

Chapter 02: Developmental, Congenital, and Childhood Diseases and Disorders

Frazier & Drzymkowski: Essentials of Human Diseases and Conditions, 6th Edition

MULTIPLE CHOICE

1. Congenital diseases
 - a. are always fatal.
 - b. are always genetic.
 - c. are always nongenetic.
 - d. can be physical or mental.

ANS: D

Congenital anomalies can be mental or physical and can vary widely in severity, from trivial to fatal.

REF: 44 OBJ: 1

2. Attempts to halt premature labor include
 - a. having the mother on complete bedrest.
 - b. using drug therapy to slow or halt contractions.
 - c. giving the mother a short course of steroids to aid fetal lung maturation.
 - d. all of the above.

ANS: D

Attempts may be made to halt premature labor by having the mother on complete bedrest or using drug therapy to slow or halt contractions. The mother is often given a short course of steroids to aid fetal lung maturation. Many times the efforts to allow the fetus more time to grow and mature in the mother's uterus are unsuccessful.

REF: 49 OBJ: 5

3. The collapse of a seemingly healthy young athlete during a strenuous sporting event or other period of stressful exercise is often caused by
 - a. patent ductus arteriosus (PDA).
 - b. hypertrophic cardiomyopathy.
 - c. respiratory distress syndrome.
 - d. atrial septal defect (ASD).

ANS: B

Tragically the first sign of hypertrophic cardiomyopathy is often the collapse of a seemingly healthy young athlete during a strenuous sporting event or other period of stressful exercise. This collapse can be followed by cardiac arrest caused by a cardiac arrhythmia. Many have no symptoms until the collapse. It is possible that some individuals experience no symptoms. Signs may be discovered during a physical examination. Symptoms, often ignored by young people, may include chest pain, **syncope**, hypertension, palpitations, or shortness of breath. Some report experiencing fatigue, shortness of breath when lying down, or reduced tolerance of activity.

REF: 59 OBJ: 6

4. Down syndrome, a genetic syndrome, is the result of the child having _____ pairs of chromosomes.
- 45
 - 46
 - 47
 - 48

ANS: C

Down syndrome (formerly called *mongolism*) is a genetic syndrome in which the individual has 47 chromosomes instead of the usual 46, resulting in a congenital form of mild to severe mental retardation that is accompanied by characteristic facial features and distinctive physical abnormalities.

REF: 60 OBJ: 6

5. The most commoncrippler of children is
- spina bifida.
 - muscular dystrophy (MD).
 - Down syndrome.
 - cerebral palsy (CP).

ANS: D

CP, the most commoncrippler of children, is a condition consisting of a group of disorders possibly involving the brain and nervous system functions that deal with movement, learning, hearing, sight, and thinking. This disorder may be congenital or acquired and bilateral or unilateral in the form of a nonprogressive paralysis that results from damage to the central nervous system (CNS).

REF: 61 OBJ: 8

6. All females of childbearing age capable of becoming pregnant are encouraged to take the recommended amount of
- calcium.
 - vitamin D.
 - folic acid.
 - iron.

ANS: C

Increased intake of folic acid is encouraged in females planning to become pregnant and during early stages of pregnancy. Because many pregnancies are unplanned, all females of childbearing age capable of becoming pregnant are encouraged to take the recommended amount of folic acid each day.

REF: 66 OBJ: 8

7. Duchenne MD
- is usually the result of trauma.
 - is progressive and causes crippling and immobility.
 - affects males and females equally.
 - is not a genetic condition.

ANS: B

MD is a progressive degeneration and weakening of the skeletal muscles in which muscle fibers are abnormally vulnerable to injury. There are several types of the disease, but all are rare. The most common and best-known type is Duchenne MD, which is diagnosed soon after birth or during early childhood, usually before the age of 5 years. MD initially affects the muscles of the shoulders, hips, thighs, and calves of the legs, causing the characteristic waddling gait and toe walking. Affected muscles sometimes look larger than normal because fat replaces atrophied muscle. The child also may have lordosis or other spinal deformities. In addition, the child has difficulty climbing stairs and running, tends to fall easily, and has difficulty getting up. As the disease progresses, it involves all the muscles, causing crippling and immobility (see Figure 2-13). **Contractures** typically develop, and the child becomes increasingly susceptible to serious pulmonary infections such as pneumonia. Also, children with Duchenne MD often are impaired mentally.

REF: 63 OBJ: 8

8. *Hyaline membrane disease* refers to
- bronchomalacia.
 - infant respiratory distress syndrome (IRDS).
 - retinopathy of prematurity (ROP).
 - necrotizing enterocolitis (NEC).

ANS: B

IRDS, or hyaline membrane disease, is similar to adult respiratory distress syndrome in that the patient suffers acute hypoxemia caused by infiltrates within the alveoli.

REF: 51 OBJ: 4

9. The symptoms and signs of anemia include
- pallor, weakness, fatigability, and listlessness.
 - an abnormal reduction of red blood cells (RBCs).
 - tachycardia, jaundice, and mental sluggishness.
 - all of the above.

ANS: D

Anemia is an abnormal reduction in the concentration of RBCs or in the hemoglobin content of circulating blood. Pallor, weakness, fatigability, and listlessness are noted initially in the anemic child or infant. Palpitations, tachycardia, cardiac enlargement, jaundice, and mental sluggishness are symptoms of severe anemia.

REF: 117 OBJ: 25

10. Childhood immunizations are important because
- they are needed to obtain a passport.
 - they help prevent colds.
 - they prevent epidemics of serious contagious diseases.
 - of all of the above.

ANS: C

Dramatic results have been achieved in pediatric medicine through routine prophylactic immunization with vaccines that build specific and prolonged protection. To prevent epidemics of contagious diseases, all U.S. states require that children have inoculations before entering school.

REF: 93 OBJ: 17

11. Some warning signs of lead poisoning include
 - a. loss of appetite and vomiting.
 - b. irritability and ataxic gait.
 - c. anemia.
 - d. all of the above.

ANS: D

Children exposed to toxic levels of lead, a poisonous metallic element, exhibit signs of lead poisoning. Some warning signs are loss of appetite, vomiting, irritability, and ataxic gait. Chronic symptoms include anemia, weakness, colic, and peripheral neuritis. Evidence of mental retardation resulting from brain damage is possible. A child with acute lead intoxication presents as a medical emergency. The child has symptoms of encephalopathy with vomiting, headache, stupor, convulsions, and coma resulting from cerebral edema (see Figure 2-50).

REF: 122 OBJ: 27

12. Which of the following statement(s) is/are true about asthma?
 - a. The bronchial tubes are hyperactive and hypersensitive.
 - b. Severe attacks require hospitalization.
 - c. It is a leading cause of absenteeism in school children.
 - d. All of the above are true.

ANS: D

Asthma is the result of hyperactive and hypersensitive bronchial tubes. The bronchial spasms of asthma can be triggered by many extrinsic (allergic) or intrinsic (nonallergic) factors, including stress, heavy exercise, infection, and inhalation of allergens or other substances. In a severe attack unresponsive to drug therapy, a condition called *status asthmaticus* may lead to fatal respiratory failure and thus the need for endotracheal intubation. The patient requires hospitalization for aggressive medical treatment and follow-up. Asthma is a chronic, reversible, obstructive disease caused by increased reactivity of the tracheobronchial tree to various stimuli. There are two major processes at work: constriction of the bronchioles and inflammation of the airway. It is a leading cause of chronic illness and school absenteeism in children.

REF: 108 OBJ: 22

13. CP is
 - a. a result of damage to the CNS.
 - b. a condition that can be cured with treatment.
 - c. a condition that primarily affects intelligence.
 - d. all of the above.

ANS: A

CP, the most commoncrippler of children, is a condition consisting of a group of disorders possibly involving the brain and nervous system functions that deal with movement, learning, hearing, sight, and thinking.

REF: 62 OBJ: 8

14. Down syndrome is
- a congenital form of mild to severe mental retardation.
 - associated with distinctive physical abnormalities and heart defects.
 - caused by a chromosomal abnormality.
 - all of the above.

ANS: D

Down syndrome, in addition to mild to severe mental retardation, is associated with heart defects and other congenital abnormalities. Typically, the infant has a small head; a skull with a flat back; a characteristic slant to the eyes; a flat nasal bridge; small, low-set ears; a small mouth with a protruding tongue; and small, weak muscles (see Figures 2-10 and 2-11). The hands are short with stubby fingers and a deep horizontal crease across the palm (simian line). There is an exaggerated space between the big and little toes. Infants with Down syndrome have an extra chromosome number 21 (**trisomy 21**). It occurs in 1 in 650 live births and more often in infants born to women more than 35 years of age (see Figure 2-12). Infants with severe Down syndrome usually are identified at or before birth; milder forms are diagnosed later. The physical characteristics may be blatantly obvious. Findings on examination of the eyes may include the presence of small white dots on the iris. A *karyotype* showing the chromosomal abnormality can confirm the diagnosis.

REF: 60 OBJ: 6

15. The most common cyanotic congenital heart defect that is actually a combination of four defects is called
- ventricular septal defect (VSD).
 - PDA.
 - tetralogy of Fallot.
 - transposition of the great arteries.

ANS: C

The most common cyanotic cardiac defect is a combination of four congenital heart defects: (1) VSD, an abnormal opening in the ventricular septum; (2) pulmonary **stenosis**, a tightening of the pulmonary valve or vessel; (3) dextroposition (displacement to the right) of the aorta, which overrides (receiving circulation from both ventricles) the VSD; and (4) right ventricular *hypertrophy*, caused by increased pressure in the ventricle.

REF: 77 OBJ: 10

16. The diagnostic investigation of a congenital heart defect may include
- physical examination and patient history.
 - radiographic studies of the chest and blood tests.
 - heart catheterization and electrocardiogram (ECG).
 - all of the above.

ANS: D

Physical examination and patient history are essential. The physician palpates the neck vessels and auscultates for blood pressure and murmurs. Diagnostic procedures depend on the initial findings and may include radiographic chest films, blood tests, cardiac catheterization, echocardiogram, and ECG. Many defects can be detected on a prenatal ultrasound. The diagnostic investigation determines the presence and severity of any structural or functional abnormality or defect.

REF: 78 OBJ: 10

17. Congenital hip dysplasia (CHD)
- causes cleft lip.
 - is a nontraumatic deformity of the foot.
 - is more common in male infants.
 - is an abnormal development of the hip joint.

ANS: D

Developmental dysplasia of the hip, previously known as *CHD*, is an abnormal development of the hip joint that ranges from an unstable joint to dislocation of the femoral head from the **acetabulum**.

REF: 80 OBJ: 11

18. The birth defect in which there is a hole in the middle of the roof of the mouth is
- cleft palate.
 - cri-du-chat syndrome.
 - Robinow's syndrome.
 - meningocele.

ANS: A

Cleft palate is a birth defect in which there is a hole in the middle of the roof of the mouth (palate).

REF: 81 OBJ: 11

19. Cryptorchidism is
- a congenital malignant tumor.
 - also known as *phimosis*.
 - the failure of the testicle(s) to descend into the scrotum.
 - Wilms' tumor.

ANS: C

Cryptorchidism is failure of one or both of the testicles to descend from the abdominal cavity into the scrotum.

REF: 82 OBJ: 12

20. Symptoms of Wilms' tumor may include
- nasal regurgitation during feeding.
 - hematuria, pain, and hypertension.
 - a shortening of the femur.
 - projectile vomiting, loss of weight, and skin rash.

ANS: B

The most common presentation is a mass in the kidney region, which is often discovered by a parent or examining physician. The mass is firm, nontender, and usually confined to one side of the body. The patient may experience other symptoms resulting from compression caused by the tumor mass, metabolic alterations due to the tumor, or metastasis. These include *hematuria*, pain in the abdomen or chest, hypertension, anemia, vomiting, intestinal obstruction, constipation, weight loss, and fever.

REF: 83

OBJ: 12

21. The disease that involves impairment of intestinal motility, which causes obstruction of the distal colon, is called
- Hirschsprung's disease.
 - pyloric stenosis.
 - cryptorchidism.
 - phimosis.

ANS: A

Hirschsprung's disease, a congenital condition, is an impairment of intestinal motility that causes obstruction of the distal colon.

REF: 86

OBJ: 13

22. Cystic fibrosis (CF), a chronic dysfunction of the exocrine glands, primarily attacks the
- nervous system.
 - lungs and digestive system.
 - blood vessels.
 - immune system.

ANS: B

Symptoms may become apparent soon after birth or may develop in childhood. The disease primarily attacks the lungs and the digestive system, producing copious thick and sticky mucus that accumulates and blocks glandular ducts.

REF: 87

OBJ: 15

23. The primary treatment of phenylketonuria (PKU) includes
- oxygen therapy.
 - a high-sodium chloride diet.
 - a protein-restricted diet.
 - surgical intervention.

ANS: C

The treatment is to place the infant on a phenylalanine-free diet that allows the infant to grow with normal brain development. Individuals with PKU must eat a diet low in phenylalanine for the rest of their lives. Because natural proteins contain phenylalanine, the patient must remain on a protein-restricted diet. Restrictions or elimination of the following food is required: meat, chicken, fish, cheese, nuts, and dairy products. The newborn cannot have breast milk with its high levels of phenylalanine. Some starchy foods, including potatoes, corn, pasta, and bread, require close monitoring. The sweetener aspartame used in diet soda and diet food is metabolized into substances including phenylalanine.

REF: 89

OBJ: 14

24. The production of copious thick and sticky mucus that accumulates and blocks glandular ducts is characteristic of
- Wilms' tumor.
 - PKU.
 - pyloric stenosis.
 - CF.

ANS: D

The disease primarily attacks the lungs and the digestive system, producing copious thick and sticky mucus that accumulates and blocks glandular ducts. The clinical effects of CF can be immense, including a dry paroxysmal cough, exercise intolerance, pneumonia, bulky diarrhea, vomiting, and bowel obstruction.

REF: 87

OBJ: 15

25. Klinefelter's syndrome and Turner's syndrome are examples of
- inherited chromosomal diseases.
 - genetic, chromosomal diseases that are not inherited.
 - chromosomal diseases that affect females only.
 - inherited diseases of no significant consequence.

ANS: B

Klinefelter's syndrome and Turner's syndrome are examples of genetic, chromosomal diseases that are not inherited. They result from nondisjunction, or the failure of a chromosome pair to separate, during *gamete* production (see Figure 2-33).

REF: 89

OBJ: 16

26. Which of the following statements is/are true about erythroblastosis fetalis?
- Antibodies in the mother's blood destroy the RBCs of the fetus.
 - It is characterized in the fetus by anemia, jaundice, and enlargement of the liver and spleen.
 - There is no destruction of fetal blood cells.
 - Both a and b are true.

ANS: D

The cause is Rh factor incompatibility. Rh factor is the antigen found on RBCs of the Rh-positive individual. The mother, through a prior pregnancy, has become sensitized to the Rh factor (Rh isoimmunization) of the fetal RBCs. When sensitized maternal blood finds its way into fetal circulation, particularly during delivery, the antibodies in the mother's blood destroy the RBCs of the fetus (see Figures 2-48 and 2-49). If an Rh-negative woman has children with an Rh-positive man, some or all of the infants will be Rh positive. During pregnancy, blood from the Rh-positive fetus may move from fetal circulation into the mother's bloodstream, where it can stimulate the mother's body to form antibodies against the Rh factor. When sufficient quantities of the antibodies pass back into the infant's circulation, the antibodies can clump and destroy Rh-positive cells, causing the symptoms of erythroblastosis fetalis. Erythroblastosis fetalis is characterized by anemia, jaundice, *kernicterus*, and enlargement of the liver and spleen. In the most severe form, called *hydrops fetalis*, the fetus or infant is in great jeopardy because of extreme *hemolysis*. If the infant survives, the condition is marked by heart failure, edema, pulmonary congestion, lethargy, seizures, and mental retardation.

REF: 119 OBJ: 26

27. Sudden infant death syndrome (SIDS) is
- most likely to occur in a sick infant.
 - not known to have any risk factors.
 - the number one cause of death of infants between ages 1 month and 12 months.
 - predictable and preventable.

ANS: C

SIDS, formerly called *crib death*, is defined officially as the sudden death of an infant under the age of 1 year for which a cause cannot be established. It is the number one cause of death among infants from 1 to 12 months of age; 1 in 2000 infants dies of SIDS.

REF: 103 OBJ: 19

28. Which of the following statements is true about lead poisoning?
- The child suffering from acute lead intoxication presents a medical emergency.
 - It is hard to detect, since lead is normally present in the blood.
 - It is a silent disease, because there are no warning signs or chronic symptoms.
 - There is no evidence of mental retardation resulting from brain damage.

ANS: A

Children exposed to toxic levels of lead, a poisonous metallic element, exhibit signs of lead poisoning. Some warning signs are loss of appetite, vomiting, irritability, and ataxic gait. Chronic symptoms include anemia, weakness, colic, and peripheral neuritis. Evidence of mental retardation resulting from brain damage is possible. A child with acute lead intoxication presents as a medical emergency. The child has symptoms of encephalopathy with vomiting, headache, stupor, convulsions, and coma resulting from cerebral edema (see Figure 2-50).

REF: 122 OBJ: 27

29. A systemic disease with superficial cutaneous lesions that begin as red macules that progress to papules then to vesicles that form crusts is
- rubeola.

- b. rubella.
- c. chickenpox.
- d. diphtheria.

ANS: C

Chickenpox is a systemic disease with superficial cutaneous lesions that begin as red macules that progress to *papules* and then finally become *vesicles* that form crusts. The lesions first are seen on the face or the trunk and then spread over the extremities; they can be distributed everywhere on the body and even have been found internally.

REF: 93

OBJ: 17

30. Pregnant women should be isolated from individuals infected with _____ to prevent perinatal infection.
- a. rubeola.
 - b. rubella.
 - c. chickenpox.
 - d. diphtheria.

ANS: B

Rubella causes great danger to the unborn children of pregnant women who contract the disease. Women of childbearing age who have not been immunized against rubella or who have not had the disease can transmit rubella to their infants if they become infected during pregnancy. When the virus is transferred to the fetus during the first trimester of pregnancy, a variety of congenital deformities, known as *congenital rubella syndrome*, occur in about 25% of births. The risk is lower if infection occurs later in pregnancy. Anomalies caused by congenital rubella syndrome include congenital cardiac disease, blindness, deafness, and mental retardation. Pregnant women should be isolated from individuals infected with rubella to prevent perinatal infection; in addition, pregnant women must not be given the rubella vaccine. The best protection against congenital rubella syndrome is universal routine immunization with live rubella vaccine during infancy or as soon as possible in adulthood.

REF: 102

OBJ: 18

31. Prompt and complete treatment of acute tonsillitis caused by group A beta-hemolytic streptococci is necessary to prevent
- a. rheumatic fever.
 - b. rheumatic heart disease.
 - c. kidney complications.
 - d. all of the above.

ANS: D

When the throat culture is positive for group A beta-hemolytic streptococci (strep throat), a full 10-day course of penicillin is given. This strict regimen is necessary to prevent rheumatic fever, rheumatic heart disease, and kidney complications. The child may need a liquid diet or saline throat irrigations if the pain is debilitating. Tonsillectomy may be recommended for chronic tonsillitis.

REF: 107

OBJ: 21

32. Serious side effects of uncontrolled diarrhea in the infant or child include

- a. intestinal obstruction and rhonchi.
- b. dehydration and electrolyte imbalance.
- c. diaper rash and Reye's syndrome.
- d. all of the above.

ANS: B

Diarrhea may be mild or severe, acute or chronic. In the infant or child, diarrhea can rapidly cause dehydration and electrolyte imbalance when fluid loss is profuse.

REF: 113 OBJ: 13

33. Which of the following statements is/are true about leukemia?
- a. It is the most common childhood malignancy.
 - b. It is a primary malignant disease of bone marrow.
 - c. The diagnosis is confirmed by microscopic examination of the bone marrow.
 - d. Both a and c are true

ANS: D

Leukemia, a cancer of blood-forming tissues, is the most common childhood malignancy. A bone marrow aspiration is examined. Chromosome analysis of the leukemic cells is performed for diagnosis because the presence of different characteristic abnormalities has prognostic value.

REF: 118 OBJ: 25

34. Causes of prematurity include
- a. incompetent cervix, bicornuate uterus, and premature rupture of membranes.
 - b. maternal physical conditions including infection and hypertension.
 - c. multiple gestation.
 - d. all of the above.

ANS: D

There are many reasons that infants enter the world before reaching the traditionally accepted gestational age of 40 weeks and thus have very low birth weights. Causes of premature labor resulting in a premature infant are an *incompetent cervix*, **bicornuate** uterus, toxic conditions, maternal infection, trauma, premature rupture of the amniotic membranes, history of previous miscarriages, multiple gestations, intrauterine fetal growth retardation, and other physical conditions of the mother, such as pregnancy-induced or chronic hypertension. Diabetes, heart disease, kidney disease, poor nutrition, substance abuse, and lack of prenatal care also contribute to the incidence of the mother giving birth to a preterm infant. In some cases, the etiology is never identified. Attempts may be made to halt premature labor by having the mother on complete bedrest or using drug therapy to slow or halt contractions. The mother is often given a short course of steroids to aid fetal lung maturation. Many times the efforts to allow the fetus more time to grow and mature in the mother's uterus are unsuccessful.

REF: 48 OBJ: 4

35. Conditions often resulting from premature birth include
- a. NEC, IRDS, and ROP.
 - b. spina bifida, hydrocephalus, and anencephaly.
 - c. MD, CF, and Down syndrome.

d. all of the above.

ANS: A

Advances in technology have made survival of low-weight and short-gestation infants possible. The prognosis for these children varies depending on gestational age, weight, and the occurrence of anomalies and developmental deficits. There are documented cases of 12-ounce and/or 22-gestational-week babies surviving. They fall into the 1% of premature babies born at that weight and gestational age. Being born before the normal prenatal development is complete, these children often have many problems to overcome (see Figure 2-5). A primary risk is a cerebral bleed, which may occur during the labor and delivery process or may result from handling after delivery. The cerebral bleed may cause the development of CP, mental functioning deficiencies, or other neurologic conditions. Another major concern is underdevelopment of the pulmonary system, including the lung tissue and the airway. Some pulmonary conditions these infants experience are IRDS, bronchopulmonary **dysplasia**, laryngomalacia, tracheomalacia, and bronchomalacia. Lack of body fat can affect the maintenance of body temperature. Any stress or increased or high supplemental O₂ flow may be responsible for ROP and possible blindness. NEC is a danger in the digestive system because of the reduced tolerance of the alimentary tract. ASD and PDA often are present because the fetal circulatory system has failed to mature.

REF: 50

OBJ: 4

36. Conjoined twins may be joined
- head to head (cranial).
 - posteriorly at the sacrum and coccyx.
 - chest to chest.
 - at all of the above points.

ANS: D

These children may be joined at different locations of the body and may share various organs. The attachment to each other may involve a small portion of tissue or may be as extensive as fusion at the head or sharing of an organ or body part. Common types or variations usually are categorized by the location and involvement of the junction through the term *pagus*, meaning fastened, included in the classification terminology. Twins with a cranial union are considered to be craniopagus. Those with anterior junction at the chest, often sharing the heart and vital portions of the chest wall and internal organs, are called thoracopagus conjoined twins. Thoracopagus is the most common form of conjoined twins. The term *pygopagus* describes those twins who are joined posteriorly at the rump. Another posterior junction occurring at the sacrum and coccyx is termed *ischiopagus*. When the connection proceeds from the breastbone to the waist, the term *omphalopagus* describes the junction. A very rare form, dicephalus, is the condition in which the individual has one body and two separate heads and necks.

REF: 47

OBJ: 1

37. The most common childhood malignancy is
- Wilms' tumor.
 - leukemia.
 - anemia.
 - neuroblastoma.

ANS: B

Leukemia, a cancer of blood-forming tissues, is the most common childhood malignancy. It is characterized by an abnormal increase in the number of immature white blood cells (WBCs) or undifferentiated blastocytes.

REF: 118 OBJ: 25

38. Causes of infantile colic may be
- sensitivity to cow's milk.
 - sensitivity to iron.
 - excessive swallowing of air during feeding process.
 - a and c.

ANS: D

The etiology of colic is unknown, although several theories have been advanced. One hypothesis suggests that improper feeding techniques may be responsible, and another theory blames overeating or the swallowing of excessive air. Sensitivity to cow's milk may be the causative factor, even for the nursing infant. In this case, the nursing mother is urged to eliminate cow's milk from her own diet. Regardless of the cause, the infant is extremely uncomfortable and cries a great deal, with sleep pattern disturbance.

REF: 112 OBJ: 13

39. Cardiac cyanotic defects include
- VSD and PDA.
 - tetralogy of Fallot and transposition of the great arteries.
 - coarctation of the aorta
 - ASD.

ANS: B

Central cyanosis is a sign that the atrial blood is not fully oxygenated. The infant will appear cyanotic with a blue tinge to the lips, tongue, and nail beds. The five main cardiac causes of central cyanosis are tetralogy of Fallot, transposition of the great arteries, truncus arteriosus, tricuspid atresia, and total anomalous pulmonary venous return. The two most common defects are discussed here. (See Evolve for discussion of the remaining defects.)

REF: 77 OBJ: 10

40. The most common fatal genetic disease is
- hypertrophic cardiomegaly.
 - CF.
 - Down syndrome.
 - tetralogy of Fallot.

ANS: B

CF, an autosomal recessive inherited disorder, is a chronic dysfunction of a gene called the *CF transmembrane conductance regulator* that affects multiple body systems. It is the most common fatal genetic disease.

REF: 87 OBJ: 15

41. Chickenpox is spread by virus being transmitted by
- bacillus entering through damaged skin.

- b. a toxin formed in respiratory mucosa.
- c. direct or indirect droplet nuclei from the respiratory tract of the infected person.
- d. group A beta-hemolytic streptococci.

ANS: C

The causative organism is the varicella-zoster virus (VZV), also known as *human herpes virus 3 (HHV-3)*, a member of the herpes virus group. The virus is transmitted by direct or indirect droplet nuclei spread from the respiratory tract of the infected person or a carrier. Fluid from the cutaneous lesions is also infectious, but dried crusty lesions are not contagious. The patient is considered contagious for 1 to 2 days before the eruptions until about 6 days after the eruptions.

REF: 93

OBJ: 17

42. Symptoms and signs of rubeola include
- a. tenderness in the neck and below the ears.
 - b. Koplik's spots on the buccal mucosa.
 - c. a violent cough ending in a high-pitched inspiratory whoop.
 - d. red macules that progress to papules and then vesicles that form crusts.

ANS: B

Early symptoms include cough, coryza, conjunctivitis, and *photophobia*. The child has a fever, followed in 3 to 7 days by a red blotchy rash. The rash starts behind the ears, hairline, and forehead and then progresses down the body (see Figure 2-38). Before the eruption of the rash, Koplik's spots can be detected on the oral mucosa as tiny white spots on a red background.

REF: 99

OBJ: 17

43. Mumps is a childhood disease that is best prevented by
- a. the measles-mumps-rubella (MMR) vaccine.
 - b. Gardasil vaccine.
 - c. broad-spectrum antibiotics.
 - d. cough medicines.

ANS: A

Childhood immunization is the best prevention. Children should receive the first immunization of MMR at the age of 12 months. The second dose should be given before the child enters school and can be given within 1 month of the initial immunization. An unimmunized person should be referred to a physician for active immunization within 48 hours of contact to prevent or alter the severity of the disease.

REF: 98

OBJ: 17

44. When a person is diagnosed with leukemia, there will be a/an
- a. increase in WBCs.
 - b. decrease in WBCs.
 - c. normal WBC count.
 - d. decrease in platelets.

ANS: A

Leukemia, a cancer of blood-forming tissues, is the most common childhood malignancy. It is characterized by an abnormal increase in the number of immature WBCs or undifferentiated blastocytes.

REF: 118 OBJ: 25

45. The most common progressive form of MD is
- occulta.
 - Duchenne.
 - Down.
 - myotonic.

ANS: B

MD is a progressive degeneration and weakening of the skeletal muscles in which muscle fibers are abnormally vulnerable to injury. There are several types of the disease, but all are rare. The most common and best-known type is Duchenne MD, which is diagnosed soon after birth or during early childhood, usually before the age of 5 years.

REF: 63 OBJ: 8

46. One way to prevent epidemics of contagious diseases is
- aspirin.
 - immunizations.
 - multivitamins.
 - cold and cough medications.

ANS: B

Although the infant acquires limited natural immunity from the mother, the growing child is vulnerable to many infectious diseases and the disabilities that they cause. Many of these communicable diseases can be prevented. Dramatic results have been achieved in pediatric medicine through routine prophylactic immunization with vaccines that build specific and prolonged protection. To prevent epidemics of contagious diseases, all U.S. states require that children have inoculations before entering school (see Figure 2-35). (See the discussion of immunity in Chapter 3.)

REF: 93 OBJ: 17

47. The procedure that allows amniotic fluid to be tested and cells to be microscopically examined for abnormal substances or chromosomal abnormalities is
- fetal monitoring.
 - amniocentesis.
 - acupuncture.
 - ultrasound studies.

ANS: B

One can diagnose congenital anomalies in a fetus by taking a fluid sample from the amniotic sac between the 15th and 18th weeks of pregnancy. This procedure, known as **amniocentesis**, allows amniotic fluid to be tested and cells to be microscopically examined for abnormal substances or chromosomal abnormalities.

REF: 45 OBJ: 2

48. Bronchiolitis, inflammation of the bronchioles, the smallest air passages of the lungs, is usually caused by
- bacteria.
 - viruses.
 - fungi.
 - protozoa.

ANS: B

Bronchiolitis is most commonly caused by infection with respiratory syncytial virus (RSV). Many other viruses can cause this disease, including parainfluenza and adenovirus.

REF: 111

OBJ: 23

TRUE/FALSE

1. Genetic disorders and syndromes are the result of an abnormal gene taking up residence on one of the 22 pairs of nonsex chromosomes.

ANS: T

Genetic syndromes are a form of congenital anomalies. Genetic information is contained in microscopic threadlike structures in the nucleus of human body cells. The genetic material contained within the genes is responsible for inheritance traits. Each cell within the human body contains 46 chromosomes arranged in 23 pairs, 22 of which are termed *homologous pairs*, and the remaining pair is the sex chromosomes.

REF: 45

OBJ: 2

2. The lungs of the neonate lack the surfactant needed to allow the alveoli to expand.

ANS: T

The lungs of the neonate lack the *surfactant* needed to allow the alveoli to expand. The surfactant normally is produced relatively late in fetal life; consequently, premature infants are at risk. The outcome of this inability of the lungs to expand is inadequate surface area for proper gas exchange and a potentially fatal lack of oxygen in the blood.

REF: 51

OBJ: 4

3. A congenital spinal defect in which no herniation of spinal cord or meninges exists is called a *myelomeningocele*.

ANS: F

Myelomeningocele (also known as *spina bifida cystica*) is a protrusion of a portion of the spinal cord and the meninges through a defect in the spinal column, usually in the lumbar region. Myelomeningocele is the most severe form of spina bifida. Because spinal nerves or the spinal cord are present in this herniation, the infant exhibits neurologic symptoms. The infant may have musculoskeletal malformation, immobile joints, or paralysis of the lower extremities. Depending on the level of the anomaly, bowel or bladder control may be affected.

REF: 64

OBJ: 7

4. In hydrocephalus, a large amount of cerebrospinal fluid (CSF) accumulates in the skull, causing increased intracranial pressure.

ANS: T

In hydrocephalus, the amount of CSF is increased greatly or its circulation is blocked, resulting in an abnormal enlargement of the head and characteristic pressure changes in the brain.

REF: 68 OBJ: 7

5. PKU is an inborn error in metabolism of amino acids causing brain damage and mental retardation when not corrected.

ANS: T

PKU is an inborn error in the metabolism of amino acids that causes brain damage and mental retardation when not corrected.

REF: 89 OBJ: 14

6. Because anemia is a symptom of various diseases, it is important to determine the cause of the anemia.

ANS: T

The cause of Reye's syndrome is unknown. However, it typically follows infection with influenza A or B viruses or chickenpox. It has been linked to the use of aspirin during these infections.

REF: 118 OBJ: 25

7. Reye's syndrome has been linked with the use of aspirin to treat chickenpox and influenza.

ANS: T

The cause of Reye's syndrome is unknown. However, it typically follows infection with influenza A or B viruses or chickenpox. It has been linked to the use of aspirin during these infections.

REF: 124 OBJ: 17

8. Rh factor incompatibility occurs when the mother, through prior pregnancy, has become sensitized to the Rh factor of the fetal RBCs.

ANS: T

The cause is Rh factor incompatibility. Rh factor is the antigen found on RBCs of the Rh-positive individual. The mother, through a prior pregnancy, has become sensitized to the Rh factor (Rh isoimmunization) of the fetal RBCs. When sensitized maternal blood finds its way into fetal circulation, particularly during delivery, the antibodies in the mother's blood destroy the RBCs of the fetus (see Figures 2-48 and 2-49).

REF: 120 OBJ: 26

9. Leukemia is characterized by an abnormal decrease in the number of immature WBCs.

ANS: F

Leukemia, a cancer of blood-forming tissues, is the most common childhood malignancy. It is characterized by an abnormal increase in the number of immature WBCs or undifferentiated blastocytes.

REF: 118

OBJ: 25

10. The infant born with fetal alcohol syndrome (FAS) may experience alcohol withdrawal shortly after birth.

ANS: T

Intrauterine exposure to sufficient levels of alcohol has been associated with fetal growth retardation, in which the infants are short and below average in weight. Facial characteristics of FAS include smaller eye openings with eyes spaced widely apart and a thin upper lip. The infant may experience growth deficiencies and CNS problems. Heart defects including ASD and VSD may be present. FAS is also associated with mental retardation. The infant may exhibit signs of alcohol withdrawal shortly after birth.

REF: 125

OBJ: 28

11. Closure of PDA may be achieved by medication or by surgical closure of the ductus.

ANS: T

PDA results when the ductus fails to functionally close. During normal fetal circulation, the patent ductus short-circuits shunting the circulation from the lungs and instead directs blood from the pulmonary trunk to the aorta. If PDA continues after birth, circulation of oxygen is compromised because this abnormal opening is a shunt that allows oxygenated blood to recirculate through the lungs (see Figure 2-22). PDA is detected during a physical examination when a classic “machinery” murmur is heard on auscultation and palpitation reveals a thrill. The infant’s growth and development may be slowed, and various signs of heart failure may be present. Closure may be attempted by drug therapy using an antiprostaglandin or ibuprofen. The other option is surgical closure of the ductus.

REF: 76

OBJ: 9

12. Tetanus is an acute, potentially deadly systemic infection characterized by painful involuntary contraction of skeletal muscles.

ANS: T

Tetanus is an acute, potentially deadly systemic infection characterized by painful involuntary contraction of skeletal muscles. The patient is extremely febrile (temperature $>101^{\circ}$ F), is irritable, and sweats profusely. He or she has a stiff neck, a tight jaw (lockjaw), spasms of the facial muscles, and difficulty swallowing. As the infection progresses, the muscles of the back and abdomen become rigid, with generalized convulsive muscle spasm (opisthotonos). These tonic spasms may cause death from asphyxiation.

REF: 102

OBJ: 17

13. NEC is an acute inflammatory process resulting from ischemic necrosis of the lining of the bronchial tubes.

ANS: F

NEC is a danger in the digestive system because of the reduced tolerance of the alimentary tract.

REF: 55

OBJ: 5

14. Epiglottitis typically strikes children between ages 3 and 7 years.

ANS: T

Epiglottitis typically strikes children between ages 3 and 7 years. The symptoms include a sore throat, croupy cough, fever, and respiratory distress caused by laryngeal obstruction. Visual inspection reveals a red and swollen epiglottis. Rapidly increasing dyspnea and drooling are the most significant signs of this critical respiratory emergency.

REF: 106

OBJ: 20

15. Asthma is a leading cause of chronic illness and school absenteeism in children.

ANS: T

Asthma is a chronic reversible obstructive disease caused by increased reactivity of the tracheobronchial tree to various stimuli. There are two major processes at work: constriction of the bronchioles and inflammation of the airway. It is a leading cause of chronic illness and school absenteeism in children.

REF: 108

OBJ: 22

16. Neural tube defects include spina bifida occulta, meningocele, and myelomeningocele.

ANS: T

Spina bifida is a group of malformations of the spine in which the posterior portion of the bony canal containing the spinal cord (usually in the lumbar region) is completely or partially absent (see Figure 2-14, A). Also called *neural tube defects*, the three different levels of the condition originate during early weeks of gestation as the spinal cord and bony canal develop. During this developmental stage, there is a failure of the posterior spinal processes to close, usually in the lumbar region. This failure of complete closure allows the **meninges** and, in severe cases, the spinal cord to herniate. Depending on the extent of the herniation and the amount of the neural tube that has herniated, various degrees of neural deficits or impairment occur. The three types of spina bifida are spina bifida occulta, meningocele, and myelomeningocele.

REF: 64

OBJ: 7

17. Down syndrome children's hands appear normal.

ANS: F

The hands are short with stubby fingers and a deep horizontal crease across the palm (simian line).

REF: 60

OBJ: 6

18. Young individuals with hypertrophic cardiomyopathy usually exhibit obvious symptoms of the disorder.

ANS: F

Tragically, the first sign is often the collapse of a seemingly healthy young athlete during a strenuous sporting event or other period of stressful exercise. This collapse can be followed by cardiac arrest caused by a cardiac arrhythmia. Many have no symptoms until the collapse. It is possible that some individuals experience no symptoms. Signs may be discovered during a physical examination. Symptoms, often ignored by young people, may include chest pain, **syncope**, hypertension, palpitations, or shortness of breath. Some report experiencing fatigue, shortness of breath when lying down, or reduced tolerance of activity.

REF: 59 OBJ: 2

19. MD affects only males.

ANS: T

As is the case for hemophilia and color blindness, the disease affects only males and generally is inherited through female carriers.

REF: 63 OBJ: 8

20. Hydrocephalus is easily diagnosed prenatally.

ANS: F

Diagnosis is made through the clinical picture, physical examination, and radiographic skull studies. Computed tomography and magnetic resonance imaging scans demonstrate the condition.

REF: 68 OBJ: 7

21. Treatment of hydrocephalus consists of placing a ventriculoatrial shunt.

ANS: T

Treatment consists of surgical intervention to place a shunt in the ventricular or subarachnoid spaces to drain off the excessive CSF. Some catheters empty into the peritoneal cavity, and other shunt catheters empty into the right atrium of the heart (see Figure 2-17). One-way valves help to shunt the excessive CSF away from the cerebrospinal canal and to maintain a normal pressure.

REF: 73 OBJ: 7

22. During fetal circulation, blood bypasses the lungs.

ANS: T

Because the fetal lungs are not functioning, this blood mostly bypasses the lungs. Most of the blood entering the right atrium by the inferior vena cava is shunted directly into the left atrium through the **foramen ovale**. There is a small valve on the left side of the atrial septum called the *septum primum*. This valve keeps blood from going back into the right atrium. The remaining fetal blood that has entered the right atrium contains a large amount of oxygen-poor blood from the superior vena cava and travels to the right ventricle and into the pulmonary trunk. The pulmonary blood vessels have a high resistance to blood flow because of the collapsed state of the lungs; thus they only allow a small amount of blood to enter the pulmonary circulation. This small amount is enough to nourish the pulmonary tissue.

REF: 82 OBJ: 10

23. Cryptorchidism may be treated with hormonal therapy.

ANS: T

The testes often descend spontaneously during the first year of life. If this does not happen by 4 years of age, the treatment is to place the undescended testes into the scrotum by either surgical manipulation (orchiopexy) or hormonal drug therapy (B-HCG or testosterone). Treatment is important because untreated cryptorchidism may lead to sterility in the adult male. There is an increased risk of testicular cancer in untreated cryptorchidism.

REF: 85 OBJ: 12

24. Symptoms of pyloric stenosis include episodes of projectile vomiting and failure to gain weight.

ANS: T

Vomiting can range from a mild regurgitation to projectile expulsion. The infant has a distended abdomen, is irritable, and often has a fever. Aspiration of vomitus into the lungs can result in pneumonia.

REF: 85 OBJ: 13

25. Abdominal distension is not a symptom of Hirschsprung's disease.

ANS: F

The symptoms and signs differ slightly depending on the age of the child experiencing an exacerbation of the condition. In the neonatal period, the newborn fails to pass **meconium** within 48 hours after birth. The infant may have bile-stained or fecal vomitus and does not want to feed. The abdomen becomes distended. After the neonatal period, the symptoms and signs include a failure to thrive, with obstinate constipation, vomiting, and abdominal distention. When the condition worsens, the infant may become feverish and may have explosive, watery diarrhea.

REF: 86 OBJ: 13

26. Turner's syndrome can be cured with proper treatment.

ANS: F

There is no cure for this genetic disorder; however, the prognosis is good if the patient has no other complicating conditions, including cardiac or kidney disorders. Moderate degrees of learning disorders are common. These females will never be able to conceive their own child because they have no ovaries to produce eggs.

REF: 92 OBJ: 16

27. Chickenpox is caused by a member of the herpes virus group.

ANS: T

The causative organism is the VZV, also known as *HHV-3*, a member of the herpes virus group. The virus is transmitted by direct or indirect droplet nuclei spread from the respiratory tract of the infected person or a carrier. Fluid from the cutaneous lesions is also infectious, but dried crusty lesions are not contagious. The patient is considered contagious for 1-2 days before the eruptions until about 6 days after the eruptions. The incubation period is 2-3 weeks.

REF: 93 OBJ: 17

28. It is recommended that all adults receive the Tdap immunization every 10 years to reduce the incidence of whooping cough.

ANS: T

Adults should have a booster inoculation to ensure that immunity still exists. Therefore, when an injection to prevent tetanus is administered to adults, the inoculation Tdap should be administered. This will also be helpful in preventing outbreaks of whooping cough.

REF: 99 OBJ: 17

29. Adenoid hyperplasia can contribute to recurrent otitis media and conductive hearing loss.

ANS: T

Adenoid hyperplasia can contribute to recurrent otitis media and conductive hearing loss resulting from obstruction of the eustachian tube.

REF: 108 OBJ: 21

30. Status asthmaticus is not considered a medical emergency.

ANS: F

In a severe asthma attack unresponsive to drug therapy, a condition called *status asthmaticus* may lead to fatal respiratory failure and thus the need for endotracheal intubation. The patient requires hospitalization for aggressive medical treatment and follow-up. Occasionally some children may develop pneumonia concurrent with the asthma attack.

REF: 110 OBJ: 22

31. Tetanus is not a possible fatal condition.

ANS: F

Tetanus is an acute, potentially deadly systemic infection characterized by painful involuntary contraction of skeletal muscles. Tetanus carries a 35% mortality rate, so prevention is important.

REF: 102 OBJ: 17

32. Helminth infestation may be detected through stool specimens.

ANS: T

The diagnosis is made by detection of eggs or worms in the anal opening by the placing of transparent adhesive tape in the perianal area. A stool specimen examined microscopically may be positive.

REF: 113 OBJ: 24

33. It is necessary to monitor urinary output of children with diarrhea.

ANS: T

Teach parents to observe urinary output for volume and color. When the child is in diapers, have parents count the number of soiled or wet diapers within a set time and report this to the office.

REF: 116 OBJ: 13

34. Tetanus carries a 35% mortality rate, so prevention is important.

ANS: T

Tetanus carries a 35% mortality rate, so prevention is important.

REF: 103 OBJ: 17

35. Congenital anomalies in a fetus can be diagnosed by taking a fluid sample from the amniotic sac between the 15th and 18th weeks of pregnancy.

ANS: T

One can diagnose congenital anomalies in a fetus by taking a fluid sample from the amniotic sac between the 15th and 18th weeks of pregnancy. This procedure, known as **amniocentesis**, allows amniotic fluid to be tested and cells to be microscopically examined for abnormal substances or chromosomal abnormalities.

REF: 45 OBJ: 1

36. An example of an abnormal substance obtained during an amniocentesis is an elevated alpha-fetoprotein (AFP) level.

ANS: T

An example of an abnormal substance is an elevated AFP level.

REF: 45 OBJ: 3

37. Treatment for bronchiolitis is supportive care.

ANS: T

Treatment is supportive care. Albuterol, racemic epinephrine, or hypertonic saline given via a nebulizer may improve respiratory symptoms. Supplemental oxygen may be required. If the patient is unable to feed, administration of intravenous fluids is necessary. In extreme cases, the patient experiences respiratory failure and must be intubated and mechanically ventilated until the infection runs its course. Even after the acute infection, the airways may be sensitive for several weeks, leading to recurrent wheeze and cough.

REF: 111

OBJ: 23