, status of the blood supply to the physis, and the treatment of the fracture. The Salter-Harris classification is based on the mechanism of injury, the relationship of the fracture line to the layers of the physis, and the prognosis with respect to subsequent growth disturbance. There are five classifications that can be remembered by the mnemonic SALTR:

- *Type I: Slipped; the fracture plane passes all the way through the growth plate, not involving bone, and the prognosis is good.*
- *Type II: Above; the fracture passes across most of the growth plate and up through the metaphysis, and the prognosis is good. Type II is by far the most common type of growth plate fracture, with up to 75% of cases falling into this category.*
- *Type III: Lower; the fracture plane passes some distance along the growth plate and down through the epiphysis. The prognosis is poorer as the proliferative and reserve zones are interrupted.*
- *Type IV: Through or transverse or together; the fracture plane passes directly through the metaphysis, growth plate and down through the epiphysis. There is a poor prognosis as the proliferative and reserve zones are interrupted.*
- *Type V: Ruined or rammed; crushing type injury that does not displace the growth plate but damages it by direct compression. These are rare and difficult to diagnose initially due to the lack of radiologic signs and are the worst prognosis, requiring anatomic reduction to prevent articular incongruity and osseous bridging across the physis.*

3.

Which of the following congenital heart diseases (CHD) causes cyanosis?

Dextro-transposition of the great arteries (d-TGA)

Atrial septal defect (ASD)

Ventricular septal defect (VSD)

Aortic stenosis

Correct answer: Dextro-transposition of the great arteries (d-TGA)

The cyanotic heart diseases in children are as follows:

- *Truncus arteriosus*
- *Transposition of the great arteries*
- *Tricuspid atresia*
- *Tetralogy of Fallot*
- *Total anomalous pulmonary venous return*

All of these diseases begin with the letter 'T' and can be remembered easily in that regard. With d-TGA, there is incomplete separation and migration of the truncus arteriosus during fetal development. The aorta arises from the right ventricle, and the pulmonary artery arises from the left ventricle. The aorta then receives the deoxygenated systemic venous blood and returns it to the systemic arteries, while the pulmonary artery receives the oxygenated pulmonary venous blood and returns it to the pulmonary circulation. Cyanosis is evident immediately after birth (within 1 hour to 1 day). Management involves referral to pediatric cardiology, and arterial switch surgery is usually performed in the first few days of life if possible.

4.

A 12-year-old female presents to the pediatric urgent care center with a temperature of 102 F (38.89 C) for 2 days, headache, and complaints of a stiff neck. On physical exam, she is positive for Kernig's sign. Her neurological exam is significant for slight nuchal rigidity, and her eye exam is significant for photophobia. You suspect meningitis and proceed with a spinal tap.

Which of the following would you expect to find in the cerebrospinal fluid (CSF) with aseptic meningitis?

Increased lymphocytes

Decreased CSF protein concentration

Positive bacterial antigen tests

Decreased glucose concentration

Correct answer: Increased lymphocytes

Aseptic meningitis is an inflammatory process of the meninges, most often characterized by acute signs and symptoms of meningeal irritation (headache, stiff neck, fever); mononuclear CSF pleocytosis (predominance of monocytes, macrophages, and/or lymphocytes); a normal or, less frequently, elevated CSF protein concentration; normal or, less often, low CSF glucose concentration; and no organisms demonstrable by Gram stain or bacterial cultures.

The most common cause is viral (not bacterial) infection. Up to 90% of cases are caused by enteroviruses and arboviruses. Symptoms vary, but headache and fever are predominating symptoms.

Again, CSF findings include increased lymphocytes, normal glucose concentration, normal or slightly elevated protein, and negative bacterial antigen tests.

5.

Which of the following is the MOST commonly diagnosed neoplasm in neonates?

Neuroblastoma

Teratoma

Wilms tumor

Soft-tissue sarcoma

Correct answer: Neuroblastoma

A neuroblastoma is a solid tumor of unknown etiology that originates from neural crest tissue along the craniospinal axis. The majority of neuroblastomas develop in the abdomen, usually in the adrenal gland. It is the most commonly diagnosed neoplasm in neonates (extracranial solid malignancy), with about 600 new cases diagnosed each year.

On rare occasions, neuroblastomas may regress without therapy (generally only those in children younger than 1 year old). Management most often involves a multidisciplinary approach, including surgical removal followed by radiation therapy or chemotherapy. The prognosis depends on the age of the child and stage of the tumor.

Teratomas, Wilms tumors, and soft-tissue sarcoma are less frequent types of neonatal neoplasms.

6.

Pelvic inflammatory disease (PID) in adolescents is often a polymicrobial infection. Of the following sexually transmitted infections (STIs), which MOST commonly cause this disease?

Gonorrhea and chlamydia

Gonorrhea and syphilis

Chlamydia and herpes simplex virus

Syphilis and herpes simplex virus

Correct answer: Gonorrhea and chlamydia

PID is an infection of a woman's reproductive organs. It is most commonly caused by chlamydia and gonorrhea but can be caused by other infections. Vaginal flora, other aerobic and anaerobic organisms, GBS, genital mycoplasma, and gram-negative bacteria are also implicated.

The CDC recommends presumptive treatment for sexually active young women if they experience pelvic or lower abdominal pain when no other cause of pain can be found. Women with PID should also be tested for HIV.

7.

Some disorders occur with greater frequency in children of certain races or whose ancestors were from specific geographic regions. One example of this phenomenon is sickle cell anemia (SCA), which occurs MOST frequently in individuals of what descent?

African

Central American

Mediterranean

Indian

Correct answer: African

Sickle cell disease has an autosomal recessive inheritance pattern. It is found most often in people of African descent but is also detected among ethnic groups from the Mediterranean, the Caribbean, Central and South America, and India. Owing to migration, it now occurs worldwide.

Sickle cell trait occurs in 8% of African Americans, and approximately 1 in 400 African Americans has sickle cell disease. This incidence exceeds that of most other serious genetic disorders in children, including cystic fibrosis and hemophilia.

8.

Urine testing and blood glucose measurements are generally sufficient to diagnose:

Type 1 diabetes mellitus (T1DM)

Type 2 diabetes mellitus (T2DM)

Obesity

Hypoglycemia

Correct answer: Type 1 diabetes mellitus (T1DM)

Type 1 diabetes mellitus (T1DM) is caused by autoimmune destruction of pancreatic beta cells thought to be triggered by a preceding environmental event in genetically susceptible individuals. The destroyed beta cells result in deficient insulin secretion, reduced biologic effectiveness, or both. Glucose levels then become elevated in the blood, and as a result, diabetes ensues.

Polyuria and urinary incontinence can be the first symptoms of diabetes mellitus and are secondary to hyperglycemia and the osmotic diuresis resulting from chronic glycosuria. Urine testing (for glucose and ketones) and blood glucose measurements are generally sufficient to make the diagnosis.

Diagnostic studies for type 2 diabetes mellitus (T2DM) include urine testing and fasting blood sample for blood glucose, HbA_{1c}, lipid panel, TSH and free T₄, and insulin level.

Children and adolescents with a BMI greater than the 85th percentile for age and gender (indicating obesity) should be screened for a number of comorbidities: prediabetes and type 2 diabetes, dyslipidemia with fasting lipid panel, AST and ALT levels, blood pressure measurement, obstructive sleep apnea, and PCOS, along with a free and total testosterone level.

Hypoglycemia requires a blood glucose measurement to diagnose, not urine testing.

9.

Which of the following theorists focused mainly on theories of moral development and socialization, emphasizing the process by which children learn the expectations and norms of their society and culture?

Kohlberg

Piaget

Erikson

Freud

Correct answer: Kohlberg

Lawrence Kohlberg was a psychologist best known for his theories of moral development and socialization, emphasizing the process by which children learn the expectations and norms of their society and culture. Kohlberg's work primarily involved male participants. Kohlberg's stages include:

- *Punishment avoidance and obedience*
- *Instrumental realistic orientation*
- *Good interpersonal relationships*
- *Maintaining social order*
- *Social contract and utilitarian orientation*
- *Universal principles*

Freud is best known for his psychoanalytic theory in finding links between the conscious mind and the body through the unconscious mind. Descriptions of the id, ego, and superego were some of his most significant contributions.

Erikson described the stages of the individual throughout the lifespan, presenting problems within each stage that the individual seeks to master.

Piaget's theory of cognitive development is about how a child constructs a mental model of the world. According to Piaget, children are born with a very basic mental structure (genetically inherited and evolved) on which all subsequent learning and knowledge is based. He emphasized how children modify themselves depending on their environmental experiences and their stage-related competency level.

10.

What is the MOST common type of seizures in children?

Febrile seizures

Petit mal seizures (absence seizures)

Myoclonic seizures

Grand mal seizures (generalized tonic-clonic seizures)

Correct answer: Febrile seizures

Febrile seizures are the most common type of seizures in childhood and occur in up to 5% of children. They occur in conjunction with a fever (38 degrees C or higher). They are brief, generalized, clonic or tonic-clonic in nature, and can be either simple or complex. Simple febrile seizures last less than 15 minutes, while complex febrile seizures may last longer than 15 minutes and can have focal attributes. Febrile seizures typically occur between the ages of 6 months and 5 years (in association with a fever) in the absence of intracranial infection.

The etiology of febrile seizures is unclear, with risk factors including family history of febrile seizures, or in those with predisposing factors, such as NICU stay >30 days, developmental delays, or day care attendance.

11.

An inheritance pattern in which a disorder is inherited in a dominant manner exclusively from the mother is:

Mitochondrial inheritance

Multifactorial inheritance

Autosomal recessive

Autosomal dominant

Correct answer: Mitochondrial inheritance

Mitochondrial inheritance is an inheritance pattern that arises from mitochondrial DNA (mtDNA). Mothers alone pass on mtDNA (i.e., matrilinear or maternal inheritance) because only egg cells contribute mitochondria to the developing embryo. Disorders that arise from mutations in mtDNA can appear in every generation, affecting both sexes, but mtDNA is inherited from the mother; thus, a mother will pass on the disease to all of her children.

Autosomal recessive inheritance is a type of single gene disorder that requires two copies of a mutated gene (one from each parent) located on one of the autosomal chromosomes (chromosome 1-22). Offspring who inherit only one abnormal gene in the pair are considered carriers.

Autosomal dominant inheritance is a type of single gene disorder characterized by the inheritance of a single copy of a mutated gene located on one of the autosomal chromosomes (chromosome 1-22). The gene mutation is passed on from only one parent but results in an inherited disorder; the paired gene from the other parent is normal. The parent passing on the gene mutation typically has the disorder.

In multifactorial inheritance, a combination of both genetic (with one or more contributing genes) and environmental factors are involved.

12.

You have an adolescent patient with costochondritis. He is experiencing localized tightness and pain to the anterior chest wall, which is exacerbated by coughing, sneezing and deep breathing.

Your key differential diagnosis is:

Rib fracture

Rheumatic disease

Clavicular fracture

Vertebral osteomyelitis

Correct answer: Rib fracture

Costochondritis is a common cause of chest pain in children and adolescents and involves an inflammatory process of one or more of the costochondral cartilages that causes localized tenderness and pain of the anterior chest wall. Most cases are idiopathic in nature, though trauma to the area and unaccustomed physical effort are factors known to cause costochondritis.

Rib fractures are the key differential diagnosis if pain is associated with an injury.

Childhood rheumatic diseases can also have complaints similar to costochondritis but generally have other characteristic physical findings. Vertebral osteomyelitis is a differential diagnosis of back pain. Clavicular fracture is not associated with costochondritis.

13.

A two-day-old male has developed a rash that has spread over much of his body. The infant was born full-term via an uncomplicated vaginal delivery to a healthy mother. The infant does not exhibit any signs of distress; he has no fever and is breastfeeding well. During your examination, you note firm, yellow-white 2-mm papules with an erythematous base on the skin clustered together over the infant's cheeks, forehead, trunk, and limbs. A histological stain demonstrates large numbers of eosinophils.

Which of the following is the MOST likely diagnosis?

Erythema toxicum

Herpes simplex virus (HSV)

Transient neonatal pustular melanosis

Milia

Correct answer: Erythema toxicum

Erythema toxicum is a benign self-limited eruption occurring primarily in healthy newborns in the first 24 to 48 hours. Up to 50% of newborns develop erythema toxicum, with a higher incidence in term than in preterm infants. This condition is characterized by yellow-white papules with a blotchy red base; it resolves within 2-3 weeks and is usually completely gone by 4 months of life. No treatment is required; the course is brief and transient.

An HSV rash is characterized by grouped vesicles on an erythematous base; the mother may have active lesions or a history of the disease. HSV is diagnosed via DFA or ELISA detection of HSV antigens, and acyclovir is given for treatment.

Transient neonatal pustular melanosis is a rash that is described best as vesicopustules that rupture easily and leave a halo of white scales around a central macule of hyperpigmentation, generally seen on the trunk, limbs, palms, and soles. There is no definitive diagnosis, and no treatment is necessary as a resolution is spontaneous.

Milia are multiple, firm, pearly, opalescent white papules scattered over the forehead, nose, and cheeks. Their intraoral counterparts are called Epstein pearls. Milia histologically represents superficial epidermal inclusion cysts filled with keratinous material associated with the developing pilosebaceous follicle. Milia exfoliate

spontaneously in most newborns over the first few weeks of life and thus, no treatment is needed.

14.

You are doing medical missions work in Bangladesh and note many of the children you see are presenting with the same chief complaint. While obtaining a history on these children, you realize that a majority of their diet consists of grains, dairy, and occasional meats. They lack fruits and vegetables in their daily diets.

What is the MOST likely chief complaint?

Bleeding gums, easy bruising, and slow wound healing

Rickets

Anorexia and night blindness

Muscle weakness, tachycardia, and confusion

Correct answer: Bleeding gums, easy bruising, and slow wound healing

These patients are likely deficient in vitamin C, which primarily comes from fruits and vegetables, especially citrus fruits, as well as broccoli, collard greens, spinach, tomatoes, potatoes, strawberries, and peppers. Scurvy may occur in patients with deficient vitamin C levels. Other signs include cracked lips, bleeding gums, slow wound healing, and easy bruising.

Rickets would most likely be caused by a diet deficient in vitamin D (sources include sunlight and fortified food products, especially milk and fish).

Anorexia and night blindness are signs of a deficit in vitamin A (sources come from liver, fish liver oils, fortified milk, eggs, red and orange vegetables, and dark green leafy vegetables).

Muscle weakness, tachycardia, and confusion would likely be clinical findings in the child with a thiamin (vitamin B1) deficiency. Dietary sources include whole grains, brewer's yeast, legumes, seeds and nuts, fortified grain products, organ meats, and lean cuts of pork.

15.

A 4-day-old, full-term male develops excessive purulent ocular discharge. Which of the following etiologies carries the GREATEST risk of morbidity in this newborn?

Neisseria gonorrhoeae

Chlamydia trachomatis

Staphylococcus aureus

Haemophilus influenzae

Correct answer: *Neisseria gonorrhoeae*

Gonococcal conjunctivitis is the most serious cause of ophthalmia neonatorum (conjunctival infection or inflammation occurring in the first month of life). It is of particular concern because it produces a hyperacute, profusely purulent conjunctivitis that can lead to corneal perforation and blindness. It is less often seen in Western countries due to chemoprophylaxis but is still an important cause of ophthalmia neonatorum.

*The other answer choices are all bacteria that can cause conjunctivitis in the newborn, but not as great of a risk as *Neisseria gonorrhoeae*.*

16.

Of the following functions, which are NOT performed by the kidneys?

Store substances

Filter substances

Reabsorb substances

Secrete substances

Correct answer: Store substances

Physiologically, the kidneys serve to filter, clear, reabsorb, and secrete substances essential to the body's metabolism. The urinary system starts to form and excrete urine at just three months of gestational age, and glomerular filtration and renal blood flow become stable by 1 to 2 years old. The kidneys mature through infancy, although all measurable variables of kidney function approach adult values between 6 and 12 months old.

The bladder stores primarily urine, allowing urination to be infrequent and voluntary.

17.

In all age groups except in the neonate, the hallmark of pneumonia is:

Fever and cough

Fever and diminished breath sounds

Fever and tachypnea

Fever and hypoxia

Correct answer: Fever and cough

The hallmark of pneumonia is fever and cough, except in the neonate where there may be an absence of cough.

Tachypnea and increased work of breathing may precede coughing. Cough, hypoxia, nasal flaring, retractions, rales, and rhonchi lung sounds are specific but not as sensitive for a pneumonia diagnosis.

Viral pneumonia tends to have an insidious onset that is associated with more wheezing than what is typically seen with bacterial pneumonia. On the other hand, lobar pneumonia (caused by pneumococcal pneumonia) generally presents with fever, cough, and decreased breath sounds in the area of the pneumonia.

18.

You are seeing a 10-year-old male for his annual physical examination. When you ask him about home life, he tells you that his parents recently divorced. Which of the following is this child's MOST LIKELY reaction compared to a younger child's reaction to divorce?

Intense, conscious anger

Acceptance of the divorce

Emotional lability

The ability to see the positive aspects of the divorce

Correct answer: Intense, conscious anger

The school-age child is most likely to exhibit conscious, intense anger in regards to the stressor of his parent's recent divorce. As a part of the process of developing relationships with others, school-age children refine their ability to identify, label, and manage their feelings. However, their experiences are limited, and their cognitive abilities are still expanding. They need help in labeling and processing complex emotions such as sadness, depression, worry, and envy. They also need help to manage those and other feelings consciously and in acceptable ways. Coping abilities are significantly affected by the availability of social supports from caregivers.

The younger child is more likely to accept the situation, but have frequent and varied emotional reactions to the divorce as they process through it. The ability to see the positive aspects is not likely to occur until much later in development.

19.

Which of the following BEST supports the diagnosis of pertussis in a child with a cough?

Prolonged course

Infiltrates seen on chest x-ray

Age of 7 years or older

Leukopenia

Correct answer: Prolonged course

In children, the illness B. pertussis generally follows a three-stage pattern:

- Catarrhal stage: 1-2 weeks with nonspecific complaints and manifestations of the common cold including low-grade fever, mild but worsening cough, coryza, sneezing. This is the most contagious stage.*
- Paroxysmal stage: 2-4 weeks with absent or minimal fever, persistent paroxysmal cough ending with an inspiratory whoop, worse at night, with vomiting, cyanosis, sweating, and exhaustion present (especially after coughing)*
- Convalescent phase: 3 weeks to 6 months; paroxysms become less frequent and less distressing, although the cough may become louder.*

Pertussis (whooping cough) is mainly a clinical diagnosis, but a prolonged course of cough along with leukocytosis (not leukopenia) with a marked lymphocytosis supports the diagnosis. Pertussis is known as the "100-day cough" in China.

Polymerase chain reaction (PCR) is the primary diagnostic test used in most commercial and state laboratories for confirmation. Chest x-rays are most often normal or have nonspecific findings, and a majority of cases occur in children younger than 5 years old.

20.

Which of the following statements is TRUE regarding attention-deficit/hyperactivity disorder (ADHD) in children and adolescents?

Children in rural areas are more likely to be diagnosed with ADHD and less likely to receive behavioral therapy

ADHD is one of the most neglected and misunderstood psychiatric conditions in the United States

Widespread social stigma is a major contributing factor as to why very few children receive an appropriate diagnosis of ADHD in the United States

ADHD affects roughly 1 in 200 children and adolescents across Europe

Correct answer: Children in rural areas are more likely to be diagnosed with ADHD and less likely to receive behavioral therapy

ADHD is one of the most commonly diagnosed disorders in childhood, and symptoms can profoundly affect the behavior of individuals across many settings in their lives. ADHD prevalence increases with the child's age and affects approximately 9-15% in school-age children. It is more frequent in males than in females, and females tend to have more problems with inattention.

The National Survey of Children's Health reports a 6.1% overall prevalence of ADHD in children 2 to 17 years of age in the United States. This study also showed that children in rural areas are more likely to be diagnosed and less likely to receive behavioral therapy for their ADHD.

ADHD has been identified as one of the most neglected and misunderstood psychiatric conditions in Europe. Although ADHD is estimated to affect approximately 1 in 20 children and adolescents across Europe, very few people received an appropriate diagnosis due to the lack of public awareness and widespread social stigma, as well as the lack of appropriate community support and resources.

21.

At what age is a child's vision close to fully developed?

12 months old

18 to 24 months old

6 to 10 months old

5 years old

Correct answer: 12 months old

Vision is close to fully developed at 12 months of age. Eye growth, however, is not completed until 10 to 13 years old.

22.

Sickle cell disease, thalassemia, and hemophilia are all examples of:

Monogenetic disorders

Chromosome disorders

Multifactorial disorders

Mitochondrial disorders

Correct answer: Monogenetic disorders

Monogenetic disorders (also referred to as single gene disorders) occur when the mutation affects a single gene. The mutation may be present on one or both chromosomes and is most often classified as dominant, recessive, or X-linked. Examples include sickle cell, thalassemia, neurofibromatosis, hemophilia, Duchene muscular dystrophy, cystic fibrosis, fragile X syndrome, PCKD, Marfan syndrome, and Tay-Sachs disease.

Chromosome disorders occur with changes in the number or structure of an entire chromosome (e.g., Down syndrome is caused by an extra copy of chromosome 21).

Multifactorial disorders result from a combination of genetic and environmental factors (e.g., neural tube defects).

Mitochondrial disorders are caused by mutations in mitochondrial DNA (nonchromosomal DNA) and are typically progressive disorders affecting the brain and muscles. They are characterized by exclusively maternal (matrilinear) transmission.

23.

Which of the following deficiencies has been linked with neurologic symptoms associated with the build-up of homocysteine?

Vitamin B12

Vitamin B1 (Thiamin)

Vitamin B2 (Riboflavin)

Vitamin B6 (Pyridoxine)

Correct answer: Vitamin B12

Vitamin B12 is essential for neurologic function, adequate red blood cell formation, and DNA synthesis. A vitamin B12 deficiency has been associated with megaloblastic anemia, sore tongue, weakness, and neurological deficits. These neurologic symptoms are often associated with the build-up of homocysteine, due to the failure of precursor B-12 dependent conversion enzymes. Because of the associated neurologic manifestations that can occur, prompt diagnosis of vitamin B12 deficiency is imperative.

Vitamin B1 (thiamin) deficiency is exhibited by muscle weakness, ataxia, confusion, anorexia, tachycardia, and heart failure in infants.

Signs of Vitamin B2 (riboflavin) deficiency are oral-buccal cavity lesions, generalized seborrheic dermatitis, scrotal and vulval skin changes, normocytic anemia, and dimness of vision.

Vitamin B6 (pyridoxine) deficiency manifests as dermatitis, anemia, convulsions, neurologic symptoms, and abdominal distress in infants. It may also be seen in combination with other B-complex vitamin deficiencies.

24.

A 10-year-old male presents to urgent care with complaints of a persistent cough for 10 days and fever of 102.5 F (39.17 C) for the past 2 days. He complains of a headache and extreme fatigue. His respirations are 24 breaths per minute, his pulse is 105 beats per minute, and O₂ saturations are at 88%. Upon auscultation of his lungs, you note diminished breath sounds in the left lower lung base.

Which of the following organisms is MOST likely to be contributing to his illness?

Mycoplasma pneumonia

Respiratory syncytial virus (RSV)

Chlamydia trachomatis

Haemophilus influenza

Correct answer: Mycoplasma pneumonia

The most common causes of community-acquired bacterial pneumonia in school-aged children are M. pneumonia, C. pneumonia, S. pneumonia, and M. tuberculosis. M. pneumonia has a slow onset. A persistent cough, malaise, and headache are clinically present, and symptoms are generally accompanied by a high fever (>102.2 F).

H. influenza can cause community-acquired pneumonia in school-aged children but is less common. C. trachomatis typically causes pneumonia in a 1- to 3-month-old child, and viral pneumonia, such as RSV, is the most common cause of pneumonia in children younger than 2 years of age.

25.

What are the four parameters included in the clinical dehydration scale (CDS), an assessment tool used to help determine pediatric management of dehydration?

General appearance, eyes, moistness of mucous membranes, presence of tears

General appearance, capillary refill time, breathing, urine output

General appearance, eyes, capillary refill time, urine output

General appearance, moistness of mucous membranes, mental status, heart rate

Correct answer: General appearance, eyes, moistness of mucous membranes, presence of tears

Dehydration is overwhelmingly the result of an infectious process, most often viral, that causes diarrhea in many cases. Children are at an increased risk due to their higher surface area-to-volume ratios, higher rate of insensible loss, and in younger children, their inability to communicate or actively replace losses.

A clinical dehydration scale (CDS) is a predictive tool regarding length of stay and need for intravenous (IV) fluids. The four parameters used for assessment are:

- *General appearance*
- *Eyes (sunken or not)*
- *Moistness of mucous membranes*
- *Presence of tears*

Capillary refill time (CRT), breathing, urine output, mental status, and heart rate are all signs and symptoms that are part of the clinical picture of the child's hydration status, but are not parameters used for the CDS predictive tool regarding management.

26.

You are assessing a 2-day-old male and observe a distended abdomen with hypoactive bowel sounds, and he has not yet passed a meconium stool. You order an x-ray of the newborn's abdomen, which reveals dilated loops of bowel. A biopsy determines the diagnosis of Hirschsprung disease.

Which of the following statements regarding Hirschsprung disease is CORRECT?

It involves an absence of ganglion cells in the bowel wall

It is often diffuse, though rare cases involve focal parts of the colon

It is common in children with achondroplasia

It results in a portion of the colon having no secretory function

Correct answer: It involves an absence of ganglion cells in the bowel wall

Hirschsprung disease is an absence of ganglion cells in the bowel wall. It is the most common cause of neonatal colon obstruction and accounts for approximately 33% of all neonatal obstructions. It is not often diffuse, rather localized to a particular area of the bowel, most often in the rectosigmoid region, resulting in a portion of the colon having no motility. It is common in children with trisomy 21 (Down syndrome), and affects males four times more commonly than females.

Clinical findings include failure to pass meconium in the first 48 hours of life, failure to thrive, poor feeding, chronic constipation, vomiting, abdominal obstruction, diarrhea, explosive bowel movements, or flatus. The infant may also present with a distended abdomen with bilious vomiting.

27.

Which of the following is a major manifestation of the revised Jones criteria used in the diagnosis of acute rheumatic fever (ARF)?

Subcutaneous nodules

Fever

Elevated acute-phase reactants

A positive throat culture

Correct answer: Subcutaneous nodules

The diagnosis of an initial attack of ARF is based on evidence of documented GAS pharyngeal infection, and the revised Jones criteria: findings of two major manifestations, or one major and two minor manifestations of ARF.

Major manifestations include:

- *Carditis*
- *Polyarthritits*
- *Chorea*
- *Erythema marginatum*
- *Subcutaneous nodules*

Minor manifestations include:

- *Clinical fever, polyarthralgia*
 - *Laboratory elevated acute phase reactants (ESR or leukocyte count)*
-

28.

Which of the following countries has the HIGHEST incidence of Kawasaki Disease (KD), the second most common childhood vasculitis?

Japan

India

United Kingdom

United States

Correct answer: Japan

KD is a medium-vessel vasculitis of childhood with a predilection for the coronary arteries. It presents as an acute febrile illness, and is the leading cause of acquired heart disease. It is the second most common childhood vasculitis, with a varying incidence from country to country.

Japan has the highest incidence of 264.8 per 100,000 in 2012 in children from 0 to 4 years. This is a 10- to 20-fold increase of incidence in Japan, as compared to the United States and United Kingdom. Children of Asian/Pacific Islander descent have the highest rate of hospitalization in the United States, pointing to the role genetics play in the disease.

29.

In a child with an initial diagnosis of type 1 diabetes mellitus (T1DM), which of the following conditions is MOST likely to occur concomitantly?

Thyroiditis

Polycystic ovary syndrome (PCOS)

Addison's disease

Precocious puberty

Correct answer: Thyroiditis

The provider should always screen for concomitant associated autoimmune conditions. About 25% of children with T1DM have thyroid autoantibodies present at the time of diagnosis, which is predictive of thyroid dysfunction. Hypothyroidism is more common, although hyperthyroidism (Graves' disease) may also be present. Currently, the American Diabetes Association (ADA) recommends screening children with T1DM annually for autoimmune thyroid disease by obtaining a TSH level.

Celiac disease also occurs more frequently in children with diabetes (1% to 6%) compared to children without, although it's not nearly as common as thyroid dysfunction. Addison's disease is found in less than 1% of patients with type 1 diabetes, and there is little correlation between precocious puberty and PCOS, and type 1 diabetes.

30.

Which of the following is NOT a likely cause of childhood nocturnal enuresis?

Infrequent voiding during the day

Familial disposition

Behavioral comorbidities

Constipation

Correct answer: Infrequent voiding during the day

Nocturnal enuresis is a common complaint in children, defined as incontinence during sleep. Primary enuresis occurs in children who have never attained dryness at night, whereas secondary enuresis occurs in children who have previously been potty trained. The diagnosis of secondary enuresis requires a minimum age of 5 years old, and one episode a month for a duration of 3 months. The cause of enuresis varies among children and can be difficult to determine.

A number of factors have been found to be associated with enuresis, including:

- *Constipation*
- *Familial disposition*
- *Neurologic developmental delay*
- *Behavioral comorbidities (strong association between enuresis and ADHD)*
- *Functional small bladder capacity*
- *Sleep disorders*
- *Stress and family disruptions*
- *Polyuria*
- *Inappropriate toilet training (especially common when parents are overly demanding or punitive of the child)*

Infrequent voiding is a clinical finding (symptom) associated with dysfunctional voiding or a problem of bladder emptying.

31.

The rule of Two/Too is a mnemonic for gathering and interpreting:

Genetic health data

Environmental health issues

Developmental milestones

Obstetric history

Correct answer: Genetic health data

Genetic red flags indicate the potential for genetic risk. One way to remember the important components to look for or ask about when taking a health history is the rule of Two/Too, a mnemonic for gathering and interpreting genetic health data.

The rule of Two/Too is as follows: Too many of something: a person is too tall, too short, too early, too young, too different, etc., or Two birth defects, two cancers, two in a family, or two generations involved.

32.

In a child with microcephaly caused by primary craniosynostosis, it is important to know in your INITIAL exam that:

Sagittal suture involvement is most common, but posterior plagiocephaly is usually benign

There is a male to female ratio of 1:2 in Caucasian children

Synostosis which requires surgery should be done as soon as possible

Computed tomography (CT) will give you the best idea of whether or not a suture is closed, and is considered the standard in evaluation of bradycephaly

Correct answer: Sagittal suture involvement is most common, but posterior plagiocephaly is usually benign

Primary or "true" craniosynostosis involves premature closure or absence of one or more cranial sutures. Primary craniosynostosis occurs in 1 per 2,000 to 2,500 births, is ethnically neutral, and can vary in type and prominence between genders. Early fusion of the sagittal suture is the most common craniosynostosis, accounting for about half of all cases.

In posterior plagiocephaly, which is pressure-related, occipital flattening is usually self-limited, benign, and due to positional pressures on the growing cranium (an infant spending more time on his back than on his tummy). Predominant causes of posterior plagiocephaly are craniosynostosis of the lambdoidal sutures, or positional suture molding (vast majority).

Synostosis treatment is often surgical, though it is not generally done until 6-12 months of age as some other, less invasive measures are tried first. Primary craniosynostosis occurs in one per 2000 to 2500 births, is ethnically neutral, and can vary in type and prominence between genders.

While CT is considered the standard for a skull shape deformity, this is an aspect of management and not part of the initial exam (never neglect to perform a thorough initial exam simply because you suspect you will be ordering a CT). Deformational plagiocephaly does not require imaging studies in most situations when the history and physical examination are diagnostic.

33.

Cardiac syncope in a child that manifests with cyanosis, pallor, and shortness of breath MOST likely originates from:

Pulmonary hypertension

Mitral stenosis

Aortic stenosis

Arrhythmias

Correct answer: Pulmonary hypertension

Syncope is the transient loss of consciousness due to a decrease in cerebral blood flow. Recovery is relatively prompt, and most episodes in children are benign. Approximately 95% of syncope is vasovagal.

However, syncope associated with cardiac causes, such as primary pulmonary hypertension, is often clinically silent until severe symptoms are present. Pulmonary hypertension is the only etiology that includes cyanosis as a major clinical manifestation, and often syncope associated with pulmonary hypertension accompanies exercise.

34.

A homeless African-American woman brings her 3-year-old daughter in to the Emergency Department. She reports that her daughter has had a fever, cough, and congestion for the past 7 days, which she has been treating with acetaminophen and ibuprofen. After a thorough history and physical examination, including a chest x-ray, you diagnose the child with pneumonia.

The MOST likely factor that caused the mother to delay bringing her daughter in for care sooner is:

Poverty

Ethnicity

Lack of education

Neglect

Correct answer: Poverty

Child health is fundamental to overall child development, and children with health insurance are more likely to have a regular source of care and access to preventive healthcare services.

Lack of health care insurance is the single strongest predictor of quality of care for children in the United States. Poverty is a primary reason why the homeless delay receiving care for themselves or family members. If the homeless person does not qualify for healthcare assistance, this can be a huge barrier to accessing care.

The other answer choices can be reasons for delaying care, but the most likely reason is poverty.

35.

Which of the following is a major clinical manifestation (Jones criteria) of acute rheumatic fever (ARF)?

Carditis

Fever

Polyarthralgia

Elevated acute-phase reactants (ESR or leukocyte count)

Correct answer: Carditis

ARF is a nonsuppurative complication following a sequela of streptococcal infection, typically 2 to 3 weeks after group A streptococcal (GAS) pharyngitis. It results in an autoimmune inflammatory process involving the joints (polyarthritis), heart (rheumatic heart disease), CNS (Sydenham chorea) and subcutaneous tissue (subcutaneous nodules and erythema marginatum). It most commonly presents between the ages of five and 15 years old. Long-term effects on tissues are generally mild except for the damage done to cardiac valves, leaving fibrosis and scarring that results in rheumatic heart disease.

The diagnosis of an initial attack of ARF is based on the following revised Jones criteria:

- *Evidence of documented GAS pharyngeal infection (culture, rapid strep antigen test, or ASO titer)*
- *Findings of two major manifestations or one major and one minor manifestation of ARF*

Major manifestations include:

- *Carditis (pancarditis, valves, pericardium, myocardium)*
- *Polyarthritis (migratory and painful)*
- *Chorea (uncoordinated jerking movements of face, hands, feet)*
- *Erythema marginatum (nonpruritic rash involving pink rings on torso and limbs)*
- *Subcutaneous nodules*

Minor manifestations include:

- *Clinical fever, polyarthralgia*

- *Laboratory elevated acute phase reactants (ESR or leukocyte count)*
-

36.

What is the gold standard for diagnosis of cystic fibrosis (CF)?

Pilocarpine iontophoresis sweat test

One or more of the following clinical features: chronic sinopulmonary disease, GI and nutritional abnormalities, salt loss syndrome, chronic metabolic alkalosis, and/or male urogenital abnormalities

Newborn screening

Pulmonary function tests

Correct answer: Pilocarpine iontophoresis sweat test

CF is a multisystem genetic disorder manifested by chronic obstructive pulmonary disease (COPD), GI disturbances, and exocrine dysfunction. It is the most common autosomal-recessive disease. It affects the sodium-chloride transport gene and impairs the lungs, exocrine pancreas, and the vas deferens.

The sweat test is considered the gold standard for diagnosing CF. The child must have one or more of the clinical features of CF before ordering the sweat test. Newborn screening (NBS) is now done in all 50 states that may measure immunoreactive trypsinogen (IRT) in the newborn's blood. If IRT is elevated, sweat testing is performed. Pulmonary function tests (PFTs) are used to follow the clinical course.

In addition, the diagnosis of CF can be made in patients with clinical features of the disease if:

- Sweat chloride is greater than 60 mmol/L*
 - The concentration of sweat chloride is in the intermediate range of 30 to 59 mmol/L for infants younger than 6 months old, or 40 to 59 mmol/L for older individuals*
 - The child has two disease-causing CFTR mutations*
-

37.

Medical care and screening with a pediatric primary care provider should be arranged for a child refugee within how many days of entry into the community?

90 days

60 days

30 days

120 days

Correct answer: 90 days

The domestic health assessment must take place within 90 days of arrival; however, within 30 days is ideal. If an applicant has a medical waiver for entry, health care should be arranged sooner than 90 days. The CDC Division of Global Migration and Quarantine is responsible for notifying state and local health departments of new arrivals who need medical treatment and/or follow-up.

The first primary care visit with a pediatric provider for immigrants, including refugees and international adoptees, should include a review of all medical records, a medical history, a developmental assessment, a psychosocial assessment, a complete physical exam and measurements, and tiered laboratory testing based on country of origin and risk factors.

38.

Which of the following statements regarding childhood type 1 and type 2 diabetes mellitus is FALSE?

Islet autoimmunity is uncommon in type 1 diabetes and present in type 2 diabetes

Type 1 diabetes can occur at any age, whereas type 2 frequently occurs when the child is 10 years old or older

Obesity is clinically found in over 90% of cases of type 2 diabetes in a child, whereas it is not related to type 1 diabetes

Insulin sensitivity is normal in type 1 diabetes and decreased in type 2 diabetes

Correct answer: Islet autoimmunity is uncommon in type 1 diabetes and present in type 2 diabetes

Type 1 diabetes is caused by the autoimmune destruction of pancreatic beta cells in the islet of Langerhans and thought to be triggered by a preceding environmental event in genetically susceptible individuals.

The prevalence of type 2 diabetes in children ages 10 to 19 years old is 0.46 per 1000 youth, increasing by approximately 30% over the period from 2001 to 2009.

Type 2 diabetes is strongly associated with environmental factors, such as obesity, sedentary lifestyles, and high-calorie, high-fat diets and is associated with decreased insulin sensitivity.

39.

The child with which of the following conditions is MOST LIKELY to also have an anxiety disorder?

Fragile X syndrome

Epilepsy

Rett Syndrome

Prader-Willi syndrome (PWS)

Correct answer: Fragile X syndrome

Fragile X syndrome is the most commonly diagnosed genetic cause of intellectual disability (ID) in males. It affects 1 in 4,000 males and 1 in 8,000 females. It is found in all racial and ethnic groups and is the result of an inheritable unstable DNA in the FMR1 gene of the X chromosome.

It is a genetic condition that causes a range of developmental delays and behavioral problems, including speech and language delays, cognitive deficits, attention difficulty, anxiety disorder, aggression, and autism.

Anxiety may be present in the other conditions listed but is not as common as with Fragile X syndrome.

40.

A 14-year-old male comes into the pediatric clinic for complaints of a rash. This patient was told by his wrestling coach that he needs a note from the doctor that he has been seen and treated for the rash, or that it is not contagious. He states that the rash is slightly itchy, but otherwise does not bother him much. He noticed it about 2 weeks ago and says it began as a single, round pink patch that was scaly with a raised border, and states he has been using an over-the-counter antifungal cream in case it was ringworm, but has not noticed much improvement. He reports not eating anything new or different that he can remember in the past several weeks. The rash is located on his back and wraps around his trunk horizontally. It is in a "Christmas tree" pattern, following the dermatome skin lines. He reports he has not had a history of illness and is an otherwise healthy adolescent. He is up-to-date on his vaccinations.

What is the MOST likely diagnosis?

Pityriasis rosea

Psoriasis

Stevens-Johnson syndrome (SJS)

Angioedema

Correct answer: Pityriasis rosea

Pityriasis rosea (meaning rose-colored flaking) is a common, mild, self-limiting rash that begins with a herald spot or patch. This herald spot occurs in 70% of presentations, and it typically presents on the trunk, upper arm, neck, or thigh. This disease most commonly occurs in adolescents, and up to 98% of cases result in lifelong immunity.

Although the etiology has not been established, it is thought that it may be caused by human herpesvirus 6 or 7 (HHV-6 or HHV-7). It is minimally contagious and generally resolves spontaneously in 6 to 12 weeks. Recurrence is common. Management involves calamine lotion, tepid baths with Aveeno, antihistamines, and emollients as needed for itching. Oral erythromycin may expedite the resolution of the eruption.

41.

Which of the following is the MOST common cardiac arrhythmia in children?

Supraventricular tachycardias (SVTs)

Atrial flutter

Complete AV block

Atrial fibrillation

Correct answer: Supraventricular tachycardias (SVTs)

Arrhythmias can manifest as a primary disorder or as a consequence of a cardiac or other systemic disorder. Sinus tachycardia is caused by predisposing factors that increase cardiac output, such as fever, anxiety, infection, pain, and dehydration.

SVTs are the most common pathologic arrhythmia in children. Diagnosis is usually made by capturing it on an ECG or by Holter monitor. Management includes vagal maneuvers, IV adenosine, or if necessary, synchronized cardioversion. Long-term management involves prevention of recurrence with beta-blockade, or digoxin therapy (as long as the patient does not have Wolff-Parkinson-White syndrome).

The other answer choices are rare in the pediatric population.

42.

You are seeing a 12-year-old female for her annual school physical exam. She has no complaints and no significant medical history. She enjoys school. Her vital signs are normal; she is 58 inches tall and weighs 150 pounds. She is at the 97th percentile for weight and the 37th percentile for height. Her BMI is 31.3, placing her above the 95th percentile for her age.

What would your next step in management be at this time regarding this patient's weight?

Obtain a fasting glucose, insulin and lipid levels, total cholesterol, and liver function tests for further evaluation of health status

Recommend eliminating sugary foods and returning for a follow-up in 6 months

Submit a referral to a comprehensive weight-loss program

Recommend a VLCD (very low-calorie diet), increased fiber intake, and encourage incorporating moderate-to-vigorous activity into her daily routine

Correct answer: Obtain a fasting glucose, insulin and lipid levels, total cholesterol, and liver function tests for further evaluation of health status

For a BMI of greater than 95% or if BMI is greater than 85% and other risk factors are present, such as family history of diabetes or cardiovascular disease, labwork should be obtained to assess for diabetes, hyperlipidemia, and metabolic syndrome in children 4 years or older.

You should also create a healthy eating plan including family-focused education, registered dietician counseling, and increased daily moderate-to-vigorous activity.

43.

A 5-month-old female with a diagnosed congenital heart defect, poor feeding, hypotonia, constipation, and recurrent otitis media is referred to a geneticist for developmental delay. Chromosome analysis is consistent with DiGeorge syndrome.

This syndrome is caused by a deletion in which chromosomal region?

22q11

7q11

5p

15q11

Correct answer: 22q11

The patient in this scenario is exhibiting characteristics of DiGeorge syndrome (also known as velocardiofacial syndrome), caused by a deletion in chromosome 22q11.

Deletions in chromosomes 7q11 are associated with Williams syndrome. Deletions in 5p are associated with Cri-du-Chat syndrome, and Prader-Willi syndrome is caused by a deletion in 15q11 (deletion in the paternally-derived chromosome 15).

44.

A 13-year-old male presents to your pediatric clinic with complaints of left-sided hip and groin pain for several weeks. Upon physical examination, you note limited abduction and extension of the left leg and external rotation of the thigh when the hip is in a flexed position. The patient's x-ray reveals slippage of the femoral epiphysis.

Which of the following is the MOST common historical finding associated with this condition?

Obesity

Leukocytosis

Diabetes mellitus

Hypothyroidism

Correct answer: Obesity

Slipped capital femoral epiphysis (SCFE) is a medical term referring to a Salter-Harris type I fracture through the proximal femoral physis, or growth plate, which results in slippage of the overlying end of the femur (epiphysis). Stress around the hip causes a shear force to be applied at the growth plate.

Normally, the head of the femur, called the capital, should sit squarely on the femoral neck. Abnormal movement along the growth plate results in the slip. The femoral epiphysis remains in the acetabulum, while the metaphysis moves in an anterior direction with external rotation.

SCFE is the most common hip disorder in adolescence. SCFEs usually cause groin pain on the affected side, but sometimes cause knee or hip/thigh pain. It generally occurs in obese adolescents with delayed skeletal maturation. SCFEs often occur in obese adolescent males, especially young African American males.

Symptoms include the gradual, progressive onset of thigh or knee pain with a painful limp and limited hip motion. Management involves a multidisciplinary approach, including consultation with an orthopedic surgeon for surgical repair.

45.

The MOST reliable method for diagnosing varicella is:

Polymerase chain reaction (PCR)

Direct fluorescent antibody (DFA)

Tzanck smear

Culture of vesicular fluid

Correct answer: Polymerase chain reaction (PCR)

As the incidence of varicella disease has decreased, many providers may be unfamiliar with the clinical presentation of the disease, especially in mild cases with few lesions. Diagnostic studies can play an important role in these instances. For both unvaccinated and vaccinated persons, the most reliable method of diagnosing VZV is the PCR (preferred) or DFA done from scrapings of a vesicle base during the first 3 to 4 days post-eruption.

Tzanck smears of lesions demonstrate multinucleated giant cells; however, these are not specific for VZV.

A positive serologic test for varicella-zoster IgM antibody is also confirmatory. The virus can be cultured from vesicular fluid, CSF, and/or biopsy of tissue, but is less sensitive than that of the PCR. The WBC count is usually within normal limits.

46.

Which of the following statements is ACCURATE about the CRAFFT questionnaire?

It is a screening instrument for substance abuse in the primary care setting

It is not appropriate for screening in adolescents

A total score of 3 or higher is a positive screen, indicating a need for further evaluation

It is best to ask these questions with a parent or caregiver present

Correct answer: It is a screening instrument for substance abuse in the primary care setting

The CRAFFT questionnaire is an appropriate screening instrument for substance abuse in the primary care setting; it is used with children under the age of 21, primarily among adolescent clinic patients. Positive responses to two or more items indicate a high likelihood for substance abuse and merits further evaluation and treatment.

Interviewing the adolescent with parents is a key strategy for obtaining information about etiologic factors and any recent changes in behavior, cognitive, emotion and overall health. However, it is essential to interview the adolescent alone as well at every visit to assess mental health and family issues (the patient needs to be alone when asking CRAFFT questions).

47.

Russell sign is an indication of which of the following psychiatric disorders, which may be seen in the pediatric population?

Bulimia

Anorexia

Oppositional defiant disorder (ODD)

Post-traumatic stress disorder (PTSD)

Correct answer: Bulimia

Bulimia is an eating disorder characterized by bingeing and purging behaviors that occur at least once a week for at least 3 months. If a patient controls caloric intake through purging via vomiting, findings may include hypertrophic salivary glands, dental erosions secondary to gastric acid irritation, and abrasions or calluses on the dorsum of the hand secondary to manual induction of vomiting. This is referred to as Russell sign, which is cuts, calluses, and/or abrasions to knuckles from inducing vomiting, and is a common finding in patients with bulimia.

48.

Which of the following statements is CORRECT regarding the diagnosis of learning disabilities?

There are correctable causes of cognitive impairments and behavioral disturbances

Learning ability assessment can be performed by assessing developmental history

Learning disabilities are best diagnosed by positive laboratory findings using tests that are validated for this purpose

Neuropsychological testing is the gold standard for diagnosing learning disabilities

Correct answer: There are correctable causes of cognitive impairments and behavioral disturbances

Correctable causes of cognitive and behavioral disturbances include seizure disorder, recent head injury, and hearing and/or vision impairments.

Developmental history alone is inadequate for diagnosis. Thorough assessment includes identification of risk factors, observation for characteristics of learning disorders, and consideration of other causes of learning problems (familial, decreased academic achievement, attention deficits, etc.). In addition, medical history is important to factor in as well.

Multiple causes of learning disabilities may be determined by laboratory findings, such as metabolic causes, toxin exposure, and chromosomal abnormalities. However, there is no single laboratory analysis for diagnosing learning disorders. Many individuals with developmental delay do not have any laboratory abnormalities. There is no gold standard for diagnosing learning disabilities; neuropsychological testing helps in confirming the diagnosis.

49.

Which of the following conditions is thought to be the MOST frequent reason for intestinal obstruction in children?

Intussusception

Hernias

Inflammatory bowel disease (IBD)

Impacted feces

Correct answer: Intussusception

Intussusception involves a section of intestine being pulled antegrade into adjacent intestine with the proximal bowel trapped in the distal segment. It is thought to be the most common cause of intestinal obstruction in children. It most often occurs between 5 and 10 months of age and is also the most common cause of intestinal obstruction in children 3 months to 6 years old, with 80% of cases occurring before 2 years of age. In younger infants, it is generally idiopathic and responds to noninvasive treatments.

Known medical factors, such as polyps, HSP, lipomas, foreign bodies, and rotavirus can be contributing factors in other children. Intussusception may be a complication of cystic fibrosis as well.

The other answer choices are less frequent reasons for pediatric intestinal obstructions.

50.

What is the MOST common pulmonary disease in a newborn?

Neonatal respiratory distress syndrome (NRDS)

Transient tachypnea of the newborn (TTN)

Bronchopulmonary dysplasia (BPD)

Respiratory syncytial virus (RSV)

Correct answer: Neonatal respiratory distress syndrome (NRDS)

NRDS, formerly known as hyaline membrane disease, is a syndrome caused by developmental insufficiency of pulmonary surfactant production and structural immaturity in the lungs. It can also be a consequence of neonatal infection. The result is alveolar atelectasis and decreased lung compliance. It is increasingly referred to as surfactant deficiency disorder (SDD).

Most cases of NRDS occur in newborns before 37 to 39 weeks. The more premature the baby is, the higher the chance of respiratory distress syndrome (RDS) after birth. NRDS can also be due to genetic problems with lung development. Antenatal steroids, postnatal surfactant, and newer ventilation techniques have reduced mortality from RDS to approximately 10%.

History and clinical findings include: sibling who had RDS, maternal diabetes, cesarean delivery or induction of labor before the baby is full-term, problems with delivery that reduce blood flow to fetus/newborn, multiple pregnancy (twins or more), and preterm, precipitous delivery.

51.

Which of the following genetic tests is used to identify and evaluate the size, shape, and number of chromosomes?

Karyotype

Fluorescence in situ hybridization (FISH)

Biochemical testing

Chromosomal microarray

Correct answer: Karyotype

Diagnostic genetic testing is used to confirm a diagnosis and is used in a symptomatic individual or in response to a positive screening test. Such tests are selected depending on the type or specific disease one is trying to confirm. The main types of diagnostic genetic testing are:

- *Karyotype*
- *FISH*
- *Biochemical testing*
- *Chromosomal microarray*
- *Molecular testing*
- *Next generation sequencing*

Karyotype testing is used to identify and evaluate the size, shape, and number of chromosomes.

FISH is used to locate and detect a specific area of a particular chromosome, including subtle missing, additional, or rearranged chromosomal material, by labeling a known chromosome sequence with fluorescent tags to see the location of genetic material.

Biochemical testing is used to study the amount, activity level, or structure of proteins and enzymes that result from gene mutations.

Chromosomal microarray is used to detect microdeletions or duplications in any of the chromosomes but not specific gene mutations.

52.

Leukemia is the most common form of childhood cancer, accounting for up to 30% of all pediatric cancers. Which of the following types of leukemia accounts for nearly 80% of childhood leukemia cases?

Acute lymphocytic leukemia (ALL)

Acute myeloid leukemia (AML)

Chronic lymphocytic leukemia (CLL)

Chronic myeloid leukemia (CML)

Correct answer: Acute lymphocytic leukemia (ALL)

Leukemias are classified according to cell type involvement (lymphocytic or nonlymphocytic) and by cellular differentiation. ALL is characterized by predominantly undifferentiated white blood cells (WBCs).

ALL accounts for about 80% of childhood leukemia cases, with a peak incidence between 2 and 6 years of age, and 56% of leukemia cases in adolescents. There have been dramatic improvements in survival for ALL over the past four decades, with outcomes approaching 90% in the latest studies.

AML is less common in children than ALL and accounts for about 15% of leukemia cases in children and 31% of those in adolescents.

53.

Which of the following statements is CORRECT in regards to the diagnosis of ADHD in children?

Caucasian children are more likely to be diagnosed with ADHD than those who are African-American, Hispanic, or of other races/ethnicities

The average age of diagnosis is 9 years old

ADHD is one of the least commonly diagnosed disorders in childhood

Brain imaging studies have identified structural and functional differences in the temporal lobes and the hypothalamus

Correct answer: Caucasian children are more likely to be diagnosed with ADHD than those who are African-American, Hispanic, or of other races/ethnicities

ADHD is one of the most commonly diagnosed disorders in childhood. Children who are African-American, Hispanic, or of other races are 69% less likely to be diagnosed with ADHD than white children.

The average age of diagnosis is 7 years old, with children who have severe ADHD diagnosed earlier. Brain imaging studies have identified structural and functional differences in the frontal lobes and subcortical structures of the brain, indicating that brain development is drastically different from normal.

54.

You are seeing a new patient today, a 12-month-old female whose family recently moved from out of state. The mother informs you that her daughter has had recurrent ear infections and upper respiratory infections, with two rounds of pneumonia since birth. In addition, she has severe eczema and seems to bruise easily. You note diffuse petechiae on the patient's body and decide to do a complete blood count (CBC), which shows thrombocytopenia.

Which of the following conditions does this patient MOST likely have?

Wiskott-Aldrich syndrome

Severe combined immunodeficiency (SCID)

DiGeorge syndrome

Selective IgA

Correct answer: Wiskott-Aldrich syndrome

Wiskott-Aldrich syndrome is an X-linked recessive, immunodeficiency disorder characterized by the clinical triad of thrombocytopenia, eczema (atopic dermatitis), and recurrent infections. In addition, bloody stools and/or hemorrhage are often present as a result of thrombocytopenia, and EBV malignancy (Burkitt lymphoma) may be part of the clinical presentation in the patient with this syndrome.

The other answer choices are also pediatric primary immunodeficiency disorders but present differently.

55.

A 7-year-old female in the first grade is brought in for a clinic appointment by her parents because they are concerned that she is not yet reading, and is, therefore, falling behind her peers. They report that she struggles to identify letters correctly, write on the lines of the paper, and use scissors. Despite reassurance from her teachers that "she is a diligent student who tries hard and will catch up," they have sought extra assistance outside of school. They also report that she has difficulties interacting with other children her age and has very few friends when compared to her 10-year-old brother. Her teacher reports that she is quiet and shy and often overshadowed by her peers.

Which of the following tests would be MOST appropriate to further assess her development?

Parent's Evaluation of Developmental Status (PEDS)

Vanderbilt Assessment Scale

Ages and Stages Questionnaire (ASQ)

Patient Health Questionnaire (PHQ)

Correct answer: Parent's Evaluation of Developmental Status (PEDS)

The PEDS is a parent interview screening/surveillance form that is designed to screen development, social-emotional, behavior, and mental health domains in the child, and identifies children at low, moderate, and high-risk for disabilities and delays. Parents are asked to complete ten questions, only taking a few minutes to complete them.

It is appropriate for use from birth to 8 years old, and available in English, Spanish, Vietnamese, and many other languages, making it readily accessible for many families whose primary language is not English.

The Vanderbilt Assessment Scale is a tool that is used for school-age children and adolescents to evaluate inattention, hyperactivity, conduct disorders, and anxiety or depression.

ASQ is used to screen for social-emotional development in the three- to 60-month-old child.

PHQ is used for both children and adults as a psychosocial screening tool for depression and suicide.

56.

A 3-day-old male presents to the pediatric clinic with a 2-day history of serosanguinous discharge, edematous bulbar, and palpebral conjunctiva to both eyes. The mother reports that she had a vaginal birth at home, with a midwife present. You decide to treat the baby for *N. gonorrhoeae* conjunctivitis because the mother is unsure if prophylactic eye ointment was given. The birth was uncomplicated. Mom denies having a (sexually transmitted infection) STI before.

In the case that the prophylaxis eye ointment was given, what is the MOST likely cause of symptoms?

Chemical conjunctivitis

Chlamydial conjunctivitis

Gonococcal conjunctivitis

Ophthalmia neonatorum

Correct answer: Chemical conjunctivitis

A prior common cause of conjunctivitis in the newborn was chemical conjunctivitis from the prophylactic administration of silver nitrate at birth, which is one reason the product is no longer recommended.

Chemical conjunctivitis usually occurs in the first 24 to 72 hours of life and frequently manifests as nonpurulent drainage and edematous bulbar and palpebral conjunctiva. It generally resolves without specific treatment within 3 to 4 days.

*Chlamydial and gonococcal conjunctivitis are two types of ophthalmia neonatorum, a neonatal infection contracted by newborns during delivery from a mother infected with either *Neisseria gonorrhoeae* or *Chlamydia trachomatis*. The U.S. Preventive Services Task Force recommends prophylactic topical 1% tetracycline or 0.5% erythromycin ointment administered within 24 hours after birth for prevention of gonococcal conjunctivitis, as this is the most serious cause of ophthalmia neonatorum owing to concerns of the bacteria causing corneal perforation and blindness.*

57.

Anorexia, growth retardation, skin changes, and immunologic abnormalities are all signs of a deficit in which of the following minerals?

Zinc

Iron

Magnesium

Calcium

Correct answer: Zinc

Zinc is a mineral that aids in cellular metabolism, growth, and repair. It is found in meats, animal products, seafood (especially oysters), and eggs. You should suspect a zinc deficiency in the child who presents with anorexia, growth retardation, skin changes, and immunologic abnormalities.

Signs of too much zinc include GI upset, vomiting, acute toxicity, and impaired immune response.

58.

The initial sign of male puberty is:

Testicular enlargement

Development of pubic hair

Change in the voice

Rapid growth in height

Correct answer: Testicular enlargement

The initial sign of puberty in males is testicular enlargement, on average occurring at 11.5 years old. The growth of the testes occurs approximately 6 months before the development of pubic hair in most males. Once puberty begins, the left testis generally hangs lower than the right. If testicular enlargement does not precede other changes, the provider should consider whether the adolescent is taking exogenous anabolic steroids.

Change in the male voice coincides with peak height velocity (PHV). Rapid growth in height (PHV) tends to occur later in puberty for boys. Boys generally lag about 2 years behind girls, but 95% have their growth spurt between 12 and 16 years old. Males can continue to grow (although minimally) well beyond their teenage years, but most males complete linear growth by age 17.

59.

Both the American Academy of Pediatrics (AAP) and Bright Futures recommend blood pressure (BP) screenings at every well-child health visit beginning at what age?

3 years old

2 years old

4 years old

5 years old

Correct answer: 3 years old

Hypertension in infants and young children is most often secondary to another disease process, most commonly renal in origin. Increased rates of obesity can cause primary hypertension in older school-age children and adolescents; however, the GU system must be considered.

The AAP and the Bright Futures Practice Guidelines recommend routine blood pressure (BP) screening at every preventive health care visit beginning at 3 years old.

60.

What is the MOST common tumor in childhood?

Acquired melanocytic nevi

Wilms tumor

Osteosarcoma

Neuroblastoma

Correct answer: Acquired melanocytic nevi

Nevi are a common finding in children. The two most common types are vascular nevi and pigmented nevi.

Acquired melanocytic nevi, a type of pigmented nevi, are the most common tumor of childhood. They are caused by an overgrowth of pigment cells and are a type of lesion that contains nevus cells (a type of melanocyte). These acquired moles arise during early childhood and are a form of benign neoplasm, while congenital moles, or congenital nevi, are considered a minor malformation and may be at a higher risk for melanoma. Acquired melanocytic nevi are benign, light brown to dark brown to black, flat, or slightly raised, occurring anywhere on the body, especially on sun-exposed areas above the waist.

The other answer choices are common malignant childhood tumors.

61.

Muscle weakness, ataxia, confusion, anorexia, tachycardia, and heart failure in infants are all considered signs of a deficiency of which of the following vitamins?

Thiamin (Vitamin B1)

Riboflavin (Vitamin B2)

Niacin (Vitamin B3)

Folate (Folacin)

Correct answer: Thiamin (Vitamin B1)

These are all indicative of a thiamin (vitamin B1) deficiency. The patient's caregiver(s) should be counseled on dietary sources to increase this vitamin in the child's diet, such as whole grains, brewer's yeast, legumes, seeds and nuts, fortified grain products, organ meats, and lean cuts of pork.

A riboflavin (vitamin B2) deficiency would manifest with oral-buccal cavity lesions, generalized seborrheic dermatitis, scrotal and vulval skin changes, normocytic anemia, and dimness of vision.

Signs of niacin (vitamin B3) deficit are dermatitis, diarrhea, inflammation of mucous membranes, and indigestion.

In severe cases of folate deficiency (vitamin B9), megaloblastic anemia is present. Other signs include glossitis, GI disturbances, increased risk of neural tube defects and growth retardation in infants of folate-deficient mothers.

62.

An 18-year-old male presents to your office with complaints of scrotal pain and dysuria. Upon physical examination, you note scrotal edema and erythema with urethral discharge. When the testes are elevated, the patient reports immediate pain relief. Additionally, the cremasteric reflex is present.

What is the MOST likely diagnosis?

Epididymitis

Testicular torsion

Hydrocele

Inguinal hernia

Correct answer: Epididymitis

*Epididymitis presents with acute scrotal pain, edema, and erythema. It is most commonly caused by *N. gonorrhoeae* or *Chlamydia trachomatis* in the sexually active adolescent, often with infection in the urethra or bladder. However, it can be caused by a viral, coliform bacterial, or tubercular infection; by chemical irritation; by anomalies of the GU tract; or by dysfunctional voiding. It is rare before puberty, but it occurs in younger boys from *E. coli* infection.*

The epididymis is hard, indurated, enlarged, and tender. The cremasteric reflex is normal (not present in older adolescents), and Prehn sign can be elicited (pain relief with scrotal elevation). The urinalysis often shows pyuria.

In testicular torsion, Prehn sign is absent (pain increases with scrotal elevation), and urine is normal. A hydrocele usually presents with painless swelling, while an inguinal hernia presents with swelling most commonly found in the inguinal area, normal urine, and no urethral discharge.

63.

You are assessing a one-day-old newborn female for primitive reflexes. Upon eliciting the Moro reflex, the infant responds asymmetrically, indicating:

Paralysis or fractured clavicle

Brainstem problem

Spinal cord lesion(s)

CNS disease or severely depressed infant

Correct answer: Paralysis or fractured clavicle

When the Moro is elicited (presenting loud noise or allowing the infant's head to drop slightly), the infant should respond symmetrically with a "startle," in which both arms abduct and fingers first extend, then flex, then both arms adduct (come back together toward each other to midline). If the arms respond asymmetrically, paralysis or fractured clavicle is possible, and further assessment/diagnostic testing is indicated. The absence of a response indicates severe brainstem problem.

Spinal cord lesions should be ruled out in the presence of an asymmetrical response to trunk incurvation (Galant reflex). CNS disease or a severely depressed infant is likely in the case of an absent rooting reflex; however, do make sure the infant is awake and alert when eliciting the rooting reflex as a sleeping infant may not respond.

64.

Of the following clinical manifestations, which is NOT considered to be a mild allergic reaction to foods in the pediatric patient?

Mild laryngeal edema/mild asthma

Localized cutaneous erythema/urticaria

Angioedema

Rhinoconjunctivitis

Correct answer: Mild laryngeal edema/mild asthma

Redness of the skin and/or rash, either localized or generalized, facial swelling, and runny or congested nose with red eyes are all clinical features of a mild allergic reaction to food. In addition, gastrointestinal symptoms are generally caused by mild reactions to offending food(s).

However, any degree of laryngeal edema or asthma is a more severe clinical manifestation of food intolerance, which may require more immediate treatment.

Height and weight should be monitored closely in children with food allergies, because food elimination and use of alternative foods may compromise nutrition and affect growth.

65.

Which of the following population groups has the HIGHEST incidence of Henoch-Schonlein Purpura (HSP), the most common vasculitis of children?

Caucasians

African Americans

Hispanics

Asian Americans

Correct answer: Caucasians

HSP is the most common vasculitis of children and is a leukoclastic vasculitis of the small vessels. For the majority of children, the prognosis is excellent. HSP can occur anytime from infancy (as early as 6 months old) to adulthood, with Caucasians having the highest incidence and African Americans the lowest incidence.

HSP is seen slightly more in males than in females and occurs more frequently in the fall and winter months, pointing to an environmental trigger, including viral infection. While an upper respiratory infection often precedes HSP, a clear association between an infectious agent and HSP has not been found.

66.

A 7-year-old female presents to clinic with a rash on her hands, wrists, armpits, forearms, and genitalia. She complains that the rash itches severely, especially at night, but she does not have a history of fever. Her younger sibling has a similar rash. The physical examination reveals assorted vesicles, pustules, and papular lesions, concentrated on the webs of her fingers, sides of her hands, and folds in the axillae.

What condition does this child MOST likely have?

Scabies

Impetigo

Pediculosis

Tinea corporis

Correct answer: Scabies

*Scabies is caused by the mite, *Sarcoptes scabiei*, a human parasite that burrows into the skin and causes intense itching. Scabies is highly infectious, and is spread through close contact and sharing of linens or clothing. Sensitization, causing intense itching, occurs approximately 3 weeks after infestation.*

Characteristic lesions include curving S-shaped burrows, especially on the webs of fingers and sides of hands, folds of wrists and armpits, forearms, elbows, belt line, buttocks, genitalia, or proximal half of foot and heel.

Diagnostic studies include a microscopic exam of scrapings from an unscratched burrow in saline or mineral oil or the burrow ink test. Management involves 5% cream permethrin for affected individuals and others who have been exposed to the patient, as well as washing linens and clothing in hot water and vacuuming the home.

67.

This childhood disease, rarely seen in the United States, is characterized by a widespread rash indicating viremia and lymphocytopenia. Upper respiratory infection symptoms, high fever, and lethargy are predominant, and a maculopapular rash begins behind the ears and on the forehead, then moves progressively downward to cover the entire body. The entire process takes approximately 3 days.

What is this disease?

Measles

Mumps

Parainfluenza

Erythema infectiosum (fifth disease)

Correct answer: Measles

Measles is similar to mumps and influenza, and causes serious illness in children. Measles has a characteristic rash, indicating viremia (the presence of virus in the blood). It is spread primarily through respiratory secretions, but also through the blood and urine of infected persons, presents with an extensive rash and is highly contagious. This disease spreads within communities where there are larger numbers of unvaccinated or under-vaccinated individuals and where herd immunity falls below a critical point.

There is no specific treatment for the virus, other than vaccination within 72 hours of exposure to those who are vaccine-eligible and who were exposed. This is the first choice to prevent or modify the infection and may be given to infants 6 to 11 months old. Immune globulin (IGIM or IGIV) given within 6 days of exposure can be administered to prevent or modify the disease in those susceptible, regardless of their measles vaccination status. However, for the vast majority of healthy patients, it is not serious and resolves without treatment (supportive treatment).

68.

Which of the following clinical findings is a criterion for the diagnosis of systemic lupus erythematosus (SLE)?

Joint involvement

Leukocytosis

Microalbuminuria

Fatigue for at least 6 months unrelieved by rest

Correct answer: Joint involvement

SLE is a chronic, systemic rheumatic disease characterized by altered immune regulation that can involve inflammation in multi-organ systems, including the blood cells, kidneys, nervous system, and skin. It is associated with inflammatory damage to target organs brought on by autoantibodies attacking self-antigens and immune dysregulation. There is a strong genetic component, and it is more prevalent in females between puberty and menopause with a 6:1 female to male predilection.

Clinical findings depend on organ involvement, and presentation may be abrupt or have a gradual, nonspecific onset. Fever, rash, fatigue, and joint pain are the most typical presentation in children, and joint involvement is the most common initial finding.

Leukopenia or lymphopenia (not leukocytosis) are frequent laboratory findings. Proteinuria and hematuria (not microalbuminuria) are hallmarks of lupus nephritis. Fatigue for at least 6 months unrelieved by rest is a criterion necessary for a diagnosis of chronic fatigue syndrome.

69.

Which of the following is the MOST commonly identified cause of bacteremia in children?

Streptococcus pneumoniae

Haemophilus influenzae

Staphylococcus aureus

Neisseria meningitidis

Correct answer: Streptococcus pneumoniae

Streptococcus pneumoniae, or Pneumococcus, is by far the most common cause of pediatric bacteremia in all age groups except newborns.

The other choices are less common causes of bacteremia. When caused by Neisseria meningitidis bacteria, the condition is known as meningococcal bacteremia (meningitis). Meningococcal bacteremia has the worst prognosis among all the answer choices.

70.

Which of the following conditions should be suspected in the newborn with sepsis that has failed to respond to antibiotic therapy, and bacterial cultures are negative?

Herpes simplex virus (HSV)

Rubella

Mumps

Parvovirus B19 (fifth disease)

Correct answer: Herpes simplex virus (HSV)

Neonatal HSV disease should always be suspected in cases of neonatal sepsis or respiratory distress. Polymerase chain reaction (PCR) assay is the diagnostic method of choice, and best detection occurs through blood or CSF analysis. Parenteral acyclovir is the treatment of choice in life-threatening illness, neonatal infection, or disease in immunocompromised patients. Suppressive oral acyclovir therapy for six months after parenteral treatment of any classification of acute neonatal disease has been shown to improve neurodevelopmental outcomes and reduce recurrence of mucocutaneous lesions. In addition, the absolute neutrophil count (ANC) should be continuously monitored during the first six months of suppressive oral acyclovir therapy and stopped if neutropenia should occur, then restarted once resolved. Any new lesion(s) suggestive of HSV should be cultured.

71.

Of the following clinical findings, which is NOT a manifestation of nipple confusion?

Breast tenderness or pain

Ineffective suckling at the breast

Breast refusal

Sore, red, or bruised maternal nipples

Correct answer: Breast tenderness or pain

Nipple confusion (also referred to as nipple preference) happens when an infant is used to taking milk from a bottle nipple and is then introduced to the breast nipple. There are different oral-motor skills utilized in bottle-feeding versus breastfeeding, and infants who have been given a bottle nipple or a pacifier may try to breastfeed using the same sucking technique as with a bottle, making it problematic to obtain appropriate nutrition and may contribute to maternal sore nipples.

The infant with nipple confusion may exhibit signs of distress and frustration, and may cry, fuss, or push away with their arm during attempts to nurse. Clinical findings include ineffective suckling at the breast, breast refusal, and sore, red, or bruised maternal nipples.

Breast tenderness or pain is associated with mastitis, not nipple confusion.

72.

It is a common saying in pediatric medicine that a majority of diagnoses are made on the basis of the:

History

Physical examination

Family assessment

Laboratory and test data

Correct answer: History

A thorough, thoughtful health history is the first and often most critical step, because it helps to focus the clinician's diagnostic reasoning and guides the physical exam. A diagnosis is largely dependent upon the patient's history. While it is important to remember that the history and physical exam often occur simultaneously, the physical exam only provides a partial view of the situation, as it is just at that particular moment in time. It is often a cloudy picture, because the body commonly responds similarly to different assaults.

It is the history of the problem - its onset, duration, progress, associated symptoms, meaning, and effects on daily living - that brings the practitioner to an understanding to choose appropriate management.

Objective data, such as laboratory and test data, as well as the family assessment, are important aspects in determining a diagnosis. But most important is the history.

73.

Which of the following are signs of insufficient caloric intake?

Height and head circumference hold steady initially, but weight begins to drop

Weight, height, and head circumference are all significantly below the norms from birth

Head circumference is not affected, but weight and height are significantly affected from birth, or shortly thereafter

A weight-to-length ratio or weight-to-height ratio below the 25th percentile

Correct answer: Height and head circumference hold steady initially, but weight begins to drop

When a child is malnourished, regardless of the cause, the nutritional insult follows a predictable course. In the early stages, the child maintains or begins losing weight. Next, the child's linear growth slows or ceases altogether. Finally, the head circumference levels off, indicating compromised brain development.

Inadequate caloric intake should be suspected in any child with a weight-to-age ratio below the 10th percentile on standardized growth and BMI charts.

74.

A 3-year-old male with a history of feeding and growth problems has been referred to a geneticist due to global developmental delays, lack of speech, acquired microcephaly, an abnormal gait (arms held high/flexed elbows), and seizure disorder. He exhibits hand flapping, bouts of involuntary laughter, a happy affect, and abnormal sleep patterns.

Which of the following syndromes is he MOST likely exhibiting?

Angelman syndrome

Cerebral palsy (CP)

Autism

Prader-Willi syndrome

Correct answer: Angelman syndrome

This child is showing signs of Angelman syndrome, a rare neurogenetic disorder that is often misdiagnosed as CP or autism due to lack of awareness. Angleman syndrome is the result of an alteration (a gain or a loss) of genetic material from 15q11.2-13 in the maternally inherited allele (most commonly caused by a deletion in the maternally-derived copy of chromosome 15). This is the same chromosome that is affected in Prader-Willi syndrome (the result of alteration in the paternally inherited allele).

Characteristics of Angelman syndrome include developmental delay, lack of speech, seizures, and disorders affecting walking and balance. In addition, these patients frequently exhibit spontaneous persistent social smiling and fits of laughter, hand flapping, abnormal sleep, and often love the water.

75.

In the 6-month-old patient being seen for a follow-up visit for management of impetigo, you note a positive Nikolsky sign, in which the superficial layers of the skin peel with light rubbing, revealing a moist red surface underneath.

Which of the following complications do you suspect with this finding?

Staphylococcal scalded skin syndrome (SSSS)

Scarlet fever

Erythema multiforme (EM)

Cellulitis

Correct answer: Staphylococcal scalded skin syndrome (SSSS)

*SSSS is a complication of impetigo and is a blistering disease that results from circulating epidermolytic toxin-producing *S. aureus*. It is the most common in neonates (Ritter disease), infants, and children younger than 5 years old. It manifests abruptly with fever, malaise, and tender erythroderma, particularly in the neck folds and axillae. A positive Nikolsky sign is a key finding in this syndrome, and involves the ability to laterally spread a blister or slough the skin with the application of light tangential pressure, and is seen in most cases. Treatment may involve hospitalization and parenteral antibiotics.*

The other options are also complications of impetigo, but they do not present with this clinical picture.

76.

Which of the following is NOT a diagnostic criterion for anorexia/bulimia?

Refusal to maintain body weight at least 75% expected for age and height

Body dysmorphism

Bingeing or purging behaviors that occur at least once a week for at least 3 months

Intense fear of weight gain and "being fat"

Correct answer: Refusal to maintain body weight at least 75% expected for age and height

Diagnostic criteria for anorexia are:

- *Refusal to maintain body weight at least 85% expected for age and height, or failure to gain weight during growth periods so that weight drops below 85% expected for age and height*
- *Intense fear of weight gain and "being fat"*
- *Body dysmorphism*
- *Binge eating/purging subtype associated with frequent purging, although bingeing episodes are rare*

Diagnostic criteria for bulimia are:

- *Consuming large quantities of food over short periods of time (<2 hours)*
 - *Losing control during binge episodes (cannot control the amount of food they eat or are shocked at amount consumed)*
 - *Engaging in repeated behaviors to lose weight (purging, excessive exercise, fasting)*
 - *Bingeing or purging behaviors that occur at least once a week for at least 3 months*
-

77.

Which of the following is the MOST common cause of seizure activity in a full-term newborn?

Hypoxia

Hydrocephalus

Hypoglycemia

Intraventricular hemorrhage (IVH)

Correct answer: Hypoxia

Hypoxic-ischemic insults can occur from a variety of causes including placental abruption, maternal hemorrhage, cord compression, mechanical injury, maternal hypertension or diabetes, and inadequate resuscitation of the infant. Brain damage results from fetal hypoxia or ischemia over an extended period, followed by metabolic and respiratory acidosis. Hypoxia is the most common cause of seizure activity in the term neonate.

The other answer choices are less common causes of seizures seen in the term infant.

78.

In congenital craniosynostosis, which suture is MOST commonly fused, accounting for about half of all cases?

Sagittal suture

Coronal suture

Metopic suture

Lambdoidal suture

Correct answer: Sagittal suture

Skull malformations may be due to primary or secondary causes. Congenital (or "true" or "primary") craniosynostosis involves premature fusion of one or more cranial sutures.

Congenital craniosynostosis occurs in 1 per 2,000 to 2,500 births, is ethnically neutral, and can vary in type and prominence between genders. The sagittal suture is most commonly fused. This is referred to as scaphocephaly or dolichocephaly. Sagittal suture fusion occurs in approximately 1 in 5000 births, and accounts for about half of all cases.

It is imperative to monitor cranial symmetry for the first year of life to detect this condition early, and if skull shape deformity is found or suspected, a CT scan is indicated for confirmation.

79.

In pediatric patients, which of the following statements is TRUE regarding osteomyelitis?

It most commonly occurs via hematogenous spread

It is less common in patients with sickle cell anemia (SCA)

It is best diagnosed via bone scans

It most frequently occurs in the short bones such as tarsals in the foot and spinal vertebrae

Correct answer: It most commonly occurs via hematogenous spread

Osteomyelitis, or bone infection, in children most commonly occurs via hematogenous spread (in the blood) with 50% of infectious cases originating from Staphylococcus aureus spread, often through puncture wounds that penetrate a bone or joint.

The long, tubular bones (femur, tibia) are most commonly affected by osteomyelitis in children and it occurs with increased frequency in pediatric patients diagnosed with sickle cell anemia (SCA). Diagnostic studies include plain film radiographs (x-rays). Bone scans are sensitive but not specific for osteomyelitis.

80.

Which of the following is the MOST severe type of urinary tract infection (UTI) seen in children?

Pyelonephritis

Cystitis

Asymptomatic bacteriuria

A complicated UTI

Correct answer: Pyelonephritis

Since young children often have limited or unusual symptoms, a high degree of suspicion must be maintained to diagnose UTI. Inflammation and/or infection can occur anywhere along the urinary tract, so a UTI must be identified based on location.

Pyelonephritis is the most severe type of UTI involving the renal parenchyma or the kidneys and must be identified and treated quickly because of the potential irreversible renal damage that can occur. It is located in the back, has an acute onset, and is associated with high fever, chills, nausea, vomiting, and, less often, diarrhea. Symptoms of cystitis may be present, including dysuria, frequency, urgency, and suprapubic pain. Costovertebral angle tenderness focuses the diagnosis to the urinary tract. A urinalysis reveals pyuria and bacteriuria, and the hemogram shows leukocytosis. Treatment is with antibiotics.

Asymptomatic bacteriuria is bacteria in the urine without any other symptoms. It is benign and does not cause renal injury. Cystitis is an infection of the bladder causing lower tract symptoms, but it does not cause fever or renal injury. A complicated UTI is defined as a UTI with fever, toxicity, and dehydration, or a UTI occurring in a child younger than 3 to 6 months old.

81.

The organism MOST commonly associated with urinary tract infection (UTI) is:

Escherichia coli

Enterobacter

Klebsiella

Pseudomonas

Correct answer: Escherichia coli

Upwards of 70% of urinary tract infections (UTIs) are caused by Escherichia coli (70%), although the other organisms listed can also cause infection.

UTI secondary to group B streptococcus (GBS) is more common in neonates.

82.

Around what age do skull sutures generally close in children?

12-24 months

18-30 months

24-36 months

36-48 months

Correct answer: 12-24 months

Sutures tend to close by 12-24 months of age, but they do not ossify until approximately 8 years of age. Often, the anterior fontanelle will persist past 1 year of age, closing on average around 18-24 months, while the posterior and lateral fontanelles will usually disappear after the first 6 to 8 weeks of life.

For this reason, the neurologic exam should always include a measurement of head circumference until the child is 2 years old (The American Academy of Pediatrics Bright Futures). In addition, the skull should be inspected for symmetry and shape.

83.

Which of the following clinical findings would MOST directly suggest a deficiency in vitamin A?

Dry skin, follicular keratosis

Rickets, delayed dentition

Scurvy, easy bruising

Underweight, growth retardation

Correct answer: Dry skin, follicular keratosis

Dry skin and follicular keratosis are often seen in vitamin A deficiency. Night blindness, corneal lesions, and an increased susceptibility to infections are other signs of deficits of this important fat-soluble vitamin. In most cases, vitamin A deficiency results from insufficient dietary intake (sources include liver, milk, eggs, green and yellow vegetables, fruits).

Rickets and delayed dentition are signs of vitamin D deficiency. A child deficient in vitamin C would display scurvy and easy bruising, while growth retardation and anorexia are indicative of too little zinc.

84.

A 14-year-old male sustained a fracture to the left radius during a fall off a trampoline. X-ray reveals extension of the fracture through the metaphysis, physis, and epiphysis of the bone. The patient has to undergo an open reduction to prevent articular incongruity and osseous bridging across the physis.

According to the Salter-Harris classification, which type of fracture is this?

Type IV

Type V

Type II

Type III

Correct answer: Type IV

The extent of the fracture is consistent with a Type IV fracture, requiring anatomic reduction, often through an open approach, for proper alignment to heal effectively.

Type II fractures involve the physis and metaphysis, with a metaphyseal fragment present on the compression side of the fracture. Type III fracture involves physeal separation with fracture through the epiphysis into the joint. Type V involves a compression or crushing injury to the physis.

85.

Which of the following statements regarding the cause of macrocephaly is TRUE?

Familial megalencephaly is the most common cause

It is due to the enlargement of the subdural space

It is always associated with increased cerebrospinal fluid (CSF) production

It is never a benign finding

Correct answer: Familial megalencephaly is the most common cause

Benign familial megalencephaly is the most common cause of mild macrocephaly in a child, particularly in the setting of otherwise normal development.

Macrocephaly is defined as a head circumference more than two standard deviations above the mean for age and gender or one that increases too rapidly. "Large" heads may simply be genetic and not clinically significant; the provider's initial evaluation should include measuring both parents' head circumferences.

Macrocephaly can also be attributed to hydrocephalus, megalencephaly (enlarged brain), subdural hematoma, tumor, thickening of the skull, or other problems.

86.

Which of the following conditions would immediately disqualify a potential athlete from school sports participation?

Fever

Shin splints

Cardiac murmur

Atlantoaxial instability

Correct answer: Fever

Estimates vary, but reports of 0.3% to 1.9% of athletes are disqualified from participation based on the findings of a pre-participation sports physical examination, and 3% to 13% require further evaluation in order to be cleared.

The majority of findings that would require further evaluation are musculoskeletal injuries (including shin splints), followed by cardiovascular symptoms or a murmur, and neurologic symptoms or complaints. Atlantoaxial instability requires further evaluation to assess the risk of spinal cord injury during participation and is commonly seen in children with Down's syndrome.

*A fever can indicate a disease state (such as infection) and would disqualify a potential athlete from sports participation until the fever has resolved. No further evaluation is needed in the case of a fever. This child will **not** be cleared for school sports participation until the fever resolves.*

87.

Which of the following pediatric patients is MOST likely to develop celiac disease?

A child with type 1 diabetes mellitus

A child with Turner syndrome

A child with IgA deficiency

A child who is 3-years-old

Correct answer: The child with type 1 diabetes mellitus

Celiac disease (CD) is an immune-mediated systemic disorder triggered by dietary exposure to wheat gluten and related proteins found in rye and barley. This condition often co-occurs with other autoimmune diseases, such as diabetes mellitus type 1, autoimmune thyroiditis, autoimmune liver disease, IgA nephropathy (not deficiency), and juvenile chronic arthritis.

The most typical presentation occurs between 6 months and 2 years old, with a female predominance of 2:1.

Celiac disease occurs less frequently in females with Turner syndrome.

88.

Prader-Willi syndrome is caused by an absence of a group of genes on the chromosome:

15

21

9

22

Correct answer: 15

Prader-Willi syndrome (PWS) is a complex genetic condition that affects many parts of the body. It is a chromosome disorder caused by the absence of a group of genes on chromosome 15. Characteristics include poor feeding and growth, hypotonia, developmental delays, and behavioral problems. Short stature and central obesity are also common in childhood.

Down syndrome is the result of an extra copy of chromosome 21, while chronic myeloid leukemia (CML) results from a translocation in which portions of chromosomes 9 and 22 are exchanged, resulting in a new, abnormal gene.

89.

Which of the following disorders is inherited in an autosomal dominant pattern?

Huntington disease

Duchene muscular dystrophy

Neural tube defects (NTDs)

Cystic fibrosis

Correct answer: Huntington disease

This type of single genetic disorder is characterized by the inheritance of a single copy of a mutated gene located on one of the autosomal chromosomes. Huntington disease is inherited in an autosomal dominant pattern.

Duchene muscular dystrophy is inherited in an X-linked recessive pattern. NTDs are inherited in a multifactorial pattern. Cystic fibrosis is inherited in an autosomal recessive pattern.

90.

Which reflex is elicited by placing an infant supine and turning the head to one side?

Asymmetric tonic neck reflex (ATNR)

Moro reflex

Stepping reflex

Galant reflex

Correct answer: Asymmetric tonic neck reflex (ATNR)

Asymmetrical tonic neck reflex (ATNR) is characterized by extension of the upper and lower extremities on the side to which the head and neck is turned, with flexion of the contralateral upper extremity (fencing posture).

An ATNR that is observed as a resting posture rather than being elicited is never normal. ATNR appears at 35 weeks gestation, is well-established by one month postnatal age, and disappears by 3 to 4 months of age in a term infant. Persistence of this reflex beyond 4 to 6 months old indicates a CNS lesion.

Moro, stepping, and Galant (trunk incurvation) reflexes are also all primitive reflexes. Moro is also known as the "startle" reflex. Galant occurs when a baby flexes toward a stimulus when gently stroked close to the spine. Stepping is when an infant mimics steps when held as though weight-bearing with feet on a hard surface.

91.

When should a child be able to stand independently for several seconds, without holding onto furniture, but may still need to hold onto something in order to walk?

12 months

9 months

15 months

6 months

Correct answer: 12 months

The ability to stand alone for 3-5 seconds is a gross motor developmental milestone that should be reached at approximately 12 months of age. Developmental assessment should be incorporated into each health supervision visit.

92.

Which of the following diagnostic genetic tests detects specific single gene mutations?

Molecular testing

Karyotype

Fluorescence in situ hybridization (FISH)

Microarray

Correct answer: Molecular testing

Molecular testing detects specific single gene mutations (e.g., deletions, insertions, and single base pair changes) known to cause single gene disorders. Molecular techniques are used to directly detect atypical sequence changes in a targeted gene or short length of DNA or to indirectly detect abnormal changes in DNA structure by identifying size variations in fragments of DNA from the targeted locus or gene.

Karyotype is used to identify and evaluate the size, shape, and number of chromosomes. FISH is used to locate and detect a specific area of a particular chromosome by labeling a known chromosome sequence with fluorescent tags to see the location of genetic material. Chromosomal microarray is used to detect microdeletions or duplications in any of the chromosomes but not specific gene mutations.

93.

The FIRST visible body change indicating puberty in females is:

Thelarche

Rapid linear growth

Development of pubic hair

Menarche

Correct answer: Thelarche

Females enter puberty sooner than boys do, progressing sequentially in the following pattern:

- *Ovaries increase in size (no visible body changes occur at this stage)*
 - *Thelarche, or breast budding, traditionally occurs between 8 and 13 years old, with the average age onset at 10.3 years. Thelarche is a result of estradiol secretion.*
 - *Rapid linear growth usually begins shortly after the onset of breast budding and reaches its peak about 1 year later*
 - *Appearance of pubic hair (adrenarche or pubarche) begins at about 11.5 years old and is related to adrenal rather than gonadal development*
 - *The first menstrual period (menarche) occurs, on average, at 12.5 years, but age of onset ranges from 9 to 15 years. Menarche occurs approximately 2.5 years after thelarche*
-

94.

Which of the following is TRUE regarding medical etiologies of autistic spectrum disorders (ASD)?

Fragile X syndrome is often associated with ASD

Exposure to mercury and/or bacterial contamination in vaccines can directly cause ASD

It is uncommon to have a co-occurring neurodevelopmental diagnosis

Advanced parental age is not a risk factor for development of ASD

Correct answer: Fragile X syndrome is often associated with ASD

ASD is a neurobiologic and neurodevelopmental disorder that affects communication and behavior beginning in the first two years of life. ASD causes impairment in social interaction with additional impairment in communication and behavior patterns.

Although the etiology of ASD is unclear, the greatest consensus lies with genetic etiology with familial inheritance patterns, and the association of certain genetic disorders.

Fragile X syndrome is the most common single gene cause of ASD. Other genetic disorders associated with ASD include neurofibromatosis, tuberous sclerosis, Angleman syndrome, and Rett syndrome.

Up to 83% of children diagnosed with ASD have one or more co-occurring neurodevelopmental diagnoses. Advanced parental age has been identified as a risk factor for ASD. There is overwhelming evidence that no such association exists between ASD and mercury exposure, or bacterial or fungal contamination in vaccines.

95.

You are examining a seven-year-old Native American female for complaints of vomiting and diarrhea over the past 12 hours; she has had 5 episodes of watery stools without the presence of mucous or blood and 4 episodes of vomiting. She has been afebrile, and her urine output has been normal. Her symptoms began a few hours after eating grilled chicken while enjoying a day outside at a local park. Several of her family members had similar symptoms. Her vital signs are stable, bowel sounds are hyperactive, and she exhibits slight abdominal tenderness upon palpation without guarding.

What is the MOST likely mechanism responsible for the patient's symptoms?

Enterotoxins

Cytotoxins

Mucosal invasion

Spore ingestion

Correct answer: Enterotoxins

Acute gastroenteritis was formerly used to describe acute diarrhea, but this term is technically a misnomer because the etiology of diarrhea does not technically involve the stomach. With acute diarrhea, there is a disruption of the normal intestinal absorptive versus secretory mechanisms of fluids and electrolytes, resulting in excessive loss of fluid into the intestinal lumen.

In children two years and older, diarrheal stooling is described as occurring four or more times in 24 hours and can last up to 14 days. Associated symptoms include abdominal pain, nausea, and vomiting. This condition is extremely common in the pediatric population, caused by a wide variety of viruses, bacteria, and parasites.

Salmonella is a common type of enterotoxigenic bacterial organism that has a reservoir in a wide variety of foods, including contaminated eggs and undercooked poultry. This foodborne illness has a brief incubation period of 1 to 3 days, while the other mechanisms listed all have longer incubation periods.

96.

A mother brings her child into the clinic due to her concerns about her child's delayed communication. Which of the following scenarios would be considered a speech delay?

A 36-month-old whose speech is intelligible to a stranger 50% of the time

A 12-month-old who babbles, says "mama" and "dada" and uses 3 other words

An 18-month-old who articulates 15 words and names two body parts

A 24-month-old who just started speaking in two-word sentences

Correct answer: A 36-month-old whose speech is intelligible to a stranger 50% of the time

A 3-year-old's speech should be understandable to a stranger 90% of the time. In addition, the child should be able to use three- to four-word sentences, name objects, give their full name when asked, and begin to relate events.

Other red flags that would warrant referral for further evaluation at 36 months of age include: unable to give their full name when asked, unable to match two colors, does not use plurals, does not know 2 or 3 prepositions, is unable to tell a story, uses unclear consonants, speech is intelligible 50% or less of the time (unintelligible speech), or is unable to speak in sentences.

The other scenarios represent typically-developing language in toddlerhood.
