Robbins Review Questions - Chapter 10

1. Neonate describes what stage of life?
    a. Ages 1-4
    b. Ages 5-14
    c. First year of life
    d. First 4 weeks of life
2. Primary errors of morphogenesis in which there is an intrinsically abnormal developmental process is defined as a
    a. Malformation
    b. Disruption
    c. Deformation
    d. Dysplasia
3. Localized or generalized compression of the developing fetus by abnormal biochemical forces is known as a
    a. Malformation
    b. Disruption
    c. Deformation
    d. Dysplasia
4. Differentiate between a malformation, disruption and deformation.
5. The most common underlying factor responsible for fetal deformation is _____.
    a. Malnutrition
    b. Uterine constraint
    c. Chromosomal defect
    d. Preeclampsia
6. Which of the following best describes a sequence in the context of fetal development?
    a. A group of unrelated anomalies occurring simultaneously without a common cause
    b. A genetically inherited pattern of multiple anomalies affecting several organ systems
    c. A cascade of anomalies triggered by a single initiating aberration
    d. A structural defect caused directly by mechanical forces on a normally developed structure
7. Oligohydramnios is an example of a _____ (malformation/disruption/deformation) leading to a congenital abnormality.
8. A constellation of congenital anomalies believed to be pathologically related but cannot be explained on the basis of a single, localized, initiating defect is a:
    a. Syndrome
    b. Malformation
    c. Deformation
    d. Sequence
9. Atresia is known as:
    a. The absence of organ and its associated primordium
    b. The absence of an opening (usually in hollow organs like esophagus or intestine)

    c. The absence of organ due to failure of growth of the existing primordium
    d. An abnormal organization of cells
10. Aplasia is known as:
    a. The absence of organ and its associated primordium
    b. The absence of an opening (usually in hollow organs like esophagus or intestine)
    c. The absence of organ due to failure of growth of the existing primordium
    d. An abnormal organization of cells
11. The majority of chromosomal malformations are not associated with congenital malformations. True or false?
12. What does TORCH stand for?
13. Drug use accounts for what percent of congenital malformations?
    a. <1%
    b. >1%
    c. <10%
    d. >10%
14. Infant describes what time period of life?
    a. Ages 1-4
    b. Ages 5-14
    c. First year of life
    d. First 4 weeks of life
15. The most common cause of congenital malformations is
    a. Genetic causes
    b. Environmental influences
    c. Multifactorial inheritance
    d. Maternal/placental infection
16. What range of weeks is the fetus most susceptible to teratogens during development?
    a. 1-3
    b. 1-6
    c. 3-6
    d. 3-9
17. What time period does an “embryo” become a “fetus”?
    a. At fertilization
    b. After 4 weeks
    c. After 8 weeks
    d. After 12 weeks
18. What gestational age is the cutoff for a fetus to be classified as premature
    a. < 36 weeks
    b. < 37 weeks
    c. < 38 weeks
    d. < 39 weeks
19. PPROM occurs before 37 weeks of fetal development. True or false?
20. Chorioamnionitis, funisitis, ureaplasma urealyticum and gonorrhea are all examples of _____ that can lead to preterm births.
    a. Intrauterine infections

b. Uterine, cervical or placental structural abnormalities
c. Multiple gestation
d. Necrotizing enterocolitis

21. Multiple gestation is a potential cause for premature birth.
    a. Singleton pregnancy
    b. Multiple gestation
    c. Full-term gestation
    d. Low birth weight due to post-term delivery
22. Pneumatosis intestinalis or gangrenous segments of ileum, cecum or right colon are typical symptoms of what GI disorder in premature births?
    a. Hyaline membrane disease
    b. Necrotizing enterocolitis
    c. Hirschsprung disease
    d. Omphalocele
23. Which of the following is an important cause of fetal growth restriction due to the inability to meet increased fetal demands during vigorous growth?
    a. Maternal diabetes
    b. Uteroplacental insufficiency
    c. Chromosomal abnormalities
    d. Congenital infections
24. What is the main deficiency in premature fetuses that contributes to respiratory distress syndrome?
    a. Alveolar macrophages
    b. Pulmonary surfactant
    c. Bronchial smooth muscle
    d. Type I pneumocytes
25. What features describe lungs from neonatal respiratory distress syndrome victims?
    a. Solid, air filled, reddish purple lungs that sink in water
    b. Solid, airless, reddish purple lungs that float in water
    c. Empty, air filled, pinkish lungs that float in water
    d. Solid, airless, reddish purple lungs that sink in water
26. Which of the following complications is associated with neonatal respiratory distress syndrome?
    a. Tetralogy of Fallot
    b. Patent ductus arteriosus
    c. Coarctation of the aorta
    d. Transposition of the great arteries
27. Neonates born at term are considered small for their gestational age if they weigh less than
    a. 1000 g
    b. 1500 g
    c. 2000 g
    d. 2500 g
28. Transcervical perinatal infections can occur due to _____. Select all that apply.
    a. Inhaling infected amniotic fluid
    b. Parasitic infection

    c. Viral infection
    d. Passing through an infected birth canal
29. Hematologic perinatal infections can occur at any time during gestation and are most commonly due to _____ and _____ infections.
30. Accumulation of edema fluid in the fetus during intrauterine growth refers to the condition of
    a. Immune hydrops
    b. Oligohydramnios
    c. Kernicterus
    d. Fetal hydrops
31. Cystic hygroma is
    a. Generalized edema of the fetus
    b. Post nuchal fluid accumulation
    c. Anemia due to hemolysis
    d. Highly viscous mucus in the respiratory tract due to decreased water secretion
32. Hemolytic disease caused by blood antigen group incompatibility (Rh) results in what disorder?
    a. Fetal hydrops
    b. Immune hydrops
    c. Nonimmune hydrops
    d. Oligohydramnios
33. What is the term that describes bilirubin binding to lipids in the brain?
    a. Hepatic encephalopathy
    b. Kernicterus
    c. Hydrops fetalis
    d. Cerebral edema
34. Turner syndrome (45, X karyotype), transplacental infection by parvovirus B19, and fetal anemia all can lead to
    a. Fetal hydrops
    b. Immune hydrops
    c. Nonimmune hydrops
    d. Oligohydramnios
35. Phenylketonuria, galactosemia and cystic fibrosis are all inborn errors of _____.
    a. Immunity
    b. Metabolism
    c. Waste clearance
    d. Respiration
36. Phenylketonuria is an autosomal recessive disorder where there is an inability to convert phenylalanine into _____.
    a. Tyrosine
    b. Alanine
    c. Methionine
    d. Glutamine

37. Why do subsequent pregnancies (i.e., not the first) with exposure of Rh-negative maternal blood to fetal Rh-positive blood result in an immune response from the mother, while this typically does not occur in first pregnancies?
    a. Maternal immunity is stronger in later pregnancies due to hormonal changes
    b. Fetal Rh status changes between pregnancies
    c. Initial exposure leads to sensitization and antibody formation, which affects future pregnancies
    d. The placenta becomes more permeable in later pregnancies
38. A strong musty or mousy body odor in infants, commonly observed in Scandinavian populations, is indicative of which disorder?
    a. Phenylketonuria
    b. Maple syrup urine disease
    c. Galactosemia
    d. Homocystinuria
39. Vomiting and diarrhea after milk ingestion, eventually leading to hepatomegaly, cataracts and CNS alterations is indicative of _____.
    a. Phenylketonuria
    b. Galactosemia
    c. Cystic fibrosis
    d. Alpha-1 antitrypsin deficiency
40. Which of the following are clinical manifestations of the most common lethal genetic disease affecting Caucasians?
    a. Progressive neurodegeneration with cherry-red spots on the macula
    b. Vaso-occlusive crises and chronic hemolytic anemia
    c. Progressive muscle weakness starting in early childhood, primarily affecting boys
    d. Chronic lung disease secondary to recurrent respiratory infections, pancreatic insufficiency, and salty sweat
41. The mutated CFTR protein in cystic fibrosis primarily affects which of the following systems through abnormal fluid secretion?
    a. Exocrine glands, respiratory tract, gastrointestinal tract, and reproductive tract
    b. Endocrine glands, lymphatic system, and central nervous system
    c. Musculoskeletal, renal, and hematopoietic systems
    d. Cardiovascular, endocrine, and integumentary systems
42. What is the major cause of death in individuals with cystic fibrosis?
    a. Cardiorespiratory complications such as chronic respiratory infections, obstructive lung disease, or cor pulmonale
    b. Liver cirrhosis from bile duct obstruction
    c. Pancreatic insufficiency leading to malnutrition
    d. Complications from diabetes mellitus
43. SIDS is the sudden death of an infant _____ that cannot be explained after thorough investigation.
    a. >1 year old
    b. <1 year old
    c. >2 years old
    d. <2 years old

44. Astrogliosis of the brainstem and cerebellum, along with histological evidence of an upper respiratory infection, are subtle and uncertain signs of which condition?
    a. Neonatal herpes simplex virus infection
    b. Congenital cytomegalovirus infection
    c. Sudden infant death syndrome (SIDS)
    d. Kernicterus
45. What is the most common type of malignant tumor origin in childhood?
    a. Hematopoietic system
    b. Epithelial tissue
    c. Mesenchymal tissue
    d. Neuroectodermal
46. What term describes focal excessive overgrowth of cells or tissues native to a site but without forming normal tissue architecture?
    a. Hyperplasia
    b. Hamartoma
    c. Heterotopia
    d. Neoplasia
47. When normal cells or tissue are present in an abnormal location (ie lung cells in the small intestine) it's described by the term _____.
    a. Hyperplasia
    b. Hamartoma
    c. Heterotopia
    d. Neoplasia
48. What is the most common tumor of infancy?
    a. Neuroblastoma
    b. Hemangioma
    c. Wilms tumor
    d. Teratoma
49. What tumor accounts for the most deaths in children < 15 years old?
    a. Leukemia
    b. Wilms tumor
    c. Hepatoblastoma
    d. Retinoblastoma
50. Which of the following best describes the histologic features of many malignant nonhematopoietic pediatric neoplasms?
    a. They resemble mature adult epithelial cells with well-formed glandular structures
    b. They are composed primarily of fibrous stromal cells with abundant eosinophilic cytoplasm
    c. They show extensive necrosis and calcification without recognizable tissue origin
    d. They often appear as sheets of small round blue cells with primitive features and organ-specific differentiation
51. Which of the following statements about pediatric teratomas is most accurate?
    a. Immature teratomas are benign and most often found in boys under 2 months of age

b. All sacrococcygeal teratomas are malignant due to yolk sac tumor components
c. Sacrococcygeal teratomas are the most common childhood teratomas and show a bimodal age distribution
d. Teratomas do not occur in midline structures such as the mediastinum or retroperitoneum

52. Where do neuroblastic tumors arise?
    a. Parasympathetic ganglia or adrenal medulla
    b. Sympathetic ganglia or adrenal cortex
    c. Sympathetic ganglia or adrenal medulla
    d. Parasympathetic ganglia or adrenal cortex
53. _____ are the most common neuroblastic tumor.
    a. Retinoblastoma
    b. Neuroblastoma
    c. Hepatoblastoma
    d. Leukemia blastoma
54. Blastema, immature stroma and tubules seen histologically are classic descriptors of a
    a. Wilms tumor
    b. Neuroblastoma
    c. Teratoma
    d. Retinoblastoma
55. Gonadal dysgenesis and nephropathy describe what syndrome?
    a. WAGR
    b. Denys-Drash
    c. Beckwith-Wiedemann
    d. Wilms
56. Beckwith-Wiedemann syndrome is distinctive in that in addition to a predisposition towards developing Wilms tumor, individuals with it have
    a. Aniridia and mental disability
    b. Hemihypertrophy and enlarged body organs
    c. Blindness and lymphatic tumors
    d. Hemangiomas on the face and scalp
57. Which of the following is not a major risk factor for premature delivery?
    a. Uterine, cervical and placental structural abnormalities
    b. Twin pregnancy
    c. Premature rupture of placental membranes
    d. Intrauterine infection
58. Which of the following correctly describes the maternal immune response to Rh+ fetal blood in an Rh- mother during initial and subsequent pregnancies?
    a. Initial exposure triggers an IgM response that does not affect the fetus; subsequent exposure triggers an IgG response that can cross the placenta and harm the fetus
    b. Initial exposure triggers an IgG response that affects the fetus; subsequent exposure triggers an IgM response with stronger effects

    c. Both initial and subsequent exposures trigger IgG responses that affect the fetus equally
    d. Initial exposure and subsequent exposure both trigger IgM responses, but only the second response causes hemolysis
59. This autosomal recessive disorder of metabolism will result in vomiting and diarrhea shortly after birth followed by jaundice and hepatomegaly. If untreated, it will cause cataracts within a few weeks and intellectual disability in 6-12 months.
    a. Phenylketonuria
    b. Mucoviscidosis
    c. Galactosemia
    d. Beckwith-Wiedemann
60. This is an inherited disorder of ion transport affecting fluid secretion in exocrine glands and in the epithelial lining of the respiratory, GI and reproductive tracts.
    a. Phenylketonuria
    b. Wilson disease
    c. Mucoviscidosis
    d. Galactosemia
61. Which of the following statements is TRUE regarding maternal phenylketonuria (PKU)?
    a. Dietary restrictions for phenylalanine are only necessary during the third trimester of pregnancy.
    b. Asymptomatic adult women with PKU do not need to follow any dietary restrictions before or during pregnancy.
    c. Women with PKU must resume a low-phenylalanine diet before conception and during pregnancy to prevent developmental defects in the fetus.
    d. Once dietary treatment is stopped in adulthood, phenylalanine levels remain stable and pose no risk to future offspring.
62. A case where an infant dies suddenly and unexpectedly while asleep, without any prior warning or distress and in which the autopsy reveals an anatomical or biochemical basis for the death, is ruled a SIDS (Sudden Infant Death Syndrome) case. True or false?
63. Which of the following is TRUE regarding hemangiomas in children?
    a. Hemangiomas typically require surgical removal because they do not regress.
    b. Hemangiomas generally remain the same size throughout childhood.
    c. Hemangiomas often spontaneously regress, even though they may initially enlarge.
    d. Hemangiomas are malignant vascular tumors that always require chemotherapy
64. In childhood, approximately ___ of neuroblastomas arise in the adrenal medulla
    a. 10%
    b. 15%
    c. 25%
    d. 40%
65. The majority of neuroblastomas occur in the adrenal gland. The next common location is
    a. Parasympathetic chain

b. Sympathetic chain
c. Brain
d. Retina

66. Wilms tumor is the most common primary _____ of childhood.
    a. Bladder tumor
    b. Liver tumor
    c. Renal tumor
    d. Gastric tumor
67. A solitary, circumscribed kidney mass that is soft, homogenous and tan-gray with foci of hemorrhage and necrosis in a 5-year-old is most likely
    a. Neuroblastoma
    b. Renal cell carcinoma
    c. Wilms tumor
    d. Oncocytoma

1. D (pg 453)
2. A (pg 453)
3. C (pg 454)
4. A malformation is a primary error of morphogenesis, in which there is an intrinsically abnormal developmental process (e.g. chromosomal defect). Disruptions result from secondary destruction of an organ or body region that was previously normal in development; thus, in contrast with malformations, disruptions arise from an extrinsic disturbance in morphogenesis. Deformations, like disruptions, represent an extrinsic disturbance of development rather than an intrinsic error of morphogenesis. A deformation is an abnormally formed structure due to mechanical forces (without which the structure would appear normal), such as the flattened face seen in Potter sequence. Disruptions are defects occurring when normal tissue is damaged or interrupted (pg 453-454)
5. B (pg 454)
6. C (pg 455)
7. Deformation (pg 454)
8. A (pg 455)
9. B (pg 454)
10. C (pg 454)
11. False (pg 455)
12. Toxoplasmosis, other (VZ, syphilis, parvovirus), Rubella, CMV, Herpes (pg 458)
13. A (pg 455)
14. C (pg 453)
15. C (pg 455)
16. D (pg 456)
17. C (pg 456)
18. B (pg 457)
19. True (pg 457)
20. A (pg 458)
21. B (pg 458)
22. C (pg 460)
23. B (pg 458)
24. B (pg 458)
25. D (pg 459)
26. B (pg 460)
27. D (pg 458)
28. A & D (pg 461)
29. Parasitic and viral (pg 461)
30. D (pg 462)
31. B (pg 462)
32. B (pg 462)
33. B (pg 463)
34. C (pg 463)
35. B (pg 464)
36. A (pg 465)
37. C (pg 462)

38. A (pg 465)
39. B (pg 463)
40. D (pg 466-467)
41. B (pg 466)
42. A (pg 472)
43. B (pg 473)
44. C (pg 474)
45. A (pg 477)
46. B (pg 475)
47. C (pg 475)
48. B (pg 475)
49. A (pg 477)
50. D (pg 477)
51. C (pg 476)
52. C (pg 477)
53. B (pg 477)
54. A (pg 482)
55. B (pg 481). WAGR and Denys-Drash have similar features and both give rise to Wilms tumor but WAGR features intellectual disability (formerly mental retardation) and a lack of irises.
56. B (pg 482)
57. C (pg 455). Premature rupture of membranes (PROM) is defined as occurring after 37 weeks (not to be confused with preterm premature rupture of membranes (PPROM)). This matters in the context of this question because a fetus delivered after 37 weeks gestational age is no longer considered premature so PROM is not a risk factor for premature delivery
58. A (pg 462)
59. C (pg 466)
60. C (pg 466)
61. C (pg 465)
62. False (pg 473). A SIDS death requires there to be no anatomical or biochemical basis for death
63. C (pg 476)
64. D (pg 478)
65. B (pg 477)
66. C (pg 480)
67. D (pg 482)