

Damjanov: Pathophysiology

Test Bank

Set 2 (questions 26-50)

MULTIPLE CHOICE

1. Which of the following hereditary diseases represents a congenital enzyme deficiency?
 - A. cystic fibrosis
 - B. sickle cell anemia
 - C. phenylketonuria
 - D. Marfan's syndrome
 - E. hereditary spherocytosis

ANS: C

All the diseases listed in this question represent a mutation of genes that encode a specific protein, but only phenylketonuria is related to a gene that encodes an enzyme (phenylalanine hydroxylase). Cystic fibrosis involves the cystic fibrosis transmembrane conductance regulator, sickle cell anemia involves hemoglobin beta gene, Marfan's syndrome involves a structural protein (fibrillin), and hereditary spherocytosis a structural protein (spectrin or ankyrin) in the cytoskeleton of red blood cells.

2. The severity of diseases inherited through mitochondrial inheritance varies and depends on the ratio of normal and abnormal mitochondria, a mixture that is called
 - A. heterochromatin
 - B. heteroplasm
 - C. heterotaxia
 - D. genetic mosaicism
 - E. dysgenesis

ANS: B

Heteroplasm, a mixture of normal and abnormal mitochondria in various tissues, accounts for the variable severity of mitochondrial diseases. Heterochromatin is clumped chromatin in the nucleus that remains clumped during mitosis. Heterotaxia is the technical term for abnormally located organs. Mosaicism is a juxtaposition of genetically distinct cells in the same tissues, as is sometimes seen in Turner's syndrome. Dysgenesis is abnormal development of organs or tissues.

3. Tumor-induced angiogenesis is promoted by
 - A. vascular endothelial growth factor (VEGF)
 - B. thrombospondin
 - C. endostatin
 - D. vasostatin
 - E. angiostatin

ANS: A

VEGF, a secretory product of many cells, promotes angiogenesis by binding to receptors on endothelial cells. VEGF promotes endothelial cell growth, migration, and vessel formation. Thrombospondin (platelet factor 4), angiostatin (cleavage product of plasminogen), endostatin (cleavage product of collagen type XVIII), and vasostatin (cleavage product of calretinin) prevent angiogenesis.

4. Ovarian cancers tend to metastasize to the lower surface of the diaphragm and the surface of the liver by
- A. local invasion
 - B. lymphatic spread
 - C. vascular spread
 - D. transcoelomic spread
 - E. venous embolism

ANS: D

Ovarian cancer has a tendency for transcoelomic spread. Such tumors detach from the primary site, float free in the abdominal cavity, and implant on the serosal surface of the liver, the intestines, or the lower surface of the diaphragm.

5. A 60-year-old man complaining of prolonged cough and expectoration for the last 6 months, fatigue, and loss of weight was admitted to the hospital for further studies. A hilar lung mass was found on x-ray, and the blood showed hypercalcemia. The most likely cause of hypercalcemia is a pulmonary
- A. adenocarcinoma
 - B. oat-cell (small cell) carcinoma
 - C. squamous cell carcinoma
 - D. carcinoid
 - E. mesothelioma

ANS: C

Hypercalcemia is most often caused by the parathyroid hormone-related protein (PTHrP), a calcium regulatory polypeptide hormone. PTHrP is normally secreted by squamous cells of the skin and can be found in the blood of patients who have squamous cell carcinoma of the bronchi.

6. A benign epithelial skin tumor composed of a frond-like outgrowth of squamous epithelium covering central fibrovascular cores is called
- A. adenoma
 - B. fibroadenoma
 - C. papilloma
 - D. rhabdomyoma
 - E. squamous cell carcinoma

ANS: C

Exophytic squamous cell tumors of the skin or transitional cell urinary bladder tumors are called papillomas (i.e., appear nipple-like; the term is derived from the Latin word *papilla*, or nipple). Adenomas and fibroadenomas are tumors composed of glandular epithelium. A rhabdomyoma is a mesenchymal tumor composed of striated muscle cells and not an epithelial neoplasm. Squamous cell carcinoma is an epithelial tumor but is malignant in contrast to papilloma.

7. Which of the following tumors is malignant?
- A. tubular adenoma
 - B. lymphoma
 - C. leiomyoma
 - D. chondroma
 - E. capillary hemangioma

ANS: B

All lymphomas are malignant, although their degree of malignancy varies. All other tumors listed here are benign. Adenoma is a benign tumor of glandular epithelium, leiomyoma of smooth muscle cells, chondroma of cartilage cells, and capillary hemangioma of blood vessels.

8. Aromatic amines found in the rubber industry enter the human body in the workplace and are converted into carcinogens in the liver. This conversion is mediated by
- A. alanine aminotransferase
 - B. aspartate aminotransferase
 - C. alkaline phosphatase
 - D. cytochrome P450 oxygenase
 - E. DNase

ANS: D

Cytochrome P450 is a group of enzymes that are essential for the metabolic conversion of many endogenous and exogenous substances in liver cells. These enzymes are involved in the conversion of some procarcinogens into proximate carcinogens. The carcinogens derived from the conversion of azo dyes in liver cells are released into the blood and reach the urinary bladder, where they cause urothelial neoplasia. Alanine aminotransferase, aspartate aminotransferase, and alkaline phosphatase belong to the clinically important enzymes forming the so-called liver function tests and are not involved in carcinogenesis. DNase is an enzyme that degrades DNA and is not related to activation of carcinogens, which takes place in the microsomal fraction of the cytoplasm.

9. Kaposi's sarcoma has been pathogenetically linked to an infection with
- A. human papilloma virus
 - B. Epstein-Barr virus
 - C. hepatitis virus B
 - D. human T-cell lymphoma/leukemia virus
 - E. herpes virus 8

ANS: E

Herpes virus 8 plays a role in the pathogenesis of Kaposi's sarcoma. Human papilloma virus causes carcinomas of the cervix, vulva, and penis. Epstein-Barr virus has been linked to Burkitt's lymphoma and nasopharyngeal carcinoma. Human T-cell lymphoma/leukemia virus is the cause of T-cell lymphoid neoplasms.

10. Asbestos has a major role in the pathogenesis of malignant tumors of the
- A. brain
 - B. thyroid

- C. pleura
- D. liver
- E. stomach

ANS: C

Asbestos has an important role in the pathogenesis of mesotheliomas, malignant tumors of the pleura. Lung cancers also occur at a higher rate following long-term asbestos exposure.

11. Alpha-fetoprotein (AFP) is found in increased concentrations in the blood of adults who have carcinoma of the
- A. esophagus
 - B. stomach
 - C. pancreas
 - D. liver
 - E. colon

ANS: D

AFP is the major fetal plasma protein produced by the fetal liver. AFP is also produced by liver cell carcinomas, and thus in adults it is a good serological marker of hepatocellular carcinoma. Germ cell tumors containing yolk sac components also secrete AFP into the blood. Other carcinomas listed in this question do not secrete AFP. Adenocarcinomas of the colon, esophagus, stomach, and pancreas produce carcinoembryonic antigen, which can also be used as a serological tumor marker.

12. Which of the following malignant tumors has a 5-year patient survival rate of less than 10% with current therapy?
- A. papillary carcinoma of the thyroid
 - B. basal cell carcinoma of the skin
 - C. Hodgkin's disease
 - D. seminoma of the testis
 - E. adenocarcinoma of the pancreas

ANS: E

Adenocarcinoma of the pancreas is almost invariably lethal and only 5% of all patients survive for 5 years. Seventy percent or more of patients who have the other four tumors listed here survive more than 5 years with modern therapy.

13. Three sons of a normal married couple had congenital immunodeficiency, but their three daughters were not affected. The boys had very low serum IgG levels, and their lymph nodes were devoid of germinal centers. No plasma cells were seen, but T lymphocytes were normal. This condition represents
- A. X-linked agammaglobulinemia of Bruton
 - B. DiGeorge's syndrome
 - C. adenosine deaminase deficiency
 - D. severe combined immunodeficiency of Swiss type
 - E. ataxia-telangiectasia

ANS: A

The male members of this kindred are affected by X-linked agammaglobulinemia (XLA), or Bruton's disease. XLA affects only males. The affected boys show signs of immunodeficiency and have very low serum IgG levels or complete agammaglobulinemia combined with a lack of B-lymphocyte maturation in lymph nodes. Pre-B cells are present but do not mature into B lymphocytes or plasma cells. T lymphocytes form normally. The other four immunodeficiency disorders are not inherited as X-linked traits and affect both males and females. DiGeorge's syndrome is a T-cell deficiency related to aplasia or hypoplasia of the thymus. Adenosine deaminase deficiency and other forms of combined immunodeficiency and ataxia-telangiectasia involve both B and T lymphocytes.

14. Human immunodeficiency virus (HIV-1), the cause of acquired immunodeficiency syndrome (AIDS), selectively infects
- A. B cells
 - B. T-helper cells
 - C. T-suppressor cells
 - D. natural killer cells
 - E. plasma cells

ANS: B

Human immunodeficiency virus selectively infects the CD4+ T-helper lymphocytes.

15. Allergic rhinitis and atopic conjunctivitis are mediated by
- A. IgA
 - B. IgE
 - C. IgG
 - D. IgD
 - E. IgM

ANS: B

Like other type I hypersensitivity reactions, allergic rhinitis (hay fever), and atopic conjunctivitis are mediated by IgE.

16. Hemolytic disease of the newborn due to the maternal-fetal blood group incompatibility represents which form of hypersensitivity?
- A. type I
 - B. type II
 - C. type III
 - D. type IV
 - E. graft-versus-host reaction

ANS: B

Hemolysis due to maternal-fetal incompatibility represents a type II hypersensitivity reaction. In classic cases the red blood cells of an Rh(+) fetus/infant are destroyed by anti-Rh antibodies in a Rh(-) mother sensitized to Rh(+) antigen in a previous pregnancy. Binding of the cytotoxic maternal antibodies to fetal red blood cells activates complement, which forms the membrane attacked complex (MAC) perforating the red blood cell membrane.

17. A 40-year-old woman had tachycardia and bouts of sweating, and was found to have hyperthyroidism due to Graves' disease. Her blood contained antibodies to
- A. Thyroid-stimulating hormone receptor
 - B. thyroglobulin
 - C. smooth antibodies
 - D. mitochondria
 - E. centromere

ANS: A

Patients with Graves' disease have antibodies to the receptor for the thyroid-stimulating hormone (TSH). These antibodies bind to thyroid follicular cells, stimulating them to produce thyroid hormones thyroxin (T_4) and triiodothyronine (T_3). Antibodies to thyroglobulin are found in Hashimoto's disease. Antibodies to smooth muscle cells are found in autoimmune hepatitis. Antibodies to mitochondria are found in primary biliary cirrhosis. Antibodies to centromere are found in scleroderma.

18. Condylomata lata, warty growths on the skin around the genitalia and on other sites, are typical of which stage or form of syphilis?
- A. primary syphilis
 - B. secondary syphilis
 - C. tertiary syphilis
 - D. latent syphilis
 - E. congenital syphilis

ANS: B

Condylomata lata are typically found in secondary syphilis, one to three months after onset of infection. Recurrence of secondary syphilis can occur in asymptomatic patients considered to have latent syphilis.

19. Granulomas of tuberculosis and gummas of syphilis contain several cell types, but gummas also contain some cells that are not found in other granulomas. Which are those cells?
- A. T lymphocytes
 - B. macrophages
 - C. multinucleated giant cells
 - D. plasma cells
 - E. fibroblasts

ANS: D

T lymphocytes, epithelioid macrophages, multinucleated giant cells, and fibroblasts are found in granulomas of tuberculosis and gummas of syphilis. Plasma cells are found only in gummas of syphilis, correlating with the antitreponemal antibodies found in most of these patients.

20. A 25-year-old woman developed septicemia accompanied by the appearance of an exfoliating rash on her palms and soles. Subsequent studies found that this toxic shock syndrome (TSS) was related to a bacterial colonization of the vaginal tampon. What is the most likely pathogen to be isolated from the tampon?
- A. *Escherichia coli*
 - B. *Salmonella typhi*

- C. *Shigella dysenteriae*
- D. *Staphylococcus aureus*
- E. *Streptococcus viridans*

ANS: D

TSS is most often caused by colonization of the vaginal tampon with TSS toxin–secreting *Staphylococcus aureus*.

21. A woman developed a well-defined, slightly raised, and indurated erythema that was hot, shiny, and tender. Most likely this is an infection of the dermis and subcutis caused by



- A. *Staphylococcus epidermidis*
- B. *Staphylococcus saprophyticus*
- C. *Streptococcus agalactiae*
- D. *Streptococcus pyogenes*
- E. *Streptococcus pneumoniae*

ANS: D

This woman has erysipelas, an infection of the dermis and subcutis, which is most likely caused by group A *Streptococcus pyogenes*. Group B *S. agalactiae* is a commensal in the vagina and may cause infection of neonates, such as meningitis or septicemia. *S. epidermidis* often infects prostheses, shunts, and catheters. *S. saprophyticus* is a rare cause of postcoital urinary tract infection in women. *S. pneumoniae* is an important cause of pneumonia, otitis media, and meningitis.

22. Diarrhea associated with enlargement of Payer's patches in the small intestine, causing ulceration of the overlying mucosa, is most likely a manifestation of infection with
- A. *Shigella dysenteriae*
 - B. *Salmonella typhi*
 - C. *Entamoeba histolytica*
 - D. *Clostridium difficile*
 - E. *Vibrio cholerae*

ANS: B

Enlargement of the lymphoid follicles in the Payer's patches and ulceration of the overlying mucosa are found typically in infections caused by *Salmonella typhi*. *Shigella* infection causes hyperemia of the colonic mucosa and focal ulceration. Intestinal amebiasis presents in the form of deep ulcers that have laterally undermined borders and are described as "flask-like." *Clostridium difficile* is the most common cause of pseudomembranous colitis. *Vibrio cholerae* causes secretory diarrhea without gross or microscopic changes in the intestinal mucosa.

23. A 23-year-old woman was bitten by a tick during a camping trip in the Rocky Mountains. Upon return from the trip, she developed fever accompanied by migratory erythema. These symptoms subsided, but then she developed pains in both arms and legs, most prominently around the knee joints. The final diagnosis was made serologically. What is the most likely cause of this disease?
- A. *Borrelia burgdorferi*
 - B. *Brucella melitensis*
 - C. *Brucella suis*
 - D. *Brucella abortus*
 - E. *Mycobacterium marinum*

ANS: A

This woman most likely has Lyme disease, a tick-borne spirochetal febrile disease that manifests with erythema migrans, arthritis, and neuromuscular disturbances. *Brucella* infections are usually acquired from farm animals, such as cows, pigs, sheep, and goats. *Mycobacterium marinum* infection is acquired from contaminated water in aquaria or swimming pools.

24. A 49-year-old man died of complications of AIDS. At autopsy, the pathologist found numerous pulmonary infarcts. Histologically, the thrombi in the branches of the pulmonary artery were permeated with broad hyphae branching at an acute angle. These hyphae also extended into the vessel wall. This infection was most likely caused by
- A. *Aspergillus fumigatus*
 - B. *Candida albicans*
 - C. *Cryptococcus neoformans*

- D. *Histoplasma capsulatum*
- E. *Blastomyces dermatitidis*

ANS: A

Aspergillus fumigatus is an opportunistic deep fungal infection that affects immunosuppressed persons. The hyphae of the fungi penetrate into the vessel wall, causing thrombi and infarcts. Other fungi listed here do not form branching hyphae and do not invade the vessel wall followed by thrombi.

25. Juvenile papillomatosis was diagnosed in the larynx of a two-year-old child. This lesion is caused by
- A. herpes simplex virus
 - B. human papilloma virus (HPV)
 - C. hepadna virus
 - D. adenovirus
 - E. parvovirus

ANS: B

Juvenile papillomatosis of the larynx is caused by (HPV), types 6 and 11. These types of HPV cause genital warts in the lower female genital tract; the transmission to the infant usually occurs during the delivery. Other viruses listed here do not form papillomas (warts).