

CHOOSE THE SINGLE BEST ANSWER FOR QUESTIONS 1 - 100.

1. Which of the following is true regarding venous thrombosis:
 - A. Most are occlusive
 - B. Most are associated with underlying endothelial injury
 - C. Usually cause infarction
 - D. Usually arise in the saphenous veins
 - E. Consist primarily of fibrin and platelets

2. In a future position, you are administering to victims of a terrorist incident involving radiation exposure from a nuclear device. One of your patients presents with vomiting of bloody material and severe diarrhea which has led to advanced dehydration and complete loss of food intake. Which of the following is correct?
 - A. He/she has received total body radiation dose of 50-100 rad
 - B. He/she will recover without medical intervention
 - C. If he/she survives, affected tissues will be restored to normal
 - D. Leukopenia and thrombocytopenia are likely
 - E. Convulsions and coma are certain to develop

3. At a relief center you encounter a two year old with a history of diarrhea. Your exam reveals listlessness, hepatomegaly and areas of cutaneous desquamation. Serum albumin is abnormally low. Which of the following is least likely?
 - A. Body weight is less than 50% for age
 - B. Lipid peroxidation
 - C. Anemia
 - D. Parasitic infection
 - E. Generalized edema

4. A female patient of yours who has experimented with dietary supplementation and "health products" from commercial outlets presents with complaints of headaches and vomiting. Liver function tests are abnormal and a liver biopsy shows necrosis but no pigment. Which of the following is correct?
- A. The findings are compatible with thiamine toxicity
 - B. The patient is at risk of hypocalcemic tetany
 - C. The patient has B₁₂ toxicity
 - D. Metastatic calcifications are likely
 - E. A pregnancy would be associated with congenital malformation(s)
5. Disease process which is most likely to be exacerbated (worsened) by a lack of Vitamin E:
- A. Collagen synthesis
 - B. Fat malabsorption
 - C. Atherosclerosis
 - D. Bleeding tendency
 - E. Osteomalacia
6. A patient of yours presents with fatigue. Lab studies demonstrate low hematocrit and low rbc count with an erythrocyte mcv of 107. The serum B₁₂ level is normal, and total body iron content is 5 grams. Which of the following is most likely?
- A. Pernicious anemia
 - B. Neural damage to the spinal cord posterior column
 - C. Iron deficiency
 - D. Marrow rbc precursors with small nuclei and abundant pink cytoplasm
 - E. Increased risk of congenital malformation in concurrent pregnancy

7. What is the most likely problem in the above patient(i.e. in question #6)?
- A. A bleeding colorectal neoplasm
 - B. Fat malabsorption
 - C. Inflammatory disease of the terminal ileum
 - D. Recent chemotherapy for a neoplasm
 - E. Autoimmune disease
8. A patient of yours has a serum iron binding capacity twice normal with a saturation of 15%. Which of the following is most likely?
- A. Plasma transferrin is increased
 - B. Serum ferritin is increased
 - C. Stainable hemosiderin in the liver is increased
 - D. The patient has excessive iron absorption
 - E. The patient has recently received multiple blood transfusions
9. A malnourished patient of yours presents with edema, dilated heart, pulmonary congestion and neuropathy. Which of the following is least likely?
- A. Confusion and disorientation
 - B. Peripheral vasoconstriction
 - C. Chronic alcoholism
 - D. Mural thrombi in the heart
 - E. Abnormal liver histology

10. Least likely to be observed in scurvy:
- A. Bone disease
 - B. Abnormal bleeding tendency
 - C. Necrosis of periventricular brain tissue
 - D. Susceptibility to oxidative cell injury
 - E. Skin manifestations
11. Which disease process is least likely to be associated with angiogenesis?
- A. Organization of a thrombus
 - B. Rheumatoid arthritis
 - C. Chronic, "benign" hypertension involving the kidney
 - D. Atherosclerosis
 - E. Invasive growth of a sarcoma
12. What is the most likely location for fibrin?
- A. The middle of keloid
 - B. The cytoplasm of an epithelioid macrophage
 - C. Myocardial hypertrophy
 - D. A hyalinized glomerulus
 - E. A wound in the substrate phase
13. A pathology report you are reading describes the presence of purulent exudate. What disease is most likely?
- A. Pericarditis in acute rheumatic fever
 - B. A thrombus
 - C. Verrucous endocarditis in SLE
 - D. Cryptococcal meningitis
 - E. Acute appendicitis

14. A patient of yours has a diagnosis of carcinoma-in-situ in a cervical biopsy. Which of the following is most likely?
- A. Neoplastic cells demonstrate minimal evidence of maturation
 - B. Regional lymph node dissection/sampling should be performed for Staging
 - C. The patient may have clinically undetectable micrometastases
 - D. Oncogenes have not been mutated or over-expressed in the neoplastic cells
 - E. Neoplastic cells exit the primary organ via the vasculature in large numbers but none has initiated a metastasis
15. Not a consequence of p53 mutation:
- A. Increased likelihood of tumor progression
 - B. Impaired ability to initiate apoptosis
 - C. Increased susceptibility to initiation
 - D. Increased binding to virus encoded protein
 - E. Decreased binding to DNA
16. Which of the following is least characteristic of a colorectal adenoma?
- A. Gross appearance of a very firm, ulcerated lesion
 - B. Lack of formation of gland like spaces
 - C. Increased nuclear to cytoplasmic ratio
 - D. Decreased amount of intracellular mucin in neoplastic cells (relative to normal colon epithelial cells)
 - E. Increased mitotic activity

17. When on a pathology rotation, you encounter a hyalinized granuloma in a lung biopsy. Which of the following statements is most likely correct?
- A. The patient is immunosuppressed
 - B. The lesion may have resulted from Candida infection
 - C. Dystrophic calcifications were present in the lesion
 - D. The patient should be treated for a fungal infection
 - E. The lung was seeded with a fungal pathogen from the blood stream
18. In a tissue biopsy from your patient, a pathologist identifies a 5-7 micron yeast with a mucin-positive capsule. Which of the following is most likely correct?
- A. It is a self limited infection
 - B. The patient has a CD4+ count of 150.
 - C. The patient is more likely to have acquired the infection in California
 - D. The organism was acquired from endogenous flora
 - E. The pathogen would cause liquefactive necrosis
19. Which of the following is least likely to be encountered during the late stages of an HIV infection?
- A. Atrophy of lymph nodes
 - B. Low serum viral titers
 - C. Lymphoma
 - D. Peripheral CD 4+ count of 100
 - E. Abnormal humoral immunity

20. Least likely cellular effect of HIV infection:
- A. Cytoplasmic HIV viral inclusions in infected cells
 - B. Lytic cell death
 - C. Induction of apoptosis
 - D. Integration into host genome
 - E. Non-productive infection (latency)
21. An otherwise normal patient of yours presents with a two week history of fatigue and pharyngitis. There is lymph node enlargement and splenomegaly. "Atypical" mononuclear cells having enlarged nuclei are present in the peripheral blood. Which of the following is most likely correct?
- A. There is likely to be infection of epithelial cells
 - B. The circulating "atypical" cells are infected with virus
 - C. The lymph nodes would show caseous necrosis
 - D. There is monoclonal B cell proliferation
 - E. Complications of this infection may occur but only if the patient was immuno suppressed
22. Over the course of 10-14 days, an immunosuppressed patient of yours develops an interstitial pneumonia diffusely involving multiple lobes. There is no clinical response to conventional antibiotics, including pentamidine. An open lung biopsy fails to reveal fungal hyphae and an AFB stain is negative. What other histologic feature within the lung was most likely?
- A. Purulent exudates
 - B. Non-budding yeast-like cysts filling the alveoli
 - C. Hyalin membranes
 - D. Caseous necrosis
 - E. Coagulation necrosis

23. Disease from which the following is least likely to represent reactivation or complications from a latent/dormant infection:
- A. CMV
 - B. Toxoplasma gondii
 - C. Mycobacterium tuberculosis
 - D. Pneumocystis carinii
 - E. EBV
24. The medical examiner may be notified of a death if:
- A. The death is natural
 - B. The death is suicide
 - C. The death is clearly accidental
 - D. The death is a suspected murder
 - E. Any of the above
25. Rigor Mortis usually lasts about:
- A. 12-24 hours
 - B. 48 hours
 - C. 3 days
 - D. One week
 - E. 6-12 hours
26. When a person dies in a fire, the medical examiner can make a determination that a dead person was alive or dead at the time of fire. This is done by:
- A. A careful examination of the clothing
 - B. The area of burns on the body
 - C. Vital reaction
 - D. A check of carbon monoxide level
 - E. Statement made by witnesses

27. The injuries that are most likely produced by a knife or sharp instrument:
- A. Brush-burn abrasions
 - B. Lacerations
 - C. Contusions
 - D. Forearm wounds in which length exceeds depth
28. After death, the rigor mortis usually become apparent at:
- A. Minutes after death
 - B. Within 1 hour after death
 - C. About 2-4 hours after death
 - D. About 8-12 hours after death
 - E. It does not begin if body is kept warm
29. A coroner:
- A. Is elected
 - B. Must be a physician
 - C. May have state-wide jurisdiction
 - D. Investigates all deaths occurring in his/her jurisdiction
30. In case of stab wounds, the most meaningful measurement that is used to determine the type of knife is:
- A. The width of the wound
 - B. The length of the wound
 - C. The depth of the wound
 - D. The sharp end of the wound
 - E. The blunt end of the wound

31. The close range gunshot wound is the gunshot wound that is fired from the gun at the distance of:
- A. Less than 10 feet
 - B. 12 inches
 - C. 28 inches
 - D. 36 inches
 - E. In contact with the skin surface
32. Which of the following is unlikely to occur in vacuolar degeneration of renal tubular epithelium due to a reduced supply of oxygen?
- A. Cell swelling from increased water content
 - B. Detachment of ribosomes from the rough endoplasmic reticulum
 - C. Clumping of nuclear chromatin
 - D. An increase in intracellular pH
33. A fibrillar substructure is characteristic of each of the following EXCEPT:
- A. Amyloid
 - B. Reduced sickle hemoglobin
 - C. Glycogen
 - D. Mallory hyalin
34. Complications of idiopathic hemochromatosis include each of the following EXCEPT:
- A. Liver cirrhosis
 - B. Cardiomyopathy
 - C. Dissecting aortic aneurysm
 - D. Liver cancer
 - E. Diabetes mellitus

35. Best definition of shock:
- A. Decreased intravascular fluid volume
 - B. Decreased cardiac output
 - C. Impaired myocardial function
 - D. Widespread organ hypo-perfusion
 - E. Low blood pressure
36. A fire broke out in the cabin of an aircraft as it was landing at Detroit Metro. All of the passengers and crew escaped with no significant cutaneous burns. However, four passengers and one flight attendant, who were among the last to exit the plane, died six days later in the hospital. The most likely cause of these deaths was:
- A. Cardiogenic shock
 - B. Overwhelming septicemia
 - C. Acute respiratory distress syndrome (ARDS) related to toxic inhalation injury
 - D. Hypovolemic shock
 - E. Chronic renal failure
37. The effect of ionizing radiation on human cells and tissues is characterized by all of the following EXCEPT:
- A. Polyploidy and/or aneuploidy
 - B. Interference with normal cell cycle progression
 - C. Fragmentation of nuclei (micronuclei)
 - D. Neovascularization of tissues
 - E. Cell death

38. Each of the following statements may be associated with ionizing radiation exposure, EXCEPT:
- A. Increased connective tissue synthesis in target tissues
 - B. A complication of high dose radiation exposure is bone marrow failure
 - C. Leiomyosarcomas are generally radioresistant
 - D. Permanent cell populations are generally radiosensitive
 - E. Irradiated tissues may show cells with tripolar mitoses
39. All of the following statements are CORRECT EXCEPT:
- A. Heavy metals (lead, mercury, cadmium, etc.) preferentially bind to the critical sulfhydryl groups of structural and functional proteins, inactivating these macromolecules
 - B. Chronic alcohol abuse has direct toxic effects on a number of organs and produces pathologic changes that are independent of nutritional factors
 - C. Adverse drug reactions (ADR) are exaggerated physiological responses to drugs administered in prescribed doses and are primarily related to such phenomena as hypersensitivity, genetic susceptibility or other idiosyncratic (poorly understood) causes.
 - D. The inhibition of globin synthesis by lead causes the accumulation of residual ribosome clusters in the cytoplasm of erythrocytes producing "basophilic stippling" of red cells in peripheral blood smears.
 - E. The hematopoietic (bone marrow) syndrome can develop following whole-body ionizing radiation exposure in the range of 200 to 500 rad (rem).

40. The Chernobyl nuclear power facility in the former USSR experienced a core rupture and a subsequent fire, which led to a significant number of deaths among reactor personnel and emergency workers. Many of these workers developed ataxia, convulsions, clinical signs of increased intracranial pressure, lapsed into coma and died within several hours of the accident. This group of deaths was most likely related to:
- A. Radiation-induced gastrointestinal (GI) syndrome
 - B. Radiation-induced central nervous system (CNS) syndrome
 - C. Radiation-induced hemopoietic (bone marrow) syndrome
 - D. Massive radiation exposure to unprotected hands and feet
 - E. Massive septicemia, as a complication of radiation-induced antibiotic-resistance
41. A 59-year-old street person was brought to the emergency department after spending several nights on a park bench in sub-zero temperatures. The toes on both feet revealed signs of severe frostbite. The cellular changes which accompany this appearance include all of the following EXCEPT:
- A. Intracellular ice crystal formation
 - B. Apoptosis
 - C. Severe tissue anoxia
 - D. Red and white cell aggregates occluding small blood vessels
 - E. Epidermal-dermal separation

42. Which one of the following pathogenetic sequences is not likely to explain a manifestation of cystic fibrosis?
- A. Impaired secretion of chloride from eccrine epithelial cells leads to decreased sodium chloride content in sweat.
 - B. Impaired secretion of chloride from bronchial epithelial cells leads to increased absorption of sodium and water from the airspace, resulting in dehydration of the mucus layer. The viscid mucus plugs airways.
 - C. Mucus plugging of pancreatic ducts leads to dilatation of the proximal duct with eventual loss of pancreatic glandular tissue and fibrosis
 - D. Fat malabsorption resulting in vitamin A deficiency contributes to the development of squamous metaplasia in duct linings
 - E. Obstruction of ducts and glands in the male reproductive tract results in azoospermia and sterility
43. Which of the following pathologic abnormalities is not likely to result from sickle cell disease in an adult?
- A. Hemosiderosis of reticuloendothelial cells
 - B. Atrophy of the spleen
 - C. Kernicterus
 - D. Infarction of the central nervous system
 - E. Anemia

44. A 13 year old boy is brought to the emergency room by his parents for acute onset of chest pain. His skin feels cold and clammy. There are numerous cutaneous xanthomas. An electrocardiogram is suggestive of a myocardial infarction.

Which one of the following laboratory findings would help confirm your clinical suspicion of a myocardial infarction?

- A. Elevation of serum creatine kinase
 - B. Elevation of serum glucose
 - C. Elevation of serum low-density lipoprotein (LDL)
 - D. Elevation of serum cholesterol
 - E. Elevation of blood pressure
45. Which one of the following laboratory findings would help to confirm your clinical suspicion of familial hypercholesterolemia?
- A. Elevation of serum creatine kinase
 - B. Elevation of serum glucose
 - C. Elevation of serum low-density lipoprotein (LDL)
 - D. Elevation of serum VLDL cholesterol tryglycerides
 - E. Elevation of blood pressure
46. Which of the following is the most likely genetic profile for this child if he does have familial hypercholesterolemia?
- A. Only the father carries the abnormal gene
 - B. Only the mother carries the abnormal gene
 - C. Both parents carry the abnormal gene
 - D. Neither parent carries the gene, i.e., the mutation is new
 - E. The child carries multiple inherited genes and has been eating too many Wendy burgers, i.e., the disease is multifactorial.

47. The child subsequently develops respiratory distress thought to be due to pulmonary edema and dies. Assuming that you have not missed any other diagnoses, which of the following sequences best summarizes this case?
- A. Elevated creatine kinase -to- myocardial infarction -to- cardiac failure -to- pulmonary edema
 - B. Hyperglycemia -to- atherosclerosis -to- coronary artery occlusion -to- myocardial infarction
 - C. Pulmonary edema -to- myocardial infarction -to- atherosclerosis -to- hypercholesterolemia
 - D. Atherosclerosis -to- coronary artery occlusion -to- myocardial infarction -to- cardiac failure -to- pulmonary edema
 - E. Hypercholesterolemia -to- atherosclerosis -to- myocardial infarction -to- coronary artery occlusion -to- pulmonary edema
48. A postmortem examination is performed on a 35 year old type 1 diabetic woman. Which of the following findings is not likely to be directly related to the diabetes?
- A. Gangrene of the left foot
 - B. Myocardial infarction
 - C. Mycroangiopathy in the retina
 - D. Leiomyomas of the uterus
 - E. Chronic pyelonephritis
49. The neuroendocrine theory of aging depends upon the concept of:
- A. The amount of energy consumed during the lifetime of the organism
 - B. A "pacemaker" center in the central nervous system
 - C. An accumulation of errors in the genetic machinery of the cell
 - D. Post-translational modifications of large molecules impairing cell function
 - E. free radicals causing progressive damage to cellular macromolecules

50. During the last millennium the maximum life span potential of humans as a whole has:
- A. Increased significantly
 - B. Stayed approximately the same
 - C. Decreased slightly
 - D. Decreased significantly
51. An elderly person with a history of chronic hypertension and atherosclerotic vascular disease is being considered for surgery that may require additional treatment with a drug whose normal route of inactivation includes urinary excretion of a conjugated metabolite. In determining the dose, the resident should probably:
- A. Use the normal dose
 - B. Consider increasing the dose to accommodate the expected decrease in hepatic function seen in elderly individuals
 - C. Count on the resilience of the elderly to overcome any adverse drug reaction
 - D. Consider decreasing the dose to accommodate the expected decrease in renal function due to the patient's underlying conditions
 - E. Give extra fluid and glucose to prevent dehydration and help wash out the drug
52. Elevated blood levels of human chorionic gonadotropin (HCG) are a useful marker for the presence of:
- A. A teratoma
 - B. A hemangioma
 - C. A choriocarcinoma
 - D. A neuroblastoma
 - E. a yolk sac carcinoma

53. The malformation in an individual with craniopagy most likely:
- A. Was inherited in the genes
 - B. Occurred during the first two weeks of gestation
 - C. Occurred during the early fetal period
 - D. Occurred during the late fetal period
 - E. Resulted from perinatal injury
54. A persistent patent ductus arteriosus is an example of:
- A. An involution failure
 - B. An atresia
 - C. An ectopia
 - D. A dysplasia
 - E. A dysraphism
55. Which one of the following abnormalities could be noted by examination of the placenta?
- A. Duodenal atresia
 - B. Cleft lip
 - C. Malrotation of the gut
 - D. Anencephaly
 - E. Amniotic bands
56. Which of the following X-linked disorders is X-linked dominant?
- A. Duchenne muscular dystrophy
 - B. Fragile X syndrome
 - C. Vitamin D resistant rickets
 - D. Hemophilia A
 - E. Hunter's disease (MPS II)

57. What is the likely cause of myocardial infarction in a patient with Hurler's disease?
- A. Accelerated atherosclerosis
 - B. Massive deposition of amyloid in the myocardium
 - C. Accumulation of dermatan or heparan sulfate in endothelial and smooth muscle cells
 - D. Persistent patent ductus arteriosus with right heart failure
 - E. Increased susceptibility to infections
58. In which of the following diseases would histologic examination of brain tissue be most likely to provide a diagnosis in an infant suspected of having the disease?
- A. Tay-Sachs
 - B. Trisomy 21
 - C. Cystic fibrosis
 - D. Marfan's
 - E. Phenylketonuria
59. A full-term infant product of an uncomplicated pregnancy and delivery and his mother return from the hospital the third postnatal day breast-feeding normally. The following day, the mother notes that the infant has developed diarrhea. On the second day, the infant begins vomiting and is brought to your attention. On examination, the infant appears ill and has hepatomegaly. Which one of the following inherited diseases is most likely in this setting?
- A. Phenylketonuria
 - B. Gaucher
 - C. Galactosemia
 - D. Niemann-Pick
 - E. Tay-Sachs

60. In which of the following tumors would Flexner-Wintersteiner rosettes most likely be found?
- A. Wilm's tumor
 - B. Squamous cell carcinoma
 - C. Teratoma
 - D. Retinoblastoma
 - E. Adenocarcinoma
61. A newborn infant develops a distended abdomen on the second-day of life and is found to have an obstruction of the small bowel. At surgery, the distended distal portion of the bowel is found to be impacted with viscid mucus and meconium (fetal stool) but no anatomic narrowing is noted at the ileocecal valve. The most likely condition accounting for this finding is:
- A. Necrotizing enterocolitis
 - B. Duodenal atresia
 - C. Deficiency of surfactant
 - D. TORCH infection in utero
 - E. Cystic fibrosis
62. A patient with neurofibromatosis type 1 states that neither of his parents had neurofibromas, cafe-au-lait spots or other known manifestations of the disease. The most likely explanation for this is:
- A. Neurofibromatosis is multifactorial
 - B. The defective enzyme was compensated for by the other allele
 - C. Neurofibromatosis is recessive
 - D. The patient has a new mutation
 - E. Amplification of trinucleotide repeat occurred in the carrier mother

63. Which one of the following diseases would not be characterized by lysosomal storage of an intermediary metabolite?
- A. Pompe's (glycogenosis type II).
 - B. Hurler's (mucopolysaccharidosis type I)
 - C. Tay-Sachs
 - D. Marfan's
 - E. Gaucher's
64. Each one of the following diseases would be most likely to cause splenomegaly?
- A. Niemann-Pick.
 - B. α 1-antitrypsin deficiency.
 - C. Cystic fibrosis.
 - D. Duchenne muscular dystrophy.
 - E. Marfan.
65. A boy presents with a muscle disease characterized by necrosis of muscle fibers and progressive fibrosis on biopsy. You are able to demonstrate absence of an important structural protein in the boy's muscle. While taking the family history, you learn that his older brother and his maternal grandfather had evidence of a similar disease. The mother's sister and brother are well. The mother's brother's two sons are tested, and the normal protein is present.

The most likely pattern of inheritance is:

- A. Autosomal dominant
- B. Autosomal recessive
- C. X-linked dominant
- D. X-linked recessive
- E. Other

66. A newborn is evaluated for the presence of a large congenital melanocytic nevus covering much of the back and upper buttock. The parents wish to know the likelihood that the infant will develop a malignant melanoma. You answer:
- A. 0%
 - B. 10%
 - C. 50%
 - D. 95%
 - E. The lesion is already malignant.

THE FOLLOWING HISTORY IS RELATED TO QUESTIONS 67 - 69.

A 22-year-old man is evaluated for unilateral hearing loss and found to have a tumor of the VIIIth cranial nerve. On examination, the man also has several flat pigmented spots over the trunk and proximal limbs. The tumor is removed without complication. Three years later, the man reappears complaining of headaches and is found to have a tumor on the opposite VIIIth cranial nerve. This is removed and is histologically identical to the previous tumor.

67. The most likely histologic diagnosis for the two tumors is:
- A. Neuroblastoma
 - B. Metastatic carcinoma
 - C. Schwannoma
 - D. Germinoma
 - E. Lisch nodule.
68. A karyotype analysis of fibroblasts from this patient is likely to show:
- A. No abnormality
 - B. An abnormal Y chromosome
 - C. An extra X chromosome
 - D. An extra chromosome 22
 - E. An 11;22 translocation
69. The patient's father had the same disease. The likelihood that the patient's brother also has the same disease as the patient is:
- A. 0%
 - B. 25%
 - C. 50%
 - D. 75%
 - E. 100%

70. A young man who survived bilateral retinoblastomas in childhood now wishes to have children. His younger brother died of osteogenic sarcoma, and his mother has been treated for breast cancer. He suspects that he has inherited an abnormal gene.

The risk of one of his children inheriting the same gene is:

- A. 0%
 - B. 7%
 - C. 25%
 - D. 50%
 - E. 99%
71. A year later, the patient presents with pain around the right orbit and is found to have a tumor involving the roof of the orbit. He has no evidence of tumor in other organs. The most likely diagnosis of the new tumor is:
- A. Wilm's tumor
 - B. Hemangioma
 - C. Osteogenic sarcoma
 - D. Acute myelogenous leukemia
 - E. Neurofibroma
72. A male infant is born prematurely at 32 weeks gestation. On examination, the infant is hydropic with an erythematous rash, skin petechiae, chorioretinitis, and a small head circumference. Ultrasound examination of the head shows dilated ventricles in the brain and periventricular calcifications.
- Which of the following tests is most likely to provide evidence for a cause for this infant's problems?
- A. Bacterial culture of the placenta for Gram-negative rods.
 - B. Measuring titers for TORCH agents in the mother.
 - C. Culturing the infant's skin for Candida.
 - D. Performing a karyotype analysis on the child and mother.
 - E. Asking the mother for a history of ethanol abuse.

73. The infant's problem most likely started:
- A. Prior to conception, i.e., it was genetic and inherited.
 - B. During the first developmental week.
 - C. In mid gestation.
 - D. At the time of birth.
 - E. In the neonatal nursery.
74. You are performing a postmortem examination of a 2-month-old male infant found dead in his crib one morning. Which one of the following findings would clinch your suspected diagnosis of SIDS?
- A. A cardiac malformation
 - B. Evidence of lysosomal storage material in reticulo-endothelial cells in the spleen
 - C. Aspirated milk in one mainstem bronchus
 - D. Broken ribs and ecchymoses over the head and trunk
 - E. A normal autopsy except for congested organs
75. Following an episode of respiratory distress syndrome requiring prolonged mechanical ventilation, a 3-month-old infant born at 26 weeks gestation, continues to have respiratory insufficiency and slowly develops evidence of right heart failure. The infant remains afebrile. These cardiorespiratory problems are most likely due to:
- A. A lung hamartoma
 - B. Bronchopulmonary dysplasia
 - C. Germinal matrix hemorrhage
 - D. Acute bronchopneumonia
 - E. Hydrops fetalis

76. A G1P0 mother with no previous prenatal care presents in labor at 38 weeks gestation. Ultrasonography shows oligohydramnios. Following breech delivery, the neonate immediately develops respiratory distress and requires ventilatory support with a respirator. On examination, his face appears flattened and the limbs appear distorted and fixed primarily in flexion.

The most likely cause of the respiratory distress is:

- A. Pulmonary hypoplasia
 - B. Deficiency of pulmonary surfactant
 - C. Bronchopulmonary dysplasia
 - D. Congenital Rubella infection
 - E. Pneumonia due to Listeria infection
77. The infant's external morphologic abnormalities are an example of:
- A. A malformation
 - B. A chromosomal abnormality
 - C. A deformation
 - D. A developmental field defect
 - E. A disruption
78. Which one of the following is least likely to have caused the abnormalities in this infant?
- A. Congenital polycystic kidney disease
 - B. Rupture of the amniotic sac several weeks prior to onset of labor
 - C. A uterus distorted by leiomyomas in the myometrium
 - D. Maternal-infant blood-group incompatibility
 - E. An unusually small uterus

Questions 79 - 82 refer to the following figure.

1. transaminase

2-oxo-isocaproic acid

2. branched chain oxo-acid
dehydrogenase

isovaleryl-CoA

isovaleric acid

3. isovaleryl-CoA
dehydrogenase

3-methylcrotonyl-CoA

methylcrotonic acid

4. methylcrotonyl-CoA
carboxylase

3-methylglutaconyl-CoA

methylglutaconic acid

5. methylglutaconyl-CoA
hydratase

3-OH-3-methylglutaryl-CoA

6. lyase

7. reductase

acetoacetic acid + acetyl-CoA

mevalonic acid

mevalonic acid

8. pathway

cholesterol

Several metabolic diseases, the branched chain organic acidurias, due to mutations causing cytoplasmic enzyme deficiencies in the catabolism of leucine, are characterized severe metabolic problems and urinary excretion of intermediary metabolites. The accompanying figure summarizes the pertinent metabolic pathways. The enzymes are numbered.

79. One metabolite, isovaleric acid, produces an odor of "sweaty feet" when excreted in the urine. Which enzyme deficiency would be most likely to produce the finding of a "sweaty feet" odor in the urine?
- A. Deficiency of enzyme 1
 - B. Deficiency of enzyme 2
 - C. Deficiency of enzyme 3
 - D. Deficiency of conversion of isovaleryl-CoA to isovaleric acid
 - E. Deficiency of enzyme 6
80. Which of the following enzyme defects would be expected to produce urinary excretion of mevalonic acid (mevalonic aciduria)?
- A. Deficiency of enzyme 1
 - B. Deficiency of enzyme 4
 - C. Deficiency of enzyme 6
 - D. Deficiency of enzyme 7
 - E. None of the above
81. In a case summary of 41 reported cases of isovaleric aciduria, it was noted that 58% were female. In some families, two or more siblings had the disorder, but the parents were normal. What is the likely pattern of inheritance?
- A. Autosomal dominant
 - B. Autosomal recessive
 - C. X-linked recessive
 - D. X-linked dominant
 - E. Chromosomal defect

82. The intervention most likely to be effective in treating an infant with isovaleric aciduria would be:
- A. Administering a cholesterol-poor diet.
 - B. Administering an inhibitor of isovaleryl-CoA dehydrogenase
 - C. Administering a leucine-poor diet
 - D. Administering a leucine-rich diet
 - E. Administering inhibitors of cholesterol synthesis

MATCHING ITEMS

DIRECTIONS: Match the Vitamin (A-F below) to the most appropriate choice 83 - 88. Each answer may be used once, more than once or not at all.

- A. Vitamin E
- B. Vitamin K
- C. Vitamin D
- D. Vitamin C
- E. Vitamin A
- F. Vitamin B₁₂

- 83. Deficiency may result from inherited genetic defect affecting synthesis
- 84. Deficiency state may be precipitated by antibiotics
- 85. Facilitates hydroxylation of proline or lysine residues
- 86. Acts as a hormone
- 87. Facilitates calcium dependent interactions involving proteins and phospholipids
- 88. Deficiency results in impaired osteoid synthesis

DIRECTIONS:

Select the alternative (A-E below) that is most closely associated with the statements numbered 89 - 93.

- A. High concentrations of oxygen
- B. Chronic lead poisoning
- C. Chronic high dose sulfur dioxide exposure
- D. Radiotherapy of an adenocarcinoma of the breast
- E. Third-degree (full thickness) cutaneous burns

- 89. A 34-year-old male presents with signs and symptoms of peripheral neuropathy
- 90. Known to enhance free radical-induced (indirect) radiation injury
- 91. Vascular and interstitial pulmonary fibrosis is potential complication
- 92. Shock and septicemia are important associated complications
- 93. Chronic bronchitis and squamous metaplasia often develop

DIRECTIONS: Match the mediator 94 - 98 to the function or characteristic for which it is most directly related (A-E below). Use each choice once only.

- A. Enhancement of phagocytosis
- B. Causes leukocytosis
- C. Pain
- D. PMN chemotaxis
- E. Derived from membrane phospholipid PMN

94. Csa

95. Bradykinin

96. C3b

97. Platelet activating factor (PAF)

98. TNF (tumor necrosis factor)

DIRECTIONS: Match each statement 99 - 102 with its corresponding intercellular accumulation from those provided (A-D below). Use each alternative once only.

- A. Excess basal laminar material
- B. Metastatic calcification
- C. Amyloid deposits
- D. Dystrophic calcification

99. May occur in caseous necrosis

100. An effect of systemic hypertension on intrarenal arterioles

101. An effect of vitamin D toxicity

102. An effect of plasma cell disorders

pictures

1. atherosclerotic aorta
2. renal tumor: Wilm=s
3. CMV renal tubular epithelium
4. HMD (arrow on membrane)
5. medulloblastoma
6. Gaucher: hepatosplenomegaly
7. renal dysplasia (Potter=s)
8. sickle cell high power spleen
9. X-dominant inheritance
10. Hunter=s: face
11. XO picture
12. cleft lip;face
13. necrotic muscle fibers :DMD
14. gross: PVL bilat white cavities
15. fetus with facial anomalies (amniotic band)
16. karyotype trisomy 18

PHOTO 41

This specimen was removed at postmortem examination of a 70-year-old man. Among the other pathologic findings found, which of the following is probably not directly or indirectly related to the disease process illustrated in the specimen.

- a. focal scarring in the myocardium.
- b. obesity.
- c. organizing thrombus in the distal femoral artery.
- d. lipofuscin accumulation in cardiac myocytes.
- e. atrophy of the kidneys.

Answer: d.

The cause of this disorder is most likely

- a. inherited by an autosomal recessive gene.
- b. due to dietary excess.
- c. multifactorial.
- d. due to hypertension.
- e. a consequence of progeria.

Answer: c.

PHOTO 12

This specimen was removed surgically from a 3-year-old male child with an enlarging abdomen and palpable left-sided abdominal mass. Given this appearance, the most likely diagnosis is

- a. dysgerminoma.
- b. Wilm=s tumor.
- c. hepatoblastoma.
- d. acute lymphocytic leukemia.
- e. omphalocele.

Answer: b.

The oncogene most likely associated with this tumor is located on chromosome

- a. 11.
- b. 22.
- c. X.
- d. 21.
- 3. 16.

Answer: a.

PHOTO 13

The following section was obtained from the kidney on postmortem examination of a macerated stillborn infant. Similar findings were noted in most other organs. In the absence of another cause, the stillbirth was most likely due to

- a. a major malformation.
- b. an umbilical cord accident.
- c. a TORCH infection.
- d. a congenital neoplasm.
- e. an inherited metabolic disease.

Answer: c.

Which of the following findings might also be expected in this circumstance?

- a. hyaline membrane disease.
- b. cystic hygroma.
- c. sacrococcygeal teratoma.
- d. chorioretinitis.
- e. lysosomal storage.

Answer: d.

PHOTO 14

This slide was taken from the lung of a 5-day-old infant born at 37 weeks gestation. The laboratory abnormality usually associated with this finding is

- a. hyperbilirubinemia.
- b. hypercholesterolemia.
- c. hypovolemia.
- d. hyperglycemia.
- e. hypoxemia.

Answer: e.

In an infant born at 26 weeks gestation, the predisposing factor for this condition is

- a. immature hepatocytes unable to conjugate bilirubin.
- b. rupture of blood vessels in the germinal matrix.
- c. immature alveolar lining cells unable to synthesize lecithin.
- d. colonization by *Clostridium difficile*.
- e. deposition of amylin.

Answer: c.

PHOTO 15

A 4-year-old girl presents with headache and irritability and is found to have a cerebellar lesion, which is biopsied. Given the histologic features illustrated, her most likely diagnosis is

- a. bacterial abscess.
- b. congenital hemangioma.
- c. lysosomal storage disease.
- d. Beckwith-Wiedemann syndrome.
- e. medulloblastoma.

Answer: e.

The expected inheritance pattern for this condition is

- a. autosomal dominant.
- b. autosomal recessive.
- c. X-linked recessive.
- d. sporadic.
- e. mitochondrial.

Answer: d.

PHOTO 16

On opening the abdomen at postmortem examination of a 1 1/2-year-old male infant who died of Gaucher=s disease, the pathology resident makes this photograph. The finding illustrated is best characterized as

- a. hypertrophy.
- b. atrophy.
- c. neoplasia.
- d. dysplasia.
- e. disruption.

Answer: a.

The finding is most likely related to

- a. chronic passive congestion due to heart failure from atherosclerotic coronary artery disease..
- b. distention of macrophages by phagocytosed glucocerebrosides.
- c. proliferation of hepatocytes.
- d. a failure of neural tube closure early in development.
- e. overeating.

Answer: b.

PHOTO 17

The section represents the typical appearance of the kidney of an infant born at 36 weeks gestation with arthrogryposis. He developed intractable respiratory failure at birth and died shortly thereafter.

The mother was noted to have oligohydramnios on examination prior to delivery. The reason for respiratory failure is most likely due to

- a. hypoplasia of lung parenchyma.
- b. prematurity.
- c. intrapartum infection of the lung.
- d. birth trauma with pulmonary hemorrhage..
- e. maceration.

Answer: a.

In this case, the arthrogryposis is an example of

- a. an inherited genetic defect.
- b. a true malformation.
- c. a deformation.
- d. a disruption.
- e. a hypoplasia.

Answer: c.

PHOTO.18

This section of spleen was taken from a 20 year old woman with a history of episodes of abdominal and joint pains, a previous stroke, and chronic anemia. She presented with acute pneumococcal pneumonia and died. The change in the erythrocytes in the section is most likely due to

- a. a lysosomal storage disease.
- b. a defect in a regulatory protein
- c. an enzyme defect.
- d. a defect in a coagulation factor.
- e. a defect in a structural protein.

Answer: e.

The pathobiologic problem most likely to result from this abnormality is

- a. thrombosis.
- b. edema.
- c. neoplasia.
- d. hypertrophy.
- e. atresia

Answer: a.

PHOTO 19

This karyotype was performed on cells obtained from amniocentesis (sampling of cells from the amniotic fluid) in a 40-year-old pregnant mother at about 16 weeks gestation along in her first pregnancy. The karyotype diagnosis is

- a. triploidy.
- b. 45, XO
- c. 47, XXY.
- d. trisomy 18.
- e. trisomy 21.

Answer: d.

The fetus is at increased risk for all of the following except

- a. intrauterine growth retardation.
- b. spontaneous abortion.
- c. erythroblastosis fetalis.
- d. prematurity.
- e. holoprosencephaly.

Answer: c.

PHOTO 20

An infant born at 36 weeks gestation with congenital heart disease and respiratory distress syndrome succumbed at age 8 weeks from intractable right heart failure. Examination of the brain was performed at autopsy. The brain weight was normal. The finding illustrated represents

- a. germinal matrix hemorrhage.
- b. holoprosencephaly.
- c. status marmoratus.
- d. anencephaly.
- e. periventricular leukomalacia.

Answer: e.

The finding was most likely due to

- a. congenital cytomegalovirus infection.
- b. hypoxic/ischemic injury.
- c. an inherited deficiency important in the formation of myelin.
- d. hyperbilirubinemia.
- e. continued myelination of scarred lesions.

Answer: b.