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FIRST EDITION

IMMUNOLOGY
Questions & Answers

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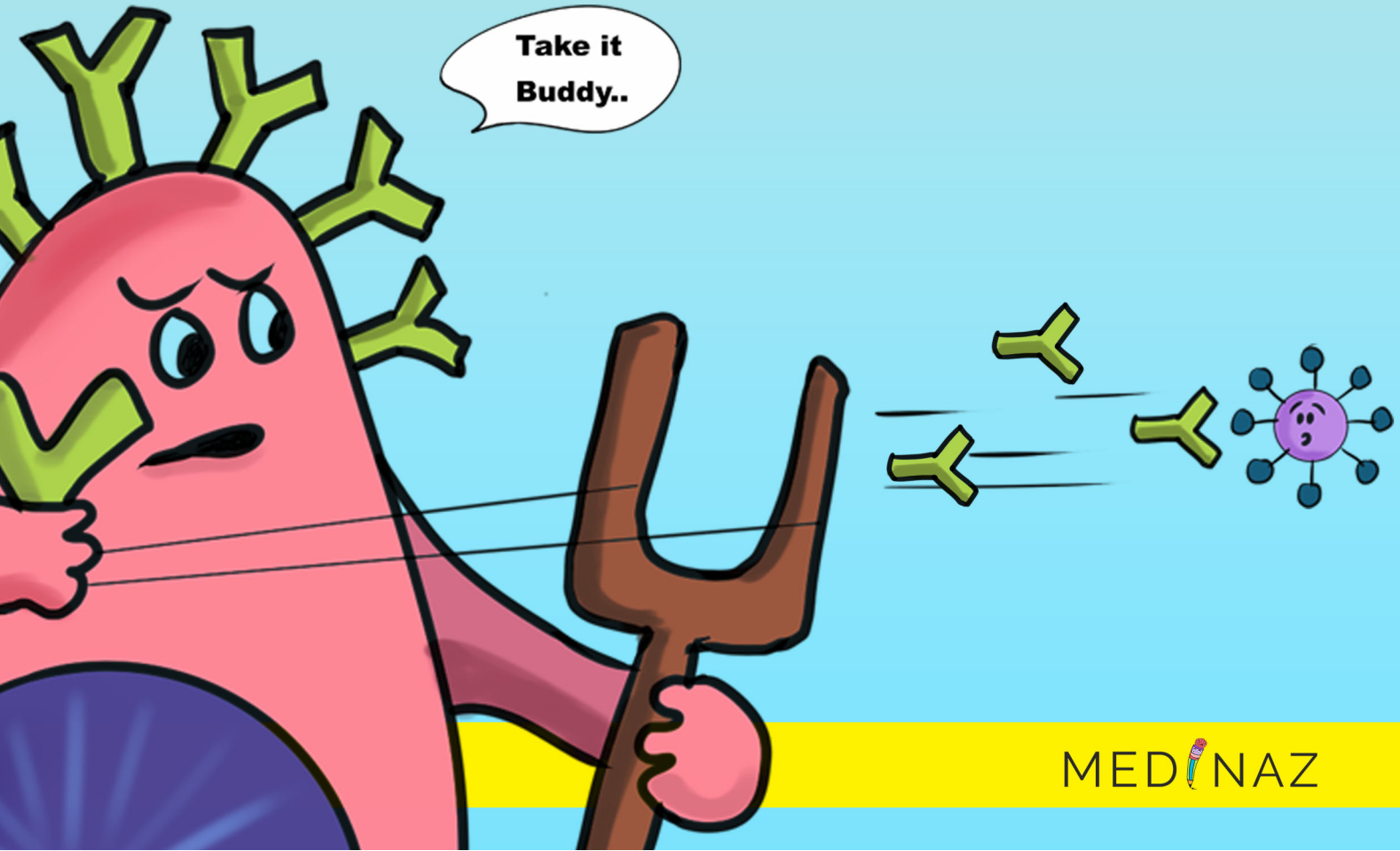
- **200+** Practice Questions & Answers
- **100** Conceptual MCQs with deatailed explanation



MEDINAZ

Immunology
Questions & Answers

Conceptual MCQs

1) A patient with rheumatoid arthritis presents to her physician and mentions that after many years without teeth problems, she has recently developed seven caries. This is a clue to her clinician that she should be evaluated for which of the following diseases?

A. Oral squamous cell carcinoma

B. Polyarteritis nodosa

C. Sjögren's syndrome

D. Systemic lupus erythematosus

E. Thyrotoxicosis

Explanation:

The correct answer is C. Rheumatoid arthritis can coexist with a variety of autoimmune diseases (including those listed in the answers), but is most frequently associated with Sjögren's syndrome.

Sjögren's syndrome is due to autoimmune involvement with subsequent scarring of the salivary and lacrimal glands, leading to dry eyes and dry mouth. Secondary effects include parotid gland enlargement, dental caries, and recurrent tracheobronchitis.

Squamous cell carcinoma of the mouth (choice A) is not associated with dryness of the mouth.

Polyarteritis nodosa (choice B) is a systemic necrotizing vasculitis. Patients present with low-grade fever,weakness, and weight loss. They may also have abdominal pain, hematuria, renal failure, hypertension, and leukocytosis.

Systemic lupus erythematosus (choice D) is an autoimmune disease characterized by vasculitis (which may produce a variety of symptoms depending on the site of the lesion), rash, renal disease, hemolytic anemia, and neurologic disturbances.

Thyrotoxicosis (choice E) produces insomnia, weight loss, tremors, heat intolerance, excessive sweating, and frequent bowel movements or diarrhea.

2) A 32-year-old, blood type A positive male receives a kidney transplant from a blood type B positive female donor with whom he had a 6-antigen HLA match. Once the kidney is anastomosed to the man's vasculature, the transplant team immediately begins to observe swelling and interstitial hemorrhage. After the surgery, the patient developed fever and leukocytosis and produced no urine. Which of the following is the most likely explanation?

A. Acute rejection due to antibody-mediated immunity

B. Acute rejection due to cell-mediated immunity

C. Chronic rejection due to cell-mediated immunity to minor HLA antigens

D. Hyperacute rejection due to lymphocyte and macrophage infiltration

E. Hyperacute rejection due to preformed ABO blood group antibodies

Explanation:

The correct answer is E. The patient is suffering from hyperacute rejection due to the preformed anti-B ABO blood group antibody found in all type A positive individuals. Hyperacute rejection occurs within minutes to a few hours of the time of transplantation, and is due to the destruction of the transplanted tissue by preformed antibodies reacting with antigens found on the transplanted tissue that activate complement and destroy the target tissue. Preformed antibodies can also be due to presensitization to a previous graft, blood transfusion, or pregnancy.

Acute rejection due to antibody-mediated immunity (choice A) is incorrect because this patient suffered from hyperacute rejection (immediate) occurring within minutes to hours, rather than days.

Acute rejection due to cell-mediated immunity (choice B) will not occur until several days or a week following transplantation. Acute rejection is due to type II and type IV reactions.

Chronic rejection, due to the presence of cell-mediated immunity to minor HLA antigens (choice C), occurs in allograft transplantation months to even years after the transplant. Chronic rejection is generally caused by both humoral and cell-mediated immunity.

An accelerated acute rejection, occurring in 3-5 days, can be caused by tissue infiltration and destruction by
presensitized T lymphocytes and macrophages (choice D) and/or antibody-dependent, cell-mediated
cytotoxicity (ADCC). Note that this is not a hyperacute reaction

3) tA 42-year-old auto mechanic has been diagnosed with end-stage renal disease. His identical twin brother has the same HLA alleles at all loci, and volunteers to donate a kidney to his brother. Which of the following terms correctly describes the proposed organ transplant?

A. Allograft
B. Autograft
C. Heterograft
D. Syngeneic graft
E. Xenograft

Explanation:

The correct answer is D. A syngeneic graft is the transfer of tissue between genetically identical individuals
(identical twins). This type of graft is usually successful.

An allograft (choice A), or homograft, is a graft between genetically different members of the same species.
This type of graft would be between two different humans and would most likely be rejected unless the recipient
is given immunosuppressive drugs.

An autograft (choice B) is a transfer of an individual's own tissue to his or her own body and is nearly always successful.

A heterograft (choice C) is the old term for a xenograft. This transfer of tissues between different species is not generally successful.

A xenograft (choice E) is a transfer of tissue between different species and is always rejected except for a few exceptions (e.g., pig heart valve).

4) Loss of which of the following classes of molecules on the surface of a tumor cell target would result in loss oftt susceptibility to killing by host immune cells?

A. CD3
B. CD4
C. CD8
D. MHC class I
E. MHC class II

Explanation:

The correct answer is D. After the MHC class I molecule has moved to the surface of the tumor cell, peptide fragments from the tumor are presented in a groove of the class I molecule. The peptide fragments are presented to cytotoxic CD8 T cells, which recognize the MHC class I molecules on the cell surface and kill the tumor cell. Loss of this molecule would therefore prevent the tumor cell from being killed.

The CD3 molecule (choice A) is a marker on all T cells. It is involved in signal transduction, but not antigen recognition. This molecule would not be on the surface of tumor cells.

The CD4 molecule (choice B) is not on the surface of a tumor cell, but it is on the surface of a CD4+ T helper lymphocyte.

The CD8 molecule (choice C) is not on the surface of a tumor cell, but it is on the surface of a CD8+ cytotoxic T lymphocyte.

MHC class II antigens (choice E) are not involved in killing of tumor cell targets. They present peptide fragments (derived from intracellular killing of extracellular organisms by macrophages) to CD4 T lymphocytes.

5) A 44-year-old white female presents with severe Raynaud's phenomenon, dysphagia, sclerodactyly, and facial and palmar telangiectasias. Which of the following autoantibodies is most likely to be present in this patient?

A. Anti-centromere antibody

B. Anti-histone antibody

C. Anti-SS-A

D. Anti-SS-B

E. Rheumatoid factor

Explanation:

The correct answer is A. The symptoms described in the question are classic for CREST syndrome. CREST syndrome is the combination of calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia. Anti-centromere antibodies are reported in a high percentage of patients with CREST, but in a small percentage of pure scleroderma patients.

Anti-histone antibody (choice B) is a marker for drug-induced lupus erythematosus.

Anti-SS-A (choice C) is a marker for Sjögren's syndrome, characterized by dry eyes and dry mouth.

Anti-SS-B (choice D) is an autoantibody directed against ribonucleoproteins, and is a marker for Sjögren's
syndrome.

Rheumatoid factor (choice E) is generally an IgM autoantibody directed against the Fc portion of IgG. RF is positive in 80% of patients with rheumatoid arthritis and may also be found in low titers in patients with chronic infections, other autoimmune diseases such as SLE and Sjögren's syndrome, or chronic pulmonary, hepatic, or renal diseases.

6) Which of the following is a major interleukin produced by CD4+ T helper 1 (TH1) lymphocytes?

A. IL-1

B. IL-2

C. IL-4

D. IL-6

E. IL-8

Explanation:

The correct answer is B. IL-2 produced by TH1 cells stimulates natural killer (NK) cells and CD8+ T cytotoxic lymphocytes and combines with IL-2 receptors on the TH1 cells to cause "self-stimulation". IL-2 is also involved in downregulating CD4+ T helper 2 lymphocytes.

IL-1 (choice A) is derived from macrophages and is a major inflammatory molecule.

IL-4 (choice C) is derived from CD4 + T helper 2 cells. It acts to downregulate CD4 + TH1 cells and allows class switching to IgE.

IL-6 (choice D) is derived from macrophages. This molecule stimulates acute phase protein production by the liver, as well as the production of other opsonizing molecules.

IL-8 (choice E) is derived from macrophages and is a major chemotactic molecule for neutrophils.

7) Maximal ventricular Na+ channel conductance occurs during which phase of the ECG?

A. P wave

B. QRS interval

C. ST interval

D. T wave

E. U wave

Explanation:

The correct answer is B. Phase 0 of the cardiac muscle action potential (AP) corresponds to the opening of voltage-dependent sodium channels, causing a transient but large increase in sodium conductance during ventricular depolarization. The shape of the QRS complex of the ECG is determined by the spread of the combined phase 0 (depolarization) of all the ventricular muscle of the heart.

The P wave (choice A) corresponds to atrial depolarization.

The ST interval (choice C) represents the time interval during which all ventricular cells are in phase 2 of their AP. Phase 2 is dominated by a high, prolonged calcium conductance through slow channels. The length of the ST interval corresponds closely to the AP duration in ventricular muscle.

The T wave (choice D) corresponds to ventricular repolarization.

The U wave (choice E) is found only occasionally in ECGs and is presumed to be due to the repolarization oft papillary muscle.

8) A 28-year-old male is brought into court for non-payment of child support. His ex-wife insists that he is the father

of her child, although he denies it. The court suggests before hearing the paternity case that various genetic

tests, including one for genetic immunoglobulin identification, be performed on the male, female, and child. Which

immunoglobulin marker would be helpful in this paternity case?

A. Allotypes

B. Idiotypes

C. IgA 2

D. IgM

E. Isotypes

Explanation:

The correct answer is A. There are genetic allotypic markers found on different immunoglobulin molecules. The best examples are kappa light chains and IgG 1, IgG 2, and IgG 3 heavy chains. These are distinctive markers that when present, can be helpful in paternity cases. If this male had these markers and the child had the same markers, then this would be presumptive evidence that the man was the father of the child. There would be other genetic tests performed in this case, because if the mother and father happened to have the same markers, then they could have come from the mother.

Idiotypes (choice B) are the immunoglobulins that have been produced in response to specific antigens (organisms) to which we have been exposed. There would probably very different idiotypes in the male, female, and child. Many of these would also be the same because of the different vaccines all would have received.

The IgA 2 (choice C) is a subclass of the immunoglobulin isotype class IgA. We all have this molecule in our serum, therefore it would not aid in paternity determination.

IgM (choice D) is one of the major isotypes of immunoglobulins that we all have in our serum. It is the first immunoglobulin formed in response to initial exposure to an organism. This is the immunoglobulin present in

the membranes of immature and mature B cells. IgM would not be helpful for determination of paternity in this case.

The isotypes (choice E) of immunoglobulin are the 5 major classes of immunoglobulin: IgG, IgA, IgM, IgD, and IgE. We all have these immunoglobulins in our serum and these would not be helpful for this paternity case.

9) Antigens processed by the exogenous antigen presentation pathway are presented in association with which of following?

A. Fc receptors

B. IgG heavy chains

C. MHC class I molecules

D. MHC class II molecules

E. T cell receptor (TCR)

Explanation:

The correct answer is D. When pathogenic organisms are phagocytized and degraded in the exogenous antigen presentation pathway, the antigenic molecules are presented on the surface of the antigen-presenting cell by MHC class II molecules to a CD4+ T lymphocyte with a specific TCR for the specific antigenic epitope.

The Fc (choice A) portion of an antibody molecule is the part of the immunoglobulin that attaches to the Fc receptors on phagocytic cell surfaces. When a Fab portion of the antibody is attached to the pathogen and the Fc attaches to the phagocytic cell surface, the phagocyte can destroy the pathogen more efficiently.

The IgG molecule (choice B) is an immunoglobulin that reacts with the antigen after it has been destroyed and presented to the T cell. The IgG immunoglobulin is never involved in antigen presentation.

In the endogenous antigen presentation pathway (eg, a virus infecting a cell), the cell would display epitopes from the virus in association with class I molecules (choice C) to the CD8+ cytotoxic T cell.

The TCR (choice E) is the area of the mature T cell that reacts with the antigen epitope that is presented by the antigen-presenting cell.

10) Loss of which of the following classes of molecules on the surface of a tumor cell target would result in loss of susceptibility to killing by host immune cells?

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C. CD8

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E. MHC class II

Explanation:

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The CD3 molecule (choice A) is a marker on all T cells. It is involved in signal transduction, but not antigen recognition. This molecule would not be on the surface of tumor cells.

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MHC class II antigens (choice E) are not involved in killing of tumor cell targets. They present peptide fragments (derived from intracellular killing of extracellular organisms by macrophages) to CD4 T lymphocytes.

11) A college student sitting in the stands at a football game suddenly begins breathing hard and complains to his friends of tightness in his chest. Minutes later, he is sweating profusely and faints. It is discovered that he had been stung by a bee. Paramedics arrive, assess the situation, then successfully treat the young man. Which one of the following drugs was most likely initially administered in this case?

A. Diphenhydramine

B. Blocking antibody

C. Cromolyn sodium

D. Epinephrine

E. Theophylline

Explanation:

The correct answer is D. This college student is experiencing a major anaphylactic reaction (type I hypersensitivity reaction) with associated bronchoconstriction and shock. Epinephrine is the treatment of choice for anaphylaxis. It will relax the smooth muscle of the respiratory tract and stimulate the heart to restore cardiac output. Epinephrine also prevents mast cell degranulation by increasing cyclic AMP levels.

Diphenhydramine (choice A), an H1 histamine receptor antagonist, is a good drug for mild allergic rhinitis, but would be ineffective in anaphylaxis.

Blocking antibody (choice B) is IgG antibody that has been produced by the patient in response to an allergen over a long period of stimulation (e.g., during desensitization by an allergist). This procedure is excellent for several different types of allergens, but not for a systemic material such as bee venom.

Cromolyn sodium (choice C) stabilizes mast cell membranes, thereby inhibiting degranulation and histamine release. This is an excellent drug if used to prevent an acute reaction to a known allergen, but it would not be used for an anaphylactic reaction.

Theophylline (choice E) inhibits phosphodiesterase, increasing cyclic AMP levels, and thereby inhibiting mast cell degranulation. This is an excellent drug for asthma and for long term allergy treatment especially inttt children, but would not be used for anaphylaxis.

12) A traveler to a foreign country develops acute lymphatic filariasis four months after his return to the United States. His symptoms include scrotal inflammation, itching, and localized scrotal swelling and tenderness of the inguinal lymph nodes. Which of the following immune mechanisms does the body employ against the live filarial worms ?

A. Antibody-dependent cell-mediated cytotoxicity

B. Anti-receptor antibodies

C. Arthus reaction

D. Complement-mediated reactions

E. Deposition of circulating antigen-antibody complexes

Explanation:

The correct answer is A. Filarial parasites have a complex life cycle beginning with transmission by mosquitoes or other arthropods. The offspring of adults (microfilariae) either circulate in the blood or migrate through the skin, often inhabiting lymphatics. Microfilariae are ingested by the arthropod vector and develop over 1 to 2
weeks into new infective larvae. The adult worms elicit an inflammatory reaction in the lymphatics, eventually leading to lymphatic obstruction and edema. The body reacts to large tissue parasites, such as filarial worms, by coating them with a thin layer of IgE molecules, which trigger eosinophil-mediated cytotoxicity (a form of Type II hypersensitivity) and release of vasoactive and spasmogenic substances from mast cells and basophils (local anaphylaxis, a form of Type I hypersensitivity).

Other forms of Type II hypersensitivity include complement-mediated reactions (choice D, e.g., the lysis of blood cells seen in transfusion reactions) and anti-receptor antibodies (choice B, e.g., muscle weakness in myasthenia gravis).

Type III hypersensitivities usually take the form of vasculitis secondary to deposition of circulating antigen-antibody complexes (choice E); the Arthus reaction (choice C) is a specific variant of these reactions in which local vasculitis induces tissue necrosis, often in the skin.

13) A 7-month-old child is hospitalized for a yeast infection that does not respond to therapy. The patient has a history of multiple, acute pyogenic infections. Physical examination reveals that the spleen and lymph nodes are not palpable. A differential WBC count shows 95% neutrophils, 1% lymphocytes, and 4% monocytes. A bone marrow biopsy contains no plasma cells or lymphocytes. A chest x-ray reveals the absence of a thymic shadow. Tonsils are absent. These findings are most consistent with

A. Bloom's syndrome

B. chronic granulomatous disease

C. severe combined immunodeficiency

D. Waldenström's macroglobulinemia

E. Wiskott-Aldrich syndrome

Explanation:

The correct answer is C. Severe combined immunodeficiency (SCID) is associated with deficiencies in both B and T cells due to a defect in differentiation of an early stem cell. Over 50% of the cases are caused by a gene defect on the X chromosome, resulting in a defective IL-2 receptor. The disease may exhibit a sex-linked or an autosomal recessive pattern of inheritance. The autosomal recessive variant is characterized by a deficiency of adenosine deaminase, which results in accumulation of metabolites that are toxic to both B and T stem cells in the bone marrow. Children usually die within the first 2 years of life with severe infections unless they receive bone marrow transplants.

Bloom's syndrome (choice A) is an autosomal recessive disorder included in the chromosomal instability group of syndromes. It is associated with small body size, immunodeficiency, light-sensitive facial erythema, and a major predisposition to cancer.

Chronic granulomatous disease (choice B) is caused by a deficiency of NADPH oxidase in neutrophils, resulting in loss of the first step of the myeloperoxidase system, and an absence of the respiratory burst. Patients are susceptible to staphylococcal infections and granulomatous infections.

Waldenström's macroglobulinemia (choice D) is a monoclonal gammopathy characterized by high serum levels of IgM and hyperviscosity complications.

Wiskott-Aldrich syndrome (choice E) is an immunodeficiency syndrome characterized by thrombocytopenia, eczema, and recurrent sinopulmonary infections. The patient has low levels of IgM and increased levels of IgG, IgA, and IgE.

14) A 27-year-old white male presents with a 3-week history of several swollen and painful toes and knees. He has a past history of conjunctivitis. He also describes some low back stiffness that is more severe in the morning. Which of the following is the most likely diagnosis?

A. Gout

B. Lyme disease

C. Reiter's syndrome

D. Rheumatoid arthritis

E. Septic arthritis

Explanation:

The correct answer is C. This is a case of Reiter's syndrome. Patients typically present with the acute onset of arthritis (usually asymmetric and additive), with involvement of new joints occurring over a period of a few days to 2 weeks. Joints of the lower extremities are the most commonly involved, but wrists and fingers can also be affected. Dactylitis (sausage digit), a diffuse swelling of a solitary finger or toe, is a distinctive feature of Reiter's arthritis and psoriatic arthritis. Tendonitis and fasciitis are common. Spinal pain and low back pain are common.

Conjunctivitis, urethritis, diarrhea, and skin lesions are also associated with Reiter's syndrome. Up to 75% of patients are HLA-B27 positive. Microorganisms which can trigger Reiter's syndrome include Shigella spp., Salmonella spp., Yersinia spp., Campylobacter jejuni, and Chlamydia trachomatis. Most patients are younger males.

Gout (choice A) usually presents as an explosive attack of acute, very painful, monarticular inflammatory arthritis. Hyperuricemia is the cardinal feature and prerequisite for gout. The first metatarsophalangeal joint is involved in over 50% of first attacks.

Lyme disease (choice B), caused by Borrelia burgdorferi, presents with a red macule or papule at the site of the tick bite. This lesion, called erythema chronicum migrans, slowly expands to form a large annular lesion with a red border and central clearing. The lesion is warm, but usually not painful. The patient also has severe headache, stiff neck, chills, arthralgias, and profound malaise and fatigue. Untreated infection is associated with development of arthritis. The large joints (e.g., knees) are usually involved with the arthritis lasting for weeks to months.

15) Rheumatoid arthritis (choice D) begins insidiously with fatigue, anorexia, generalized weakness, and vague musculoskeletal symptoms leading up to the appearance of synovitis. Pain in the affected joints, aggravated by movement, is the most common manifestation of established rheumatoid arthritis. Generalized stiffness is frequent and is usually greatest after periods of inactivity. Morning stiffness of greater than 1 hour in duration is very characteristic. Rheumatoid arthritis is more common in females. The metacarpophalangeal and proximal interphalangeal joints of the hands are characteristically involved.

Septic arthritis (choice E) is caused by a variety of microorganisms, including Neisseria gonorrhoeae and Staphylococcus aureus. Hematogenous spread is the most common route in all age groups. 90% of patients present with involvement of a single joint, usually the knee. The usual presentation is moderate-to-severe pain, effusion, muscle spasm, and decreased range of motion. Peripheral leukocytosis and a left shift are common. Disseminated gonococcal infections present as fever, chills, rash, and articular symptoms. Papules progressing to hemorrhagic pustules develop on the trunk and extensor surfaces of the distal extremities. Migratory arthritis and tenosynovitis of multiple joints is common.
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16) A researcher develops a specific antibody to the complement component C3b. Assume that intravenous administration of the antibody prevents the biological effects of C3b. Administration of the antibody would be expected to interfere with which of the following biological functions?

A. Decreased appetite

B. Fever

C. Increased collagen synthesis by fibroblasts

D. Increased leukocyte adherence to endothelium

E. Opsonization to facilitate phagocytosis

Explanation:

The correct answer is E. C3b acts as an opsonin, along with IgG, to facilitate phagocytosis. None of the other functions listed in the answer choices is attributable to C3b. Instead, all of the other answer choices are functions of the cytokines interleukin-1 (IL-1) and tumor necrosis factor (TNF). Tumor necrosis factor is produced by macrophages and activated T-cells, while many different cell types (including macrophages) produce IL-1.

Decreased appetite (choice A) is typical during an acute illness (such responses are called acute phase reactions). Other acute phase reactions include fever (choice B), increased sleep, shock, leukocytosis, and increased serum acute phase proteins.

Fibroblasts proliferate and increase their synthesis of collagen (choice C), collagenase, protease, and prostaglandin E in response to IL-1 and TNF.

Effects of IL-1 and TNF on endothelium include increased synthesis of mediators (prostaglandins, IL-1, IL- 8, platelet-derived growth factor), increased leukocyte adherence (choice D), and increased procoagulant activity with decreased anticoagulant activity.

17) A 22-year-old female comes to the sexually transmitted disease (STD) clinic for her first visit. She tells the nurse practitioner that she has had four different sexual partners in the last six months and only one of them used a condom. She also admits that she used IV drugs on several occasions two years ago. She notes fever, weight loss, lack of appetite, and periodic difficulty breathing over the past few months. She has an HIV test performed, which is positive. The physician decides to do a confirmatory test for HIV. Which one of the following tests would the physician order?

A. ELISA (Enzyme-linked immunosorbent assay)

B. FACS (Fluorescence activated cell sorting)

C. RAST (Radioallergosorbent Test)

D. RID (Radial immunodiffusion)

E. Western blot

Explanation:

The correct answer is E. The Western blot is the most appropriate test for confirmation of HIV infection. It identifies several different antibodies against HIV (anti- gp120, -gp41, -p24, and -p17).

The initial HIV test this patient had was most likely an ELISA. The ELISA (choice A) can be used to detect p24 antibody in the patient, but is not as specific as the Western blot.

Fluorescence activated cell sorting (choice B) is a technique used to separate and count specific numbers and types of cells in a sample. An example of this would be to count the number of B cells and T cells in a specific blood sample.

RAST testing (choice C) is used to determine the level of specific IgE present in a patient that reacts with a specific allergen that has been applied to a disk or glass bead.

Radial immunodiffusion (choice D) is an excellent test used for quantitation of immunoglobulin levels in patients. This is used to determine the IgG, IgM and IgA levels in patient's serum. This test cannot be used to measure levels of IgD or IgE because these two immunoglobulins are at such low levels a more sensitive test such as RIA
(radioimmunoassay) or EIA (enzyme-linked immunoassay) must be used.

18) A 38-year-old woman complains of cold, painful fingertips, as well as difficulty swallowing and indigestion. Physical examination is remarkable for a thickened, shiny epidermis over the entire body, with restriction of movement of the extremities, particularly the fingers, which appear claw-like. Which of the following autoantibodies is likely to be found in this patient's serum?

A. Anti-DNA topoisomerase I (anti-Scl-70)

B. Anti-double-stranded DNA (ds DNA)

C. Anti-IgG

D. Anti-Sm

E. Anti-SS-A

Explanation:

The correct answer is A. This patient is suffering from systemic sclerosis, also called scleroderma. Antibodies to topoisomerase I (anti-Scl-70) occur in up to 70% of patients with diffuse systemic sclerosis, but only rarely in other disorders. Systemic sclerosis is characterized initially by excessive fibrosis and edema of the skin,

especially the hands and fingers, producing sclerodactyly (characteristic changes in the fingers, which resemble claws). Raynaud's phenomenon is common. The diffuse type of systemic sclerosis generally spreads to include visceral organs such as the esophagus (producing dysphagia), the lungs (producing pulmonary fibrosis), the heart (leading to heart failure or arrhythmia), and the kidneys (renal failure causes 50% of scleroderma deaths).

Females are affected more than males (3:1 ratio). A more restricted variant of systemic sclerosis with a somewhat more benign course is CREST syndrome (Calcinosis, Raynaud's syndrome, Esophageal dysmotility, Sclerodactyly, and Telangiectasia), characterized by the presence of anti-centromere antibodies (although 10% of CREST patients will have anti-topoisomerase antibody as well).

Anti-ds DNA (choice B) is characteristic of systemic lupus erythematosus, but is not common in patients with systemic sclerosis.

Rheumatoid factor is an autoantibody directed against IgG (choice C). It is found in patients with rheumatoid arthritis.

Anti-Sm (Smith antigen; choice D) is also characteristic of SLE rather than systemic sclerosis.

Anti-SS-A (choice E) is typically seen in Sjögren's syndrome (although it may also be seen in SLE).

19) A 4-year-old boy presents to the emergency room with muscle spasms. His past medical history is significant for recurrent infections and neonatal seizures. Evaluation of his serum electrolytes reveals hypocalcemia. This patient would be most susceptible to which of the following diseases?

A. Chickenpox

B. Diphtheria

C. Gas gangrene

D. Gonorrhea

E. Tetanus

Explanation:

The correct answer is A. This boy has DiGeorge's syndrome, as evidenced by his tetany (muscle spasms) due to hypocalcemia and his history of recurrent infections and neonatal seizures. The syndrome occurs because of an embryonic failure in the development of the third and fourth pharyngeal pouches. Patients have both hypoplastic parathyroids (producing hypocalcemia) and thymuses (producing T-cell deficiency and recurrent infections). Since cell-mediated immunity (which depends on T cells) is important in defense against infections caused by intracellular pathogens (such as viruses), patients with DiGeorge's are particularly susceptible to viral infections, such as chickenpox (varicella). They also have trouble with fungal pathogens (e.g., Candida) and mycobacteria.

Note that you should be able to recognize other clues to the diagnosis, including: congenital cardiac defects, esophageal atresia, bifid uvula, short philtrum, hypertelorism, antimongoloid palpebral slant, mandibular hypoplasia, and low-set ears.

Diphtheria (choice B) is caused by Corynebacterium diphtheriae, which produces disease by the elaboration of a very potent exotoxin. Therefore, humoral immunity (antitoxin), which is not usually compromised in DiGeorge's patients, is essential for defense against the organism. (Note that the C. diphtheriae exotoxin acts by causing the ADP-ribosylation of elongation factor-2 of eukaryotic cells, thereby inhibiting protein synthesis). The disease can be avoided by immunization with diphtheria toxoid.

Gas gangrene (choice C) is caused by Clostridium perfringens, which produces a potent alpha toxin that injures cell membranes. Therefore, humoral immunity would again play a predominant role in defense against this organism. Note that the disease occurs in wounds and would not be expected in an uninjured 4-year-old boy.

Gonorrhea (choice D) is caused by Neisseria gonorrhoeae and would not be expected in a 4-year-old boy unless there was evidence of sexual abuse. Virulence factors of this organism include pili, cell wall endotoxin and outer membrane, and secretory IgA protease. Antibody responses, neutrophils, and complement are of prime importance in defense against gonococcal infections.

Tetanus (choice E) is caused by Clostridium tetani and serves as a tricky distracter, as you might have quickly associated the patient's muscle spasms with this answer choice. (This is why it is important to read the question stem carefully before prematurely jumping to the responses). C. tetani, which gains entry through deept wounds, produces tetanus toxin (exotoxin) and can be prevented by immunization with tetanus toxoid.

20) A 30-year-old woman presents to a physician with a prominent rash over her nose and cheeks. She also has complaints of fever, malaise, and muscle soreness of several months duration. Serologic studies demonstrate positive ANA with autoantibodies to double-stranded DNA. This patient's probable condition is associated with which of the following HLA type(s)?

A. HLA-A3

B. HLA-B27

C. HLA-DR2 and HLA-DR3

D. HLA-DR3 and HLA-DR4

E. HLA-DR4

Explanation:

The correct answer is C. The disease is systemic lupus erythematosus, which is an autoimmune disorder associated with HLA-DR2 and HLA-DR3. The presentation described in the question stem is classic; patients without the characteristic malar or "butterfly" rash are much harder to diagnose because their complaints are initially typically very vague.

Associate HLA-A3 (choice A) with primary hemochromatosis.

Associate HLA-B27 (choice B) with psoriasis, ankylosing spondylitis, inflammatory bowel disease, and Reiter's syndrome.

Type I diabetes is associated with both HLA-DR3 and HLA-DR4 (choice D).

Rheumatoid arthritis is associated with HLA-DR4 (choice E).

21) A 47-year-old male presents with declining renal function characterized by oliguria, elevated blood urea nitrogen and creatinine, and hematuria. He also complains of nasal congestion and epistaxis. Review of systems is notable for occasional cough and hemoptysis. Examination shows mucosal ulceration and nasal septal perforation, but no polyps. Which of the following serum markers would likely be present in this case?

A. Anti-centromere antibody

B. Anti-Ro

C. Anti-SS-B

D. c-ANCA (cytoplasmic antinuclear cytoplasmic antibody)

E. Decreased erythrocyte sedimentation rate (ESR)

Explanation:

The correct answer is D. This patient has Wegener's granulomatosis, which is characterized by renal involvement, severe upper respiratory tract symptoms, and pulmonary involvement. Other organ systems may also be involved. The renal syndrome is a crescentic rapidly progressive glomerulonephritis leading to renal failure. The upper respiratory tract findings include sinus pain and drainage, and purulent or bloody nasal discharge with or without nasal ulcerations. Nasal septal perforation may follow. Pulmonary involvement may be clinically silent with only infiltrates present on x-ray, or it may present as cough and hemoptysis. c-ANCA is a marker for Wegener's granulomatosis, present in a high percentage of patients.

Anti-centromere antibody (choice A) is associated with approximately 90% of cases with CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal motility syndrome, sclerodactyly, and telangiectasia) which is also called limited scleroderma.

Anti-Ro (choice B) is also called anti-SS-A and is associated with Sjögren's syndrome (70-95%).

Anti-SS-B (choice C) is associated with Sjögren's syndrome (60-90%).

Decreased ESR (choice E) is not a marker of Wegener's. Instead, a markedly elevated ESR is seen. Additionally, mild anemia, thrombocytosis, leukocytosis, mild hypergammaglobulinemia (IgA), and mildly elevated rheumatoid factor are seen in this disorder.

22) A 57-year-old woman with a history of hypertension and arthritis is referred to a rheumatologist for evaluation. A complete blood count (CBC) is normal, and a mini-chem panel shows no electrolyte abnormalities. Her erythrocyte sedimentation rate (ESR) is elevated, and an antinuclear antibody test (ANA) is positive. Further antibody studies are performed, and the results are shown below.

Anti-histones	high titer
Anti-double stranded DNA	not detected
Anti-single stranded DNA	not detected
Anti-SSA	not detected
Anti-SSB	not detected
Anti-SCI-70	not detected
Anti-Smith	not detected
Anit-centromere	not detected
Anti-RNP	not detected

Which of the following diseases is suggested by these results?

A. CREST syndrome

B. Diffuse form of scleroderma

C. Drug-induced lupus

D. Sjögren's syndrome

E. Systemic lupus erythematosus

Explanation:

The correct answer is C. The single finding of high autoantibody titers to histones, without any other autoantibodies, is characteristic of drug-induced lupus. The most commonly implicated drugs are procainamide, hydralazine (given for hypertension), and isoniazid. Patients typically have milder disease than in SLE, and tend to have arthritis, pleuro-pericardial involvement, and, less commonly, rash. CNS and renal disease are not usually observed.

CREST syndrome (choice A) is a milder variant of scleroderma characterized by calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and telangiectasia. Anti-centromere antibodies are diagnostic.

The diffuse form of scleroderma (choice B), also known as systemic sclerosis, causes fibrosis of the skin and internal viscera. This disorder is characterized by anti-SCI-70 and often low titers of many other autoantibodies.

Sjögren's syndrome (choice D) is characterized by dry eyes and dry mouth. Sjögren's syndrome in isolation is characteristically positive for anti-SSA and anti-SSB. If it accompanies rheumatoid arthritis, anti-RNP will be positive as well.

Systemic lupus erythematosus (choice E) is a multisystem disorder that is distinguished from drug-induced lupus by the presence of a wide variety of autoantibodies, including anti-double stranded DNA (anti-dsDNA).

23) A 24-year-old woman with a history of allergic rhinitis is involved in an automobile accident and sustains a splenic laceration. She undergoes abdominal surgery and is then transfused with four units of blood of the appropriate ABO and Rh type. As the transfusion progresses, she becomes rapidly hypotensive and develops airway edema, consistent with anaphylaxis. Which of the following pre-existing conditions best accounts for these symptoms?

A. AIDS

B. C1 esterase inhibitor deficiency

C. DiGeorge syndrome

D. Selective IgA deficiency

E. Wiskott-Aldrich syndrome

Explanation:

The correct answer is D. Patients with selective IgA deficiency may have circulating antibodies to IgA. Fatal anaphylaxis may ensue if they are transfused with blood products with serum containing IgA, although many patients with selective IgA deficiency are asymptomatic and never diagnosed. Symptomatic patients may have recurrent sinopulmonary infections and diarrhea, and also have an increased incidence of autoimmune and allergic diseases.

AIDS (choice A) predisposes for infections and neoplasms, but not anaphylaxis.

C1 esterase inhibitor deficiency (choice B) is an autosomal dominant disease characterized by recurrent attacks of colic and episodes of laryngeal edema, without pruritus or urticarial lesions. This disorder is also known as hereditary angioedema.

DiGeorge syndrome (choice C) is characterized by thymic aplasia and sometimes, hypoparathyroidism. The disorder is due to abnormal development of the third and fourth pharyngeal pouches.

Wiskott-Aldrich syndrome (choice E) is a form of immunodeficiency associated with thrombocytopenia and eczema.

24) An 8-month-old boy baby is evaluated because of repeated episodes of pneumococcal pneumonia. Serum studies demonstrate very low levels of IgM, IgG, and IgA. This patient's condition is thought to be related to a deficiency of which of the following proteins?

A. Adenosine deaminase

B. Class III MHC gene

C. Gamma chain of the IL-2 receptor

D. Purine nucleotide phosphorylase

E. Tyrosine kinase

Explanation:

The correct answer is E. The patient in the question stem has X-linked (Bruton's) agammaglobulinemia, which is now thought to be due to a deficiency in a tyrosine kinase, leading to a B cell maturation arrest at the pre-B cell level.

Selective IgA deficiency has been linked to defective class III MHC genes (choice B).

Severe combined immunodeficiency is apparently a heterogeneous disease, and different subgroups have been linked to abnormalities of adenosine deaminase (choice A), the gamma chain of the IL-2 receptor (choice C), and purine nucleotide phosphorylase (choice D).

25) An 18-year-old high school senior presents to her doctor with tender lymph nodes in her neck on the left side. She has no significant past medical history. Two weeks ago, she updated her vaccines in preparation for college. A lymph node biopsy is performed, which shows benign paracortical expansion and scattered multinucleated giant cells with eosinophilic cytoplasmic and nuclear inclusion bodies. Which of the following vaccines is most likely responsible for this woman's lymphadenitis?

A. Hepatitis B

B. Measles

C. Rubella

D. Smallpox

E. Tetanus

Explanation:

The correct answer is B. The Warthin-Finkeldey (WF) giant cell described above is pathognomonic for measles or the live attenuated measles vaccine. Most giant cells are composed of histiocytes, but the WF giant cell is created by fusion of lymphocytes. Although postvaccinal lymphadenitis may be seen with different vaccines, the usual reaction is immunoblastic proliferation within the paracortical regions of a hyperplastic lymph node.

Hepatitis B (choice A), rubella (choice C), and tetanus (choice E) are rarely associated with postvaccinial lymphadenitis.

Smallpox (choice D) is classically followed by tender regional adenopathy, one to several weeks following immunization.
There are no associated giant cells.

26) A 21-year-old college student from Connecticut with a past history of Lyme disease presents with chronic pain and swelling in his right knee. He states that he has had problems with the knee for the past two years. Which of the following HLA alleles would you expect to be present in this individual?

A. HLA-B9

B. HLA-B17

C. HLA-B27

D. HLA-DR3

E. HLA-DR4

Explanation:

The correct answer is E. Approximately 60% of patients in the United States who contract Lyme disease, but are not treated with antibiotics, will develop frank arthritis. The pattern typically consists of intermittent attacks of oligoarticular arthritis in large joints (especially knees) lasting for weeks to months in a given joint. Patients
with persistent arthritis have a higher frequently of HLA-DR4 class II MHC complex than patients with brief Lymet arthritis or normal controls.

HLA-B9 (choice A), and HLA-B17 (choice B) are not thought to be associated with Lyme disease arthritis.

HLA-DR3 (choice D) is associated with a variety of disorders, but not Lyme disease arthritis.

HLA-B27 (choice C), although associated with reactive arthritis, is not associated with Lyme disease arthritis.

27) A 7-year-old girl is walking across a vacant lot and steps on a nail. The next day, her foot is sore and the wound appears inflamed. During these early stages of infection, which of the following compounds exert the most powerful chemotactic effect on neutrophils, causing them to migrate into the inflamed area?

A. C5a and IL-8

B. IL-1 and tumor necrosis factor

C. LTC4 and LTD4

D. PGI2 and PGD2

E. Thromboxane and platelet activating factor

Explanation:

The correct answer is A. The most important chemotactic factors for neutrophils are the complement factor C5a and the interleukin IL-8.

The cytokines IL-1 and tumor necrosis factor (choice B) have complex, similar actions, including stimulation of production of many acute-phase reactions, stimulation of fibroblasts, and stimulation of endothelium.

Leukotrienes LTC4 and LTD4(choice C) cause increased vascular permeability.

Prostaglandins PGI2 and PGD2(choice D) mediate vasodilation and pain.

Thromboxane and platelet activating factor (choice E) induce platelet changes.

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28) One year after orthotopic liver transplantation for hepatitis C and cirrhosis, a 53-year-old man develops rising transaminase and bilirubin levels. In order to minimize chronic rejection injury to hepatic endothelial cells, immunosuppressive therapy is aimed at down-regulating which of the following components of the immune response?

A. Autoantibody production

B. Complement protein synthesis

C. HLA antigen expression

D. Mast cell degranulation

E. T-lymphocyte activity

Explanation:

The correct answer is E. Chronic rejection of any solid organ entails cellular injury to endothelial cells, resulting in intimal proliferation, fibrosis, and eventually ischemic injury to the graft. Immunosuppressive therapy is directed at controlling lymphocyte activity and minimizing cellular rejection.

Autoantibodies (choice A) are not involved in organ transplant rejection. The antibodies produced are alloantibodies directed only to the graft, but not to the host.

Complement proteins (choice B) are involved in the humoral component of acute rejection, and complement binding to alloantibodies increases graft damage. Complement protein production, however, is not affected by immunosuppressive therapy.

HLA antigen expression (choice C) is central to recognition of foreign cells in grafted tissue. HLA antigens are expressed constitutively by all normal cells, and immunosuppression does not affect their production.

Mast cell degranulation (choice D) is a component of the anaphylactic response (Type I hypersensitivity). Graft rejection is a Type IV hypersensitivity response, and does not involve mast cell degranulation.

29) A 40-year-old woman with systemic sclerosis presents to her physician with malaise. Physical examination is remarkable for jaundice, and serum chemistry studies demonstrate moderately elevated serum alkaline phosphatase, while aspartate aminotransferase (AST) and alanine aminotransferase (ALT) are only minimally elevated. Presence of which of the following autoantibodies would be most helpful in elucidating the likely etiology of the woman's liver disease?

A. Anti-double-stranded DNA

B. Anti-mitochondrial antibodies

C. Anti-phospholipid antibodies

D. Anti-self IgG

E. Anti-smooth muscle antibodies

Explanation:

The correct answer is B. Primary biliary cirrhosis is strongly associated with systemic sclerosis (scleroderma). This disorder characteristically affects intrahepatic bile ducts more than the hepatic parenchyma, at least in the earlier stages, and consequently causes a disproportionate increase in serum alkaline phosphatase compared to AST and ALT. The most distinctive markers for primary biliary cirrhosis are anti-mitochondrial antibodies, especially the M2 subtype. Primary biliary cirrhosis is also associated with a variety of other diseases, such as Sjögren's syndrome, rheumatoid arthritis, thyroiditis, celiac disease, and glomerulonephritis.

Anti-double-stranded DNA (choice A) and anti-phospholipid antibodies (choice C) are markers for systemic lupus erythematosus.

Anti-self IgG (choice D; also known as rheumatoid factor) is a marker for rheumatoid arthritis.

Anti-smooth muscle antibodies (choice E) are seen in autoimmune hepatitis, which is not as strongly associated with scleroderma as primary biliary cirrhosis.

30) Which of the following events occurs first in the differentiation sequence of human B cells in the bone marrow?

A. Cytoplasmic mu chains present in the B cell

B. Immunoglobulin heavy chain rearrangement

C. Immunoglobulin light chain rearrangement

D. Surface IgD and IgM present on the B cell

E. Surface IgM present on the B cell

Explanation:

The correct answer is B. The first event that occurs in the pre-B cell (progenitor) is the gene rearrangement of the heavy chain. The D gene and J gene recombination event occurs first, followed by V recombination with the D-J region.

The cytoplasmic mu chains (choice A) are the result of immunoglobulin heavy chain rearrangement, the production of the messenger RNA from this rearrangement, and the ribosomal synthesis of the protein mu chain. This is the second event that occurs in the B-cell maturational sequence.

Light chain rearrangement (choice C) occurs when recombination events occur with the V gene and J gene from the light chain germ line. After this recombination has occurred, and the messenger RNA for this germ line has produced the light chain protein, the light chains and heavy chains form.

The complete IgM molecule and IgD molecules (choice D) are present on the surface of only the mature B cells.This event is the last event to occur during the differentiation and development of B cells in the bone marrow.

The complete IgM molecule (choice E) is present on the surface of the immature B cell. This event is one of thet last events to occur in the differentiation and development of B cells.

31) A 4-year-old boy is seen by his pediatrician for epistaxis. The patient has a history of multiple bacterial and viral respiratory tract infections and eczema. An uncle had similar problems. Physical examination is remarkable for multiple petechial lesions on the skin and mucous membranes. Serum IgE is increased, and platelets are decreased. Which of the following is the most likely diagnosis?

A. Acquired hypogammaglobulinemia

B. Ataxia telangiectasia

C. DiGeorge syndrome

D. Selective IgA deficiency

E. Wiskott-Aldrich syndrome

Explanation:

The correct answer is E. Wiskott-Aldrich syndrome is an X-linked condition characterized by eczema, thrombocytopenia, and repeated infections. Affected children may present with bleeding and often succumb to complications of bleeding, infection, or lymphoreticular malignancy. The platelets are small, have a shortened half-life, and appear to be deficient in surface sialophorin (CD43). Splenectomy can correct the thrombocytopenia, but not the immune defect. Serum IgM is usually decreased, while IgE is frequently increased. Mutations in the Wiskott-Aldrich serum protein (WASP) gene on the short arm of the X chromosome are responsible for this disease.

Acquired hypogammaglobulinemia (choice A) is a disease of adults characterized by normal numbers of B cells but low immunoglobulin production.

Ataxia telangiectasia (choice B) is an autosomal recessive disease characterized by progressive cerebellar dysfunction, telangiectasias, and a variable immunodeficiency.

DiGeorge syndrome (choice C) is a developmental malformation leading to thymic aplasia and, sometimes, hypoparathyroidism.

Selective IgA deficiency (choice D) is a relatively common condition characterized by low levels of IgA.

32) A 34-year-old woman presents with fatigue, malaise, and swollen, tender joints. Physical examination is significant for a maculopapular eruption over sun-exposed areas, including the face. Examination of a peripheral blood smear reveals mild thrombocytopenia. Which of the following autoantibodies, if present, would be most specific for the diagnosis of the patient's disorder?

A. Anti-centromere antibody

B. Anti-IgG antibody

C. Antinuclear antibody

D. Anti-Sm (Smith antigen) antibody

E. Anti-SS-A (Ro) antibody

Explanation:

The correct answer is D. The patient described probably has systemic lupus erythematosus (SLE). This is a systemic disorder that often presents with fatigue, malaise, fever, gastrointestinal symptoms, arthralgias, and myalgias. Hematologic abnormalities include anemia of chronic disease, hemolytic anemia, leukopenia, lymphocytopenia, and thrombocytopenia. A circulating anticoagulant may prolong the APTT (activated partial thromboplastin time). Cutaneous manifestations include a malar rash and a generalized maculopapular eruption, both of which are photosensitive. Antibodies to the Smith antigen (core proteins of small ribonucleoproteins found in the nucleus) are present in only 20-30% of patients with SLE, but are quite specific for the disease, occurring only rarely in other autoimmune diseases.

Anti-centromere antibody (choice A) is specific for the CREST (Calcinosis, Raynaud's syndrome, Esophageal dysfunction, Sclerodactyly, and Telangiectasia) variant of progressive systemic sclerosis (scleroderma).

Rheumatoid factor is actually an autoantibody directed against the Fc portion of the IgG molecule (choice B). It is found in more than two-thirds of patients with rheumatoid arthritis.

The majority of patients with SLE (around 95%) develop antinuclear antibodies (ANA; choice C), so this test is quite sensitive, but not very specific for SLE. ANA occur in other inflammatory disorders, autoimmune diseases, viral diseases, and in a number of normal individuals. Antibodies to double-stranded DNA are more specific for SLE, but are not included as an answer choice.

Anti-SS-A antigen (choice E) refers to antibodies to certain ribonucleoproteins, which are fairly specific for Sjögren's syndrome.

33) A four-year-old boy is brought to the pediatrician because of several "boils" on his arm. His mother tells the physician that the boy has had similar lesions on several previous occasions that were treated successfully with antibiotics. She denies any history of eczema or typical childhood illnesses such as measles or chicken pox. The child has had all of his immunizations. Laboratory examination reveals a normal complete blood count, immunoglobulin levels, B cell and T cell counts, and complement levels. Serum calcium and parathyroid hormone levels are also normal. The nitroblue tetrazolium test is negative. Which of the following diagnoses is most consistent with these data?

A. Bruton's agammaglobulinemia

B. Chronic granulomatous disease

C. DiGeorge syndrome

D. SCID (severe combined immunodeficiency disease)

E. Wiskott-Aldrich syndrome

Explanation:

The correct answer is B. The fact that the boy had several different infections with pyogenic bacteria requiring antibiotics suggests an inability of phagocytes to kill bacteria. The nitroblue tetrazolium test (NBT) for reactive oxygen intermediates was negative, indicating that the boy suffers from chronic granulomatous disease (CGD).

CGD is most often due to a defect in NADPH oxidase, which is necessary for leukocyte hydrogen peroxide production.

In Bruton's agammaglobulinemia (choice A), patients have very low levels of circulating immunoglobulins. There is a virtual absence of B cells, but pre-B cells are present. These patients also experience frequent pyogenic bacterial infections, but the boy in the question had normal immunoglobulin levels and normal B cell counts.

DiGeorge syndrome (choice C) results when the 3rd and 4th pharyngeal pouches fail to develop in normal fashion and the individual is missing the thymus gland and parathyroid glands. Hypocalcemia, low parathyroid levels and a T cell abnormality are typical.

In SCID (severe combined immunodeficiency disease) (choice D) both B cells and T cells may be absent, or if present, may not function properly. In this patient, the B and T cell counts were normal. The immunoglobulin levels were normal, so the B cells were functioning, and the patient was successfully immunized, so his T cells were functioning.

Wiskott-Aldrich syndrome (choice E), an immune deficiency disease that develops in the first year of life, is characterized by pyogenic infections, eczema, and thrombocytopenia.

34) A 45-year-old man presents to a physician with complaints of double vision and ptosis. The patient has noticed that these problems are minor in the early morning, but become progressively more severe during the course of the day. Symptoms markedly improve after a test dose of edrophonium. This condition is usually related to autoantibodies directed against which of the following?

A. Acetylcholine receptor

B. Double-stranded DNA

C. Neutrophil cytoplasmic proteins

D. SS-A (Ro)

E. TSH receptor

Explanation:

The correct answer is A. The disease is myasthenia gravis, which is the result of autoantibodies directed against the acetylcholine receptor at the neuromuscular junction. Many patients with myasthenia gravis have related thymic hyperplasia or thymoma.

Antibodies to double-stranded DNA (choice B) are a feature of systemic lupus erythematosus, a collagen-vascular disease.

Antibodies to neutrophil cytoplasmic proteins (choice C), called antineutrophil cytoplasmic autoantibodies (ANCA), are a feature of Wegener's granulomatosis.

Antibodies to SS-A (choice D) are a feature of Sjögren's syndrome, characterized by autoimmune inflammation of the lacrimal and salivary glands

Antibodies to the TSH receptor (choice E) are a feature of Graves' disease. Stimulation of the receptor by the autoantibodies is responsible for producing a hyperthyroid state.

35) A child stepped on a piece of glass 1 day ago, and an active inflammatory reaction is occurring in her wound, with large numbers of neutrophils attracted to the inflammation site. Which of the following is the major chemotactic factor responsible for attracting neutrophils?

A. C3b

B. C5a

C. IgM

D. IL-2

E. Lysozyme

Explanation:

The correct answer is B. In active inflammation, the complement system has been activated and C5a is being produced. C5a is a strong chemoattractant to neutrophils and other phagocytic cells.

C3b (choice A) is an excellent opsonin of pathogenic organisms; when an organism is coated with C3b, it is more easily phagocytized. C3b is formed via the classic and alternative complement pathways.

IgM (choice C) is the first immunoglobulin produced in the primary immune response. IgM cannot cross the placenta, but it is a powerful activator of complement; elevated levels in the newborn are associated with an acute infection with a pathogen.

IL-2 (choice D) is a powerful interleukin that stimulates T helper 1 cells. It also stimulates natural killer cells and T cytotoxic CD8 lymphocytes, but is not chemotactic for neutrophils.

Lysozyme (choice E) is a material present in tears, mucous, vaginal secretions, and other body fluids. It is active against the peptidoglycan of bacterial cell walls, splitting the backbone structure of the peptidoglycan (N-acetylglucosamine and N-acetyl muramic acid polymers).

36) A 65-year-old woman is evaluated for symmetrical swelling of the proximal phalangeal joints. Physical examination also reveals large subcutaneous nodules over the extensor surfaces of both arms. Autoantibodies directed against which of the following antigens would most likely be demonstrated by serum studies?

A. Acetylcholine receptor

B. Double stranded DNA

C. Histones

D. IgG

E. Ribonucleoprotein

Explanation:

The correct answer is D. The disease is rheumatoid arthritis, and the autoantibody is rheumatoid factor, whicht is usually an IgM or IgG (or less commonly IgA) directed against the constant region of autologous IgG.

Autoantibody directed against acetylcholine receptor (choice A) is a feature of myasthenia gravis.

Autoantibody directed against double stranded DNA (choice B) is a feature of systemic lupus erythematosus.

Autoantibody directed against histones (choice C) is a feature of drug-induced lupus.

Autoantibody directed against ribonucleoprotein (choice E) is a feature of mixed connective tissue disease.

37) Which of the following is the most important costimulatory signal provided to a T cell from an antigen-presenting cell?

A. B7 molecules interacting with CD 28

B. B7 molecules interacting with LFA- 1

C. ICAM-I interacting with LFA-1

D. LFA-3 interacting with CD 28

E. MHC class II interacting with T cell receptor

Explanation:

The correct answer is A. The B7 molecule on the cell surface of the antigen-presenting cell reacts with the CD 28 molecule on the T cell surface for maximal costimulatory signals.

The B7 molecule on the surface of the antigen presenting cell reacts only with CD 28 and does not react with LFA-1 (choice B) adhesion molecule.

The ICAM- I on the surface of an antigen presenting cell reacts with the LFA-I (choice C) on the surface of a T cell for the purpose of cell-to-cell adhesion and does not function for costimulation.

The LFA-3 (CD58) is an adhesion molecule on the surface of an antigen presenting cell. It does not react with a CD28 (choice D) costimulatory molecule on the T cell surface.

The MHC class II molecule with its epitope does interact with a specific T cell receptor (TCR) (choice E), but this is not termed costimulatory. However, the interaction does stimulate the T cell to produce interleukins for further cell division.

38) Cytotoxic T cells induced by infection with virus A will kill target cells

A. from the same host infected with any virus

B. infected by virus A and identical at class I MHC loci to the cytotoxic T cells

C. infected by virus A and identical at class II MHC loci to the cytotoxic T cells

D. infected with any virus and identical at class I MHC loci to the cytotoxic cells

E. infected with any virus and identical at class II MHC loci to the cytotoxic cells

Explanation:

The correct answer is B. The CD8+ cytotoxic T cells have antigen specific T-cell receptors (TCR) on their membranes that will recognize and bind to self class I antigens. Since the viral peptides are presented as a complex with the self class I antigens, the CD8+ cells can now recognize and react to the virus A peptides.t Remember that class I antigens are expressed on all nucleated cells and platelets.

39) Which of the following genes involved in the synthesis of immunoglobulins are linked on a single chromosome?

A. C gene for gamma chain and C gene for alpha chain

B. C gene for gamma chain and C gene for kappa chain

C. V gene for kappa chain and C gene for the epsilon chain

D. V gene for lambda chain and C gene for kappa chain

E. V gene for lambda chain and V gene for heavy chain

Explanation:

The correct answer is A. The genes for the synthesis of the entire heavy chain are present on human chromosome 14. Of the options given above, the only genes that are present on one chromosome are for the C (constant) regions of the heavy chains. Remember, the heavy chains determine the identity of the immunoglobulin isotypes: IgG, IgM, IgA, IgD, and IgE.

Choice B: The C gene for the gamma heavy chain is on chromosome 14 and the C gene for the kappa light chain gene is on chromosome 2.

Choice C: The V gene for the kappa light chain gene is on chromosome 2. The C gene for the epsilon heavy chain is on chromosome 14.

Choice D: The V gene for the lambda light chain gene is on chromosome 22. The C gene for the kappa light chain gene is on chromosome 2.

Choice E: The V gene for the lambda light chain gene is on chromosome 22 and the V gene for the heavy chain gene is on chromosome 14.

40) What is the role of the macrophage during antibody formation?

A. Activation of cytotoxic CD8 T cells

B. Delayed hypersensitivity reaction

C. Lysis of virus-infected cells

D. Processing antigen and presenting it to T helper CD4 cells

E. Synthesis of immunoglobulin

Explanation:

The correct answer is D. The macrophage phagocytizes exogenous antigens (for example a bacterium), degrading the antigen into small epitopes and presenting them, on MHC class II molecules on its surface, to CD4 T helper cells.

Macrophages do not activate cytotoxic CD 8 T lymphocytes (choice A). The major activator of cytotoxic CD 8 T lymphocytes is IL-2 from CD4+ THl cells.

Delayed hypersensitivity reactions (choice B) are the results of CD4+ THl cells. These cells do not produce antibody. They secrete gamma interferon and interleukin 2 (IL-2), stimulating more cells to become involved in the delayed hypersensitivity reaction.

The cells that participate in lysis of virus infected cells (choice C) are cytotoxic CD8+ T lymphocytes that react with MHC class I molecules containing epitopes of the virus from the infected cell. Macrophages do not participate in this activity.

Macrophages never synthesize antibody (choice E). B cells initially produce antibody, then are converted to plasma cells or memory B cells. The stimulus for the production of this antibody comes from T helper cells that were stimulated by epitopes presented to them by macrophages.

41) A 60-year-old female presents with progressive tightening and hardening of the skin on her arms and face that has recently caused disfigurement and difficulty performing manual activities. You suspect an autoimmune disorder, and order an antinuclear antibody panel. The results show antitopoisomerase I and no anticentromere antibodies. You advise the patient that, if left untreated, her disease will most likely

A. convert to a dermatologic malignancy

B. progress to involve other skin surfaces

C. progress to potentially fatal systemic fibrosis

D. remit completely

E. stabilize, with residual dermal fibrosis

Explanation:

The correct answer is C. This patient has scleroderma (or systemic sclerosis), an autoimmune connective tissue disorder. The skin is most frequently involved in this disease and is characterized by excessive tissue fibrosis. There is evidence for both an immunologic and vascular etiology to the disease. Almost all patients with scleroderma have antinuclear antibodies. Those with the antitopoisomerase antibody usually develop diffuse systemic sclerosis, and they usually die from consequences of systemic disease such as pulmonary fibrosis or malignant hypertension.

Scleroderma does not predispose to dermatologic malignancies (choice A).

Progression to involve other skin surfaces (choice B) without visceral involvement suggests limited systemic sclerosis, associated with an anticentromere antibody. 96% of patients with another limited form of systemic sclerosis, the CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, telangiectasias), also have an anticentromere antibody.

Scleroderma is a slowly progressive disease that will not stop or spontaneously reverse on its own (choices D and E).

42) A 60-year-old alcoholic male with a long history of cigarette smoking is brought to the emergency department after being found behind the neighborhood bar at 4 AM in freezing weather. On arrival, he is lethargic and experiences a shaking chill. His heart rate is 106, his breathing is labored with diffuse rales, and his temperature is 102.5 degrees Fahrenheit. His sputum is blood tinged, containing numerous gram-positive cocci which are identified as Streptococcus pneumoniae. The man is treated with penicillin and his condition improves over the next few days. Which of the following immune effector mechanisms was most important in completely clearing this infection?

A. ADCC (Antibody dependent cell cytotoxicity)

B. Complement mediated opsonization

C. Cytotoxic T cell lymphocytes

D. LAK cells

E. Natural killer cells

Explanation:

The correct answer is B. One of the most efficient mechanisms for eliminating extracellular pathogenic bacteria is by opsonization and phagocytosis by macrophages. The IgG and IgM antibody produced in response to the organism reacts with the capsular structure, stimulating the activation of the classical pathway of the complement system. This pathway produces large amounts of C3b that coat the organism, preparing it for phagocytosis.

ADCC cells (choice A) are actually natural killer (NK) cells that find virally infected cells and tumor cells that have been coated with antibody and react with and destroy them. These cells do not destroy antibody-coated bacteria, only body cells that are coated with antibody.

Cytotoxic T lymphocytes (choice C) react only with cells that have antigen epitopes presented in association with class I MHC molecules. An example would be a virus epitope from a virally infected cell presented by class I molecules on the surface of the cell.

LAK cells (choice D) are NK cells that have been activated by IL-2. They are considered to be superactivated NK cells.

The NK cells (choice E) are cells of the innate immune system that destroy virally infected cells or tumor cells.t This does not involve antibody and it does not involve extracellular pathogens.

43) A small 9-month-old male with a history of recurrent pyogenic infections is seen in a clinic. Immunoglobulin levels and a CBC are performed. The CBC is normal except for slight neutropenia and thrombocytopenia. Determination of immunoglobulin levels indicates elevated IgM, but deficiencies of IgG and IgA. The underlying defect involves which of the following molecules?

A. CD40 ligand (CD40L) on the T cell

B. CD40 molecule on the B cell

C. Gamma interferon

D. Interleukin-2 (IL-2)

E. Interleukin-3 (IL-3)

Explanation:

The correct answer is A. Patient's with hyper-IgM syndrome (HIGM) experience very little, if any, isotype switching. The B cells in these patients cannot undergo the switch from IgM to IgG, IgA, or IgE that normally occurs during B-cell maturation. When B cells undergo isotype switching they require two factors: IL-4, which binds to a specific receptor on the B cell, and the CD40 molecule on the B-cell surface, which binds to the CD40 ligand (CD40L) on an activated T-cell surface. The deficiency is due to mutations in the CD40L. This immunodeficiency results in patients who are IgG- and IgA-deficient, but synthesize large amounts of polyclonal IgM. Affected individuals are susceptible to pyogenic infections, and often form IgM autoantibodies to neutrophils, platelets, or tissue antigens. The disease is inherited as an X-linked recessive in 70% of the cases.

The problem is due to mutations in the CD40L, not the CD40 molecule on the B cell (choice B).

Gamma-interferon (choice C) is primarily a type I helper T cell (TH1) cytokine, although it does inhibit the TH2 lineage response to specific antigens.

A decrease in IL-2 (choice D) would inhibit the cell-mediated immune response and the patient would be susceptible to infection with intracellular microorganisms.

IL-3 (choice E) is considered a growth factor for hematopoietic stem cells and mast cells. The patient had a normal CBC except for slight neutropenia and thrombocytopenia.

44) A deficiency of the complement protein C4 would inhibit which of the following complement activities?

A. Completion of the classic pathway to the splitting of C3

B. Formation of C3b for opsonization

C. Formation of C5 convertase via the alternative pathway

D. Formation of C5a for chemotactic attractant for neutrophils

E. Formation of the membrane attack complex

Explanation:

The correct answer is A. The classic complement pathway involves C1, C4, and C2 to the point of splitting C3.C3 is then split to yield C3b and C3a. A deficiency of C4 would have no effect on any of the other answer choices listed.

The complement protein C3 can be split into C3a and C3b (choice B) using the alternative pathway. The additional proteins required in this pathway would be factors B and D and properdin.

The C5 convertase enzyme can be formed in the alternative pathway (choice C) without using C4.

The C5 molecule could be split into C5a (choice D) and C5b using the alternative pathway, without the involvement of C4.

The membrane attack complex (choice E; C5b, C6, C7, C8, and polymers of C9) lyses the pathogenic cell. This process does not require the alternative pathway and would not require C4.

Anti-centromere
High titer
Anti-Scl-70
Not detected
Anti-Pol-I
Not detected
Anti-RNP
Not detected
Anti-dsDNA
Not detected

Anti-centromere
High titer
Anti-Scl-70
Not detected
Anti-Pol-I
Not detected
Anti-RNP
Not detected
Anti-dsDNA
Not detected

45) Which of the listed diseases is most strongly suggested by the above antibody studies?

A. CREST

B. Diffuse scleroderma

C. Mixed connective tissue disease

D. Sjögren's syndrome

E. Systemic lupus erythematosus

Explanation:

The correct answer is A. Anti-centromere antibodies are a marker for the CREST (calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasias) variant of scleroderma. The markers for diffuse scleroderma (choice B), anti-Scl-70 and anti-Pol-I, are usually negative in CREST syndrome, as are most other autoantibodies.

The marker for mixed connective tissue disease (choice C) is anti-RNP.

The markers for Sjögren's syndrome (choice D) are anti-SSA, anti-SSB, and anti-RAP.

Systemic lupus erythematosus (choice E) typically produces a large number of autoantibodies, of which anti-dsDNA and anti-Sm are the most distinctive.

46) What is the role of class II MHC proteins on donor cells in the process of graft rejection?

A. They are recognized by helper T cells, which then activate cytotoxic T cells to kill the donor cells

B. They are the receptors for interleukin-2, which is produced by macrophages when they attack the donor cells

C. They cause the release of perforins to lyse the cells

D. They induce IgE, which mediates graft rejection

E. They induce the production of blocking antibodies that protect the graft

Explanation:

The correct answer is A. Class II MHC proteins are expressed on the surfaces of macrophages, dendritic cells, and B cells; this complex of molecules is recognized by CD 4+ helper T cells. The T cells of the transplant recipient recognize allogeneic MHC molecules on the surface of an antigen-presenting cell of the donor. It is thought that interstitial dendritic cells of the donor are the most important immunogens because not only do they express class I and II HLA molecules, but they are also endowed with co-stimulatory molecules. CD 8+ cytotoxic T cells recognize the class I molecules. CD 4+ cells proliferate as Th1 cells and produce interleukin 2, which causes differentiation of the CD 8 cells. The CD 8+ cytotoxic cells of the recipient then cause lysis of the donor cells.

Interleukin-2 (choice B) activates T cells by binding to high-affinity IL-2 receptors (IL-2R).

Perforins (choice C) are produced by CD 8+ cytotoxic lymphocytes as they bind to Class I MHC molecules. The perforins damage the donor cell membranes, resulting in lysis.

IgE-mediated reactions (choice D) are not associated with graft rejection.

Blocking antibodies (choice E) are employed as a form of immunosuppressive therapy. Antilymphocyte globulins and monoclonal anti-T cell antibodies (monoclonal anti-CD3) are used to inhibit rejection of the graft. This process does not involve class II MHC proteins on donor cells.

47) A 33-year-old single mother of two young children visits her physician because of an oral ulcer. A review of systems is significant for fatigue, myalgia, and joint pain. Laboratory results demonstrate leukopenia, and a high-titered antinuclear antibody. A speckled staining pattern due to anti-Sm is seen with immunofluorescence; urinary protein is elevated. Which of the following is the most likely diagnosis?

A. Generalized fatigue

B. Goodpasture's syndrome

C. Mixed connective tissue disease

D. Scleroderma

E. Systemic lupus erythematosus

Explanation:

The correct answer is E. Systemic lupus erythematosus (SLE) is a prototype connective tissue disease. The diagnosis requires four criteria to be met from a list of eleven possible criteria: malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, serositis, renal disorder, neurologic disorder, hematologic disorder, immunologic disorder, and antinuclear antibody. This patient also has anti-Sm, which is pathognomonic for SLE,
but is only found in 30% of the affected patients. Antinuclear antibodies (ANA) are present in 95-100% of cases of SLE; anti-double-stranded DNA is found in 70% of the cases.

Generalized fatigue (choice A) due to being a single working mother of two children could well be a possibility, but the presence of the other criteria make SLE more likely.

Goodpasture's syndrome (choice B) is characterized by linear disposition of immunoglobulin, and often C3, along the glomerular basement membrane (GBM). Glomerulonephritis, pulmonary hemorrhage, and occasionally idiopathic pulmonary hemosiderosis occur.

Mixed connective tissue disease (choice C) is an overlap syndrome characterized by a combination of clinical features similar to those of SLE, scleroderma, polymyositis, and rheumatoid arthritis. These patients generally have a positive ANA in virtually 100% of the cases.

High titer anti-ribonucleoprotein (RNP) antibodies may be present, generating a speckled ANA pattern. Anti-RNP is not pathognomonic for mixed connective tissue disease, since it can be found in low titers in 30% of the patients with SLE.

Scleroderma (choice D) is characterized by thickening of the skin caused by swelling and thickening of fibrous tissue, with eventual atrophy of the epidermis. ANA are often associated with the disease, but the staining pattern is generally nucleolar.

48) A superantigen is a bacterial product that

A. binds to B7 and CD28 costimulatory molecules

B. binds to the β chain of TCR and MHC class II molecules of APC stimulating T cell activation

C. binds to the CD4 + molecule causing T cell activation

D. is presented by macrophages to a larger-than-normal number of T helper CD4 + lymphocytes

E. stimulates massive amounts of IgG synthesis because of its large size

Explanation:

The correct answer is B. A superantigen, such as TSST- 1 or staphylococcal enterotoxin, cross-links the variable domain of the TCR β chain to the MHC class II molecule and specifically induces massive T cell activation.

The superantigen does not bind the B7 and CD28 costimulatory molecules (choice A). Instead, the
costimulatory molecules bind to each other to stimulate the reaction between the antigen-presenting cell and T
cell.

The superantigen does not bind the CD 4 molecules (choice C) but instead binds on the other side of the TCR
receptor complex.

The term superantigen has nothing to do with the antigen being presented by macrophages to T cells (choice
D).

The term superantigen has nothing to do with its size or its ability to stimulate antibody production (choice E).
The term superantigen is used because of its unusual ability to create massive T cell activation by the unique
type of binding.

49) The blood from an 8-year-old boy was analyzed by flow cytometry. The exact number of B cells was counted.

Which of the following cell surface markers was likely used to identify the B cells in this blood sample ?

A. CD3

B. CD4

C. CD8

D. CD19

E. CD56

Explanation:

The correct answer is D. The best markers for identification of B cells are CD19, CD20, and CD21. The CD21 marker is a receptor for EBV (Epstein-Barr Virus).

The CD3 marker (choice A) is present on all T cells with either a CD4 or CD8 marker. This is the marker that is used to identify total T cell count in a blood sample. The CD3 marker is used for signal transduction in the different T cells.

The CD4 marker (choice B) is used to identify T helper cells. These are the cells that recognize exogenous peptides presented on MHC class II molecules by macrophages. CD4+ T helper cells are also involved in cell-mediated delayed hypersensitivity, production of cytokines for stimulation of antibody production by B cells,

and stimulation of macrophages.

The CD8 marker (choice C) is used to identify cytotoxic T cells. These are the cells that recognize viral epitopes attached to the MHC class I molecules of a virally infected cell.

The CD56 marker (choice E) is used to identify NK(natural killer) cells. These cells are important in innate host defense, specializing in killing virally infected cells and tumor cells by secreting granzymes and perforins.

50) Which of the following is an example of a type II hypersensitivity?

A. A patient with berylliosis

B. A patient with heat intolerance, sinus tachycardia, and proptosis of the eyes

C. Eczematous reaction on the dorsum of the foot in a patient who washed his socks in a new detergent

D. Glomerulonephritis in a patient with systemic lupus erythematosus

E. Wheal and flare reactions and vesicles at multiple sites on the lower legs in a patient attacked by fire ants

Explanation:

The correct answer is B. Type II hypersensitivity is mediated by antibodies directed toward antigens that are present on the surface of cells or other tissue components. The antigen may be intrinsic to the cell membrane or may take the form of an exogenous antigen that is adsorbed to the cell surface. The patient described in choice B has Graves disease, which is an autoimmune form of hyperthyroidism produced by autoantibodies directed against the TSH (thyroid stimulating hormone) receptor. These antibodies are called LATS (long-acting thyroid stimulator) and stimulate thyroid function, resulting in the release of thyroid hormones.

In berylliosis (choice A), noncaseating granulomas typically are present in the lungs and hilar lymph nodes. This is a form of type IV hypersensitivity.

An eczematous reaction (choice C) associated with washing clothes in a new detergent may either represent type IV hypersensitivity or a non-immune reaction associated with direct toxicity from some component of the soap penetrating the skin.

Glomerulonephritis in systemic lupus erythematosus (choice D) is due to the deposition of antigens in the glomerular basement membrane with the resultant formation of antigen-antibody complexes. These complexes activate the complement cascade, which causes neutrophils to enter the area and produce tissue damage.

Wheal and flare reactions (choice E) are cutaneous manifestations of type I hypersensitivity.

51) T cells that have a low affinity for MHC class I molecules differentiate in the thymus to become which type of cell?

A. CD 8 + cytotoxic lymphocyte

B. Gamma-delta T cell

C. Natural killer cell

D. T helper 1 cell

E. T helper 2 cell

Explanation:

The correct answer is A. CD8+ cytotoxic T lymphocytes are positively selected in the thymus because they have low affinity for MHC class I molecules. If they had possessed high affinity for the MHC class I molecules, they would have been eliminated (negative selection) because of the danger of autoimmune disease. Also, cells with no affinity for MHC class I molecules would be eliminated.

The gamma-delta designation in a T cell (choice B) refers to type of receptor on the cell. Most T-cell receptors are alpha-beta receptors, but some are of a different isotype termed gamma-delta.

Natural killer cells (choice C) are large granular lymphocytes that are part of the innate immune response.
Natural killer cell function does not depend on MHC class I or class II molecules; it simply kills tumor cells or virally infected cells.

T helper 1 (choice D) and T helper 2 cells (choice E) would have a low affinity for class II MHC molecules in order to survive in the thymus. If they had no affinity or strong affinity for these molecules, they would have been eliminated.

52) A 36-year-old farmer has been exposed to poison ivy on several different occasions and usually develops very severe skin lesions. He enrolls in an immunological study at an urban medical center. A flow cytometric measurement of T cells reveals values within the normal range. An increased serum concentration of which of the following cytokines would decrease the likelihood of a delayed-type hypersensitivity reaction in this individual?

A. Gamma interferon

B. IL-2

C. IL-4

D. IL-8

E. IL-10

Explanation:

The correct answer is E. The IL-10 cytokine is produced by T helper 2 (TH2) cells and inhibits T helper I (TH1) cells. Since the cytokines from TH1 cells stimulate cell-mediated immunity and delayed hypersensitivity, an increased level of IL-10 would decrease the likelihood of a delayed type hypersensitivity reaction. Other cytokines from TH2 cells stimulate B cells to produce antibody.

The gamma interferon cytokine (choice A) from T helper I cells inhibits TH2 cells. It also activates NK cells and activates macrophages.

The IL-2 cytokine (choice B) stimulates TH1 subset, CD8 T cytotoxic cells, and activates NK cells. It is one of the most active cytokines and is involved in many other reactions.

The IL-4 cytokine (choice C) from TH2 cells stimulates B cells to produce antibody, inhibit macrophages, and stimulates class switching from IgG isotype to IgE isotype.

The IL-8 cytokine (choice D) is produced by macrophages and is chemotactic for neutrophils. This cytokine is most important in stimulating an inflammatory reaction and attraction of neutrophils to the site.

53) A 41-year-old patient informs her physician that in her childhood, she experienced two bouts of rheumatic fever. Although she appears to be well at present, which of the following sequelae of rheumatic fever is most likely to present as a chronic disease in her later years?

A. Arthritis

B. Fibrinous pericarditis

C. Mitral valve disease

D. Myocarditis

E. Neurological disease

Explanation:

The correct answer is C. After an initial attack of rheumatic fever, an affected individual is at increased risk for developing recurrent rheumatic disease after each pharyngeal infection by beta-hemolytic streptococci. Any one of the manifestations of rheumatic disease can reoccur, but will usually resolve. In marked contrast, the damage to the heart valves caused by rheumatic fever can lead to fibrous scarring and deformity, producing valvular dysfunction (particularly mitral stenosis), which may lead to heart failure in late adulthood.

The arthritis (choice A) produced by rheumatic fever is transitory, and resolves after the attack has run its course. There are no long-term complications of the arthritis.

Fibrinous pericarditis (choice B), like almost any fibrinous inflammatory response, is an acute process.
Although the patient may develop some scarring of the pericardium, this is generally not sufficient to produce long-lasting consequences.

Myocardial involvement (choice D) by rheumatic fever is typically seen as Aschoff bodies, collections of fibrinoid necrosis, lymphocytes, plasma cells, and histiocytes within the myocardium. When the event resolves, the Aschoff bodies will be replaced by discrete fibrous scars. No appreciable myocardial dysfunction ensues.

Sydenham's chorea, the neurologic manifestation of rheumatic fever, leaves no lasting neurological effects (choice E).

54) A formula-fed 1-month-old boy is exposed to his sister, who has chickenpox. He does not develop signs of varicella. His mother had the infection 5 years ago. Which class of immunoglobulins did he acquire from his mother in utero that protected him from this virus?

A. IgA

B. IgD

C. IgE

D. IgG

E. IgM

Explanation:

The correct answer is D. This baby is exhibiting passive immunity acquired from his mother in utero. IgG is the only class of immunoglobulins that can cross the placenta. As such, IgG molecules diffuse into the fetal circulation, providing immunity. This circulating maternal IgG protects the newborn during the first 4-6 months of life. Note that IgG is also capable of opsonization and complement activation (a feature shared with IgM).

IgA (choice A) functions in the secretory immune response. The secretory form of this immunoglobulin (sIgA) is found in tears, colostrum, saliva, breast milk, and other secretions. It is produced by the plasma cells in the lamina propria of the GI and respiratory tracts.

IgD (choice B) functions as a cell surface antigen receptor on undifferentiated B cells.

IgE (choice C) is involved in the allergic response and immediate hypersensitivity reactions. The Fc region of IgE binds to the surface of basophils and mast cells. Antigen binding to two IgE molecules leads to mast cell degranulation and the release of leukotrienes, histamine, eosinophil chemotactic factors, and heparin.

IgM (choice E) is the first antibody detected in serum after exposure to antigen. IgM circulates as a pentamer and thus has five Fc regions. This structure makes it especially effective in fixing complement. Isohemagglutinins, rheumatoid factors, and heterophile antibodies are all IgM.

55) Administration of the DPT vaccine (diphtheria toxoid, pertussis products, and tetanus toxoid) would stimulate
which of the following types of immunity?

A. Adoptive

B. Artificial active

C. Artificial passive

D. Natural active

E. Natural passive

Explanation:

The correct answer is B. Administration of the DPT vaccine stimulates the innate immune system to produce antibody and memory cells against this mixture. Active immunity is when we produce our own antibody. Artificial refers to the fact that the stimulus was the vaccination with the antigens in question.

Adoptive immunity (choice A) involves the patient receiving cells from another host who had been stimulated to produce their products.

Artificial passive immunity (choice C) refers to the immunity produced by receiving an injection of antibody. An example is the administration of immune globulin directed against hepatitis A after an individual had been exposed to it.

Active immunity (choice D) means that we are stimulated to produce our own antibodies. The term natural active refers to the fact that we received the stimulus (antigen) by a natural means, such as exposure to the organism.

Natural passive immunity (choice E) refers to the immunity a fetus receives via the placenta. The term passive means that the fetus received the antibody from another source and did not make it itself.

56) A full-term baby boy is delivered after an uneventful pregnancy, and is well for the first 2 years of his life. He receives all his immunizations without any complications. Starting around his 2nd birthday, the mother begins to note frequent upper respiratory tract infections, and the child is hospitalized three times for pneumonia. Laboratory testing would most likely reveal a deficiency of which of the following immunoglobulins in this child?

A. IgA

B. IgD

C. IgG

D. IgM

Explanation:

The correct answer is A. Selective IgA deficiency (<5 mg/dL) is the most common of all the primary immunodeficiency diseases. The incidence reported in the US has ranged from 1:250 to 1:1000. IgA has two subclasses, IgA1 and IgA2. IgA1 predominates in the serum, while IgA2 predominates in mucosal secretions as a dimer bound together by a J chain with a secretory piece attached. Recurrent bacterial and viral infections of the respiratory tract can be attributed to a lack of secretory IgA (sIgA), the predominant immunoglobulin of the mucosal immune system.

IgD (choice B) has not been given any particular function other than to act as a receptor on the B cell. It can be found in very low levels in serum.

IgG (choice C) is the major immunoglobulin found in the humoral immune response. A patient with a low IgG will experience pyogenic infections.

IgM (choice D) is found in the early response to an antigen. If the patient was deficient in IgM he would have also been characteristically low in IgG and would have experienced recurrent pyogenic infection, usually commencing by the age of 5-6 months.

57) Which of the following class II antigens would be most likely to play a contributing role in hay fever?

A. DR2

B. DR3

C. DR4

D. DR5

E. DR7

Explanation:

The correct answer is A. Hay fever is closely associated with DR2 (relative risk 19). DR2 is also associated with narcolepsy and multiple sclerosis.

DR3 (choice B) is associated with Goodpasture's syndrome, celiac sprue, type 1 diabetes, and systemic lupus erythematosus.

DR4 (choice C) is associated with pemphigus vulgaris, rheumatoid arthritis, and type 1 diabetes.

DR5 (choice D) is associated with pernicious anemia and juvenile rheumatoid arthritis.

DR7 (choice E) is associated with steroid responsive nephrotic syndrome.

58) A 46-year-old woman presents with complaints of feeling as if she has "sand in her eyes" and reports difficulty swallowing such foods as crackers or toast. Which of the following pairs of tests would likely yield positive results in this patient?

A. Anti-centromere antibody and rheumatoid factor

B. Anti-Scl-70 antibody and anti-Smith antibody

C. Anti-Smith antibody and anti-double stranded DNA antibody

D. Rheumatoid factor and anti-double stranded DNA

E. Rheumatoid factor and anti-SS-A antibody

Explanation:

The correct answer is E. This patient has Sjögren's syndrome, which is an autoimmune disease characterized by lymphocytic infiltration of exocrine glands resulting in dry mouth (xerostomia) and dry eyes (keratoconjunctivitis sicca). Patients have an increased risk of malignant lymphoma. Autoantibodies produced include anti-Ro (SS-A), anti-La (SS-B), antinuclear antibodies, and rheumatoid factor.

Choice A: Anti-centromere antibodies are a very specific marker for CREST syndrome. Rheumatoid factor is usually positive in rheumatoid arthritis but may be seen in low titers in patients with other autoimmune diseases and chronic inflammatory conditions.

Choice B: Anti-Scl-70 antibodies are seen in patients with scleroderma. Anti-Smith antibody is seen in systemic lupus erythematosus.

Choice C: Anti-Smith antibody and anti-double stranded DNA antibody are associated with systemic lupus erythematosus.

Choice D: Rheumatoid factor is seen in approximately 80% of patients with rheumatoid arthritis and in low titers in other autoimmune disorders. Anti-double stranded DNA (anti-dsDNA) is seen in patients with SLE.

59) A 12-year-old girl presents with a skin abscess. The causative organism is found to be Staphylococcus aureus. Over the past year, she has had several similar abscesses, as well as two bouts of aspergillosis. Which of the following is the most likely explanation for her repeated infections?

A. Defective chemotactic response

B. Defective NADPH oxidase

C. Deficiency of C5

D. IgA deficiency

E. Thymic hypoplasia

Explanation:

The correct answer is B. A history of recurrent infection with S. aureus and S. aspergillus suggests a diagnosis of chronic granulomatous disease, which is caused by a defect in the NADPH oxidase of neutrophils, resulting in an inability to generate toxic oxygen metabolites following phagocytosis. Patients become susceptible to certain opportunistic infections, particularly those caused by S. aureus, S. aspergillus, Nocardia, and Salmonella.

Defective chemotactic response (choice A), seen in diseases such as Chédiak-Higashi syndrome and lazy-leukocyte syndrome, would make patients more susceptible to infections by bacteria such as Staphylococci and Streptococci.

Deficiency of C5 (choice C), a complement component, would make patients prone to infection by gram-negative bacteria, especially Neisseria species.

IgA deficiency (choice D) represents the most common immunodeficiency and would present with increased predisposition to autoimmune disorders, respiratory infections, and milk allergy.

Thymic hypoplasia (choice E) is caused by defective development of the 3rd and 4th pharyngeal pouches. The result is decreased T-cell levels, making patients more prone to viral and fungal infections. They also will exhibit hypocalcemia and other signs of parathyroid insufficiency.

60)A 4-month-old male presents with twitching of the facial muscles. He has previously been seen for several severe episodes of Candida infections. On examination, the child has low-set ears, hypertelorism, and a shortened philtrum. What additional findings would be likely in this individual?

A. Absent thymic shadow on chest x-ray

B. Decreased alpha-fetoprotein

C. Decreased IgA levels

D. Elevated IgM levels

E. Prominent telangiectasias around the eyes

Explanation:

The correct answer is A. The clinical findings describe DiGeorge syndrome. Patients clinically present with tetany (usually first noted in the facial muscles) due to hypocalcemia secondary to hypoparathyroidism. The thymus is absent, as are the parathyroid glands, due to failure of development of the 3rd and 4th pharyngeal pouches. Recurrent infections due to defective cellular immunity and abnormal facies are additional features.

Decreased alpha-fetoprotein (choice B) is an amniotic fluid marker for Down's syndrome. Down's syndrome patients have abnormal immune responses that predispose them to serious infections (particularly of the lungs) and to thyroid autoimmune disease. However, there is no defect of the parathyroid glands.

Decreased IgA levels (choice C) describes selective IgA deficiency, which is the most common hereditary immunodeficiency. The syndrome is due to a failure of heavy-chain gene switching in B cells.

Elevated IgM (choice D) is seen in hyper-IgM syndrome. Patients have a high concentration of IgM and normal numbers of T and B cells, but low levels of IgG, IgA, and IgE. Helper T cells have a defect in the surface protein CD40 ligand that interacts with CD40 on the B-cell surface. This results in an inability of the B cell to switch from the production of IgM to other classes of antibodies.

Prominent telangiectasias around the eyes (choice E) are seen as part of the ataxia-telangiectasia syndrome. This is an autosomal recessive disorder, and is also referred to as a chromosomal breakage syndrome.

Ataxia-telangiectasia is associated with increased numbers of translocations, especially involving the T-cell receptor loci; the gene for this disorder has been mapped to chromosome 11. Patients have an increased incidence of malignancy.

61) Anti-ribonucleoprotein (anti-RNP) - high titer
Rheumatoid factor (RF) - low titer
Anti-single stranded DNA (anti-ssDNA) - low titer
Anti-double-stranded DNA (anti-dsDNA) - not detected
Anti-Smith antigen (anti-Sm) - not detected
Anti-SCI-70 - not detected

Which of the listed diseases is suggested by the antibody studies above?

A. CREST syndrome

B. Diffuse scleroderma

C. Drug-induced lupus

D. Mixed connective tissue disease

E. Systemic lupus erythematosus

Explanation:

The correct answer is D. Mixed connective tissue disease (MCTD) is clinically an overlap autoimmune disorder, including joint pain, myalgias, pleurisy, esophageal dysmotility, and skin disease. Interestingly, the condition is usually characterized by high antibody titers to ribonucleoprotein (RNP), which serves as a disease marker, and the condition is apparently immunologically distinct from other connective tissue disorders. Low titer RF and anti-ssDNA are common features of MCTD, but other autoantibodies are less commonly observed.

Anti-centromere antibody is the distinctive marker for the CREST (choice A) variant of scleroderma, characterized by calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia.

Anti-SCI-70 is the distinctive marker for diffuse scleroderma (choice B), or systemic sclerosis, a multisystem disorder in which fibrosis of the skin, blood vessels, and viscera occurs.

High titers of anti-histone antibody without other autoantibodies is the distinctive marker for drug-induced lupus
(choice C). This disorder is particularly associated with administration of hydralazine and procainamide.

High titers of anti-dsDNA is the distinctive marker for systemic lupus erythematosus (choice E).

62) A 32-year-old female who is 6 months pregnant presents for prenatal care. A routine evaluation is performed, including testing for HIV antibody. The patient is reported to be negative for RPR, but positive for HIV antibody by the enzyme-linked immunoassay (EIA). The HIV Western blot is positive for antibody to the p24 antigen. The patient should be counseled

A. that she and her baby were both infected with HIV

B. that she had a false-positive HIV EIA

C. that she is negative for HIV since the RPR was negative

D. that she is positive for the HIV virus

E. to have an HIV polymerase chain reaction (PCR) test performed

Explanation:

The correct answer is E. A patient who is HIV EIA-positive must always have the result confirmed by a confirmatory assay (e.g., HIV Western blot). The HIV Western blot is considered positive when the patient demonstrates the presence of antibody to at least two of three important HIV antigens, which are gp120, gp41, and p24. If no reaction is observed, then the patient is considered negative, but any reaction that is not consistent with a positive is reported as indeterminate. Therefore, this patient is considered indeterminate. The physician can wait 6 months and retest by Western blot; if the results are identical, then the patient is reported as negative, or the patient can be tested by another confirmatory test such as the PCR assay. A negative PCR in this situation would classify this patient as negative; however, it would be wise to retest the patient in 3-6 months if she had risk factors.

Approximately 30% of the babies from untreated and 8% from treated HIV-positive mothers will be infected. One cannot conclude from the available data that both she and her baby are infected (choice A).

It is possible that this patient had a false-positive HIV EIA assay, but with the present data, it is impossible to know if the patient is in the early stages of seroconversion or if the test result is a false positive (choice B).

RPR (choice C) is a test for syphilis (and not a very specific one at that), not a test for HIV.

Because this patient's Western blot was indeterminate, a confirmatory test (e.g., Western blot or PCR) must be performed to determine whether she is truly positive for the HIV virus (choice D).

63) A 29-year-old Mexican American woman receives an intradermal tuberculin injection and later develops an indurated, erythematous papule 12 mm in diameter. This reaction is an example of which of the following?

A. Antibody-dependent cell-mediated cytotoxicity

B. Local anaphylaxis

C. T-cell mediated cytotoxicity

D. Type III hypersensitivity

E. Type IV hypersensitivity

Explanation:

The correct answer is E. The tuberculin reaction is an example of delayed-type hypersensitivity (a form of Type IV hypersensitivity) in which the bulk of the tissue damage is done by macrophages that are stimulated by a few previously sensitized CD4+ memory T-cells recognizing antigens presented by the macrophages. In contrast, in T-cell mediated cytotoxicity (choice C, another form of Type IV sensitivity) the damage is done by CD8+ cytotoxic T-cells that recognize "foreign" cell surface antigens and directly lyse targeted cells.

Antibody-dependent cell-mediated cytotoxicity reactions (choice A, a form of Type II hypersensitivity) involves cells coated with a thin layer of antibody that triggers attack by cells (monocytes, neutrophils, eosinophils, and natural killer cells) that can bind to Fc receptors.

Local anaphylaxis (choice B, a form of Type I hypersensitivity) is due to the release of vasoactive substances by mast cells and basophils stimulated by memory (CD4+) T-cells reacting to antigen.

Type III (choice D) hypersensitivity is due to deposition of circulating antigen-antibody complexes, often in small blood vessels.

64) Which of the following could prevent an allergen from reacting with a specific IgE molecule present on the mast
cell membrane?

A. Antihistamine

B. Blocking antibody

C. Cromolyn sodium

D. Epinephrine

E. Theophylline

Explanation:

The correct answer is B. Blocking antibody is generally an IgG antibody against the allergen. It is induced in the allergic patient by administering small amounts of allergen over a period of time. When the person is again exposed to the allergen, the IgG reacts with the allergen before it can reach the IgE-coated mast cell.

Antihistamines (choice A) would block histamine receptors but would not react with the allergen before it could reach the IgE-coated mast cell.

Cromolyn sodium (choice C) is a drug that stabilizes mast cell membranes, thus inhibiting degranulation, but would have no effect on allergen binding to IgE on mast cells.

Epinephrine (choice D) is the mainstay of therapy in severe cases of immediate hypersensitivity (anaphylaxis). It increases intracellular cyclic AMP, thus decreasing mast cell degranulation, and causes smooth muscle dilation in the airways. Epinephrine would have no effect on the binding of the allergen to IgE on mast cells.

Cyclic AMP is degraded in cells by the enzyme phosphodiesterase. Theophylline (choice E) inhibits this enzyme, thereby increasing mast cell cyclic AMP levels and making degranulation less likely without affecting the binding of the allergen to IgE on mast cells.

65)

1. Degranulation of mast cells and basophils occurs when allergen binds to two adjacent IgE antibodies on the membrane of the cell

2. Interleukin 4 released by CD4 helper T cells causes activated B cells to switch from making IgM to IgE

3. Processed allergen is associated with a class II molecule on an antigen presenting cell and recognized by
CD4+ TH2 cells

4. Prostaglandins and leukotrienes are released

5. The allergen is phagocytized and processed by a macrophage or a dendritic cell

Based on the numbered list above, which of the following is the correct sequence of events in the Type I hypersensitivity reaction?

A. 1,2,3,5,4

B. 1,4,3,5,2

C. 1,5,2,3,4

D. 5,3,1,4,2

E. 5,3,2,1,4

Explanation:

The correct answer is E. This question illustrates an important point. An allergen or antigen must first be processed by an antigen-presenting cell in an unsensitized host before the CD 4+ T helper cells can react to it.
These cells of the TH2 subclass then produce a variety of cytokines including interleukin 4. Interleukin 4 causes the heavy chain switching in the B cells from IgM to IgE. The IgE binds to the surface of mast cells or basophils by the Fc fragment. When the patient is re-exposed to the sensitizing allergen, the IgE binds to the allergen and causes degranulation of the mast cell or basophil.

66) Which of the following cell surface markers is required for lysis of IgG-coated target cells (antibody-dependent, cell-mediated cytotoxicity, or ADCC) by natural killer cells?

A. CD3

B. CD16

C. CD19

D. CD21

E. CD56

Explanation:

The correct answer is B. CD16 is a cell surface marker used to identify natural killer (NK) cells (lymphocytes lacking most T- and B-cell markers). CD16 is an Fc receptor for IgG, allowing the NK cells to bind to the coated target cell during ADCC, facilitating lysis.

CD3 (choice A) is a five-polypeptide cluster that represents the non-variable part of the T-cell receptor complex. The variable part is able to rearrange itself to adapt to specific antigens. Natural killer cells are CD3-negative.

CD19 (choice C) is a B-cell marker. It is a signal-transducing molecule that is expressed in early B-cell differentiation. NK cells are negative for CD19.

C21 (choice D) is also a B-cell marker. It is a complement receptor, and is also the same receptor the Epstein-Barr virus uses to bind to cells during infection.

CD56 (choice E) is a natural killer cell marker, but is not involved with ADCC.

67) A 15-month-old boy with recurrent episodes of ear, skin, and respiratory tract infections is found to have extremely low levels of serum IgG. Which of the following findings would support a diagnosis of common variable immunodeficiency in this child?

A. Absent germinal centers in lymph nodes

B. Depressed levels of IgA and IgM

C. Depressed numbers of circulating B lymphocytes

D. Hypocalcemia

E. Reversal of CD4:CD8 ratio

Explanation:

The correct answer is B. Common variable immunodeficiency is a disease caused by abnormal B-cell differentiation, characterized by decreased numbers of plasma cells and inadequate immunoglobulin production. The majority of patients with common variable immunodeficiency have low levels of all antibody classes.

Lymph node architecture is unaltered in common variable immunodeficiency, although plasma cells are not present in the medullary cords. In contrast, normal germinal centers are absent (choice A) in X-linked agammaglobulinemia and some cases of severe combined immunodeficiency disease.

Patients with common variable immunodeficiency have normal numbers of circulating B lymphocytes. B-cell deficiency (choice C) is more typical of X-linked agammaglobulinemia.

Hypocalcemia (choice D) and immunodeficiency are features seen in DiGeorge syndrome, in which the thymus and parathyroids are absent due to failure of development of the pharyngeal pouches. The parathyroids and thymus are normal in common variable immunodeficiency.

The CD4:CD8 ratio is reversed (choice E) in HIV disease, due to selective destruction of CD4+ T cells. T cells are not affected by common variable immunodeficiency.

68) A 24-year-old man presents with complaints of itching on his arms and face. Physical examination reveals well-circumscribed wheals with raised, erythematous borders and blanched centers. Which form of hypersensitivity is this patient probably exhibiting?

A. Acute serum sickness (Type III)

B. Antibody-dependent cell-mediated cytotoxicity (Type II)

C. Anti-receptor antibodies (Type II)

D. Delayed type hypersensitivity (Type IV)

E. Immediate type hypersensitivity (Type I)

Explanation:

The correct answer is E. Urticaria (hives) is a good example of a local anaphylaxis reaction, which is classified as a Type I hypersensitivity reaction. Type I hypersensitivity reactions involve preformed Ig E antibody bound to mast cells or basophils, which release vasoactive and spasmogenic substances when they react with antigens.

Acute serum sickness (choice A) is now uncommon but was formerly seen when animal sera were used for passive immunization.

The eosinophil-mediated cytotoxicity against parasites is an example of antibody-dependent cell-mediated cytotoxicity (choice B).

Myasthenia gravis is an example of a disease caused by anti-receptor antibodies (choice C).

The tuberculin (PPD) reaction used to test for tuberculosis exposure is an example of delayed-type hypersensitivity (choice D).

69) A 47-year-old woman presents to a physician with finger stiffness. Physical examination demonstrates marked thickening of the skin, most striking on the hand, which is limiting finger mobility. The physical examination also reveals multiple small telangiectasias and several hard nodules on the buttocks. Questioning of the patient reveals recent difficulty swallowing and skin color changes when the hands are exposed to cold. Autoantibody formation to which of the following substances is most strongly associated with this patient's condition?

A. Centromeres

B. Double-stranded DNA

C. Glomerular basement membrane

D. Scl 70

E. Smith antigen

Explanation:

The correct answer is A. The patient has the CREST variant of scleroderma, which is associated with anti-centromere antibody. The CREST syndrome comprises calcinosis (the hard calcified, subcutaneous nodules of the buttocks), Raynaud's phenomenon (the skin color changes in response to cold), esophageal dysmotility (difficulty swallowing), sclerodactyly (scleroderma involving the fingers), and telangiectasia (small vascular lesions of the skin). The CREST variant usually has a more benign course than the systemic variant of scleroderma.

Anti-double-stranded DNA (choice B) and anti-Smith antigen (choice E) are associated most strongly with systemic lupus erythematosus.

Anti-glomerular basement membrane (choice C) is associated with Goodpasture's disease.

Anti-Scl 70 (choice D) is associated with the systemic form of scleroderma.

70) A 54-year-old man presents with complaints of shortness of breath, a sore tongue, and a pins-and-needles sensation in his feet. Laboratory examination reveals macrocytosis, anemia, and hypersegmented neutrophils. Antibodies to intrinsic factor are detected in the patient's serum. Which of the following class II antigens would be most likely to play a contributing role in the etiology of this patient's disease?

A. DR2

B. DR3

C. DR4

D. DR5

E. DR7

Explanation:

The correct answer is D. The disease with autoantibodies to intrinsic factor is pernicious anemia (atrophic gastritis and megaloblastic anemia secondary to vitamin B12 deficiency). Pernicious anemia is associated with the DR5 class II antigen (relative risk 5); DR5 is also associated with juvenile rheumatoid arthritis.

DR2 (choice A) is associated with allergy, multiple sclerosis, and narcolepsy.

DR3 (choice B) is associated with Goodpasture's syndrome, celiac sprue, Type 1 diabetes mellitus, and systemic lupus erythematosus.

DR4 (choice C) is associated with pemphigus vulgaris, rheumatoid arthritis, and Type 1 diabetes mellitus.

DR7 (choice E) is associated with steroid-responsive nephrotic syndrome.

71) A 35-year-old male presents to an infectious disease specialist with recurrent infections with encapsulated bacterial organisms. The history indicates that these infections have become apparent in the last 6 months.Laboratory findings indicates that the total immunoglobulin level exceeded 900 mg/dL with a low CH50 (hemolytic complement) level. Which of the following is the correct diagnosis?

A. Acquired hypogammaglobulinemia (common variable hypogammaglobulinemia)

B. C3 deficiency

C. Hyper-IgM syndrome

D. Wiskott-Aldrich syndrome

E. X-linked infantile hypogammaglobulinemia

Explanation:

The correct answer is B. Individuals with C3 deficiency have an increased susceptibility to recurrent bacterial infections, especially with encapsulated bacteria. This susceptibility illustrates the important role of C3 as an opsonin. C3 deficiency is often not detected until later life.

Acquired hypogammaglobulinemia, also known as common variable hypogammaglobulinemia, (choice A) is ruled out by the fact that the total immunoglobulin level is greater than 300 mg/dL.

In hyper-IgM syndrome (choice C) there is very little, if any, isotype switching, resulting in patients who are IgG and IgA deficient, but synthesize large amounts of polyclonal IgM. These patients are susceptible to pyogenic infections, and thus the disorder is detected very early in life.

In Wiskott-Aldrich syndrome (choice D), a combined immunodeficiency syndrome, immune abnormalities are apparent at birth. Patients usually have low IgM levels with elevated levels of serum IgA and IgE; recurrent pyogenic infections, eczema, and thrombocytopenia are characteristic

Patients with X-linked infantile hypogammaglobulinemia (choice E) are detected in the first 4-8 months of life and have total immunoglobulin levels less than 200 mg/dL. Recurrent pyogenic infections are characteristic of this disorder.

72) A 45-year-old homeless man has a chronic cough, a cavitary lesion of the lung, and is sputum positive for acid-fast bacilli. Which of the following is the principal form of defense by which the patient's body fights this infection?

A. Antibody-mediated phagocytosis

B. Cell-mediated immunity

C. IgA-mediated hypersensitivity

D. IgE-mediated hypersensitivity

E. Neutrophil ingestion of bacteria

Explanation:

The correct answer is B. The principal host defense in mycobacterial infections (such as this patient's tuberculosis) is cell-mediated immunity, which causes formation of granulomas. Unfortunately, in tuberculosis and in many other infectious diseases characterized by granuloma formation, the organisms may persist intracellularly for years in the granulomas, only to be a source of activation of the infection up to decades later.

While antibody-mediated phagocytosis (choice A) is a major host defense against many bacteria, it is not the principal defense against Mycobacteria.

IgA-mediated hypersensitivity (choice C) is not involved in the body's defense against Mycobacteria.

IgE-mediated hypersensitivity (choice D) is not involved in the body's defense against Mycobacteria. It is important in allergic reactions.

Neutrophil ingestion of bacteria (choice E) is a major host defense against bacteria, but is not the principal defense against Mycobacteria.

73) Which marker or markers is present on B cells and could be used to specifically identify such cells in a flow cytometric analyzer?

A. CD 3

B. CD 8

C. CD 14

D. CD 16 and CD 56

E. CD 19 and CD 20

Explanation:

The correct answer is E. B cells have IgM and IgD on their surfaces, which are excellent markers for B cells. In addition, B cells have CD 19, 20, and 21 markers that aid in their identification.

The CD 3 (choice A) marker is present on all T cells.

The CD 8 (choice B) marker is present on cytotoxic T lymphocytes.

The CD 14 (choice C) marker is used to identify macrophages.

The CD 16 and CD 56 (choice D) markers are present on natural killer (NK) cells.

74) A 26-year-old systems analyst presents for evaluation of a bee sting allergy. He describes an episode in which he was stung on the forearm by a bee and, within 5 minutes, experienced pruritus, urticaria, and mild wheezing. The effector cell in this type of hypersensitivity is a(n)

A. eosinophil

B. mast cell

C. megakaryocyte

D. neutrophil

E. TH1 CD4+ lymphocyte

Explanation:

The correct answer is B. This patient is experiencing the early phase of type I hypersensitivity, characterized by pruritus and watery discharge from the nose, bronchospasm, mucus secretion in the airways, and a wheal-and-flare response with pruritus in the skin. The mechanism of hypersensitivity involves prior sensitization of a population of TH2 cells that produced cytokines, including interleukin-4. The interleukin-4 causes B cells to switch their heavy chain class from IgM to IgE. The IgE molecules then attach to mast cells or basophils. With subsequent antigen (allergen) challenge, the mast cells degranulate and release mediators, including histamine, which produces the anaphylactic response.

Eosinophils (choice A) are involved in type I hypersensitivity reactions as well as type II antibody-dependent cell cytotoxicity reactions directed against parasites. Eosinophils enter the area of the reaction due to the release of eosinophil chemotactic factor (eotaxin) and the beta-chemokine RANTES released from TH2 CD4+ cells and mast cells.

Eosinophils are recruited into the tissues as part of the late-phase reaction of type I hypersensitivity.

Their survival in tissue is dependent upon IL-3, IL-5, and granulocyte-monocyte colony stimulating factor (GM-CSF) released from TH2 cells.

Megakaryocytes (choice C) are bone marrow cells that produces platelets. They are not involved in type I hypersensitivity.

Neutrophils (choice D) are not a cell type that is key to the development of type I hypersensitivity.

TH1 CD 4+ lymphocytes (choice E) are associated with delayed type hypersensitivity reactions such as contact dermatitis involving exposure to poison ivy or poison oak.

75) A 3-year-old male who is small for his age presents with a history of pyogenic infections. Physical examination is remarkable for a high fever, hepatosplenomegaly, and inguinal and cervical lymphadenopathy. A culture of a purulent discharge from an abscess grows out Staphylococcus aureus. Immunoglobulin and complement levels are normal. The boy received all of the standard immunizations without any adverse effects. The boy's immune deficiency most likely involves which of the following?

A. B cells

B. Chemotaxis

C. IgG subclass 2

D. Phagocytic cells

E. T cells

Explanation:

The correct answer is D. This patient has chronic granulomatous disease (CGD), which is associated with a defective intracellular respiratory burst in phagocytes. CGD consists of a group of heterogenous disorders of oxidative metabolism affecting the pathways required for hydrogen peroxide production by phagocytic cells. It is inherited as an X-linked or autosomal recessive trait. Normally, stimulated phagocytic cells undergo a respiratory burst consisting of increased oxygen consumption leading to the generation of intracellular hydrogen peroxide and superoxide radicals.

The hydrogen peroxide and superoxide radicals are required for the killing of ingested intracellular organisms. The reaction is catalyzed by an NADPH oxidase that appears to be defective in the phagocytes of patients with CGD; the enzyme defect appears to be due to inherited mutations in the genes encoding cytochrome subunits. In CGD patients, the engulfment process by the phagocytic cells is normal; however, the pathogenic organism will not be killed, but will persist within the cell.

The patients suffer from infections with organisms that are normally considered of low virulence (e.g., Staphylococcus aureus, Aspergillus, Candida, Escherichia coli, and Serratia marcescens). The nitroblue tetrazolium (NBT) test is used to screen for CGD.

B cells (choice A) are responsible for antibody-mediated immunity, and the immunoglobulin levels in this patient were normal.

Chemotaxis (choice B) is important in the migration of the phagocytic cell toward the site of infection, not in intracellular killing.

IgG subclass 2 (choice C) is the most important immunoglobulin in the protection against encapsulated organisms.

T cells (choice E) are important in the host response to viruses, fungi, and intracellular bacterial organisms.

The patient was immunized normally (including the live, attenuated MMR vaccine), making T-cell dysfunction unlikely.

76) A 20-year-old healthy married Caucasian male undergoes a splenectomy following a severe motorcycle accident.

He is started on a prophylactic course of antibiotics following surgery, which he discontinues 3 months later. 9 months after the splenectomy, he presents to the emergency room with shortness of breath, pleuritic chest pain, chills and fever, and is diagnosed with lobar pneumonia. Which of the following is the most likely organism?

A. Chlamydia pneumoniae

B. Pneumocystis carinii

C. Pseudomonas aeruginosa

D. Staphylococcus aureus

E. Streptococcus pneumoniae

Explanation:

The correct answer is E. The spleen is very effective in the removal of encapsulated organisms such as Streptococcus pneumoniae, Haemophilus influenzae and Neisseria meningitidis. The capsular polysaccharides of S. pneumoniae are highly antigenic and type-specific. Type-specific anticapsular antibodies to these T-independent antigens result in effective opsonization and host recovery.

In untreated S. pneumoniae infections, recovery clearly is due to opsonizing antibody. Splenectomized patients may have low levels of IgG2 and IgM because splenic lymphocytes are the major producers of these antibodies, which provide protection against encapsulated organisms. Splenectomy therefore renders patients extremely susceptible to infections with encapsulated organisms; immunization against S. pneumoniae is therefore recommended.

Chlamydia pneumoniae(choice A) causes a community-acquired pneumonia. However, the patient has no reason for increased susceptibility to this organism (the organism is not encapsulated).

Pneumocystis carinii (choice B) is common in patients with acquired immune deficiency. A clinical and social history would quickly indicate the likelihood of such an infection. Pneumocystis is currently thought to be a fungus, although previously it was believed to be a protozoan.

Pseudomonas aeruginosa(choice C) is typically a cause of hospital-acquired pneumonia (and is not encapsulated).

Staphylococcus aureus (choice D) is typically a cause of hospital-acquired pneumonia (and is not encapsulated).

77) A Mexican immigrant presents with a 3-month history of weight loss, night sweats, a productive cough with blood-tinged sputum, anorexia, general malaise, and a low grade fever. A PPD skin test shows > 10 mm of induration. If the area of induration were biopsied, which of the following type of reactive cells would be found?

A. B lymphocyte

B. Eosinophil

C. Mast cell

D. Neutrophil

E. T lymphocyte

Explanation:

The correct answer is E. The CD4+ population of T lymphocytes, specifically TH1 cells, are responsible for the delayed hypersensitivity reaction seen with a skin test in a previously sensitized patient. The clinical pattern in the test question is classic for reactivation or adult-type tuberculosis. One of the risk factors to consider in evaluating a patient for tuberculosis is a foreign-born patient who has recently immigrated to the U.S. A 10 mm reaction on a skin test is considered positive if the person is in a high-incidence group (foreign born, medically underserved, low-income population, or resident of a long-term care facility).

B lymphocytes (choice A) are involved in humoral immune reactions. Antibody production is not a feature of tuberculosis hypersensitivity.

Eosinophils (choice B) are important in type I hypersensitivity reactions and in immune mediated responses to parasitic infections (ADCC). They are not associated with tuberculosis hypersensitivity.

Mast cells (choice C) are tissue cells which are involved in type I hypersensitivity reactions. They have surface receptors for the Fc fragment of the IgE molecule.

Neutrophils(choice D) are associated with acute inflammatory reactions. They are considered a "non-specific" cell in that they do not interact with an antigen. The neutrophil is not a classic cell type seen in tuberculosis hypersensitivity.

78) A child has a history of recurrent infections with organisms having polysaccharide antigens (i.e., Streptococcus pneumoniae and Hemophilus influenzae). This susceptibility can be explained by a deficiency of

A. C3 nephritic factor

B. C5

C. IgG subclass 2

D. myeloperoxidase in phagocytic cells

E. secretory IgA

Explanation:

The correct answer is C. IgG is the predominant antibody in the secondary immune response. IgG subclass 2 is directed against polysaccharide antigens and is involved in the host defense against encapsulated bacteria.

C3 nephritic factor (choice A) is an IgG autoantibody that binds to C3 convertase, making it resistant to inactivation. This leads to persistently low serum complement levels and is associated with Type II membranoproliferative glomerulonephritis.

C5 (choice B) is a component of the complement system. C5a is an anaphylatoxin that effects vasodilatation in acute inflammation. It is also chemotactic for neutrophils and monocytes and increases the expression of adhesion molecules. A deficiency of C5a would affect the acute inflammatory response against any microorganism or foreign substance.

Myeloperoxidase in phagocytic cells (choice D) is an element of the oxygen-dependent pathway present in phagocytic cells that effectively kills bacterial cells. The hydrogen peroxide-halide complex is considered the most efficient bactericidal system in neutrophils. Chronic granulomatous disease is associated with a deficiency of NADPH oxidase, which converts molecular oxygen to superoxide (the first step in the myeloperoxidase system). Patients are susceptible to granulomatous infections and staphylococcal infections.

Secretory IgA (choice E) is the immunoglobulin associated with mucous membranes. Selective IgA deficiency is the most common hereditary immunodeficiency. In this disorder, there is failure of the B cell to switch the heavy chain class from IgM to IgA. Patients have an increased incidence of sinopulmonary infections, diarrhea, allergies, and autoimmune diseases.

79) Which of the following is the primary opsonin in the complement system?

A. C1q

B. C3b

C. C5

D. C5a

E. Factor B

Explanation:

The correct answer is B. C3b is the most critical molecule in both the classical and alternative complement pathways. C3, the most abundant protein of all the complement proteins, is cleaved into C3a and C3b. C3b attaches to bacterial surfaces for opsonization by phagocytes. C3a binds to mast cells and basophils, activating them and producing histamine release.

C1q (choice A) is a complement component in the classical pathway. It binds to the constant heavy domain of an IgG molecule that has reacted with the bacterial surface epitope. C1q is not involved in opsonization as it simply functions as an enzyme in the early complement cascade.

C5 (choice C) is a protein, that once split into C5b, initiates the assembly of the membrane attack complex.
This complex consists of C5b, C6, C7, C8, and polymerization of C9, and is responsible for lysis of the bacteria. C5a is a strong chemotactic molecule.

This fragment is the result of C5 being split by the C5 convertases of both pathways. This C5a (choice D) is a strong chemotactic factor for neutrophils and results in stimulating the inflammatory response.

Factor B (choice E) is an activator protein of the alternative pathway. It combines with C3b to form C3bBb. This C3bBb is the C3 convertase of the alternative pathway.

80) A 27-year-old woman presents to the emergency department complaining of wheezing and swelling of the lips. The symptoms began acutely just after eating a chicken and nut dish at a nearby Chinese restaurant. She also complains of tightness in her chest and difficulty breathing. On examination, she has a temperature of 36.8 C, a heart rate of 110/min, a blood pressure of 90/50 mm Hg, and a respiratory rate of 24/min. She is anxious and in mild respiratory distress. There is diffuse facial erythema, with swelling of the lips and tongue. Wheezing is noted bilaterally, and oxygen saturation is 92%. Which of the following cytokines is necessary for production of the antibody that mediates this response?

A. Interferon-γ

B. Interleukin 1

C. Interleukin 2

D. Interleukin 3

E. Interleukin 4

F. Interleukin 5

G. Interleukin 6

Explanation:

The correct answer is E. This is a case of immediate (type I) hypersensitivity mediated by IgE, mast cells, and basophils. The cytokine that mediates the isotype switch to IgE in B lymphocytes is interleukin 4, produced by T-helper-2 cells.

Interferon-γ(choice A) is a cytokine that activates macrophages and would not be involved in this response.

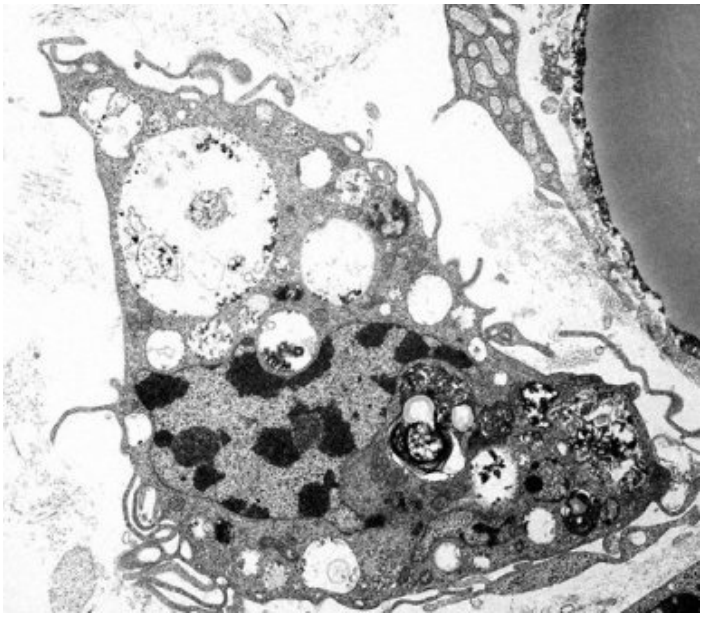
Interleukin 1 (choice B) is an endogenous pyrogen involved in the production of the acute phase reaction of systemic inflammation.

Interleukin 2 (choice C) is the cytokine that induces T cell proliferation.

Interleukin 3 (choice D) is a hematopoietic colony stimulating factor.

Interleukin 5 (choice F) is an eosinophil stimulating factor.

Interleukin 6 (choice G) is another endogenous pyrogen and a stimulator of B cells, but not an inducer of isotype switch to IgE production.



81) Which of the following characteristics most strongly suggests that the cell depicted above is a phagocytically active macrophage?

A. Immunocytochemical detection of collagenase

B. Microvilli-covered surface

C. Presence of receptors for IgG and complement

D. Presence of secondary lysosomes throughout cytoplasm

E. Shape of the nucleus

Explanation:

The correct answer is C. Macrophages interact with a segment of the heavy chains of the IgG molecule, the Fc domain. Thus, the macrophage receptors for IgG are called Fc receptors. Complement receptors are also found on the macrophage surface. Phagocytosis occurs more readily when both of these components are present.

Neutrophils and eosinophils have similar receptors: neutrophils assist macrophages in destruction of bacteria,

while eosinophils respond to parasitic infections.

Collagenase can be demonstrated immunologically (choice A) in a number of cells including eosinophils, mast cells, osteoclasts, and fibroblasts.

Microvilli (choice B) or pseudopodial extensions are common in macrophages, but also may be found on other cells such as fibroblasts and epithelial cells that phagocytize material.

Secondary lysosomes (choice D) are a feature of macrophages, but may also be found in fibroblasts and epithelial cells.

The shape of the nucleus (choice E) has little to do with phagocytic activity.

82) A 3-year-old male presents with a skin rash and epistaxis. He has had several, severe sinopulmonary infections.

A careful history reveals that his maternal uncle died of bleeding complications following an emergency cholecystectomy. What additional findings are likely in this case?

A. A CD4/CD8 ratio of < 1.5:1

B. Cerebellar ataxia

C. Elevated platelet count and high serum IgG, IgA, and IgE levels

D. Low platelet count and low serum IgG levels

E. Low platelet count and low serum IgM levels

Explanation:

The correct answer is E. This clinical scenario is typical for Wiskott-Aldrich syndrome. This is a sex-linked recessive disorder presenting with the triad of thrombocytopenia, eczema, and recurrent sinopulmonary infections. Serum IgM levels are low but IgG, IgA, and IgE levels are increased. Patients have a defective response to polysaccharide antigens, which is due to a cytoskeletal defect in T cells that inhibits their binding to B cells.

A CD4/CD8 ratio of < 1.5:1 (choice A) is the pattern seen in AIDS due to selective tropism of the CD4+ T-helper cell population.

Cerebellar ataxia (choice B) is part of the ataxia-telangiectasia syndrome. The ataxia develops between age 2 and 5. The defect is associated with a DNA repair enzyme deficiency.

Elevated platelet count and increased serum levels of IgG, IgA, and IgE (choice C) is not associated with a particular syndrome. Polyclonal gammopathies result in an increase in immunoglobulin of more than one class.

This benign alteration is frequently seen in viral or bacterial infections. Thrombocythemia may be associated with a myeloproliferative syndrome or with a secondary reactive process.

Low platelet count and low serum IgG (choice D) are not associated with any particular disorder.

83) A 4-year-old child presents to a physician with purpura. Questioning of the mother reveals that the child also has a history of eczema and recurrent pneumococcal pneumonia. Blood studies demonstrate thrombocytopenia and a selective decrease in IgM. Patients with this disorder have a 12% chance of developing which of the following potentially fatal malignancies?

A. Astrocytoma

B. Chronic myelogenous leukemia

C. Colon carcinoma

D. Non-Hodgkin's lymphoma

E. Wilms' tumor

Explanation:

The correct answer is D. The patient has Wiskott-Aldrich syndrome, which is an X-linked partial combined immunodeficiency disorder. It presents clinically as the triad of thrombocytopenic purpura, eczema, and recurrent opportunistic infections with organisms having polysaccharide capsules, such as Streptococcus pneumoniae. The patients characteristically have decreased serum IgM and T-cell deficits (which appear later in life). Most patients die before the second decade of life, either from infection or from non-Hodgkin's lymphoma. Bone marrow transplantation has had some therapeutic success in these patients.

None of the other neoplasms listed occurs with greater frequency in patients with Wiskott-Aldrich syndrome.

84) A 20-year-old college student working part-time in a pediatric AIDS clinic develops a viral exanthem with a rash. Her blood is drawn and tested for specific antibodies to varicella-zoster (chickenpox). Anti-varicella immunoglobulin belonging to which of the following antibody classes would indicate that she is immune to chickenpox?

A. IgA

B. IgD

C. IgE

D. IgG

E. IgM

Explanation:

The correct answer is D. The immunoglobulin that represents past exposure to an antigen is IgG. It is the predominant antibody in the secondary response and is an important defense against bacteria and viruses. In the secondary response, a much larger amount of IgG antibody is produced than IgM, and the levels of IgG tend to persist for a much longer time than in the primary response.

IgA (choice A), the main immunoglobulin found in secretions, prevents the attachment of bacteria and viruses to mucosal surfaces. It does not bind complement.

IgD (choice B) probably functions as an antigen receptor. It is found on the surface of many B lymphocytes and is found in small amounts in serum.

IgE (choice C) is the primary antibody in type I hypersensitivity. It mediates the anaphylactic response and also participates in host defenses against parasites (antibody-dependent cell-mediated cytotoxicity-ADCC).

IgM (choice E) is the chief immunoglobulin produced in the primary response to an antigen. It is also produced in the secondary response, but in lesser amounts than IgG. It exists as a pentamer, which affords a high degree of avidity of binding to antigens. It is produced by the fetus in certain infections, but does not cross the placenta. It does bind complement.

85) A 12-year-old girl has a temperature of 102.5 F and a sore throat. Two days later, she develops a diffuse erythematous rash and is taken to her pediatrician. On physical examination, there is circumoral pallor, and anerythematous rash with areas of desquamation is noted. The myocardial damage that can follow this infection is produced in a manner similar to the damage associated with which of the following disorders?

A. Atopic allergy

B. Contact dermatitis

C. Graft-vs.-host disease

D. Graves disease

E. Idiopathic thrombocytopenic purpura

F. Myasthenia gravis

G. Rheumatoid arthritis

H. Serum sickness

I. Systemic lupus erythematosus

Explanation:

The correct answer is E. This is a case of rheumatic fever, which is an immunologically mediated sequela to Streptococcus pyogenes pharyngitis. It is a type II cytotoxic hypersensitivity, involving antibodies that bind to cardiac tissue, activate complement, and thereby cause cell destruction. It is therefore most similar to idiopathic thrombocytopenic purpura, which is also a form of type II cytotoxic hypersensitivity, in this case mediated by antibodies against platelets producing complement fixation and causing the clotting dyscrasia.

Atopic allergy (choice A) is a form of type I hypersensitivity, mediated by IgE antibodies and basophils and mast cells.

Contact dermatitis (choice B) is a form of type IV hypersensitivity mediated by T cells and macrophages.

Graft-vs.-host disease (choice C) is a form of type IV hypersensitivity mediated by T cells and macrophages.

Graves disease (choice D) is a form of type II hypersensitivity, but it is NOT cytotoxic in its action. Instead, antibodies to the TSH receptors on thyroid cells cause overstimulation of the gland and its eventual exhaustion.

Myasthenia gravis (choice F) is a form of type II hypersensitivity, but NOT of the cytotoxic variety. In this case,antibodies to the acetylcholine receptors on neurons diminish neurotransmission.

Rheumatoid arthritis (choice G) is a form of type III hypersensitivity, caused by immune complex deposition in joints and subsequent activation of complement.

Serum sickness (choice H) is a form of type III hypersensitivity, caused by immune complex deposition.

Systemic lupus erythematosus (choice I) is a form of type III hypersensitivity, caused by immune complex deposition.

86) A 32-year-old medical technician had a history of acute eczematous dermatitis on her hands and wrist in the distribution of the latex gloves she wore. The skin of her hands was dry, crusted, and thickened. The eczematous reaction cleared after a 2-week vacation. After 72 hours back on the job, the eczematous dermatitis returned and continued to grow worse. Which of the following characterizes the technician's reaction to the latex gloves?

A. Irritant dermatitis

B. Type I reaction

C. Type II reaction

D. Type III reaction

E. Type IV reaction

Explanation:

The correct answer is E. Sensitization to latex has become a major healthcare problem. Local skin irritations are common but more severe allergic reactions occur, up to and including rare anaphylactic reactions that are occasionally fatal. The immune responses to latex are immediate-type hypersensitivity (type I) reactions, expressing themselves in minutes, and/or delayed-type hypersensitivity (type IV) reactions, which will express themselves in 48-72 hours. The type I reactions are due to the IgE-mediated sensitivity to latex proteins while the type IV reactions are due to a cell-mediated response to the chemicals that are added in the processing of latex. The type IV response in this circumstance would be referred to as contact dermatitis.

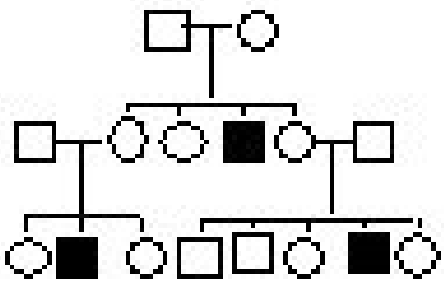
Irritant dermatitis (choice A) can be observed in the early stages of sensitization and can be due to sweating, rubbing, and residual soap. The timing of the reaction after 72 hours points to the type IV reaction rather than an irritant dermatitis.

A type I reaction (choice B) would have been apparent in minutes with characteristic rhinitis, conjunctivitis, urticaria, asthma, angioedema, or anaphylaxis after she put on her latex gloves.

A type II reaction (choice C) is a cytolytic response mediated by an antibody. The type II reaction is observed in the context of hemolytic anemias, thrombocytopenia, neutropenia, etc.

A type III reaction (choice D) is an immune-complex response that sets into motion an inflammatory response.

87) PEDIGREE



A couple brings their son in to a specialty clinic for evaluation of recurrent bacterial infections involving the respiratory tract. Other family members have a similar disorder, as noted in the pedigree above. Which of the following is the most likely diagnosis?

A. Bruton's agammaglobulinemia

B. Common variable immunodeficiency

C. DiGeorge syndrome

D. Hereditary angioedema

E. Isolated IgA deficiency

Explanation:

The correct answer is A. Several immunodeficiency disorders have X-linked genetics, including Bruton's agammaglobulinemia, Wiskott-Aldrich syndrome, and some cases of severe combined immunodeficiency diseases. Bruton's agammaglobulinemia is characterized by recurrent respiratory infections caused by pyogenic organisms.

Common variable immunodeficiency (choice B) is a relatively common, but probably heterogeneous, group of acquired and familial diseases. It is distinct from X-linked (Bruton's) agammaglobulinemia.

DiGeorge syndrome (choice C) is due to a developmental malformation of the third and fourth pharyngeal pouches leading to failure of the thymus, and sometimes the parathyroids, to develop. Defective cellular immunity and abnormalities of calcium metabolism are typical.

Hereditary angioedema (choice D) is a usually recessive genetic disease caused by deficiency of C1 esterase inhibitor.

Isolated IgA deficiency (choice E) can be acquired or genetic, but is not usually X-linked.

88) Which of the following cytokines is associated with the development of cell-mediated immunity?

A. IL-3

B. IL-4

C. IL-5

D. IL-10

E. IL-I2

Explanation:

The correct answer is E. Gamma interferon from T helper 1 CD4 + lymphocytes and IL-12 from macrophages are major stimulators of the development of a cell-mediated immune response to intracellular organisms such as tuberculosis, leishmaniasis, histoplasmosis, and many other organisms.

IL-3 (choice A) is a cell growth factor produced by thymic epithelial cells and T cells to regulate different T cell differentiation processes.

IL-4 (choice B) is produced by T helper 2 CD4 + lymphocytes and is involved in class switching to IgE. It also downregulates T helper 1 CD4 + lymphocytes.

IL-5 (choice C) is produced by T helper 2 CD4 + lymphocytes and is involved in IgA production, eosinophil differentiation, and B cell differentiation.

IL-10 (choice D) is produced by T helper 2 CD4 + lymphocytes. It downregulates the cell-mediated immune system.

89) A 25-year-old female presents with pain and tenosynovitis of the wrists and ankles, and arthralgias of other joints. She notes two prior episodes similar to the present one. She just had her menstrual period during the previous week. Physical examination reveals ulcerated lesions overlying the wrists and ankles. These symptoms are likely due to deficiency of which of the following?

A. C1 esterase inhibitor

B. Ciliary function

C. Complement (C6-C8) components

D. Endothelial adhesion molecules

E. Eosinophils

Explanation:

The correct answer is C. This patient has disseminated gonococcemia. Gonococcal arthritis and tenosynovitis typically involve both the upper and lower extremities equally. Vesicular skin lesions are characteristic of disseminated gonococcal disease. Females are at particular risk of gonococcemia during menstruation, since sloughing of the endometrium allows access to the blood supply, necrotic tissue enhances the growth of Neisseria gonorrhoeae, and there is an alteration of the pH. Patients who have a C6-C8 deficiency have an increased risk of disseminated gonococcemia and tend to have multiple episodes. These patients are also at risk for bacteremia from Neisseria meningitidis.

C1 esterase inhibitor deficiency (choice A) can occur as an autosomal dominant disorder or is acquired. Patients have angioedema without urticaria. The syndrome is also associated with recurrent attacks of colic and episodes of laryngeal edema.

Ciliary dysfunction (choice B) is a marker of Kartagener's syndrome (immotile cilia syndrome). The syndrome includes infertility, bronchiectasis, sinusitis, and situs inversus. It is an autosomal recessive disorder caused by abnormalities in the dynein arm of the cilia.

Endothelial adhesion molecule deficiency (choice D), or beta2 integrin deficiency, is characterized by failure of neutrophils to express CD18 integrins on their surface. Patients have impaired phagocyte adherence, aggregation, chemotaxis, and phagocytosis of C3b-coated particles. Clinically, there is delayed separation of the umbilical cord, sustained agranulocytosis, recurrent infections of skin and mucosa, gingivitis, and periodontal disease.

Eosinophil deficiency (choice E) or eosinopenia occurs with stressors such as acute bacterial infection and following administration of glucocorticoids. There is no known adverse effect of eosinopenia.

90) A pregnant 38-year-old mother of two presents at the office concerned about her pregnancy. She is Rh-negative and her husband is Rh-positive. Both of her children are also Rh-negative. She has had two spontaneous abortions and carried a third pregnancy to term, but the child died at birth, diagnosed with erythroblastosis fetalis. The test of choice to determine the presence of circulating anti-Rh antibody in the mother is a(n)

A. direct Coombs test to measure IgG anti-Rh antibody

B. direct Coombs test to measure IgM anti-Rh antibody

C. indirect Coombs test to measure IgG anti-Rh antibody

D. indirect Coombs test to measure IgM anti-Rh antibody

E. Ouchterlony test to measure precipitin IgG anti-Rh antibody

Explanation:

The correct answer is C. The indirect Coombs test would be the test of choice to detect the presence of IgG (warm agglutinin) anti-Rh antibody circulating in the mother's blood. Serum is collected from the mother and mixed with Rh-positive red blood cells, followed by incubation with complement. If the mother had circulating IgG anti-Rh, it would bind to the Rh antigen on the Rh-positive red blood cells, fixing the complement and lysing the target cells. A titer of greater than or equal to 1:8 is considered diagnostic.

The direct Coombs test (choices A and B) would be used to determine if anti-IgG or anti-C3 antisera can agglutinate the baby's RBCs. This would not be useful for determining whether circulating IgG antibody to the Rh factor is present in the serum from the mother.

An indirect Coombs test to measure IgM anti-Rh antibody (choice D) would not be informative, since IgM anti-Rh antibody will not cross the placenta.

The Ouchterlony test, or double-diffusion precipitin reaction (choice E), is used to determine if two antigens are identical, unrelated, or related but not identical.

91) An IgG2 molecule is composed of which of the following?

A. One alpha, one gamma2, and two kappa chains

B. One gamma1 chain and two kappa chains

C. Two gamma1 chains and one kappa and one lambda chain

D. Two gamma1 chains and two kappa chains

E. Two gamma2 chains and two kappa chains

Explanation:

The correct answer is E. IgG molecules contain two gamma heavy chains of a given subtype and two light chains (either kappa or lambda). The 2 in IgG2 indicates the subclass to which the molecule belongs. IgG2 contains two gamma2 chains (since a given B cell can only form one type of heavy chain).

The IgG molecule will contain either two kappa chains or two lambda chains, but never one of each (choice C).

A given cell produces immunoglobulin molecules with a single type of heavy chain (compare with choice A).

IgG molecules with gamma1 chains (choices B, C, and D) would be of the IgG1 subclass.

92) A 42-year-old Nigerian in the United States visiting his brother comes to the clinic with complaints of several months of weight loss, night sweats, sputum production, and occasional spitting up blood. His physician places a tuberculin skin test and the results are positive. This positive tuberculin test indicates which of the following?

A. A cell-mediated immune response has occurred

B. A humoral immune response to TB is positive

C. The B and T cell systems are functional

D. The B cell system is functional

E. The phagocytic neutrophilic system is functional

Explanation:

The correct answer is A. The tuberculin skin test is a delayed type hypersensitivity test. This type IV hypersensitivity is a cell-mediated immune response involving CD4 T helper 1 (TH1) lymphocytes. This individual is TB positive, and should now have cultures and a chest x-ray to confirm an active case of tuberculosis.

Elimination of tuberculosis, like all intracellular bacterial infections, is not dependent on a humoral (antibody) response (choice B). This patient could have antibody to tuberculosis, but recovery is entirely dependent on cell-mediated immunity.

The positive tuberculin test is a result of functioning CD4 TH1 cells, but does not depend on a functional B cell system (compare with choice C).

The B cell is involved in the production of immunoglobulins. This disease is dependent on cell mediated (T cell immunity) and not B cell production of immunoglobulins (antibody) (compare with choice D).

The positive tuberculin test does not involve phagocytic neutrophils (compare with choice E). The primary cell involved is the CD4 TH1 lymphocyte.

93) A 6-year-old boy with a sore throat, cough, and fever is noted to have a marked elevation in his serum C-reactive protein. This finding indicates which of the following?

A. Developing autoimmune reaction

B. Ineffective immune response

C. Meningitis

D. Non-specific inflammation

E. Respiratory compromise

Explanation:

The correct answer is D. C-reactive protein is one of the most commonly measured acute-phase reactants, which are a group of serum proteins showing a rapid increase in serum concentration in response to any inflammatory process. This finding is entirely non-specific–it only indicates a recent inflammatory process.

An autoimmune reaction (choice A), which is certainly a concern with streptococcal pharyngitis, is suggested by the development of a rising ASO (antistreptolysin O) titer weeks after the illness. Acute phase reactants are not specific to autoimmune processes.

Increases in C-reactive protein indicate a healthy immune response to an infective pathogen. An ineffective immune response (choice B) would not elicit acute-phase reactions.

Meningitis (choice C) cannot be diagnosed from a blood test. The diagnosis of meningitis requires the appropriate clinical signs (neck stiffness, mental status changes) and a positive lumbar puncture.

Respiratory compromise (choice E) produces changes in arterial blood gases and blood pH. Acute-phase reactants do not reflect respiratory status.

94) Which of the following substances takes part in the nonoxidative killing pathway by which pathogenic bacteria can be killed?

A. Hydrogen peroxide

B. Hypochlorous acid

C. Lysozyme

D. Myeloperoxidase

E. Superoxide ions

Explanation:

The correct answer is C. Lysozyme is present in tears, saliva, mucus, vaginal secretions, and several other body fluids. This material lyses the peptidoglycan layer of the cell wall of bacteria without participation of any of the elements of the oxidative killing pathway.

Hydrogen peroxide (choice A) is formed in the oxidative killing pathway from the superoxide ions. This material is also very toxic to bacteria pathogens.

Hypochlorous acid (choice B) is formed from hydrogen peroxide and halide ions in the presence of myeloperoxidase.

Myeloperoxidase (choice D) is an enzyme present in the oxidative killing pathway that breaks down hydrogen peroxide.

Superoxide ions (choice E) are formed by NADPH oxidase in the first reaction of the oxidative killing pathway. These superoxide ions are extremely toxic to bacterial organisms.

95) An alert pediatric intern notices that a neonate with dysmorphic facies is twitching abnormally. As he watches, the baby experiences a seizure. Stat laboratories indicate a glucose of 90, serum sodium of 140, serum potassium of 4.2 and serum calcium of 3.9. Over the next several months, the child is admitted to the hospital twice for Candida infections, and once for a viral exanthem. Which of the following is the most likely diagnosis?

A. Ataxia telangiectasia

B. Bruton's hypogammaglobulinemia

C. DiGeorge syndrome

D. Severe combined immunodeficiency

E. Wiskott-Aldrich syndrome

Explanation:

The correct answer is C. DiGeorge syndrome is due to fetal malformation of epithelial elements of the third and fourth pharyngeal pouches, leading to thymic aplasia or severe hypoplasia and sometimes, absence of the parathyroids. Clinically, the condition is classified as a selective T-cell deficiency, but varying degrees of deficiency of antibody production may also be seen. Affected infants are susceptible to fungal and viral infections, and may exhibit tetany due to severe hypocalcemia.

In autosomal recessive ataxia telangiectasia (choice A), progressive cerebellar ataxia is accompanied by multiple telangiectasias on exposed skin and a variable, usually moderate, immunodeficiency that may involve both antibody production and cell-mediated immunity. Chronic or recurrent sinus and pulmonary infections result in bronchiectasis.

In Bruton's (X-linked) agammaglobulinemia (choice B), children have normal numbers of circulating T cells with very few circulating B cells and only tiny amounts of circulating antibody. Affected individuals have recurrent bacterial infections beginning late in the first year of life.

Severe combined immunodeficiency (choice D) is actually a cluster of several diseases with variable genetics characterized by severely deficient T cell functions and variable (often depressed) antibody production. Multiple, sometimes simultaneous, infections with viruses, bacteria, and fungi occur.

Wiskott-Aldrich syndrome (choice E) is an X-linked disease with thrombocytopenia, lymphopenia, and decreased T cell function. Lymphoid malignancy (e.g., acute lymphocytic leukemia) may occur.

96) A 38-year-old woman with systemic lupus erythematosus (SLE) is seen by her family physician. On her last visit, he sent blood to the laboratory for an antinuclear antibody panel. Based on the results, he recommends that the patient be seen by a nephrologist. A high titer of autoantibodies directed against which of the following antigens most likely prompted his decision?

A. Double-stranded DNA (dsDNA)

B. Histones

C. Ribonucleoprotein (RNP)

D. Smith antigen (Sm)

E. Single-stranded DNA (ssDNA)

Explanation:

The correct answer is A. Antibodies to double-stranded DNA, unlike most other autoantibodies, are considered to have prognostic significance in SLE. High titers of this antibody are associated with the development or progression of lupus nephritis. Lupus nephritis can occur in the absence of any demonstrable abnormalities on urinalysis; renal biopsy is helpful in such cases to document disease progression.

Anti-histone antibodies (choice B) can be seen as one of several autoantibodies in SLE or as an isolated finding in drug-induced lupus.

High titers of anti-RNP (choice C) specifically suggests mixed connective tissue disease rather than lupus, although 30-40% of lupus patients will have this antibody.

Anti-Sm (choice D) is specific for SLE, but does not predict outcome.

Anti-ssDNA autoantibodies (choice E) are a nonspecific feature of SLE and can be seen in other autoimmune diseases.

97) A 28-year-old mother gives birth to her first child. The father is homozygous Rh D positive and the mother is homozygous Rh D negative. The baby is born without any complications, but the mother is not given RhoGAM (anti-Rh IgG) following the delivery. Eighteen months later she delivers another child, who is anemic, slightly jaundiced, and has an enlarged spleen and liver. Which type of hypersensitivity best describes this condition?

A. Atopic disease

B. Cytotoxic disease

C. Delayed hypersensitivity

D. Immediate hypersensitivity

E. Immune complex disease

Explanation:

The correct answer is B. This is an example of type II hypersensitivity, in which antibody is produced against cells or receptors in the body. In this type of cytotoxic disease, the mother produced antibody against the Rh D positive cells to which she was exposed upon delivery of her first child. This child must be Rh D positive,
because the father is homozygous Rh D positive. The Rh D cells are foreign to the mother since she is Rh D negative, and so she mounted an antibody response against the antigen. The second child also must have been Rh D positive, and the mother's antibody crossed the placenta resulting in the disease.

Atopic disease (choice A) is a Type I immediate hypersensitivity reaction that occurs in minutes following a second exposure to the offending allergen. The antigen could be ragweed, tree pollen, grass pollen, animal fur, or food products.

Type IV hypersensitivity is delayed type hypersensitivity (choice C), developing in response to viral, fungal, and intracellular bacterial antigens. It involves CD4 T helper lymphocytes, but not antibody.

Immediate hypersensitivity (choice D) is a Type I reaction that occurs in minutes following a second exposure to the offending allergen. Common antigens include ragweed, tree pollen, grass pollen, animal fur, or food products.

Immune complex disease (choice E) is a Type III hypersensitivity reaction. It involves antibody and complement,
but in the form of antigen-antibody-complement complexes. Rheumatoid arthritis, systemic lupus erythematous, and serum sickness are examples of type III hypersensitivity

98) A 21-year-old man presents with cough, fever, and hemoptysis. Blood tests show significantly elevated BUN and creatinine. Immunofluorescent microscopy reveals a diffuse linear pattern of fluorescence along the basement membranes of alveolar septa and glomerular capillaries. Which type of hypersensitivity is associated with this disease?

A. Type I

B. Type II

C. Type III

D. Type IV

Explanation:

The correct answer is B. This patient has a USMLE-favorite disease: Goodpasture's syndrome, which affects both the renal and pulmonary systems. In the kidney, it causes a rapidly progressive glomerulonephritis caused by antibodies directed against a collagen component of the glomerular basement membrane (anti-GBM antibody––a classic clue to this diagnosis). These antibodies create a linear pattern on immunofluorescence. Note that they are also active against the basement membrane of respiratory alveoli, accounting for the pulmonary component of the disease.

Autoimmune reactions such as those found in Goodpasture's, certain drug allergies, blood transfusion reactions, and hemolytic disease of the newborn are classified as Type II hypersensitivities (antibody-mediated cytotoxicity). IgG or IgM antibody reacts with membrane-associated antigen on the surface of cells, causing activation of the complement cascade and, ultimately, cell destruction.

Type I (choice A) (immediate, atopic, or anaphylactic) reactions require an initial (sensitizing) exposure to an antigen. Upon re-exposure to the antigen, cross-linking of IgE receptors occurs on the surface of basophils and mast cells. The mast cells then release a variety of mediators, including histamine. Clinical syndromes include asthma, atopic dermatitis, eczema, and allergic rhinitis. Hives ("wheal and flare") are characteristic of Type I hypersensitivity.

Type III (choice C) hypersensitivity (immune complex-mediated hypersensitivity) is caused by antibodies to foreign antigens. Immune complexes of IgG or IgM with the antigen activate complement, resulting in the generation of C3b, which promotes neutrophil adherence to blood vessel walls. The complexes also generate C3a and C5a (anaphylatoxins), which lead to inflammation and tissue destruction. The hallmark signs of Type III sickness occur 7-14 days after exposure to the offending antigen and include urticaria, angioedema, fever, chills, malaise, and glomerulonephritis. Clinical syndromes include serum sickness (e.g., penicillin, streptomycin, sulfonamide, phenylbutazone hypersensitivity) and the Arthus response. Immune complexes are also observed in lupus (SLE). Type III glomerulonephritis (e.g., poststreptococcal glomerulonephritis) is characterized by a "lumpy bumpy" appearance on immunofluorescence using tagged antibody specific for immunoglobulin or complement.

Type IV (choice D) is also known as delayed-type hypersensitivity (DTH). Unlike the other types, which are mediated by antibody, DTH depends on TDTH cells that have been sensitized to a particular antigen. T cells react with antigen in association with MHC class I gene products and release lymphokines. Examples include tuberculin skin sensitivity and contact dermatitis (e.g., poison ivy rash).

99) Which of the following enzymes does the neutrophil use to initiate the production of toxic oxygen compounds that kill bacteria?

A. Hydrogen peroxide

B. Myeloperoxidase

C. NADPH oxidase

D. Superoxide

E. Superoxide dismutase

Explanation:

The correct answer is C. The first step in killing bacteria is the production of superoxide ion, O2- , by the action of NADPH oxidase on NADPH and O2. The superoxide is then converted to hydrogen peroxide, either spontaneously or through the action of superoxide dismutase. The hydrogen peroxide can also be converted to the toxic HOCl. radical by the action of myeloperoxidase.

Hydrogen peroxide(choice A) is not an enzyme.

Myeloperoxidase (choice B) is part of the killing pathway, but is active later in the sequence, when H2O2 is converted in the presence of Cl- to the HOCl. radical.

Superoxide (choice D) is not an enzyme.

Superoxide dismutase (choice E) is part of the killing pathway, but is active later in the sequence, whent superoxide is converted to hydrogen peroxide.

100) Liver biopsy in a patient with cirrhosis demonstrates markedly increased iron stores. The patient has no known history of hemolytic anemia or other potential causes of a secondary increase in hepatic iron stores. This patient's primary disorder is most strongly associated with which of the following HLA alleles?

A. HLA-A3

B. HLA-B27

C. HLA-DR2

D. HLA-DR3

E. HLA-DR4

Explanation:

The correct answer is A. The disease is primary hemochromatosis, in which abnormal iron accumulation occurs in many different organs. The liver may become cirrhotic, and the patient may become diabetic due to pancreatic damage. Heart failure may occur due to myocardial dysfunction, and the skin can take on a bronze
discoloration. In fact, the old term for hemochromatosis was "bronze diabetes." Primary hemochromatosis is associated with the HLA-A3 allele.

HLA-B27 (choice B) is associated with psoriasis, ankylosing spondylitis, inflammatory bowel disease, and Reiter's syndrome.

HLA-DR2 (choice C), together with HLA-DR3 (choice D), is associated with systemic lupus erythematosus.

HLA-DR3 (choice D) is additionally associated with Sjögren's syndrome, chronic active hepatitis, and type 1 diabetes mellitus (with HLA-DR4, choice E).

In addition to being associated with type 1 diabetes, HLA-DR4 (choice E) is also associated with rheumatoid arthritis.

Practice Questions & Answers

1. Which of the following features is not shared between ‘T cells’ and ‘B cells’?
a) Antigen Specific Receptors
b) Class I MHC Expression
c) Positive selection during development
d) All of the above

2. CD4 is not important for which of the following?
a) Antibody production
b) Cytotoxicity of T cells
c) Memory B cells
d) Opsonisation
e) None

3. Type 1 MHC presents peptide antigen to T cell, so that peptide binding site is formed by?
a) Alfa and Beta chain
b) Distal domain alfa 1 and 2
c) Alfa and Beta microglobulin
d) Proximal domain alfa 1 and 2

4. Function of CD4 is all except:
a) Memory
b) Immunoglobulin production
c) Activation of macrophages
d) Cytotoxicity

5. A super-antigen is a bacterial product that
a) Binds to B7 and CD28 co-stimulatory molecules
b) Binds to the beta chain of TCR and MHC class II molecules of APC stimulating T cell activation
c) Binds to the CD4+ molecule causing T cell activation
d) Is presented by macrophages to a larger than normal number of T helper CD4+ lymphocytes

6. Memory T cells can be identified by using the following marker:
a) CD45RA
b) CD45RB
c) CD45RC
d) CD45RO

7. All of the following statements about NK cells are true except:
a) They are derived from large granular cells
b) They comprise about 5% of human peripheral lymphoid cells
c) They are MHC restricted cytotoxic cells
d) They express IgG Fc receptors

1) C 2) E 3) B 4) B 5) B 6) D 7) C

8. The following feature is common to both cytotoxic T-cells and NK cells:
a) Synthesize antibody
b) Require antibodies to be present for action
c) Effective against virus infected cells
d) Recognize antigen in association with HLA class II markers

9. MHC restriction to antigen presentation is not done for:
a) Killing of viruses by cytotoxic cells
b) Killing of bacteria by helper cells
c) T cell activation in autoimmunity
d) Graft rejection

10. Most potent stimulator of naïve T cell is:
a) Mature dendritic cell
b) Follicular dendritic cell
c) Macrophages
d) B cell

11. Natural killer cells attacks which of the following cells:
a) Cells which express MHC1
b) Cells which are not able to express MHC1
c) MHC cells which express MHC2
d) Cells which are not able to express MHC

12. Toll like receptors, recognize bacterial products and stimulates immune response by:
a) Perforin and granzyme mediated apoptosis
b) FADD ligand apoptosis
c) Transcription of nuclear factor mediated by N-FxB which recruits cytokines
d) Cyclin

13. The following interleukin is characteristically produced in a TH1 response:
a) IL-2
b) IL-4
c) IL-5
d) IL-10

14. CD-95 has a major role in:
a) Apoptosis
b) Cell necrosis
c) Interferon activation
d) Proteolysis

8) C 9) D 10) A 11) B 12) C 13) A 14) A

15. Which of the following chemical mediators of inflammation is an example of a C-X-C or alpha Chemokine?
a) Lipoxin LXA4
b) Interleukin IL-8
c) Interleukin IL-6
d) Monocyte Chemo-attractant protein MCP-1

16. The complement is fixed best by which of the following immunoglobulins:
a) IgG
b) IgM
c) IgA
d) IgD

17. Antigen presenting cells are which of the following:
a) Astrocytes
b) Endothelial cells
c) Epithelial cells
d) Langerhan's cells

18. Antigen presenting cells are:
a) Langerhan's cell
b) Macrophage
c) Cytotoxic T cells
d) Hlper T cells
e) B-lymphocyte

19. Perforins are produced by:
a) Cytotoxic T cells
b) Suppressor T cells
c) Memory helper T cells
d) Plasma cells
e) NK cells

20. Cell surface molecules involved in peripheral tolerance induction are:
a) B7 and CD28
b) CD40 and CD40L
c) CD34 and CD51
d) B7 and CD3

21. Marker for B-Lymphocyte:
a) CD34
b) CD33
c) CD19
d) CD20
e) CD22

15) B 16) B 17) BCD 18) A BE19) A 20) A 21) CDE

23. Antigen presenting cells present in skin are called
a) Langerhan's cells
b) Kupffer's cells
c) Microglia
d) Melanocytes

24. Plasma cells
a) Contain nucleus
b) Helps in the formation of antibody
c) Are deficient in cytoplasm
d) Are derived from T-cells

25. The normal ratio of CD4 to CD8 is
a) 1:1
b) 2:1
c) 8:1
d) 10:1

26. CD4 cells is used to identify which of the following
a) MHC I
b) MHC II
c) T cells
d) B cells

27. CD3 is marker for:
a) Monocyte
b) T cell
c) B cell
d) None

28. Which of the following is not true about innate immunity?
a) It is present prior to antigenic exposure
b) It is relatively non-specific
c) Memory is seen
d) It is the first line of defence

29. Which one of the listed receptors is the type of receptor on leukocytes that binds to pathogen-associated molecular patterns (PAMPs) and mediates immune response to bacterial lipopolysaccharide?
a) Cytokine
b) Receptor
c) G-protein-coupled receptor
d) Mannose receptor
e) Toll-like receptor

23) A 24) B 25) B 26) C 27) B 28) C 29) D

30. Immunity against cancer cells:
a) Basophils
b) Eosinophils
c) NK cells
d) Neutrophils

31. NK cell express:
a) CD15, CD55
b) CD16, CD56
c) CD16, CD57
d) CD21, CD66

32. NK cell CD marker is:
a) 16
b) 60
c) 32
d) 25

33. The following interleukin is characteristically produced in a TH1 response?
a) IL-2
b) IL-4
c) IL-5
d) IL-10

34. Most potent stimulator of Naïve T-cells:
a) Mature dendritic cells
b) Follicular dendritic cells
c) Macrophages
d) B-cell

35. Which of the following immune cells have the expression of CD8 on their surface?
a) T-cells
b) B-cells
c) Null cells
d) Macrophages

36. Kupffer cells are found in:
a) Heart
b) Lungs
c) Liver
d) Spleen

30) C 31) B 32) A 33) A 34) A 35) A 36) C

37. Birbeck granules are present in:
a) Merkel cell
b) Langerhan's cell
c) Langhan's cell
d) Melanocyte

38. Macroglobulin is derived from:
a) B cells
b) T cells
c) Both
d) Natural killer cells

39. Which of the following is not true regarding Ig# antibodies?
a) It mediates release of histamine and other chemical mediators
b) It is the primary antibody involved in allergic reactions
c) It is involved in anti-parasitic immune responses
d) May cross the placenta and fix complement

40. Which of the following immunoglobulin does bot fix complement?
a) IgA
b) IgG
c) IgM
d) IgE

41. B cells are located in which region of lymph nodes:
a) Paracortical region
b) Cortical follicles
c) Medullar sinuses
d) Subcapsular region

42. Plasma cells produce specific antibodies by
a) Isotope selection
b) Class selection
c) Isotope selection
d) Clonal selection

43. Surface Immunoglobulin is found in which cell?
a) T-cell
b) B-cell
c) NK cell
d) Plasma cell

37) B 38) A 39) D 40) D 41) B 42) D 43) B

44. MHC class III genes encode:
a) Complement component C3
b) Tumor necrosis factor
c) Interleukin 2
d) Beta 2 microglobulin

45. The HLA class III region genes are important elements in:
a) Transplant rejection phenomenon
b) Governing susceptibility to autoimmune diseases
c) Immune surveillance
d) Antigen presentation and elimination

46. HLA is located on:
a) Long arm of chromosome 6
b) Long arm of chromosome 3
c) Short arm of chromosome 6
d) Short arm of chromosome 3

47. HLA B27 is positive in:
a) Ankylosing spondylitis
b) Rheumatoid arthritis
c) SLE
d) Behcet syndrome

48. Mixed lymphocyte culture is used to identify:
a) MHC class I antigen
b) MHC class II antigen
c) B lymphocytes
d) T helper cells

49. HLA typing is useful in:
a) Disputed paternity
b) Thanatology
c) Organ transplant
d) Dactylography

50. True about MHC-class II:
a) Not involved in innate immunity
b) Cytotoxic T-cell involved
c) Present in nucleated cells
d) Present in B-cells

44) B 45) B 46) C 47) A 48) B 49) AC 50) ACD

51. MHC-II positive cells are all except:
a) B cells
b) T cells
c) Macrophages
d) RBCs
e) Platelets

52. True about MHC:
a) Transplantation reaction
b) Autoimmune disease
c) Immunosuppression
d) Involved in T cell function
e) Situated at long arm of chromosome 6

53. Epitope binding floor of the MHC molecule consists of
a) Alpha helices
b) Beta plleated structure
c) Alpha and Beta-1 chain
d) Beta-2 microglobulin

54. HLA B27 is bot seen in which of the following?
a) Ankylosing spondylitis
b) Reiter's syndrome
c) Rheumatoid arthritis
d) Psoriatic arthritis

55. THE ROLE PLAYED BY Major Histocompatibility Complex 1 and 2:
a) Transduce the signal to T cells following antigen recognition
b) Mediate immunogenic class switching
c) Present antigens for recognition by T cell antigen receptors
d) Enhance the secretion of cytokines

56. MHC class I are present on all except:
a) Platelets
b) All nucleated cells
c) RBCs
d) WBCs

57. Major histocompatibility complex class I is seen on which of the following cell?
a) Macrophages only
b) All body cells
c) B cell only
d) All blood cells except erythrocytes

51) BDE 52) ABD 53) AC 54) C 55) C 56) C 57) D

58. Antigen presented along with HLA class II stimulate
a) CD8 cell
b) CD4 cell
c) CD2 cell
d) CD19 cell

59. Which of the following is having a 90% association with HLA B27?
a) Ankylosing spondylitis
b) Rheumatoid arthritis
c) Psoriasis
d) Reiter syndrome

60. Which of the following is the function of MHC I and II?
a) Signal transduction in T cells
b) Antibody class switching
c) Antigen presentation to T cells
d) Increase the secretion of cytokines

61. Antigen presenting cells are all except:
a) M-cells
b) Thymocytes
c) Macrophages
d) Langerhans cells

62. MHC-2 protein is present in all except:
a) Cortical macrophages
b) Medullary macrophages
c) Cortical epithelial cells
d) Medullary epithelial cells

63. HLA-Cw6 is associated with:
a) Myasthenia gravis
b) Behcets disease
c) Pemphigus vulgaris
d) Psoriasis vulgaris

64. HLA class II is linked with which of the following?
a) Graft rejection
b) Graft versus host disease
c) Killing of virus infected cells
d) Susceptibility to autoimmune diseases

58) B 59) A 60) C 61) A 62) B 63) D 64) B

65. HLA association with myasthenia gravis is:
a) HLA-B27
b) HLA-B51
c) HLA-B47
d) HLA-B8

66. HLA associated with pustular psoriasis is:
a) HLA CW6
b) HLA B13
c) HLA B27
d) HLA B17

67. To rule out rheumatoid arthritis, most important among the following is:
a) HLA DR8
b) HLA DR4
c) HLA DQ1
d) HLA B27

68. What type of hypersensitivity reaction is seen in myasthenia gravis?
a) Type 1 hypersensitivity reaction
b) Type 2 hypersensitivity reaction
c) Type 3 hypersensitivity reaction
d) Type 4 hypersensitivity reaction

69. Hemolytic disease of new born is an example of:
a) Type 3 hypersensitivity reaction
b) Type 2 hypersensitivity reaction
c) Arthus reaction
d) Type 4 hypersensitivity reaction

70. Raji cell assays are used to quantitate:
a) Complement levels
b) Immune complexes
c) T cells
d) Interferon levels

71. Hypersensitivity pneumonitis is classically a/an:
a) Allergic reaction
b) Type II hypersensitivity
c) Immune complex mediated hypersensitivity
d) Cell mediated hypersensitivity

65) D 66) C 67) B 68) B 69) B 70) B 71) C

72. The immunoglobulin involved in type I hypersensitivity reaction is:
a) IgE
b) IgM
c) IgA
d) IgG

73. Arthus reaction is what type of hypersensitivity reaction:
a) Localized immune complex
b) Ag-Ab reaction
c) Complement mediated
d) Ab mediated

74. A 40 years old man has chronic cough with fever for several months. The chest radiograph reveals a diffuse reticulonodular pattern. Microscopically on transbronchial biopsy there are focal areas of inflammation containing epithelioid cell granuloma, Langhans giant cells, and lymphocytes. These findings are typical for which of the following type of hypersensitivity immunologic responses:
a) Type I
b) Type II
c) Type III
d) Type IV

75. Ram Devi presented with generalized edema sweating and flushing tachycardia and fever after bee sting. This is:
a) T cell mediated cytotoxicity
b) IgE mediated reaction
c) IgG mediated reaction
d) IgA mediated hypersensitivity reaction

76. Example of type IV Hypersensitivity is/are:
a) Farmer's lung
b) Contact hypersensitivity
c) Immediate hypersensitivity
d) Myasthenia gravis

77. Example of type II hypersensitivity is/are:
a) Blood transfusion reaction
b) Arthus reaction
c) Hay fever
d) Post-streptococcal glomerulonephritis

72) A 73) A 74) D 75) B 76) B 77) A

78. Which of the following diseases is/are mediated through complement activation:
a) Atopic dematitis
b) Graft versus Host disease
c) Photoallergy
d) Necrotizing vasculitis
e) Urticaria

79. Which of following statements is not true about Mycobacterium tuberculosis infections?
a) M. tuberculosis leads to development of delayed hypersensitivity
b) Lymphocytes are the primary cells infected by M. tuberculosis
c) Positive tuberculin test signifies cell mediated hypersensitivity
d) Tuberculin test does not differentiated between infection and disease.

80. A man after consuming sea food develops rashes. It is due to:
a) IgE mediated response
b) Complement activation
c) Cell mediated response
d) None of the above

81. Granuloma in sarcoidosis is called
a) Hard sore
b) Soft sore
c) Hard tubercle
d) Caseating granuloma

82. Myasthenia gravis may be associated with
a) Thymoma
b) Systemic lupus erythematosus
c) Hyperthyroidism
d) All of the above

83. Which of the following type of hypersensitivity reaction is found in blood transfusion reaction?
a) Anaphylactic type
b) Cytotoxic type
c) Type III hypersensitivity
d) Cell mediated hypersensitivity

84. Which of the following type of hypersensitivity reactions occurs in Farmer's lung?
a) Type I
b) Type II
c) Type III
d) Type IV

78) D 79) B 80) A 81) A 82) D 83) B 84) C

85. Tuberculin test positivity indicates:
a) Good humoral immunity
b) Infection with mycobacterium
c) Good cell mediated immunity
d) None

86. Cell mediated immunity is:
a) Type I
b) Type II
c) Type III
d) Type IV

87. Antibody found in patients with myasthenia gravis is directed against
a) Acetylcholine
b) Acetylcholine receptors
c) Acetylcholine vesicles in nerve terminal
d) Actin-myosin complex of the muscle

88. Myasthenia gravis is most commonly associated with which of the following?
a) Thymoma
b) Thymic carcinoma
c) Thymic hyperplasia
d) Lymphoma

89. Patient has been given penicillin 48 hours ago, with no history of drug allergy. Now he develops wheeze and hemolysis. Antibody for penicillin is positive. Type of hypersensitivity is which of the following:
a) Type I
b) Type II
c) Type III
d) Type IV

90. IgE receptors are present on:
a) Mast cells
b) NK cells
c) B cells
d) Histiocytes

91. A 45-year old patient presents with history of fever, night sweats and weight loss. On X-ray a mass in apical lobe of lung is seen. On histopathology it was found to have caseous necrosis. What is the likely underlying process involved?
a) Enzymatic degeneration
b) Hypersensitivity reaction with modified macrophages, lymphocytes and giant cells
c) Acute decrease in blood supply

85) C 86) D 87) B 88) C 89) B 90) A 91) B

92. Serum sickness is:
a) Type 1 hypersensitivity reaction
b) Type 2 hypersensitivity reaction
c) Type 3 hypersensitivity reaction
d) Type 4 hypersensitivity reaction

93. Which of the following is true about serum sickness?
a) Type 2 hypersensitivity
b) Can lead to leukocytoclastic vasculitis
c) Hyper complement enemia
d) Can occur due to homologus antigen

94. Centre of tubercular granuloma is formed by:
a) T-lymphocytes
b) B-lymphocytes
c) Langhan's giant cells
d) Necrotic zone

95. Hyperacute rejection is due to:
a) Preformed antibodies
b) Cytotoxic T-lymphocyte mediated injury
c) Circulating macrophage mediated injury
d) Endothelitis caused by donor antibodies

96. All are affected in Graft-Versus host reaction except:
a) Skin
b) GIT
c) Liver
d) Lung

97. Performed antibodies cause:
a) Hyperacute rejection
b) Acute rejection
c) Chronic rejection
d) Acute humoral rejection

98. True about graft versus host disease is:
a) Associated with solid organ transplantation
b) Graft must contains immunocompetent T cell
c) It is seen in immunosuppressed persons
d) Also called as Runt disease in animals

92) C 93) B 94) D 95) A 96) D 97) A 98) ABCD

99. Acute humoral renal transplant rejection is characterized by the following, except:
a) Presence of anti-donor antibodies
b) Interstitial and tubular mononuclear cell infiltrate
c) Necrotizing vasculitis
d) Acute cortical necrosis

100. Transfer of the graft of different species are called as:
a) Isograft
b) Allograft
c) Homograft
d) Xenograft

101. Acute graft versus host disease reaction occurs in all except:
a) Liver
b) Adrenal
c) Gut
d) Skin

102. Principal cause of death in renal transplant patient is:
a) Uraemia
b) Malignancy
c) Rejection
d) Infection

103. Performed antibodies cause:
a) Hyperacute rejection
b) Acute rejection
c) Chronic rejection
d) Acute humoral rejection

104. Which of these complement factor is a marker of humoral rejection?
a) C3d
b) C5a
c) C3b
d) C4d

105. Method of prevention of GVHD in bone marrow transplantation is:
a) T-cell removal
b) Prior immune suppression
c) Post procedure immune suppression
d) All of the above

99)B 100) D 101) B 102) D 103) A 104) D 105) D

106. True about adult autologous stem cell transplant are all except:
a) Used in the treatment of leukemia
b) Stem cells are collected directly form the bone marrow
c) G-CSF is given to expand the number of stem cells
d) It allows high dose of chemotherapy

107. Number of criteria for HLA matching are:
a) 10
b) 4
c) 16
d) 22

108. The commonest type of graft rejection is:
a) Hyperacute rejection
b) Acute rejection
c) Chronic rejection
d) All are equal in incidence

109. Organ with least chance of rejection is:
a) Blood
b) Liver
c) Kidney
d) Heart

110. Runt disease is associated with:
a) Acute rejection
b) Hyperacute rejection
c) Chronic rejection
d) Graft versus host disease

111. Prior immune suppression is not helpful in which type of graft rejection:
a) Acute refection
b) Hyperacute rejection
c) Chronic rejection
d) None of the above

112. Which of the following is true about GVHD?
a) Occurs when host is immunocompetent
b) Occurs when donor cells are immunocompetent
c) Most common organ involved is lung
d) Most common in renal transplant

106) B 107) A 108) B 109) B 110) D 111) B 112) B

113. Micro cytotoxicity is used for:
a) Tissue typing
b) Drug allergy
c) Infection
d) Susceptibility
e) Substance toxicity

114. Acute graft rejection occurs within:
a) 3 hours
b) 3 days
c) 3 months
d) 3 years

115. Autoimmunity in EBV infection is the result of :
a) Molecular mimicry
b) Polyclonal B cell activation
c) Expressing sequestrated antigens
d) Antgenic cross reactivity

116. A 14yrs old girl on exposure to cold has pallor of extremities followed by pain and cyanosis. In later ages of life she is prone to develop?
a) Systemic lupus erythematosus
b) Scleroderma
c) Rheumatoid arthritis
d) Histiocytosis

117. Which is not autoimmune disease?
a) Systemic Lupus Erythematosus
b) Grave's Disease
c) Myasthenia Gravis
d) Sickle cell disease

118. Which among the following is seen in antiphospholipid antibody syndrome?
a) Beta 2 microglobulin antibody
b) Anti-nuclear antibody
c) Anti-centromere antibody
d) Anti-glycoprotein antibody

119. Necrotizing lymphadenitis is seen in:
a) Kimura disease
b) Kikuchi Fujimoto disease
c) Hodgkin disease
d) Castelman disease

113) A 114) C 115) B 116) B 117) D 118) D 119) B

120. Wire loop lesions are seen in:
a) SLE
b) Diabetic nephropathy
c) Benign nephrosclerosis
d) Wegener's granulomatosis

121. Tissue from rat used for detection of antinuclear antibodies?
a) Kidney
b) Brain
c) Stomach
d) Liver

122. Which is not found in a case of AIDS?
a) Perivascular giant
b) Cell
c) Vacuolization
d) Inclusion bodies
e) Microglial nodule

123. A person present with recurrent swelling on face and lips due to emotional stress/ likely cause is:
a) C1esterase inhibitor deficiency
b) Allergy
c) Anaphylaxis
d) None of the above

124. All of the following statements are true about Wiskott Aldrich syndrome except:
a) It is an autosomal recessive disorder
b) There is failure of aggregation of platelets in response to agonists
c) Thrombocytopenia
d) Is seen
e) Patient presents with eczema

125. Hematoxylin bodies seen in:
a) SLE
b) PAN
c) Rheumatoid arthritis
d) Wegener's granulomatosis

126. Wire loop lesions are often characteristic for the following class of lupus nephritis:
a) Mesangial proliferative glomerulonephritis (WHO class II)
b) Focal proliferative glomerulonephritis (WHO class III)
c) Diffuse proliferative glomerulonephritis (WHO class IV)
d) Membranous glomerulonephritis (WHO class V)

120) A 121) D 122) C 123) A 124) A 125) A 126) C

127. A renal biopsy from a 56 years old woman with progressive renal failure for the past 3 years shows glomerular and vascular deposition of pink amorphous material. It shows apple-green birefringence under polarized light after Congo red staining. These deposits are positive for lambda light chains. The person is most likely to suffer from:
a) Rheumatoid arthritis
b) Tuberculosis
c) Systemic lupus erythematosus
d) Multiple myeloma

128. A young lady presented with bilateral nodular lesions on shins. She was also found to have bilateral hilar lymphadenopathy on chest X-ray mantoux test reveals indurations of 5 mms. Skin biopsy would reveal:
a) Non caseating granuloma
b) Vasculitis
c) Caseating granuloma
d) Malignant cells

129. Anti ds-DNA antibodies are commonly seen in:
a) SLE
b) Scleroderma
c) PAN
d) Dermatomyositis
e) Rheumatoid arthritis

130. Low complement levels seen in:
a) PSGN
b) MPGN
c) Good pasture's syndrome
d) Wegner's graulomatosis
e) Infective endocarditis.

131. Which is seen in Chediak-Higashi syndrome?
a) Leucocytosis
b) Neutropenia
c) Defective microbial killing
d) Presence of large granules in neutrophil
e) Immunodeficiency

132. Adenosine deaminase deficiency is seen in:
a) Severe combined immunodeficiency
b) Wiskott Aldrich Syndrome
c) Agammaglobulinemia as HIV

127) D 128) A 129) A 130) ABE 131) BCDE 132) A

133. True about alpha-1 antitrypsin deficiency is/are:
a) Autosomal dominant
b) Pulmonary emphysema
c) Diastase resistant hepatic cells
d) Hepatic cells are orcein stain positive
e) Associated with berry aneurysm

134. All are true regarding Hyper IgE syndrome except:
a) Inheritance is as a single locus autosomal dominant trait with variable expression
b) Coarse facial features
c) Recurrent staphylococcal abscesses involving skin, lungs
d) High serum IgE with low IgG, IgA and IgM

135. All are true about Wiskott-Aldrich Syndrome except:
a) Bloody diarrhea during infancy
b) Low IgM and elevated IgA and IgE
c) Large size platelets
d) Atopic dermatitis

136. Diagnosis of X linked Agammaglobulinemia should be suspected if:
a) Absent tonsils and no palpable lymph nodes on physical examination
b) Female sex
c) High isohemagglutinins titers
d) Low CD3

137. Which of the following cell types is not a target for initiation and maintenance of HIV infection?
a) CD4 T cell
b) Macrophage
c) Dendritic cell
d) Neutrophil

138. All of the following are found in SLE except:
a) Oral ulcers
b) Psychosis
c) Discoid rash
d) Leucocytosis

139. Which of the following immunoglobulin is absent in Ataxia telangiectasia:
a) IgG
b) IgM
c) IgA
d) IgD

133) BC 134) D 135) C 136) A 137) D 138) D 139) C

140. Scl-70 antibody is characteristic of:
a) Systemic lupus erythematosus
b) Scleroderma
c) Dermatomyositis
d) Sjogren's syndrome

141. LE cell phenomenon is seen in:
a) Lymphocyte
b) Neutrophil
c) Monocyte
d) Eosinophil

142. Most sensitive test for screening of "Systemic Lupus Erythematosus"
a) LE PHENOMENON
b) RHEUMATOID FACTOR
c) Anti-Nuclear Factor (ANF)
d) Double stranded DNA test

143. According to WHO, the feature of class II lupus is:
a) Transient proteinuria
b) Massive proteinuris
c) Hematuria
d) RBC casts

144. ANCA antibody with peripheral rim distribution is indicative of:
a) Anti-histone antibody
b) Anti-smith antibody
c) Anti-double stranded DNA antibody
d) Anti-double stranded RNA antibody

145. Basic pathology in cystic fibrosis is:
a) Defect in the transport of chloride across epithelia
b) Defect in the transport of sodium across epithelia
c) Defect in the transport of potassium across epithelia
d) Defect in the transport of bicarbonate across epithelia

146. Besbuer Boeck Schaumann disease is also called as:
a) Sarcoidosis
b) Crohn's disease
c) Whipple's disease
d) Hodgkin's disease

140) B 141) B 142) C 143) C 144) C 145) A 146) A

147. Most common viral antigen used for diagnosis of HIV in blood before transfusion is:
a) p24
b) p17
c) p7
d) p14

148. Most common vascular tumor in AIDS patients is:
a) Kaposi's sarcoma
b) Angiosarcoma
c) Lymphangioma
d) Lymphoma

149. Which is not an autoimmune disease?
a) Syphilis
b) SLE
c) Systemic sclerosis
d) RA

150. Bilateral parotid gland enlargement is seen in all except:
a) Sarcoidosis
b) Sjogren's syndrome
c) SLE
d) Viral infections

151. Sarcoidosis does not involve
a) Brain
b) Heart
c) Lung
d) Kidney

152. Characteristic of SLE of kidney is:
a) Focal sclerosis
b) Focal necrosis
c) Wire loop lesions
d) Diffuse glomerulosclerosis

153. Libman-Sacks endocarditis is seen in:
a) Rheumatoid arthritis
b) SLE
c) Infective endocarditis
d) Nonbacterial thrombotic endocarditis

147) A 148) A 149) A 150) C 151) A 152) C 153) B

154. Chediak-Higashi syndrome is due to defect in:
a) Opsonisation
b) Chemotaxis
c) LAD
d) Extracellular microbicidal killing

155. Anti-double stranded DNA is highly specific for:
a) Systemic sclerosis
b) SLE
c) Polymyositis
d) Rheumatic sclerosis

156. Anti-topoisomerase I is marker of:
a) Systemic sclerosis
b) Classic polyarteritis nodosa
c) Nephrotic syndrome
d) Rheumatoid arthritis

157. An 8-year old boy presents with sarcoidosis. Which of the following is correct?
a) Hilar lymphadenopathy with perihilar calcification
b) Basal infiltrates
c) Rubbery lymph nodes
d) Egg-shell-calcification

158. Most common site for lymphoma in AIDS patients is:
a) CNS lesions
b) Spleen
c) Thymus
d) Abdomen

159. All are true about histological features of Kaposi's sarcoma except:
a) Microscopically lesion similar to granulation tissue
b) Dilated and irregular blood vessels with interspersed infiltrate of lymphocyte and plasma cells
c) Atypical blood vessels have solid spindle cell appearance
d) Nodule is the initial lesion of Kaposi's sarcoma

160. HIV affects which of the following most commonly?
a) Helper cells
b) Suppressor cell
c) RBCs
d) Platelets

154) B 155) B 156) A 157) A 158) A 159) D 160) A

161. Which of the following lesion/conditions shows most specific anatomic changes in HIV infection?
a) Lymph nodes
b) Opportunistic infections
c) CNS lesions
d) Kaposi's sarcoma (blood vessels)

162. Which of the following autoantibody is least likely associated with SLE?
a) Anti-ds DNA
b) Anti Sm
c) Anti-topoisomerase
d) Anti-histone

163. Which of the following autoantibody is specific for SLE?
a) Ds DNA
b) Anti-RO
c) Anti-centromere
d) Anti-topoisomerase

164. Regarding severe combined immunodeficiency disease, which of the following statement is true?
a) Adenosine deaminase deficiency
b) Decreased circulating lymphocytes
c) NADPH oxidase deficiency
d) C1 esterase deficiency

165. Which of the following is a finding in lymphoid tissues in individuals with common variable hypogammaglobulinemia?
a) Decreased B cell count
b) Increased B cell count
c) Normal B cell count
d) Absent B cells

166. Thymic hypoplasia is seen in which of the following?
a) Wiskott-Alsrich syndrome
b) Digeorge syndrome
c) IgA deficiency
d) Agammaglobulinemia

167. Onion peel appearance of splenic capsule is seen in:
a) SLE
b) Scleroderma
c) Rheumatoid arthritis
d) Sjogren syndrome

161) C 162) C 163) A 164) A 165) C 166) B 167) A

168. Following is not a feature of AIDS related lymphadenopathy:
a) Florid reactive hyperplasia
b) Follicle lysis
c) Haematoxylin bodies
d) Collection of monocytoid B-cells in sinuses

169. Most common CNS neoplasm in HIV patient is:
a) Meduloblastoma
b) Astrocytoma
c) Primary CNS lymphoma
d) Ependymoma

170. A false negative tuberculin reaction may be obtained in all of the following situations except:
a) Children previously tested with tuberculin test
b) Post=measles test
c) Corticosteroid therapy
d) Miliary tuberculosis

171. Risk of HIV transmission is not seen with:
a) Whole blood
b) Platelets
c) Plasma derived hepatitis B vaccine
d) Leucocytes

172. All of the following methods are used for the diagnosis
a) DNA-PCR
b) Viral culture
c) HIV ELISA
d) P24 antigen assay

173. Mantoux test reading of less than 5mm indicates:
a) Tuberculous infections
b) Disseminated TB
c) Susceptibility to TB
d) Immunity to TB

174. Epitope spreading refers to:
a) A type of mechanism of spread of malignant tumors
b) One type of mechanism of HIV dissemination
c) A mechanism for the persistence and evolution of autoimmune disease
d) One of the mechanisms of apoptosis

168) C 169) C 170) A 171) C 172) C 173) B 174) C

175. Heerfordt's syndrome consists of fever, parotid enlargement, facial palsy and
a) Arthralgia
b) Bilateral hilar adenopathy
c) Erythema nodosum
d) Anterior uveitis

176. HIV affects CD4 cells by which protein?
a) Gp 120
b) Gp 41
c) CCR5
d) CXCR4

177. Treatment for Asymptomatic HIV is done when CD4 count is below
a) 200
b) 350
c) 400
d) 500

178. The poly-arthritic condition that is NOT common in males
a) Gout
b) Psoriatic arthritis
c) Ankylosing spondylitis
d) Systemic lupus erythematosus

179. Hodgkins lymphoma caused for by:
a) EBV
b) CMV
c) HHV6
d) HHV8

180. Which of these is an immune-privileged site?
a) Area postrema
b) Loop of Henle
c) Optic nerve
d) Seminiferous tubules

181. Which of the following is not an autoimmune disorder?
a) Ulcerative colitis
b) Grave's disease
c) Rheumatoid arthritis
d) SLE

175) D 176) A 177) B 178) D 179) A 180) D 181) A

182. Autoimmunity is caused by all except:
a) Infections
b) Expression of cryptic antigens
c) Negative selection of T-cells in the thymus
d) Inappropriate expression of the MHC proteins

183. Which of the following is not seen in SLE affected kidneys?
a) Focal glomerulonephritis
b) Diffuse glomerulonephritis
c) Membranous glomerulonephritis
d) Lipoid nephrosis

184. Anti RO(SSA) antibodies are seen in:
a) Systemic sclerosis
b) Subacute cutaneous lupus
c) Myasthenia gravis
d) Mixed connective tissue disorder

185. Secondary amyloidosis is associated with:
a) Aβ
b) AL
c) AA
d) APrP

186. A 60 year old female is suffering from renal failure and is on hemodialysis since last 8 years. She developed carpal tunnel syndrome. Which of the following finding will be associated?
a) AL
b) AA
c) ATTR
d) β2 microgloculin

187. The best investigation for the diagnosis of amyloidosis is:
a) Colonoscopy
b) Rectal biopsy
c) Upper GI endoscopy
d) CT scan

188. Which type of amyloidosis is caused by mutations in transthyrentin gene?
a) Familial Mediterranean fever
b) Familial amyloidosis
c) Polyneuropathy
d) Prion protein associated amyloidosis

182) C 183) D 184) B 185) C 186) D 187) B 188) B

189. In Hemodialysis associated amyloidosis, which of the following is seen:
a) Transthyretin
b) β2 Microglobulin
c) SAA
d) α2 Microglobulin

190. Bone marrow in AL amyloidosis shows:
a) Bone marrow plasmacytosis
b) Granulomatous reaction
c) Fibrosis
d) Giant cell formation

191. A diabetic patient is undergoing dialysis. Aspiration done around the knee joint would show:
a) A beta 2 microglobulin
b) AA
c) AL
d) Lactoferin

192. What is the best method for confirming amyloidosis?
a) Colonoscopy
b) Sigmoidoscopy
c) Rectal biopsy
d) Tongue biopsy

193. Neointimal hyperplasia causes vascular graft failure as a result of hypertrophy of:
a) Endothelial cells
b) Collagen fibers
c) Smooth muscle cells
d) Elastic fibers

194. Which one of the following stains is specific for Amyloid?
a) Periodic Acid Schiff (PAS)
b) Alizarin red
c) Congo red
d) Von-Kossa

195. In amyloidosis Beta pleated sheet will be seen in:
a) X-ray crystallography
b) Electron microscope
c) Spiral electron microscope
d) Congo red stain

189) B 190) A 191) A 192) C 193) C 194) C 195) A

196. A 50 year old presented with signs and symptoms of restrictive heart disease. A right ventricular endomyocardial biopsy revealed deposition of extracellular eosinophilic hyaline material. On transmission electron microscopy, this material is most likely to reveal the presence of:
a) Nonbranching filaments of indefinite length
b) Cross banded fibers with 67m periodicity
c) Weibel Palade bodies
d) Concentric whorls of lamellar structures

197. Amyloid deposits stain positively with all of the following except:
a) Congo red
b) Crystal violet
c) Methanamine silver
d) Thioflavin T

198. On electron microscopy amyloid characteristically exhibits:
a) Beta-pleated sheat
b) Hyaline globules
c) 7.5-10nm fibrils
d) 20-25nm fibrils

199. Familial amyloidotic polyneuropathy is due to amyloidosis of nerves caused by deposition of:
a) Amyloid associated protein
b) Mutant calcitonin
c) Mutant transthyretin
d) Normal transthyretin

200. Lardaceous spleen is due to deposition of amyloid in:
a) Sinusoids of red pulp
b) White pulp
c) Pencillary artery
d) Splenic tranbeculae

201. What are the stains used for Amyloid?
a) Thioflavin
b) Congo red
c) Eosin
d) Auramine
e) Rhodamine

202. Gingival biopsy is useful in the diagnosis of:
a) Sarcoidosis
b) Amyloidosis
c) Histoplasmosis
d) Scurvy

196) A 197) C 198) C 199) C 200) A 201) AB 202) B

203. Amyloid is:
a) Mucopolysaccharide
b) Lipoprotein
c) Glycoprotein
d) Intermediate filament

204. Serum amyloid associated protein is found in:
a) Alzheimer's disease
b) Chronic inflammatory states
c) Chronic renal failure
d) Malignant hypertension

205. Most common site of biopsy in amyloidosis:
a) Liver
b) Spleen
c) Kidney
d) Lung

206. Currently matched pairs in amyloidosis are:
a) Multiple myeloma-light chain
b) Chronic inflammation-AA
c) Cardiac-ATTR
d) Neural-Beta-2 microglobulin

207. A diabetic patient is undergoing dialysis. Aspiration done around the knee joint would show:
a) A-β2 Microglobulin
b) AA
c) AL
d) Lactoferrin

208. Amyloidosis is most commonly seen in:
a) Maturity onset DM
b) Type 1 DM
c) Type 2 DM
d) Equally seen with all forms of DM

209. Which of the following is the most serious organ involvement in amyloidosis?
a) Cardiac tissue
b) Renal tissue
c) Splenic tissue
d) Hepatic tissue

203) C 204) B 205) C 206) ABC 207) A 208) C 209) B

210. Which type of Amyloidosis is caused by mutation of the transthyretin protein?
a) Familial Mediterranean fever
b) Familial amyloidotic polyneuropathy
c) Dialysis associated amyloidosis
d) Prion protein associated amyloidosis

211. Cause of death in amyloidosis involving kidney:
a) Cardiac failure
b) Renal failure
c) Sepsis
d) Liver failure

212. Secondary amyloidosis complicates which of the following:
a) Pneumonia
b) Chronic glomerulonephritis
c) Irritable bowel syndrome
d) Chronic osteomyelitis

213. On Congo-red staining, amyloid is seen as:
a) Dark brown color
b) Blue color
c) Brilliant pink color
d) Khaki color

214. Lardaseous spleen is due to deposition of amyloid in:
a) Sinusoids of red pulp
b) White pulp
c) Pencillary artery
d) Splenic trabeculae

215. Which of the following is the chemical nature of Hemodialysis associated with amyloid?
a) AA
b) AL
c) Beta-2-Microglobulin
d) ATTR

210) B 211) B 212) D 213) C 214) A 215) C

216. Familial amyloidosis is seen in:
a) Alzheimer's disease
b) Senile cardiac amyloidosis
c) Renal amyloidosis
d) Splenic amyloidosis

217. Excessive accumulation of which hormone protein causes organ dysfunction:
a) Growth hormone
b) Prolactin
c) Calcitonin
d) Parathormone

218. Serum amyloid associated protein is increased in:
a) Alzheimer's disease
b) Ankylosing spondylitis
c) Chronic renal failure
d) Malignant hypertension

216) B 217) C 218) B

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