1. A 9-month-old infant presents with weight loss, fever, irritability, and petechial rash for 1 week. He has had multiple ear infections in the past and has a draining right ear. He has seborrhea on his scalp. He shows signs of dehydration on examination. Radiograph of the chest shows multiple pulmonary infiltrates. Which of the following is a true statement regarding this infant’s condition?

- a. A short course of methylprednisolone along with intravenous antibiotics is indicated
- b. A bone scan will be diagnostic of the condition
- c. The petechial rash is always due to thrombocytopenia
- d. Diagnostic workup includes urine osmolality
- e. The prognosis is excellent with appropriate therapy

Langerhan’s cell histiocytosis

- Variable presentation: Skeleton is involved in 80% (osteolytic lesions and pathological fractures)
- Destruction of mastoid causes chronic draining ears
- Skin (50%): seborrheic dermatitis, petechial hemorrhage even in the absence of thrombocytopenia
- Lymphadenopathy (30%), hepato splenomegaly (20%), pulmonary infiltrates (15%) and exophthalmos
- Pituitary dysfunction-DI, Failure to thrive
- Skin biopsy is diagnostic and prognosis is good for single system disease
- Systemic manifestations: fever, malaise and irritability
2. An 8 year-old girl presents with a history of fever, joint pain, and rash for three days. On examination she has swollen ankle and knee joints and a pericardial rub is heard on auscultation. Which of the following is the most appropriate next step in the management of this girl?

- a. Obtain a throat culture before administering antibiotics
- b. Administer intravenous immunoglobulin therapy
- c. Administer high dose methylprednisolone
- d. Perform bedside knee joint aspiration and send aspirates for bacteriologic cultures
- e. Consult a cardiologist for pericardiocentesis

Acute Rheumatic fever

- Group A beta hemolytic streptococci
- Pancarditis in 40-80% patients
- Polyarthritis: exquisitely tender joints, migratory and affects several different joints
- Sydenham chorea: may occur after many months of pharyngitis. Emotional lability is frequently present. Chorea may present as increasing clumsiness.
- Erythema marginatum: Rare, pink macule with blanching in the center and can be serpiginous
- Subcutaneous nodules: pea-sized on the extensor surface of limbs, firm and non tender
- Jones criteria

Jones Criteria

- Major: Carditis, polyarthritis, erythema marginatum, chorea and subcutaneous nodules
- Minor: Fever, arthralgia, elevated acute phase reactants, prolonged PR interval
- Plus evidence of preceding group A strep infection: Culture, rapid antigen or rising antibody titers
3. An 11 year-old boy presents with a history of fever, joint pain, and the rash. He also complains of abdominal pain and a swollen scrotum. He had a streptococcal throat infection a week ago. The following is true regarding his condition:

- a. He has scarlet fever
- b. His platelet count is likely be normal
- c. Broad spectrum antibiotic therapy is indicated
- d. He has Kawasaki’s disease
- e. Bone marrow aspirate will be diagnostic

HSP

- Typically follows URI
- IgA mediated vasculitis of small vessels
- Lesions occur in crops
- Arthritis with edema of joints
- Edema and bleeding in the GI tract can cause severe abdominal pain
- Renal involvement (25-50%): membranous glomerulonephritis
- CNS involvement is rare
- Indication for steroids: GI and CNS complications

4. A 14 year-old adolescent girl presents with exertional dyspnea and palpitations. Examination reveals a heart murmur. Each of the following is appropriate regarding her management, EXCEPT:

- a. She has a 1 in 4 (25%) chance of having an offspring with the similar condition
- b. She needs an ophthalmologic evaluation
- c. She requires endocarditis prophylaxis for dental and invasive surgical procedures
- d. She should receive beta blocker (propranolol) therapy
- e. A urinary cyanide nitroprusside test may help in the diagnosis
Marfan syndrome

- Autosomal dominant: 1/10000 live births with 30% spontaneous mutations
- Fibrillin gene
- Skeletal: dolichostenomelia, pectus excavatum, and scoliosis
- Cardiac: aortic root dilatation, MVP
- Ocular: lens dislocation, myopia and iridodendritis
- Lab should include negative urinary cyanide nitroprusside test
- Beta blocker therapy to slow the progression of aortic root dilatation. Endocarditis prophylaxis is indicated

5. A 13 month-old infant presents to the emergency department with a 6-days history of fever, rash, conjunctivitis, fissured lips, joint swelling, lymphadenopathy, and irritability. She had received her measles-mumps-rubella vaccine 7 days ago. The next most appropriate step is to:

- a. Reassure the parents that her symptoms are due to a reaction to the vaccine
- b. Administer intravenous penicillin G
- c. Administer high-dose methylprednisolone therapy
- d. Admit her and perform a lumbar puncture to rule out bacterial meningitis
- e. Consult a pediatric cardiologist for an echocardiogram

Kawasaki disease

- Diagnosis is made clinically. Next logical step is echo cardiography
- 20% of untreated patients develop cardiac complications
- Myocarditis and decreased ventricular function is 50% of patients
- Aseptic meningitis, cholecystitis and sterile pyuria
- Elevated inflammatory markers
- Diagnostic criteria
6. A 12 month-old infant is admitted for chronic enteritis, failure to thrive, and eczematous lesions over the perioral and perianal areas. Which of the following is true regarding her condition?

- a. Treatment with high-dose zinc with close monitoring of levels is indicated
- b. She may have an elevated serum alkaline phosphatase levels
- c. Therapy with acyclovir is indicated
- d. Gluten free diet should be introduced slowly
- e. It is inherited as an autosomal dominant pattern

Acrodermatitis enteropathica

- Autosomal recessive
- Vesiculo bullous lesions, chronic enteritis and malnutrition
- Also seen in secondary Zinc deficiency
- Diagnostic features: low serum Zinc and Alkaline phosphatase
- Dermatatis herpetiformis is in the DD and is typically associated with Celiac disease

7. A 16 year-old high school student presents with 1-week history of abdominal pain, vomiting, and loss of appetite. On examination she has scleral icterus and is mildly dehydrated. Blood work reveals a Hb 6gm/dL, HCT 21, WBC12000/mm³, Platelets 260000/mm³, and reticulocytes 32%. Liver profile show elevated liver enzymes. Urine analysis is positive for glycosuria, aminoaciduria, and phosphaturia. The next most appropriate step is:

- a. Reassure her that she has infectious hepatitis and arrange for a follow up in 2 weeks
- b. Admit her to the hospital and start N-acetyl cysteine therapy
- c. Order an urgent abdominal ultrasound examination of her abdomen
- d. Perform slit lamp examination of her eyes
- e. Order a bone marrow aspiration
Wilson disease

- Autosomal recessive (Chromosome 13)
- Hepatic involvement in young and neurological manifestations in older patients
- Ceruloplasmin is low and elevated urinary copper excretion/ more after D-penicillamine therapy
- Neurological, hepatic, renal and hemolytic anemia
- KF is almost always present in patients with neurological symptoms

8. A 5 year-old child presents for a pre-kindergarten physical examination. The mother of the child is concerned because the child doesn't play with his friends and tires easily. Further history reveals that he first walked at 22 months of age. The following are all true regarding his condition EXCEPT:

- a. He is at risk to develop intellectual impairment
- b. There is a 50% chance that his son will be affected with the same condition
- c. He is at risk to develop heart failure
- d. His mother may not be a carrier of the disease
- e. Further work up, including muscle biopsy, is indicated

DMD

- Most common hereditary (X-linked recessive) neuromuscular disease
- Delayed walking is an indication to check serum CK levels
- Gowers sign is evident by the age of 3 and trendelenburg gait develops afterwards
- Relentless progression of muscle weakness and wheelchair bound by the age of 10
- Cardiomyopathy is a constant feature
- Intellectual impairment occur in all patients
- EMG shows myopathic features but Muscle biopsy is diagnostic
9. A 12-year-old young man who has recently immigrated from Eastern Europe presents with a one-month history of intermittent fevers and a painful rash on his shins. He also has lost 20 pounds in the past month. Which of the following should be obtained/performed as part of the initial work-up?

- a. A cardiology workup for rheumatic fever
- b. Urinalysis for glycosuria
- c. A tuberculin skin test
- d. An allergy panel
- e. Barium enema

Erythema nodosum

- The nodules are shiny red and painful
- Hypersensitivity reaction
- Tuberculosis, IBD, strep infections, sarcoidosis, histoplasmosis, and yersinia
- Drugs: sulfa, phenytoin and OCPs

10. A 16-year-old female presents to the clinic with fever, headache, and an erythematous malar facial rash. She tires easily and has arthralgia. The most appropriate management at this point would be to:

- a. Prescribe oral antibiotics and hydrocortisone locally for the rash
- b. Order blood work for antinuclear antibody (ANA)
- c. Arrange for a cranial CT scan
- d. Start intravenous acyclovir
- e. Start tetracycline therapy
SLE

- ANA is present in 80% and is a screening test
- ANA is positive in SLE, JRA, scleroderma, mononucleosis, and chronic active hepatitis
- Antibody to double stranded DNA
- Anti-Rho antibody in SLE mom can cause heart block in the neonate

11. A 12 year-old young lady is seen for intractable acne. She has been treated with multiple topical therapies without benefit. Further history reveals that she has a sibling with a seizure disorder. On physical examination she has a hypopigmented patch on her trunk. Of the following, the next most appropriate plan of action for this girl would be to:

- a. Treat her with oral tetracycline therapy
- b. Refer her to a dermatologist
- c. Obtain a renal ultrasound
- d. Order karyotyping
- e. Treat her with anticonvulsant therapy

12. An 11 year-old girl is seen for fever, sore throat and rash. Which of the following is true regarding her diagnosis and management?

- a. Treatment with ampicillin might make the rash worse
- b. Household contacts of the child should receive Rifampicin prophylaxis
- c. An ECG and an ASO titer should be obtained
- d. High-dose aspirin and immunoglobulin therapy is indicated
- e. None of the above
Varicella

- Progressive varicella: visceral organ involvement, coagulopathy, and severe hemorrhagic vesicles
- Patients present in severe abdominal pain and appearance of hemorrhagic vesicles
- Immuno-compromised patients: Congenital cellular immune deficiency, malignancy and recent chemotherapy
- Can occur in patients who receive corticosteroids including inhaled steroids
- Management is supportive

13. The parents of a 5-year-old boy are concerned regarding his headache, which doesn’t seem to get better with acetaminophen. The boy has growth hormone deficiency and receives growth hormone therapy regularly. He is fully immunized and takes regular vitamin supplementation. There are no focal neurological signs. The following is true statements regarding his condition EXCEPT:

• a. A lumbar puncture with opening pressure measurement is indicated after obtaining a cranial CT scan
• b. The condition may be due to hypervitaminosis A
• c. The growth hormone therapy has to be discontinued
• d. Repeated lumbar puncture may be required
• e. Acetazolamide therapy may be helpful

Pseudotumor cerebri

- Alteration in CSF production, absorption
- Metabolic disorders: galactosemia, hypoparathyroidism, hypophosphatasia
- Prolonged corticosteroid and growth hormone therapy
- Hypervitaminosis A, obesity, contraceptive pills
- Infections: roseola infantum, chronic otitis, GBS,
- Polycythemia, Iron deficiency anemia, SVC obstruction
14. A 4 year-old boy has a history of fever for 4 days. He was evaluated at a community emergency department two days ago and was diagnosed with otitis media and prescribed trimethoprim-sulfamethoxazole. Today, he developed a rash. On physical examination, he has cool extremities and exhibits altered mental status. The most important initial step in management should include:

- a. Obtaining an urgent CT scan of the head
- b. Obtaining rapid intravenous access and administer rapid fluid bolus
- c. Administering intravenous steroids
- d. Performing a lumbar puncture to rule out meningitis
- e. Administering trimethoprim-sulfamethoxazole intravenously

Steven Johnson syndrome

- Muco-cutaneous lesions, vesiculo bullous lesions and areas of denudation
- Fluid loss equivalent to burns patient
- Mycoplasma pneumoniae, NSAID, anticoagulants and sulfas
- Supportive management