Board Simulation Session: Allergy and Immunology

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OBJECTIVES

- Question and answer format to review allergy and immunology topics for pediatric board exam and clinical practice including:
  - Asthma and other respiratory conditions
  - Allergic rhinitis
  - Immunodeficiency
  - Adverse reactions to medications, stinging insects and latex
  - Food allergy

Question 1

The parents of an 8 year old girl with asthma, atopic dermatitis and positive skin tests to dust mites, tree and grass pollens ask you what is the likelihood that she will outgrow her asthma.

What is the BEST response?

A. Less than 20% of children with asthma outgrow symptoms by adulthood.
B. Children with asthma and multiple inhalant allergies are less likely to outgrow asthma than children with asthma alone.
C. Children with exercise induced asthma rarely outgrow asthma.
D. Children with asthma & atopic dermatitis are more likely to outgrow asthma than children with asthma alone.
E. You cannot predict if she will outgrow asthma.
Question 1

- Children with asthma and atopy such as allergic rhinitis and atopic dermatitis have a poorer prognosis for persistent asthma.
- Roughly 60% of all children who have asthma will have resolution of asthma by adulthood.
- Other risk factors for persistence of asthma into adulthood include:
  - a maternal or paternal history of asthma.
  - history of severe asthma with multiple asthma exacerbations.

Roughly 60% of all children who have asthma will have resolution of asthma by adulthood.

Question 2

Which test is MOST helpful to evaluate cell-mediated immunity in a 3 yr old child with recurrent infections, chronic diarrhea, and failure to thrive?

A. Quantitative immunoglobulin measurement (IgA, IgG, IgM)
B. Give immunizations and then measure specific antibody titers 6 weeks later
C. Nitroblue tetrazolium dye reduction test
D. PPD and control delayed skin tests
E. Measure complement levels

Evaluation of immune system

- Humoral
  - Isohemagglutinins (ages 1 or older with blood type A, B)
  - Serum immunoglobulins (IgA, IgG, IgM)
  - Measure response to vaccines (diphtheria, tetanus, H. influenza)

- Cell-mediated
  - DTH response to previously encountered antigens (tetanus, mumps, Candida, Trichophyton)
  - T lymphocyte counts by flow cytometry
  - Lymphocyte stimulation testing

- Phagocytes
  - Nitroblue tetrazolium dye test or DHR 123 to measure neutrophil respiratory burst

- Complement
  - Complement 50 measures classical pathway
  - Low levels from delay in specimen processing, early complement deficiency (C1-C4) or late complement deficiency (C5-C9)

Question 3

An 8 year old boy initiated a 10 day treatment with oral amoxicillin 12 days ago. 2 days ago, he developed fever, malaise, arthralgia of multiple joints, dark urine, and urticaria.

What would be the treatment of choice for this condition?

A. Amoxicillin-clavulanic acid 80 mg/kg/day for 10 days
B. Oral diphenhydramine 1 mg/kg every 6 hrs prn
C. Prednisone 1-2 mg/kg/day for 5 days, then taper
D. Stop all medications, admit and observe

Question 3

- Serum sickness is a classic example of a type III hypersensitivity reaction
  - mediated by immunoglobulin G (IgG) or immunoglobulin M (IgM) antibody-antigen complexes
  - complexes can be deposited in various organs and activate complement
  - Serum complement levels (C3 and C4) often decreased and ESR is usually elevated
  - Less common symptoms include mild proteinuria, hemoglobinuria, and microscopic hematuria
  - Prolonged burst/taper of oral corticosteroids is treatment of choice, plus supportive treatment with analgesics and antihistamines
  - Serum sickness cannot be prevented by desensitization or by pretreatment with corticosteroids

Question 4

A 15 month old female has been hospitalized for several serious infections secondary to *Staphylococcus aureus* including osteomyelitis, hepatic abscess and pneumonia with pneumatocele formation.

What is the MOST LIKELY diagnosis?

A. Wiskott-Aldrich Syndrome
B. Selective IgA deficiency
C. Leukocyte adhesion deficiency
D. Chronic granulomatous disease (CGD)
Chronic granulomatous disease (CGD)

- Phagocytic disorder
- Occurs in 1 in 250,000 to 1 in 500,000 live births in US
- Defect: inability to kill intracellular catalase-positive bacteria and fungi after phagocytosis (Staph aureus, Candida albicans, Salmonella sp, Aspergillus sp, Serratia marcescens, Nocardia, Burkholderia cepacia)
- Recurrent infections with Staphylococci and a wide variety of other organisms suggests a phagocytic defect
- Presents with recurrent lymphadenitis, hepatic abscesses, recurrent skin infections, osteomyelitis
- Abscess formation is characteristic

Wiskott-Aldrich Syndrome

- X-linked recessive syndrome
- TRIAD: recurrent infections, eczema & thrombocytopenia

Selective IgA deficiency

- Most common inherited immunodeficiency
- Clinical presentation varies from asymptomatic to recurrent sinopulmonary infections

Leukocyte adhesion deficiency

- Associated with recurrent infections WITHOUT abscess formation
- Leukocytes lack the ability to adhere to blood vessel walls and to migrate to the site of infection
- Leukocytosis
- Abscess formation is unusual

Question 4

You are asked to see a 15 year old female who has acute onset of bilateral flank pain and hematuria. She is receiving an extended course of IV antibiotics for a Staph infection.

What is the most likely cause of these symptoms?

A. Adverse reaction to vancomycin.
B. Adverse reaction to methicillin.
C. Adverse reaction to cefazolin.
D. Renal stone.
Interstitial nephritis and antibiotics: can occur with any class of drug, but notably common with methicillin.

Question 5

A 5 year old male is admitted with his third episode of bacterial meningitis. Each time, the culture has grown *Neisseria meningitidis*. Which of the following tests is most likely to find the underlying immune deficiency responsible for these recurrent infections?

- A. Nitroblue tetrazolium dye reduction test
- B. Quantitative immunoglobulins
- C. Flow cytometry for adhesion molecules
- D. CH50

Question 6

Children with deficiencies of certain complement components, especially C6-C9 present with recurrent systemic *Neisseria* infections. CH50 test is the best single routine test for measuring complement system function.
Question 7

A 16 year old girl presents with a 3 month history of daily urticaria with pruritus and occasional episodes of lip and eyelid swelling. What is the most likely cause for her urticaria and angioedema?

A. Idiopathic
B. Hidden food allergy
C. Contact allergy to dust mites or family pet
D. Underlying autoimmune disease
E. C1 inhibitor deficiency

Question 7

- Chronic urticaria is defined as urticaria lasting for more than 6 wks
- Over 90% of pediatric patients with chronic urticaria are classified as idiopathic, with no underlying allergy or other identifiable condition
- Autoimmune disorders can rarely cause chronic urticaria
- Hereditary angioedema is characterized by isolated episodes of painful swelling, and is unlikely to have associated urticaria or pruritus

Question 8

A 12 yr old girl with known ragweed hay fever symptoms in the Fall season complains of mouth itching when eating a particular food. Which food is most likely?

A. Peanut
B. Hazel nut
C. Watermelon
D. Apple
E. Corn
Question 8
The pollen-food oral allergy syndrome causes oral allergy symptoms when patients with certain pollen allergies consume fresh fruits or vegetables, due to a cross-reactivity between allergenic proteins. For tree pollen allergic patients, this occurs with foods like apple, pear, plum, and cherry. For ragweed allergic patients, this occurs with bananas and melons.

Question 9
You are seeing a 10 year old boy in the late spring who has a 4 week history of sneezing, nasal congestion and rhinorrhea. You suspect he has allergic rhinitis.

What medication would be MOST effective in treating his chronic nasal congestion?
A. Oral decongestant
B. Intranasal corticosteroid
C. Intranasal antihistamine
D. Oral antihistamine

Medications for chronic AR include antihistamines (oral and intranasal), oral leukotriene receptor antagonists, and intranasal corticosteroids.
Guidelines from American Academy of Allergy, Asthma and Immunology recommend intranasal corticosteroids as first-line therapy.
Comparisons between intranasal corticosteroids and either oral antihistamines or oral leukotriene receptor antagonists demonstrate that intranasal corticosteroids are most effective for nasal symptoms.
Approved for use in patients as young as 2 years old.
Oral antihistamines and oral leukotriene receptor antagonists improve symptoms of allergic rhinitis compared with placebo.

Question 10

A 15 year old patient with spina bifida had an acute episode of hypotension, bronchospasm and diffuse erythema 45 minutes after starting a surgical procedure during which she received general anesthesia.

What is the MOST LIKELY causative agent for this reaction?

A. Antibiotic she received 1 day prior to surgery
B. Anesthetic inhalant
C. Colloid intravenous fluid
D. Iodine cleansing solution used prior to the surgery
E. Latex rubber

The most common agents responsible for intraoperative anaphylaxis are muscle relaxants, latex, antibiotics and anesthesia induction agents. Most episodes of surgical anaphylaxis occur during induction when muscle relaxants, sedatives and opiates are given. Other agents which can be associated with intraoperative anaphylaxis include colloids, opioids and radiocontrast media though these usually account for less than 10% of intraoperative anaphylaxis events.

Type I IgE mediated hypersensitivity reactions to antibiotics generally do not have a delayed onset.

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Latex
- Incidence of latex sensitivity ranges from 1-6%.
- Approximately 220 cases of anaphylaxis and 3 deaths per year are due to latex allergy.
- Reactions often occur during maintenance anesthesia with a delay of 30 to 60 minutes or more prior to the onset of symptoms.
- HIGH RISK: those with an increased exposure to latex:
  - spina bifida (50%)
  - chronic bladder care with latex catheters
  - health care workers (10%)
  - multiple surgical procedures
  - atopic patients

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Question 11

The rash most closely associated with celiac disease (gluten-sensitive enteropathy) is:

A. Atopic dermatitis.
B. Erythema marginatum.
C. Erythema multiforme.
D. Pyoderma gangrenosum.
E. Dermatitis herpetiformis.

Question 11

- Atopic dermatitis is associated with allergic diseases, especially food allergy, and can also be seen in certain immunodeficiencies, like Wiskott-Aldrich syndrome.
- Erythema multiforme is most commonly associated with certain drug allergy reactions and infections.
- Erythema marginatum is seen in Lyme disease.
- Pyoderma gangrenosum can be seen with inflammatory bowel disease.
- Dermatitis herpetiformis is associated with celiac disease.

Question 12

A 12 month old infant was diagnosed with X-linked agammaglobulinemia (XLA), a recessive B cell immune deficiency, after suffering multiple sinopulmonary tract infections such as otitis media, sinusitis and pneumonia.

What is an appropriate treatment for him?

A. Plasmapheresis and broad spectrum antibiotics
B. Intravenous immunoglobulin (IVIG)
C. Prophylactic antibiotics
D. Enzyme replacement therapy
**Question 12**

- **XLA**
  - Clinical presentation:
    - Often delayed until 4-6 months due to maternal IgG
    - Lymphoid tissues reduced in size
    - Absence of B cells
    - Total absence or marked deficiency of serum immunoglobulins
  - Defect: Bruton’s tyrosine kinase (Btk) gene, a cytoplasmic tyrosine kinase critical for early B-cell development
  - Autosomal recessive defects have been described with similar presentation to XLA
  - Treatment: broad-spectrum antibiotics and IVIG helpful in preventing severe acute bacterial infections and bronchiectasis

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**Question 13**

A 2 week old boy is admitted to the hospital for seizures and found to have hypocalcemia. Physical exam is significant for hypertelorism, mandible hypoplasia and a loud systolic murmur.

What immunodeficiency should you suspect?

A. Ataxia-telangiectasia  
B. X-linked hyper-IgM syndrome  
C. DiGeorge Syndrome  
D. Severe combined immunodeficiency (SCID)
DiGeorge syndrome
- caused by embryonic dysmorphogenesis of the 3rd & 4th pharyngeal pouches
- associated with deletions in chromosome 22q11
- variable presentation for immunodeficiency
- characterized by
  - absent or diminished thymus development
  - hypocalcemia
  - cardiac malformations of the aortic arch
  - facial dysmorphism including hypertelorism, short philtrum, low-set ears, mandibular hypoplasia, cleft palate hypoparathyroidism
- treatment options include bone marrow or thymic tissue transplant.

Ataxia-telangiectasia
- Autosomal recessive
- Neurologic, cutaneous and endocrine abnormalities
- Defect: mutations in ATM gene which affects protein involved in DNA repair
- Characteristics:
  - early childhood with regression in motor milestones
  - development of telangiectasias between 3-4 years old
  - Recurrent sinopulmonary infections with encapsulated bacteria
  - Immunologic findings: decreased IgE, variably decreased IgG and T-cell deficiency
  - TRIAD: immunodeficiency, cerebellar ataxia and oculocutaneous telangiectasias
  - Increased risk of malignancy and radiation sensitivity

X-linked Hyper IgM syndrome
- no isotype switching from IgM to other immunoglobulins
- susceptible to infections with encapsulated bacteria
- defect: gene encoding CD40 ligand on T cells

SCID
- profound combined defect in antibody and cell-mediated immune response (B and T cell)
- susceptible to bacterial, viral and fungal infections including CMV, Candida sp., Pneumocystis jiroveci
- several forms, all with absence of functioning T lymphocytes
- characteristics:
  - Present during infancy
  - Failure to thrive, diarrhea and recurrent opportunistic infections
  - Lymphopenia (absolute count below 2500-3000 cells/mm^3)
- Treatment: bone marrow or stem cell transplantation
A 14 year female with persistent asthma has an elevated total serum IgE level over 2000ng/mL, a history of transient pulmonary infiltrates on chest radiographs, and now has proximal bronchiectasis on a computed tomograph (CT).

What test would BEST help diagnose her underlying disorder?

A. Skin test to Aspergillus species or A fumigatus
B. Skin test to cow’s milk protein
C. Sweat test
D. Lung biopsy for non-caseating granulomas
E. Sputum culture for Histoplasma capsulatum

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Allergic bronchopulmonary aspergillosis (ABPA) complicates chronic pulmonary disease in approximately 10% of children with asthma or cystic fibrosis.

Colonization with A. fumigatus produces an exaggerated IgG and IgE response.

Results in recurrent bronchospasm and bronchiectasis.

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Criteria for diagnosis of ABPA include:
- Asthma
- An elevated total serum IgE concentration above 1000ng/mL
- Positive skin tests to Aspergillus species or specific serum IgE and/or IgG to A fumigatus
- Infiltrates on chest radiograph
- Proximal bronchiectasis

The goal of treatment of ABPA:
- Treat exacerbations
- Prevent or minimize bronchiectasis
Question 15

A 17 year old boy has a history of an adverse reaction to radiocontrast media (RCM) 2 years ago which included diffuse pruritus and urticaria accompanied by hypotension.

Which statement is TRUE regarding his adverse reaction to RCM?

A. History of this type of reaction is an absolute contraindication to further administration of RCM.
B. He should avoid RCM and iodine, such as in iodine cleaning solutions and shellfish.
C. He should receive high-osmolar, ionic RCM for the next procedure.
D. He should receive antihistamines and prednisone prior to next procedure with RCM.

Question 15

Radiocontrast media (RCM) reactions are most likely mediated by a non-immunoglobulin E (IgE) mechanism:
- Mechanism is different from anaphylaxis which is IgE mediated
- Clinical symptoms are similar since both reactions involve degranulation of mast cells and histamine release
  - Pruritus
  - Urticaria and angioedema
  - Hypotension and syncope
- "anaphylactoid" likely by direct activation and degranulation by RCM

Question 15

Risk factors for RCM reactions include
- patients with asthma and allergic diseases
- use of beta-blockers
- previous reaction to RCM

High risk patients should receive prophylaxis to reduce risk to less than 1%
- prednisone
- antihistamines
- low-osmolar and non-ionic RCM
Question 16
While a 14 year old patient with spina bifida is waiting to see you for a well child visit, she decides to have something to eat. Within 10 minutes, she starts experiencing diffuse hives, shortness of breath and wheezing.

What food do you think she may have eaten?
A. Strawberries or raspberries
B. A banana or kiwi fruit
C. An orange or apple
D. A peach or nectarine

Latex-fruit syndrome:
- Result of cross-reactivity between natural rubber latex proteins and food proteins
  - Bananas
  - Avocados
  - Kiwi fruit
  - Chestnuts
- How common?
  - 30-50% of latex allergic are sensitive to some plant-derived foods

Question 17
A 10 year old boy comes to your office for an evaluation of pruritus and a rash. The rash has been present for the past 10 days along with mild symptoms of an upper respiratory infection. Physical exam is significant for an uncomfortable boy who is scratching throughout the exam, and his skin is remarkable for diffuse urticaria. He has never experienced urticaria previously.

What is the MOST LIKELY cause of his urticaria?
A. Food allergy
B. Allergy to airborne pollen
C. Viral infection
D. Contact reaction to a new soap
E. Pet allergy
Question 17

- Acute urticaria is often caused by an allergic, or immunoglobulin E (IgE), mediated mechanism
- Common triggers include
  - Foods
  - Medications
  - Inhalant allergens
  - Infections
  - Autoimmune or collagen vascular disease
  - Physical factors such as cold, heat or pressure

The duration of the reaction is helpful in determining a possible etiology
- Reactions to foods, medications, inhalants and soaps are generally self limited and resolve once the offending agent is eliminated
- Viral and autoimmune etiologies often cause more prolonged courses

Question 18

You are seeing a 10 month old boy with severe eczema who recently moved to your town. His mother mentions that he has had recurrent infections requiring antibiotics, and that he has a history of easy bruising and purple rashes.

What is the MOST likely immunodeficiency?
A. X-linked hyper-IgM syndrome
B. Complement deficiency
C. Wiskott-Aldrich syndrome
D. Leukocyte adhesion deficiency
**Wiskott-Aldrich Syndrome**
- X-linked recessive
- TRIAD: recurrent infections, eczema & thrombocytopenia
- Often present before age 1
- Symptoms: prolonged bleeding after circumcision or bloody diarrhea during infancy, recurrent sinopulmonary infections with encapsulated bacteria
- May develop autoimmune disease or lymphoma
- Defect: gene encoding WAS protein involved in actin polymerization
- Treatment options
  - IV or SQ immunoglobulin replacement
  - Prophylactic antibiotics
  - HLA identical bone marrow transplant

**Question 18**


**Question 18**

- X-linked hyper-IgM syndrome
  - no isotype switching from IgM to other immunoglobulins
  - susceptible to infections with encapsulated bacteria
- Complement deficiency
  - autosomal recessive
  - early (C1-C4): recurrent sinopulmonary infections due to encapsulated bacteria, often associated with increased incidence of systemic autoimmune disease
  - late (C5-C9): recurrent Neisseria infections and increased risk of meningitis
- Screening test: CH50, measures complement activity

**Question 18**


**Question 19**

Regarding food allergy cross-reactivity, which of the following statements are true?

A. Most children with cow milk allergy can tolerate goat milk.
B. Children with peanut allergy are more likely to tolerate tree nuts than other legumes.
C. Shellfish allergy creates a higher risk for radiocontrast reactions compared to other allergies.
D. Patients with pollen-food oral allergy syndrome react only to the raw fruit or vegetable, not if it is cooked.
Question 19

- Peanut allergy shows cross-allergenicity to legumes in <10% of patients, while over 25% have tree nut allergy.
- Goat milk is allergenic for approx 90% of children with cow milk allergy.
- Shellfish allergy is caused by IgE-mediated sensitivity to shellfish protein, while radiocontrast reactions are dependent on the osmolarity of the contrast reagent.
- Pollen-food oral allergy syndrome is a cross reaction between pollen allergens and heat-labile proteins of fruits or vegetables.

Question 20

A 12 year old patient with severe asthma was admitted to the hospital for an acute asthma exacerbation and has been treated with a variety of medications including albuterol over the past 24 hours. You are concerned that he might have symptoms related to albuterol toxicity.

Which of the following is NOT a side effect of inhaled albuterol?

A. Muscle tremor
B. Hypotension
C. Hypokalemia
D. Tachycardia
E. Nervousness

Question 20

- The clinical manifestations of toxicity to inhaled beta-adrenergic agonists includes
  - Hypokalemia
  - Tachycardia & palpitations
  - Muscle tremors & muscle cramps
  - Hypertension (not hypotension)
  - Headache
  - Nervousness & insomnia
  - Dizziness
You are seeing a 3 year old girl and her 5 year old brother for a well child check up. Both have food allergies. Her mother asks you what is the likelihood that they will outgrow their food allergies.

What is the BEST response to her question?
A. Most children do not outgrow food allergies
B. Children with egg allergy are not likely to outgrow the allergy
C. Children with peanut allergy are not likely to outgrow the allergy
D. Children with soy allergy are not likely to outgrow the allergy

Approximately 85% of children with milk, egg, soy and wheat food allergies outgrow the allergy by 3-5 years of age.

Peanut allergy:
- Children with peanut allergy are not likely to outgrow it (20%)
- 25-50% of patients with peanut allergy also allergic to tree nuts
- Tree nuts may contain trace amounts of peanut since these foods are frequently processed together
- Avoid unrefined (cold pressed or crude) and gourmet peanut oils
  - Unrefined peanut oils may contain significant amounts of peanut protein BUT refined (hot-pressed) peanut oils do not generally not

What is the BEST test for diagnosis of chronic granulomatous disease (CGD)?
A. Immunoglobulin levels
B. Nitroblue tetrazolium test
C. Absolute neutrophil count
D. Absolute T and B cell count
E. None of the above

References:
Question 22

- Diagnosis of CGD
  - Diagnosed by demonstrating an inability of neutrophils to undergo a respiratory burst and generate superoxide ions after phagocytosis
  - Generation of superoxide ions can be demonstrated by measurement of reduction of nitroblue tetrazolium dye (NBT)
  - Newer technique: Dihydrorhodamine reductase assay with flow cytometry

Question 23

A 17 year old male is receiving allergy shots in your general pediatric clinic for allergic rhinitis. Approximately 30 minutes after his shot, he has erythema at the injection site. His vital signs include BP for 120/70, HR 80, RR 15 and oxygen sat of 97% on RA. Aside from a raised, quarter-sized area of erythema on his upper right arm at the injection site, his physical exam is normal.

What should you do for this patient?
A. Give intramuscular epinephrine
B. Recommend that he stop his allergy shots
C. Observe the patient for 1 hour
D. Send the patient home

Recommendations for administering immunotherapy

- Administer in a medically supervised setting
- Mandatory observation of 30 minutes after each shot
- Avoid immunotherapy in patients on beta-blockers or with unstable asthma
- When might an anaphylactic reaction occur?
  - Occurs 1 in 200 injections
  - During first year of buildup
  - Peak pollen seasons
  - Symptoms often occur within 30 minutes of shot
An 18 year old boy with a history of allergic rhinitis had wheezing, generalized urticaria and documented hypotension after a honeybee sting 8 weeks ago.

After referral to the local allergist for venom skin testing, venom immunotherapy was recommended.

Your patient wants to know what his chances are of having another life-threatening reaction in the next 5 years if he successfully completes venom immunotherapy.

The BEST answer to his question is:

A. Venom immunotherapy will be approximately 95% effective in preventing a future systemic reaction.
B. Venom immunotherapy will be approximately 67% effective in preventing a future systemic reaction.
C. Since he has allergic rhinitis, venom immunotherapy will be less than 60% effective in preventing a future systemic reaction.
D. Since he had a severe anaphylactic reaction with hypotension, venom immunotherapy will be less than 40% effective in preventing a future systemic reaction.
E. You cannot predict how effective venom immunotherapy will be in reducing the chance of another life-threatening reaction.

Venom immunotherapy (VIT)
- 95% effective in preventing systemic reactions in stinging-insect-sensitive patients
- Guidelines for length of VIT
  - consider discontinuation of venom IT after 5 years
  - 10% chance of a systemic reaction with each subsequent sting

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Question 25

Your are seeing an 6 month old infant with a recent history of treatment for gingivitis. His history is significant for separation of the umbilical cord at 8 weeks of age.

What immunodeficiency should you suspect?

A. X-linked hyper-IgM
B. Chediak-Higashi syndrome
C. Leukocyte adhesion deficiency
D. Complement deficiency

Leukocyte adhesion deficiency (type I)
- Leukocytes lack the ability to adhere to blood vessel walls and to migrate to the site of infection
- Characteristics:
  - delayed separation of umbilical cord (LAD I)
  - leukocytosis
  - gingivitis and periodontitis
  - recurrent infections of skin, lungs, GI tract and perirectal area
  - often necrotic WITHOUT abscess formation
  - organisms: S. aureus and gram negative bacilli
- Defect: AR mutation in CD18, adhesion molecule that prevents or decreases expression of beta-2 integrins
- Diagnosis: flow cytometry to show the decrease of absence of CD18

Chediak-Higashi syndrome
- autosomal recessive disorder with recurrent infections and partial oculocutaneous albinism
- characteristic feature: giant lysosomal granules in neutrophils

X-linked hyper-IgM syndrome
- no isotype switching from IgM to other immunoglobulins
- susceptible to infections with encapsulated bacteria

Complement deficiency
- autosomal recessive
  - early (C1-C4): recurrent sinopulmonary infections due to encapsulated bacteria, often associated with increased incidence of systemic autoimmune disease
  - late (C5-C9): recurrent Neisseria infections and increased risk of meningitis
- Screen: CH50, measures complement activity

References:
GOOD LUCK!!