Question 1

A 36-year-old high school teacher with a 10-year history of SLE presents to a local emergency room with painful swelling of her left calf and pleuritic right-sided chest pain.

Her past medical history is notable for episodes of transient thrombocytopenia responding to corticosteroid therapy. She has had cutaneous lesions, Raynaud's phenomenon, and mild proteinuria with otherwise normal renal function. She has two children, ages four and six, and has had three first trimester miscarriages. She has never used oral contraceptive pills, does not smoke, and exercises regularly. There is no family history of systemic lupus erythematosus, other systemic rheumatic disease, or thrombotic disorders.

The physical examination reveals a blood pressure of 110/60 mmHg, pulse of 86 per minute, respirations of 20, and oxygen saturation breathing room air is 92 percent. There is mild bilateral malar erythema and livedo reticularis over both thighs. A pleural friction rub is present posteriorly on the right. Heart sounds are normal. The left calf is swollen and tender to palpation.

Laboratory studies include normal complete blood count, serum creatinine, and liver function tests. Chest radiograph is normal.

Ventilation/perfusion lung scanning reveals a high probability of pulmonary embolism.

The prothrombin time is normal. Lupus anticoagulant activity using the activated partial thromboplastin time as the screening test is normal.

Antiphospholipid antibodies (ELISA):
IgG = 71 GPL (<20 GPL)
IgM = 40 MPL (<20 MPL)

You initiate therapy with heparin. The optimal regimen relating to oral anticoagulation therapy for this patient is?

A. The administration of warfarin for three to six months
B. Life-long warfarin to achieve an INR of 2.0 to 2.5
C. Warfarin for three months, followed by life-long daily aspirin
D. The administration of warfarin for 12 months
E. Life-long warfarin to achieve an INR of 3.0 or higher

Question 2

A 24-year-old woman with a history of limited scleroderma is seen for a follow-up visit. She initially developed Raynaud's phenomena followed by the onset of painful pitting of the distal digital tufts of several fingertips.

Which of the following statements regarding the pathogenesis of primary and secondary Raynaud's phenomena is most accurate?

A. In primary Raynaud's phenomena, the aberrant adrenergic response is mediated primarily by alpha-1 adrenergic receptors.
B. Increased activity of nitric oxide in secondary Raynaud's phenomena results in enhanced smooth muscle contraction and platelet aggregation, thereby contributing to the disease process.
C. Endothelial dysfunction is likely a late event in patients with progressive systemic sclerosis and Raynaud's phenomena, reflecting cumulative damage incurred by multiple anoxic events.
D. The potent vasoconstrictor, endothelin-1, is over-expressed in patients with scleroderma and secondary Raynaud's phenomena.

Question 3

A 63-year-old nursing home resident is referred to you for management of the eosinophilic-myalgia syndrome (EMS). She developed this syndrome years ago, following the use of the dietary supplement L-tryptophan to improve her sleep and mood. Features included a low-grade fever, fatigue, a nonproductive cough, and diffuse muscle pains. The eosinophil count ranged as high as 8,000 cells per uL. She has been unable to walk without assistance for the past year and was transferred to this facility for long-term care. The internist admitting the patient is puzzled by her findings; she had considered EMS to be a form of scleroderma.

Upon reviewing the various organ systems that may be involved in EMS, which one of the following features would have been expected to occur in this patient during the initial illness?

A. Pitting edema of the extremities
B. Abnormal electromyographic studies
C. Respiratory failure
D. Synovitis
E. Significant heart disease

Question 4

A 70-year-old woman with a 30-year history of rheumatoid arthritis presents with progressive right lateral thigh pain radiating toward the knee. The pain began abruptly two weeks ago when she lifted a box of books in the basement.

The physical exam demonstrates normal range of motion of both hips; the right lateral thigh is normal to palpation, but there is slightly diminished sensation to pinprick and light touch. The deep tendon reflexes are 2+ at both knees and ankles, and proximal and distal leg strength is intact. An electromyogram (EMG) and nerve conduction study (NCS) are normal.

You suspect the diagnosis of meralgia paresthetica. Which of the following statements regarding this condition in this patient is correct?

A. This disorder is due to a lesion involving the obturator nerve.
B. This diagnosis is not consistent with this patient's findings.
C. In more advanced cases, there is a fixed sensory loss over the lateral thigh.
D. Because of her RA, she has a greater risk for developing this condition.
E. The normal EMG/NCS (nerve conduction study) rules out this diagnosis.

Question 5

A 47-year-old woman with a two-year history of systemic sclerosis with diffuse scleroderma is admitted to the hospital following a seizure at home. Upon arrival, her blood pressure was 190/130 mmHg. She was confused, but without focal neurologic deficit. Fundoscopic examination revealed papilledema. There was thickening of the skin over the face, chest, forearms, thighs, calves, and feet.

Her serum creatinine was 2.4 mg/dL [212.1 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L]). Urinalysis reveals 1+ proteinuria, but no cells or casts. The hemoglobin is 10.2 g/dL [6.3 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L]) and schistocytes are noted on the smear.

Intravenous phenytoin and nitroprusside were started. You see her in consultation a few hours later. On your advice, the nitroprusside is discontinued and intravenous enalaprilat is started, 1.25 mg every six hours.

Two days later, her blood pressure is 160/118 mmHg. She is alert and oriented. The hemoglobin has declined to 9.2

g/dL [5.7 mmol/L], and the creatinine is 4.0 mg/dL [353.6 mmol/L]. The renal consultant recommends that enalaprilat be discontinued and that hydralazine and minoxidil should be started.

What is the best course of action?

A. Plasmapheresis
B. Increase the enalaprilat dose
C. Add hydralazine to enalaprilat
D. Maintain current therapy
E. Follow recommendations of the nephrologist

Question 6

A 69-year-old woman with a 25-year history of rheumatoid arthritis is brought by her daughter for evaluation. The daughter is concerned about her mother's recent functional decline, in particular her poor gait. In the past she has been able to ambulate with the use of a cane, but for the past three months she has required a walker. She finds her mother holding onto the furniture when she walks from room to room. The patient is aware of this decline. She denies any new joint pain or discomfort. She also denies muscle pain, numbness, or dysesthesia.

Her medical history reveals RA treated in the past with methotrexate and multiple other DMARDs without benefit. She has undergone bilateral knee and right total hip replacements within the past decade. Current medications include prednisone 5 mq qd, ibuprofen 600 mg tid, and lovastatin 10 mg qd for hyperlipidemia.

Physical examination reveals a frail, elderly woman, who is unsteady on her feet.

Joint examination shows subluxation of the MCP joints and ulnar deviation bilaterally. Hips and knees show full range of motion. Feet show pes planus deformity and valgus angulation at the ankles. There is no active synovitis.

Motor strength is 4/5 in the proximal muscles of both upper and lower extremities. Deep tendon reflexes are 4+ in the upper extremities and lower extremities with a few beats of unsustained clonus at the ankles. Sensory exam demonstrates reduction in toe proprioception. She cannot walk heel to toe without assistance.

What would you do next?

A. Check CK
B. Recommend discontinuation of prednisone and initiation of an anti-TNF therapy
C. Order electromyelography (EMG) and nerve conduction studies
D. Order MRI of cervical spine
E. Order MRI of lumbar spine

Question 7

A 44-year-old man with a squamous cell carcinoma of the neck received three cycles of chemotherapy consisting of bleomycin, methotrexate, and cisplatin. Six months later he developed digital ischemia affecting all of his fingers, progressing over the course of a few months to gangrenous changes at multiple digital tips. He also complained of mild dyspnea. A rheumatology consultation was requested.

All of the following findings may be observed on physical and laboratory examination EXCEPT:

A. Sclerodactyly
B. Interstitial lung disease
C. Raynaud's phenomenon
D. Positive ANA

Question 8

A 66-year-old man with longstanding polyarticular gout is seen for an acute flare. On examination, you note 12 swollen and painful joints. Because of a history of prior gastrointestinal bleeding with the use of nonsteroidal antiinflammatory drugs and a history of diabetes mellitus, you decide to initiate therapy with colchicine.

Which of the following statements regarding colchicine is true?

A. Long-term use of colchicine can cause a mitochondrial myopathy.
B. Colchicine is less effective in treating attacks of pseudogout than gout.
C. Intravenous colchicine in a bolus of 1 mg can be administered every three hours until the attack has resolved.
D. Suppression of the influx of inflammatory cells into synovium may in part underlie the prophylactic action of colchicine.
E. In patients with poor intravenous access, colchicine can be administered intramuscularly.

Question 9

A 15-year-old boy is referred to you regarding further management of his Henoch-Schönlein purpura. He developed an acute illness characterized by polyarthralgias involving the knees and ankles and a purpuric skin rash over the legs. A urinalysis showed mild proteinuria and occasional red cell casts. Two stool specimens were positive for occult blood. Other lab studies including the complete blood count, liver function studies, and creatinine were normal. No specific therapy was ordered and he improved slowly over the next four weeks. He is here today with his parents, who seek your opinion regarding the long-term prognosis of their son's condition.

All of the following statements regarding the prognosis of Henoch-Schönlein purpura (HSP) are true EXCEPT?

A. Complete recovery occurs in about 90 percent of adults and children.
B. Recurrences are rarely seen.
C. The renal prognosis is excellent in most patients.
D. Recurrences are more likely to occur in patients with nephritis.
E. All of the manifestations of active HSP usually resolve spontaneously.

Question 10

A 53-year-old previously healthy attorney is admitted to the hospital for acute shortness of breath and bloody sputum. She describes the abrupt onset of cough and bloody sputum 24 hours earlier. She denies any fever, chills, skin rash, joint or muscle pain, recent respiratory tract symptoms, or recent travel.

Past history is noted for excellent health. She is an avid marathon runner. She has never smoked or used illicit drugs. She does not take any medications.

Physical exam reveals a temperature of 99.9°F (37.7°C), respiratory rate is 22, pulse is 100, and her blood pressure is 120/90 mmHg.

Except for bibasilar rales at the bases of the lungs, the exam is otherwise normal.

Laboratory results:

Hemoglobin = 11.5 g/dL [7.1 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 12,300/mm3 with 75 percent polys 22 lymphs 3 mononuclear (normal = 4,300 to 10,800/mm3)
Platelets = 340,000/mm3 (normal = 130,000 to 400,000/mm3)

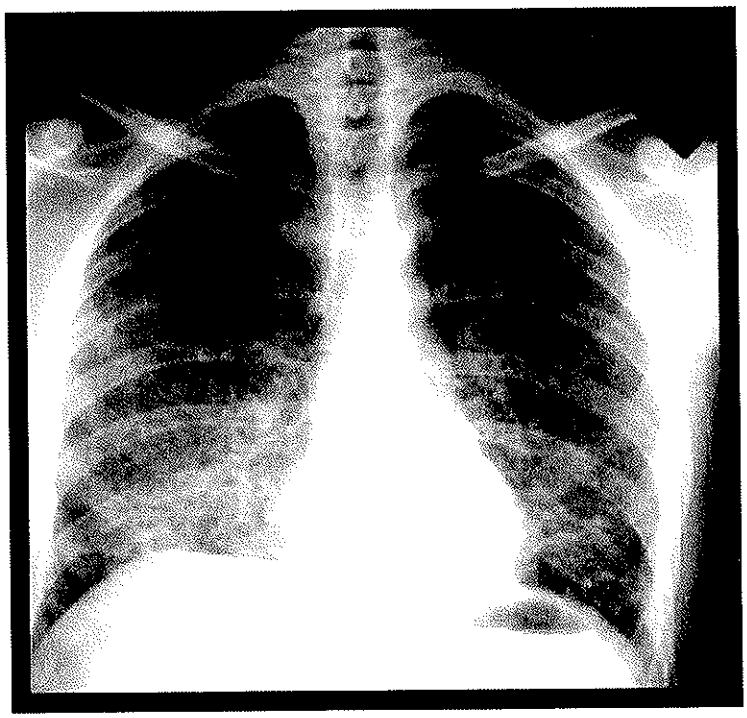
ESR = 35 mm/hr (normal = 0 to 30 mm/hr)
Creatinine = 1.1 mg/dL [97.2 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
AST = 34 IU/L (normal = 35 IU/L)
ALT = 24 U/L (normal = 35 IU/L)

ANA: 1:640
ANCA: negative
anti-GBM: negative

Other serologies are pending

Arterial blood gas (room air): pH 7.37, pO2 65, pCO2 31
Urinalysis: Trace protein, no cells or casts

A chest radiograph is taken (see figure). Lung biopsy is performed, which shows bland hemorrhage.



Courtesy of Marvin I Schwarz, MD.

What is the most likely diagnosis?

A. Wegener's granulomatosis
B. Churg Strauss syndrome
C. Systemic lupus erythematosus
D. Microscopic polyangiitis
E. Polyarteritis nodosa

Question 11

A 52-year-old man with longstanding, seropositive rheumatoid arthritis presents to you for a second opinion regarding his treatment. His disease has been severe, resulting in multiple joint deformities and requiring bilateral knee arthroplasties and bilateral metatarsal head resections.

In the past two years, he has had recurrent ulcers on his legs, two episodes of bacterial pneumonia, and a soft tissue abscess. These events occurred while receiving myochrysine 50 mg IM every two weeks. The myochrysine was dis-

continued, and methotrexate was initiated at a dose of 10 mg per week and increased over the next six months to 20 mg per week.

His laboratory studies included the following:

Hemoglobin = 10.2 g/dL [6.3 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 1,500/mm3 with 45 percent neutrophils and 35 percent lymphocytes (normal = 4,300 to 10,800/mm3)
Platelets = 133,000/mm3 (normal = 130,000 to 400,000/mm3)
Serum creatinine = 1.2 mg/dL [106.1 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
Liver function studies: normal
Rheumatoid factor = 1200 IU/mL

Methotrexate has brought his arthritis under control, but has failed to improve the appearance or size of the ulcers. Two weeks before his appointment with you he developed a soft tissue abscess over his buttock.

On physical examination his blood pressure is 130/76 mmHg, pulse is 68/min, respiratory rate is 14/min, and temperature is 98.8°F (37.4°C). General physical examination reveals a debilitated man. There is splenomegaly without lymphadenopathy. There is no active synovitis in any of the peripheral joints. However, there is reduced shoulder abduction bilaterally, flexion contractures of the elbows, MCP joint subluxation in both hands, and significant valgus deformity of both feet with pes planus and healed surgical scars over the MTP heads.

There are numerous rheumatoid nodules on the elbows and hands. There are three clean ulcers over the lower extremities, each measuring 2 to 3 cm in diameter. There is a well-circumscribed abscess over the right buttock measuring 3 x 5 cm.

His current laboratory data include the following:

Hemoglobin = 10.0 g/dL [6.2 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
MCV = 94 fL (normal = 80 to 96 fL)
WBC = 1,500 with 50 percent neutrophils and 35 percent lymphocytes (normal = 4,300 to 10,000/mm3)
Platelets = 105,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 45 mm/hr (normal = 0 to 20 mm/hr)
Serum creatinine = 1.2 mg/dL [106.1 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
Liver function studies: normal

All of the following may be useful therapeutic options EXCEPT?

A. Methotrexate 25 mg per week given parenterally
B. Granulocyte-stimulating colony factor (G-CSF)
C. Etanercept
D. Splenectomy

Question 12

A 35-year-old, African-American, married woman with two young children presents with a four-week history of sore throat, fever, malaise, and joint pain. She describes pain in elbows, wrists, and knees. The pain seems to skip from one joint to the next. Currently, the right wrist is most painful. She has been taking naproxen 400 mg daily with acetaminophen 2,000 mg daily with partial relief.

She denies any skin rash, history of sexually transmitted diseases, vaginal discharge, irregular menses, or recent sexual activity. Her last menstrual period was 10 days ago. She recalls having growing pains as a child that forced her to miss school for one month. Her family history reveals that both her parents died of cancer two years ago.

On physical examination, she appears somewhat toxic. Her temperature is 102°F (39.2°C), pulse is 130/min, respiratory rate is 15/min, and her blood pressure is 122/78 mmHg.

There is no conjunctivitis, facial rash, or skin lesions. There is an erythematous soft palate with tonsillar enlarge-

ment. There are a few small, tender cervical lymph nodes. Chest and cardiac examinations are unremarkable. There is no organomegaly.

Joint examination reveals tenderness of the right radiocarpal joint and tenosynovial swelling of the right 3rd and 4th extensor tendons.

Aspiration of the right wrist did not yield fluid.

Preliminary laboratory results reveal:

Hemoglobin = 12 g/dL [7.4 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 10,000 cells/mm3 with 70 polymorphonuclear leukocytes (normal = 4,300 to 10,800/mm3)
Platelets = 250,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 55 mm/hr (normal = 0 to 20 mm/hr)
ANA = 1:160
Urinalysis: normal

Which option is the most appropriate?

A. Obtain cultures of blood, pharynx, vagina, and rectum and begin doxycycline 100 mg twice daily for 10 days
B. Obtain throat culture and antistreptolysin O titer and begin penicillin V 250 mg TID for 10 days and aspirin
C. Obtain cultures of blood, pharynx, vagina, and rectum and begin ceftriaxone, intravenously, 1 g daily for 10 days
D. Test for serum antibodies to parvovirus (IgM and IgG) and treat with ibuprofen 800 mg tid
E. Obtain an anti-ds DNA antibody titer, serum C3, C4 and begin prednisone 20 mg per day

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Question 13

A 70-year-old man admitted to the Gastroenterology Service is referred to you for consultation regarding polyarthralgias, weight loss, and diarrhea. He has noted pain in his joints for the past two years, primarily the knees, shoulders, and elbows. He has 30 minutes of morning stiffness. He denies joint swelling. More recently, he has developed abdominal pain and diarrhea with a 20-pound (9 kg) weight loss.

Past medical history is notable for a 30-pack per year smoking history. There is no history of foreign travel or exposure to pets. Medications include ibuprofen 600 mg tid and atenolol 50 mg qd for hypertension.

Physical examination reveals a cachectic-looking male. Blood pressure is 115/68 mmHg, pulse is 66, respiratory rate is 14, and temperature is 98.6°F (37.0°C).

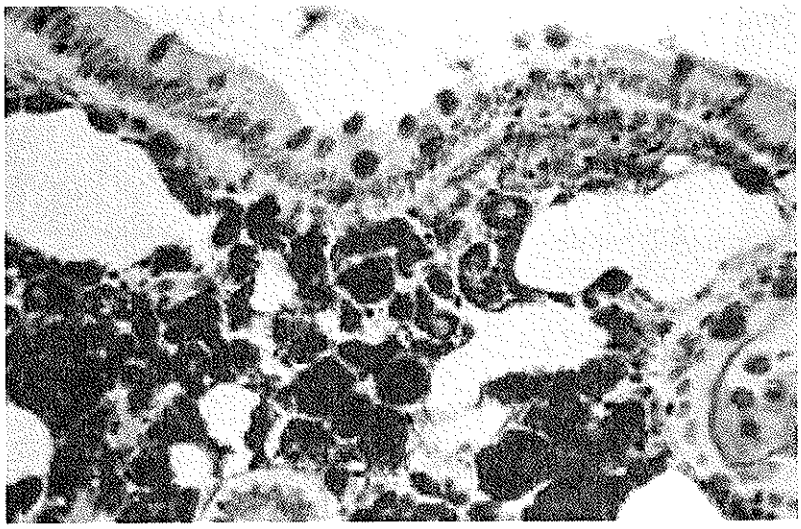
The chest and cardiovascular exams are unremarkable. The joint exam is notable for full range of motion in all joints, but with pain on motion of the shoulders, elbows, and knees. There are no effusions. The abdomen is slightly distended with some epigastric discomfort on palpation.

Laboratory results:

Hemoglobin = 11.0 g/dL [6.8 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 6,800 with 55 percent neutrophils, 40 percent lymphocytes (normal = 4,300 to 10,800/mm3)
Platelets = 400,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 40 mm/hr (normal = 0 to 20 mm/hr)
AST = 34 IU/L (normal = 35 IU/L)
ALT = 36 IU/L (normal = 35 IU/L)
Alk phosphatase = 117 IU/L [2.0 µkat/L] (normal = 30 to 120 IU/L [0.5 to 2.0 nkat/L])
TSH = 3.7 mU/L (normal = 0.4 to 5 mU/L)

Blood cultures X 2 negative.
Stool cultures X 2 negative for ova or parasites.

A PAS stain of the small biopsy is performed (show figure).



Courtesy of Robert Odze, MD.

Which one of the following statements regarding this disorder is true?

A. His gastrointestinal tract symptoms are the most frequently observed features in this condition.
B. He should be treated with tetracycline.
C. He is likely to be HLA-B27 positive.
D. Articular symptoms usually develop following the onset of gastrointestinal complaints.
E. The lack of an inflammatory response in affected tissues is a striking feature of this condition.

Question 14

A 62-year-old man with a 30-year history of erosive, seropositive, nodular rheumatoid arthritis was doing well until two months ago when he developed numbness and burning of both feet. Over the past two weeks, he has been tripping over the rugs in his home and feels that his feet are "slapping" the ground when he walks. He has been unable to dorsiflex his right wrist. At the same time, he noticed a reddish discoloration of some fingernails and the pulps of all his toes. Two of his toes have since turned black. He has lost 15 pounds (6.75 kg) over the past three months due to the loss of appetite and low-grade fevers.

Six months ago, he underwent coronary angiography for chest pain that revealed single-vessel disease. He was advised to quit smoking, but continues to smoke two packs per day. His past history is otherwise negative.

Current medications are ibuprofen 800 mg TID and aurothioglucose 50 mg IM biweekly both unchanged for the past four years.

Laboratory studies reveal a normocytic, normochromic anemia.

Hemoglobin = 10.1 g/dL [6.3 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
ESR = 120 mm/hr (normal = 0 to 20 mm/hr)
Rheumatoid factor = 788 IU/L

Serum cryoglobulins, total hemolytic complement, creatinine concentrations, and urinalysis are normal.

Which of the following disorders most likely explains this patient's recent problem?

A. Secondary amyloidosis

B. Buerger's disease
C. Rheumatoid vasculitis
D. Cholesterol emboli
E. Gold neuropathy

Question 15

A 49-year-old man presents with a four-month history of myalgias and headaches. The onset of the pain occurred a few days after he spent an afternoon chopping firewood. He states that his pain is worse on the right side of his neck and extends to the right posterior shoulder and the right mid-back. Headache occurs frequently and usually involves the right side of the head. He obtains partial relief with the use of a heating pad. Multiple medications including NSAIDs, muscle relaxants, and narcotics have been ineffective. He says he has become depressed because of the severe constant nature of the pain. His past medical history is notable for hypercholesterolemia for which he takes pravastatin.

Physical exam is remarkable for tenderness over the right occipital area; palpation of this region results in a pain sensation radiating to the right temple. He has multiple tender muscular sites, all of which are on the right side including the right upper, mid, and lower trapezius, the right deltoid and subacromial regions, and the right paralumbar regions. There is no evidence of left sided muscle tenderness. Neck has slow but complete range of motion. Peripheral joints show full active range of motion without bogginess, redness, or warmth. Active shoulder internal and external rotation against resistance does not produce pain. Shoulder impingement tests are unremarkable. Neurological exam (including pinprick, motor strength, and deep tendon reflexes of the upper extremities) is normal. The complete blood count, liver function studies, and ESR are all normal. Radiographs of the cervical and thoracic spine are normal.

This patient's most likely diagnosis is?

A. Fibromyalgia
B. Cervical radiculopathy
C. "Statin" myopathy
D. Regional myofascial pain syndrome
E. Polymyalgia rheumatica

Question 16

A 54-year-old Caucasian woman is referred to you by an otolaryngologist for further evaluation of Sjögren's syndrome. She has a one-year history of recurrent upper airway and sinus infections. A diagnosis of primary Sjögren's syndrome was made on the basis of a labial gland biopsy.

Laboratory evaluation reveals:

anti-Ro/SSA = 118 IU
anti-La/SSB = 88 IU

Which of the following statements about anti-Ro/SSA and anti-La/SSB antibodies is correct?

A. Among women with the following features: give birth to children with congenital complete heart block; are positive for anti-Ro/SSA and anti-La/SSB antibodies; and are initially without evidence for SLE or other connective tissue disease, the great majority eventually develop SLE or Sjögren's syndrome on follow-up.
B. High titers of anti-SSA/Ro autoantibodies are associated with a high incidence of extraglandular features of Sjögren's syndrome.
C. In patients with SLE or Sjögren's syndrome, it is common to encounter sera that contain anti-La/SSB activity without demonstrable antibodies to Ro/SSA.
D. There is no linkage between the production of anti-Ro/SSA antibodies, C4 complement component deficiency,

and the HLA-DR3 genotype.

Question 17

A 42-year-old cook at an Asian restaurant is referred for evaluation of intense myalgias and weakness. The pain began about four weeks ago affecting both arms and legs. He describes excellent health until six weeks earlier, when he developed a productive cough and fever, which was treated with a week of erythromycin. Cough and fever resolved but shortly thereafter he noted some nausea and diarrhea lasting one week.

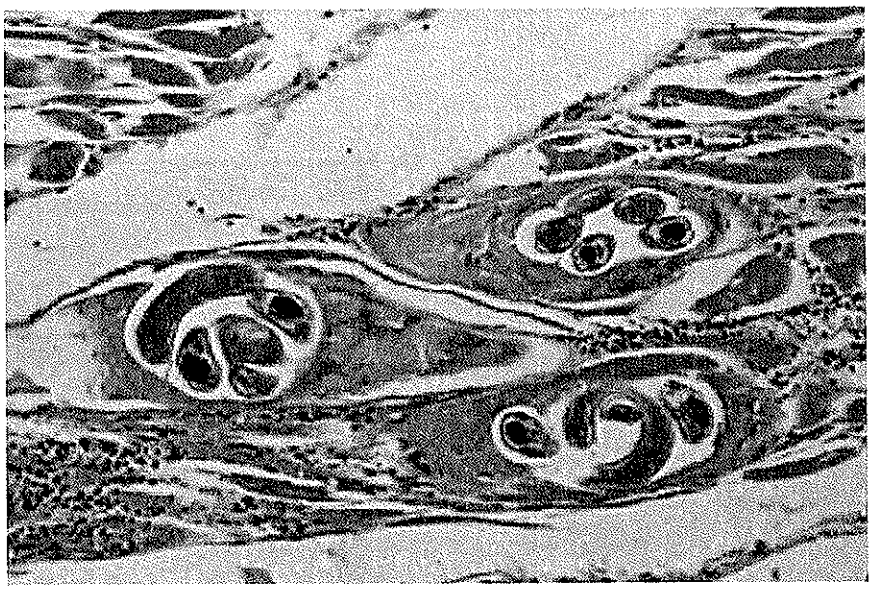
Physical examination reveals an uncomfortable appearing man with normal temperature. Lungs are clear. Heart rate and rhythm are normal, no murmur, gallop, or rub are noted. There is good muscle strength, but moderate muscle tenderness.

Past medical history is notable for hypercholesterolemia and gout, which are treated with lovastatin 20 mg/day and colchicine 0.6 mg BID, respectively. He drinks one to two glasses of plum wine per day, uses marijuana occasionally, but denies use of "hard drugs," specifically cocaine, amphetamines, and heroin.

Laboratory results:

Hemoglobin = 13.4 g/dL [8.3 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 14,900/mm3 (normal = 4,300 to 10,800/mm3) with 40 percent neutrophils, 30 percent lymphocytes, and 18 percent eosinophils
CK = 1150 U/L [19.2 µkat/L] (normal = 60 to 400 U/L [1.0 to 6.7 µkat/L])
AST = 66 U/L (normal = 35 U/L)
ALT = 52 U/L (normal = 35 U/L)
LDH = 870 U/L [14.5 µkat/L] (normal = 110 to 210 U/L [1.8 to 3.5 µkat/L])

A muscle biopsy of the deltoid muscle is shown (see figure).



Courtesy of Peter F Weller, MD.

Which of the following statements regarding this condition is true?

A. The disappearance of eosinophilia is a good prognostic sign.
B. Cardiac involvement is the most frequent cause of death.
C. Involvement of diaphragmatic muscles is rarely seen.
D. Central nervous system involvement is the most common cause of death.
E. Treatment with intravenous immune globulin is highly effective.

Question 18

A 45-year-old man is referred because of back pain. It is a dull ache intermittent in nature. However, he is most concerned about loss of height. In the last five years, he has shrunk by 2 inches (5 cm). He denies trauma, alcohol use, or chronic medical problems.

His review of systems is unremarkable. The physical examination is normal except for a moderate kyphosis.

Laboratory evaluation reveals normal blood count, serum calcium, serum phosphorus, and serum protein electrophoresis. Lateral chest x-ray shows multiple compression fractures.

To evaluate this patient further, all of the following studies could be helpful EXCEPT?

A. Serum vitamin D
B. Serum TSH
C. Serum testosterone
D. Urinary excretion of cross-linked N-telopeptides (NTX)
E. Bone mineral density

Question 19

A 74-year-old man with a history of rheumatoid arthritis and total knee replacement five years earlier is admitted to the hospital with fever and swelling of his prosthetic knee. Radiographs of the knee show evidence of lucency around the borders of the prosthesis, and synovial fluid obtained from the knee has a leukocyte count of 120,000 WBC/mm3.

His health is otherwise good with no history of cardiac or pulmonary disease.

Which of the following approaches is most likely to result in a successful outcome?

A. Intravenous antibiotics for three weeks with daily aspirations until synovial fluid accumulation subsides, followed by six weeks of oral antibiotic therapy
B. Intravenous antibiotics and surgical debridement of underlying bone with replacement of a new prosthesis during the same operation
C. Intravenous antibiotics continued for six weeks, arthroscopic debridement, and repeat arthroscopy with cultures at six weeks
D. Intravenous antibiotics until fever subsides, followed by indefinite suppressive oral antibiotic therapy
E. Removal of the infected prosthesis and replacement arthroplasty after six weeks of intravenous antibiotics and repeat negative joint fluid cultures

Question 20

An 83-year-old woman is referred to you for management of her osteoporosis. She presented two weeks ago to the Emergency Room with the acute onset of pain at the lower thoracic spine after lifting a turkey from her oven. The pain has been persistent and incapacitating. Muscle relaxants and narcotic analgesics have provided limited benefit.

She reached menopause at age 50 and never started hormone replacement therapy. There is no history of cigarette or alcohol use. She has no history of corticosteroid use.

The examination is remarkable for her stooped posture, unsteady gait, and vertebral percussion tenderness over T12.

Laboratory testing reveals a normal CBC, ESR, serum protein electrophoresis, and urinalysis. PTH and vitamin D levels are normal.

Films of the thoracic spine demonstrate compression deformity of the T10 and T12 and the L2 vertebral bodies.

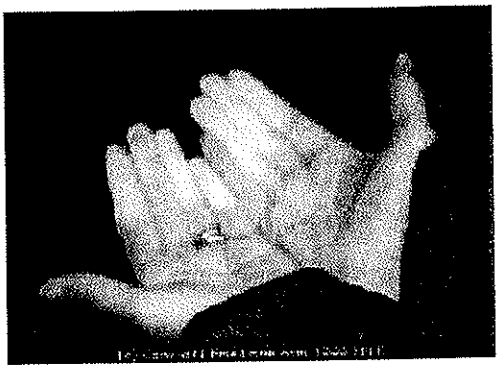
You suggest initiation of therapy with nasal calcitonin. The patient's daughter takes alendronate and wants to know what the benefits and drawbacks are for each of these drugs.

Which of the following statements regarding nasal calcitonin and alendronate is correct?

A. Nasal calcitonin use is contraindicated in women taking estrogen replacement therapy.
B. Nasal calcitonin is an effective therapy for the prevention of osteoporosis in premenopausal women.
C. Hypocalcemia with alendronate use is a common side effect.
D. Nasal calcitonin provides effective analgesic relief in patients with vertebral compression fractures.
E. Tachyphylaxis to nasal calcitonin does not occur.

Question 21

A 30-year-old actor is referred to you for treatment of her new onset Raynaud's phenomenon (see figure). For the past four years she has been taking propranolol 25 mg BID for management of stage fright. She says that she is fearful of gaining weight so she smokes 5 cigarettes per day and uses diet pills "when necessary."



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You would recommend all of the following options for the treatment of her Raynaud's phenomenon EXCEPT?

A. Encourage her to stop smoking
B. Discontinue propranolol
C. Prescribe nifedipine
D. Discontinue diet pills
E. Begin behavioral therapy with relaxation

Question 22

A 42-year-old nurse recently developed explosive-onset rheumatoid arthritis with polyarticular disease, nodules, and a positive rheumatoid factor. She has a 10-year history of asthma, which has periodically required therapy with corticosteroids. Two years ago, following the ingestion of aspirin for a headache, she had a particularly bad flare of her asthma associated with an episode of angioedema. Although she had taken aspirin and other non-steroidal medications (NSAIDs) without problems prior to this episode, she has subsequently avoided all aspirin-related medications. She has no history of nasal polyposis.

Which of the following statements regarding the potential use of NSAIDs and the risk of inducing an asthma flare in this patient is correct?

A. Any NSAID including aspirin could be used with minimal risk of a future episode.
B. Any NSAID except aspirin could be used, as there is no crossover reaction.
C. Non-acetylated salicylates are generally not associated with aspirin-induced asthma.
D. Cyclooxygenase-2 selective inhibitors would be the safest alternative agents.
E. An asthma exacerbation would be expected with all aspirins and NSAIDs.

Question 23

A 60-year-old Caucasian woman presents with worsening numbness in her feet and hands. She has a longstanding history of fibromyalgia and feels that her fatigue and myalgia have worsened over the past six months. She complains of a gritty sensation in her eyes, although she is capable of crying. She notes a dry mouth and recently has required care for three dental caries.

Current medication is amitriptyline 20 mg qhs. There is a history of significant alcohol use.

Physical examination reveals pale conjunctivae, slight enlargement of the parotid glands bilaterally, and diminished oral salivary pool. There are crackles at the bases of both lungs. Neurologic exam reveals a decrease in pinprick sensation in a "stocking-glove" distribution. There is no muscle weakness. All 18 fibromyalgia trigger points are tender. There is no joint synovitis.

Laboratory data reveals:

Hemoglobin = 13.6 g/dL [8.4 mmol/L] (normal =12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 8,700/mm3 (normal = 4,300 to 10,800/mm3)
Platelets = 320,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 42 (normal = 0 to 30 mm/h)
AST = 86 (normal = 0 to 35 IU/L)
ALT = 51 (normal = 0 to 35 IU/L)
CPK = 250 U/L [4.2 µkat/L] (normal = 40 to 150 U/L [0.67 to 2.5 µkat/L])

ANA, RF, anti-Ro and anti-La antibodies are negative.
Urinalysis is normal.
Serum protein electrophoresis: polyclonal hypergammaglobulinemia; no monoclonal spikes

Electromyography-nerve conduction (EMG) study reveals evidence of a sensory polyneuropathy of an axonal type with no evidence of myositis. Chest radiograph reveals mild increase of the interstitial markings at the lung bases.

You suspect a diagnosis of Sjögren's syndrome.

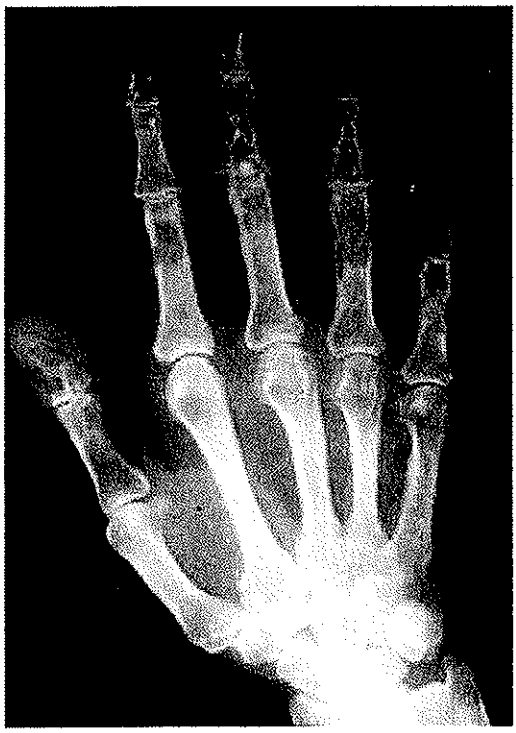
Which is the most appropriate diagnostic plan?

A. Sural nerve biopsy
B. Schirmer test, Rose Bengal stain, and formal ophthalmologic evaluation
C. Bronchoscopy with bronchoalveolar lavage (BAL) and transbronchial biopsy (TBBx) of the lung
D. No diagnostic procedure necessary, the clinical presentation is sufficient for the diagnosis of Sjögren's syndrome
E. Biopsy of a labial salivary gland

Question 24

A 28-year-old, African-American woman presents with a six-month history of pain and swelling of several proximal interphalangeal joints of her right hand. Over the past year, she has had three episodes of pain and redness of the right eye treated with glucocorticoid ophthalmic drops and prednisone.

Radiographs of the hands are taken (see figure).



Which of the following statements regarding this condition is false?

A. Bone involvement is unusual in the absence of infiltrative skin lesions.
B. Bone lesions are rarely asymptomatic.
C. Involvement of the skull may be associated with painless nodules of the scalp.
D. Involvement of long bones may resemble Paget's disease on imaging studies such as bone scans and plain radiographs.
E. Sclerotic lesions are rare in this disorder.

Question 25

A 45-year-old Caucasian woman presents to you for a second opinion. She was in her usual state of good health until two years ago, when she developed pain and numbness in the fingers following exposure to cold. She recalls that one or more of her fingers would turn blue during these episodes and that the sensation and color changes would subside after about 15 minutes if she warmed her hands.

A year later, she developed fatigue, heartburn, and low-grade fevers. These symptoms were accompanied by morning stiffness, pain, and swelling of the hands, wrists, and knees. She reports that her "lupus test" was positive at that time, and she was treated with hydroxychloroquine and diclofenac with the ultimate resolution of her symptoms.

Three months ago, she developed weakness in her arms and legs to the extent that she was having difficulty rising from a chair and combing her hair in the morning. She was treated with prednisone 60 mg/day with partial resolution of her symptoms. Her weakness returned with a reduction of the prednisone dose to 30 mg per day. She complains of intermittent facial numbness. She takes hydralazine for hypertension.

On physical examination there is sclerodactyly and proximal interphalangeal joint flexion contractures. The proximal upper and lower extremity strength is 3+/5; grip and calf strength are 5/5. There is reduced sensation to pinprick over both cheeks.

The cardiac, pulmonary, and abdominal exams are unremarkable.

Nailfold microscopy reveals dilated capillary loops and significant dropout or loss of capillaries.

Laboratory studies reveal:

Creatinine = 1.1 mg/dL [97.2 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
Creatine kinase = 1560 IU/L [26.0 µkat/L] (normal = 40 to 150 IU/L [0.67 to 2.5 µkat/L])
ESR = 83 mm/hr (normal = 0 to 30 mm/hr)
ANA: 1:2560
Antibodies to U1-RNP: 39 U

Urinalysis: normal

You suspect that she has mixed connective tissue disease (MCTD). Which of the following statements regarding MCTD in this patient is CORRECT?

A. A renal biopsy would likely show an interstitial nephritis.
B. Her exposure to hydralazine has induced MCTD.
C. She has trigeminal neuropathy, which is the most common central nervous system (CNS) manifestation of this disease.
D. A muscle biopsy is required to distinguish MCTD from polymyositis.
E. This patient should undergo a thorough evaluation for malignancy.

Question 26

A 55-year-old woman presents for a second opinion regarding treatment of osteoporosis. Results of a recent bone densitometry test include T-scores of -1.8 for the lumbar spine and -1.6 for the total hip.

The patient underwent hysterectomy at age 35 for menorrhagia. She has been on estrogen replacement since that time. Other medical problems include hypertension and hypothyroidism. Medications include hydrochlorothiazide 12.5 mg/day, calcium 1200 mg/day, vitamin D 800 IU/day, conjugated estrogen 0.625 mg/day, and levothyroxine 0.15 mg/day.

Physical examination is unremarkable.

Laboratory studies include normal CBC and electrolytes.

Other laboratory studies reveal:

Creatinine = 0.9 mg/dL [80 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
Alkaline phosphatase = 98 IU/L [1.6 nkat/L] (normal = 30 to 120 IU/L [0.5 to 2.0 nkat/L])
TSH = 0.1 IU/mL (normal = 0.4 to 5 IU/mL)

Which of the following therapeutic options should you select?

A. Increase the dose of hydrochlorothiazide
B. Begin alendronate
C. Increase daily dose of calcium
D. Increase daily dose of vitamin D
E. Reduce the dose of levothyroxine

Question 27

You are asked to see a 38-year-old woman who has recently been diagnosed with rheumatoid arthritis. She has three children and an identical twin sister. She wants to know the likelihood of her sister and/or her children developing rheumatoid arthritis. You happen to be an investigator at a major university in charge of a genetics laboratory. You agree to test the patient, her children, and her sister.

Select the correct statement regarding genetic susceptibility in rheumatoid arthritis.

A. The major class II histocompatibility gene, DR2, is highly associated with rheumatoid arthritis.
B. A short amino acid sequence known as the "shared epitope" conveys increased susceptibility to rheumatoid arthritis and can be contained in DR4, DR1, or DR6 molecules.
C. The risk of developing more severe articular disease, and for developing extraarticular disease, is not influenced

by the "dosage" of shared epitopes.
D. The shared epitope conveys susceptibility to a similar degree in Caucasians and African Americans.
E. The shared epitope is contained in the alpha chain of the HLA-DR4 molecule.

Question 28

A 32-year-old woman is referred to you for another opinion regarding her underlying diagnosis. Over the past year, she has developed a systemic illness characterized by episodes of polyarthritis, hemoptysis, renal insufficiency, and leg numbness. The arthritis is often transient, polyarticular, and symmetric primarily involving the hands and feet. The two episodes of hemoptysis have been associated with transient pulmonary infiltrates. Renal dysfunction includes a serum creatinine rising from a baseline of 0.7 mg/dL [61.9 mmol/L] one year ago, to 1.6 mg/dL [141.4 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L]) today. The urinalysis has intermittently demonstrated the presence of red cells and occasional red blood cell casts. She has a constant sensation of bilateral leg numbness in a stocking distribution.

Her previous rheumatologist has sent along copies of the most recent laboratory studies. These include:

WBC = 8,800/mm3 with normal differential (normal = 4,300 to 10,800/mm3)
Hemoglobin = 12.9 g/dL [8.0 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
Platelets = 330,000/mm3 (normal = 130,000 to 400,000/mm3)
AST = 28 U/L (normal = 0 to 35 U/L)
ALT = 27 U/L (normal = 0 to 35 U/L)
Albumin = 3.6 g/dL [36 g/L] (normal = 3.1 to 4.3 g/dL [31 to 43 g/L])
Urinalysis = 3 to 5 RBC/HPF, no casts, trace protein

ANA = 1:320
Anti-ds DNA, Ro, La, Sm: negative
CH50: normal

C-ANCA is "markedly positive" but no quantification is given. You have asked for further details. The results of chest radiographs and EMG study are not available.

Which of the following conditions does this patient most likely have?

A. SLE
B. Wegener's granulomatosis
C. Takayasu's arteritis
D. Polyarteritis nodosa
E. Churg Strauss syndrome

Question 29

An internist colleague of yours consults with you over the telephone regarding his 79-year-old mother who abruptly became blind in one eye two weeks ago. She had developed jaw claudication two months earlier, which she was told was due to poorly fitted dentures. She developed the abrupt onset of a partial field defect in one eye. Her ophthalmologist noted an ischemic optic neuropathy. The ESR was 43 mm/hr (normal = 0 to 30 mm/hr).

The patient has a history of myocardial infarction four years ago. She has had a prior vertebral compression fracture.

Current medications include:

Atenolol 50 mg qd
ASA 81 mg qd
Alendronate 70 mg q week

She was tentatively diagnosed giant cell arteritis (GCA) and started on prednisone 60 mg qd for the past two days pending a temporal artery biopsy. Your colleague is concerned about his mother taking such a high dose of prednisone and seeks your advice.

You tell him:

A. His mother does not have giant cell arteritis because the fundoscopic findings are not consistent with GCA
B. His mother probably has GCA and should follow the treatment plan outlined
C. His mother probably has GCA and should start methotrexate as a steroid sparing agent
D. His mother should seek another ophthalmologic opinion
E. His mother should await the results of the temporal artery biopsy before beginning prednisone

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Question 30

A 52-year-old woman with chronic seropositive rheumatoid arthritis (RA) is evaluated for an exacerbation of pain and swelling in her right knee and left ankle. She had been stable until 10 days ago, when she noted that her knee and ankle were more swollen and tender. She has had a fever to 102°F (38.9°C) and shaking chills for the past two days. Her treatment regimen consists of methotrexate 20 mg weekly and prednisone 5 mg qd.

On exam, she has chronic rheumatoid deformities without tenderness or swelling except for marked swelling, tenderness, and erythema of the right knee and left ankle.

Arthrocentesis of the knee reveals 30 cc of cloudy fluid with a synovial fluid leukocyte count of 65,000 cells/mm3. You suspect a superimposed bacterial arthritis.

Which of the following statements regarding this patient's condition is TRUE?

A. Synovial fluid culture is positive in less than half of the patients with a septic arthritis.
B. A false positive Gram stain can be obtained because precipitated crystal violet and mucin in the synovial fluid can mimic gram-positive cocci.
C. A depressed synovial fluid glucose and elevated lactic acid concentration are sensitive tests for the diagnosis of septic arthritis.
D. Blood cultures are positive more often than synovial fluid cultures in patients with suspected bacterial arthritis.
E. The synovial fluid is usually purulent with an average leukocyte count of 20,000 to 50,000 cells/mm3.

Question 31

A 30-year-old woman was examined by a rheumatologist one year ago for a history of migratory polyarthralgias. At that time, physical examination was negative except for poor muscle tone; tests for rheumatoid factor and antinuclear antibodies were negative; and the erythrocyte sedimentation rate (ESR) was 20 mm/hr (normal = 0 to 20 mm/hr). She was begun on a NSAID and subsequently improved.

The patient now presents with periodic swollen joints, a photosensitive facial rash, small bald spots on the back of her head, fevers, poor appetite, 20-pound weight loss, and peripheral edema. She has no previous history of gastrointestinal or renal disease, hepatitis, nausea, vomiting, headaches, rashes, hair loss, chest pain, or dyspnea.

On physical examination, her vital signs include a blood pressure of 110/70 mmHg, heart rate of 88/min, temperature of 99.4°F (37.7°C), and a weight of 115 pounds (52 kg). There are no rashes, but her skin is dry. There is no lymphadenopathy; her chest is clear, and the heart is not enlarged. She has a grade 1/6 blowing systolic murmur heard at the base of the heart, without radiation. Abdominal examination reveals vague mild abdominal tenderness. There is no splenomegaly, but the liver can be felt one finger-breadth below the right costal margin on deep inspiration. Muscle tone is poor throughout. Joint examination is normal. She has 3+ pitting edema extending from 5 inches below her knees to her feet.

Laboratory results:

WBC = 6,000/mm3 (normal = 4,300 to 10,800/mm3)
Hematocrit = 30 percent (normal = 37 to 48 percent)
Urinalysis = 2+ proteinuria
RBC = 5 to 8/high powered field
WBC = 4 to 6/high powered field
24-hour urine = 1.5 g protein
Antinuclear antibodies (ANA) = 1:640
anti-DNA = 1:320
CH50 complement = 25 mg/dL (normal >150 mg/dL)
C3 complement = 25 mg/dL (normal = 83 to150 mg/dL)
Serum albumin = 1.5 g/dL (normal = 3.5 to 5.5 g/dL)
AST and ALT: normal
pANCA: weakly positive
X-rays and CT of abdomen: negative

What is your diagnosis?

A. SLE with nephrotic syndrome
B. SLE with protein losing enteropathy
C. Malnutrition
D. SLE with liver dysfunction

Question 32

A 67-year-old man presents with a two-day history of acute swelling, pain, and redness of his left knee. He recalls a similar episode of pain three years ago involving both wrists and the knees. He was seen in a local walk-in clinic and was told that he had rheumatoid arthritis (RA).

Past history is notable for low back pain and stiffness with prolonged standing or sitting.

Examination reveals moderate swelling, tenderness to palpation, and mild warmth and erythema of the left knee, with pain on flexion beyond 90 degrees.

Synovial fluid analysis reveals 20,000 leukocytes, 92 percent neutrophils; bacterial cultures are negative. Compensated polarized light microscopy demonstrates numerous, weakly positive, birefringent rhomboidal crystals.

Anteroposterior x-rays of the knees show an area of linear calcification in the tibiofemoral joint space bilaterally.

All of the following statements regarding this patient's crystal-induced disorder are true EXCEPT?

A. It is unlikely to be the cause of his low back pain.
B. A serum iron and total iron binding capacity should be ordered.
C. A serum calcium and phosphorus should be ordered.
D. If he has the familial form of this condition, one would expect that one of his parents has this condition as well.
E. This condition may mimic RA in a minority of patients

Question 33

A 36-year-old software engineer is referred to you for evaluation of joint achiness and skin rash. He was well until six weeks earlier when he noted the onset of joint pain involving the hands, knees, ankles, and feet. The symptoms have not responded to ibuprofen or naproxen. He developed a skin rash over the legs three weeks ago and has noticed an increased level of fatigue. He denies fever, chills, or sweats.

There is no past history of medical disorders, prior surgeries, or blood transfusions. He denies recreational drug use.

On examination, he has joint tenderness involving the small joints of both hands, the wrists, knees, and feet bilaterally. There is no swelling. There is palpable purpura involving the skin over the lower extremities.

Laboratory results:

CBC: normal
ESR = 45 mm/hr (normal = 0 to 15 mm/hr)
ALT = 94 IU/L (normal = 0 to 35 IU/L)
AST = 87 IU/L (normal = 0 to 35 IU/L)
ANA = 1:40 speckled
RF = 285 U
C3 = 50 mg/dL (normal = 55 to 120 mg/dL)
C4 = 6 mg/dL (normal = 20 to 50 mg/dL)

A skin biopsy is reported to show "PAS-positive microthrombi occluding multiple vessels."

What is this patient's most likely diagnosis?

A. Hepatitis B
B. Systemic lupus erythematosus (SLE)
C. Hepatitis C (HCV)
D. Rheumatoid arthritis (RA)

Question 34

A previously healthy, 22-year-old college student had an upper respiratory tract infection two weeks ago and was treated with oral penicillin for one week. Yesterday, he developed bilateral ankle pain and swelling along with raised purpuric lesions over his lower extremities.

Laboratory results:

Serum creatinine = 3.0 mg/dL [265.2 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
BUN = 46 mg/dL [16.4 mmol/L] (normal = 10 to 20 mg/dL [3.6 to 7.1 mmol/L])
AST = 22 IU/L (normal = 0 to 35 IU/L)
ALT = 31 IU/L (normal = 0 to 35 IU/L)
ANA = 1:80

Urinalysis: 4+ proteinuria, 2+ RBCs, several RBC casts/high power field

The most likely histopathology on renal biopsy would be?

A. Mesangial proliferation with IgA deposition on immunofluorescence
B. Crescentic glomerulonephritis with IgA deposition on immunofluorescence
C. Crescentic glomerulonephritis with negative immunofluorescence and no deposits on electron microscopy
D. Proliferative glomerulonephritis with IgG and complement on immunofluorescence and subendothelial deposits and subepithelial "humps" on electron microscopy

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Question 35

A 54-year-old attorney is admitted because of a one-week history of an abrupt onset of fever, skin rash, and polyarthritis. She complains of pain in the knees, ankles, and hands. She denies any recent travel or exposure to pets.

Her only medication is an oral contraceptive agent. A skin biopsy demonstrates a dense neutrophilic infiltrate without vasculitis. You suspect a diagnosis of Sweet's syndrome.

Which of the following statements regarding this condition in this patient is correct?

A. Once her condition has improved, she does not require further follow-up.
B. You should initiate anti-tumor necrosis factor therapy (TNF) because TNF is causative in this disorder.
C. Minocycline and oral contraceptive use can predispose to the development of Sweet's syndrome.
D. She is unlikely to have Sweet's syndrome because there is a striking male predominance.
E. Her skin biopsy does not show the characteristic histopathology of Sweet's syndrome.

Question 36

A 35-year-old woman presents with low-grade fever and fatigue over a period of several months. Recently, she has noted easy fatigue of her extremities.

Physical examination reveals a chronically ill-appearing woman. Her temperature is 99.8°F (38.0°C), blood pressure is 150/90 mmHg (left arm), 166/96 mmHg (right arm), and pulse is 76/min. Bruits are audible over the left subclavian and carotid arteries, but the cardiopulmonary examination is normal. Arteriography of the aortic arch, subclavian, and carotid arteries reveals multiple abnormalities consisting of smooth-walled taperings alternating with a few areas of dilation.

Which of the following statements about this patient's condition is true?

A. CT angiography may be more sensitive than conventional angiography in establishing the diagnosis and may be used to monitor treatment.
B. Autoantibodies such as ANCA or antiphospholipid can be detected in the sera of patients with this disease.
C. Hypertension, retinopathy, and mitral regurgitation are all poor prognostic signs.
D. Most patients respond to single drug therapy using corticosteroids.
E. The associated synovitis typically involves the small joints of the hands.

Question 37

A 41-year-old woman is seen for follow-up care of her rheumatoid arthritis (RA). She has had seropositive RA for two years complicated by secondary Sjögren's syndrome. Her joint symptoms are well controlled with methotrexate and etanercept. Two months ago, the patient began using an artificial tear preparation for symptomatic relief of dry eyes. She now complains of increased bilateral ocular irritation. She denies photophobia or decreased visual acuity.

Eye examination reveals slight diffuse erythema of the sclerae bilaterally. The lower conjunctival lids are unremarkable in appearance. No ocular discharge is noted.

Which is the best course of management?

A. Change the artificial tears preparation to one containing no preservatives
B. Discontinue artificial tears and treat blepharitis with lid scrubs with a sterile eyelid cleanser and antibiotics
C. Change artificial tears preparation to one containing acetylcysteine
D. Begin ophthalmic steroid drops and refer the patient for ophthalmologic evaluation to rule out episcleritis or scleritis
E. Begin ocular antibiotic drops

Question 38

You are asked to consult on a 79-year-old man admitted to the medical service for management of congestive heart failure. Following aggressive diuresis, the serum urate was measured at 12.6 mg/dL [749.4 µmol/L] (normal = 2.5 to 8.0 mg/dL [150 to 480 µmol/L]). He denies any prior personal or family history of gout. He has never passed a kidney stone. The patient's intern asks you whether initiation of allopurinol is appropriate, since the patient is going to be discharged home on chronic diuretic therapy.

Which of the following statements regarding this patient's diuretic-induced hyperuricemia is true?

A. The distal tubule is the major site of urate handling in the kidney.
B. Urate retention will occur regardless of whether the diuretic-induced fluid losses are replaced.
C. In patients with hypertension, the degree of urate retention noted with diuretic use is dose-dependent.
D. Treatment of asymptomatic hyperuricemia greater than 12 mg/dL would be indicated to prevent uric acid nephropathy.
E. Treatment of asymptomatic hyperuricemia greater than 12 mg/dL would be indicated to prevent gouty arthritis.

Question 39

A 44-year-old woman with seropositive rheumatoid arthritis (RA) of seven years' duration presents for a follow-up visit. She had been treated with methotrexate 22.5 mg per week with excellent relief of symptoms. She has no morning stiffness, joint pain, or swelling. However, she is concerned about the recent development of new, small, nontender nodules over several fingers. They are beginning to interfere with hand function. Her father had a history of gout and she recalls that he had similar appearing nodules. She read some information about nodules on the Internet, which suggests that the diagnosis of rheumatic fever should be considered. She denies a recent history of fever or sore throat.

The joint exam was notable for synovial thickening at both wrists, the MCP and scattered PIP joints, and over the olecranon surfaces. There were six small, nontender nodules, measuring 4 mm or less over the fingers.

All of the following statements regarding nodule formation in this patient are correct EXCEPT?

A. Methotrexate has exacerbated rheumatoid nodule formation despite effectively suppressing synovial inflammation.
B. Rheumatic fever nodules are typically firm, tender, and may last for months.
C. In RA, nodule formation may be mediated via the activation of adenosine A1 receptors by methotrexate.
D. Similar to rheumatoid nodules, gouty tophi can involve the olecranon surface.
E. Rheumatoid nodules are almost always associated with positive rheumatoid factor.

Question 40

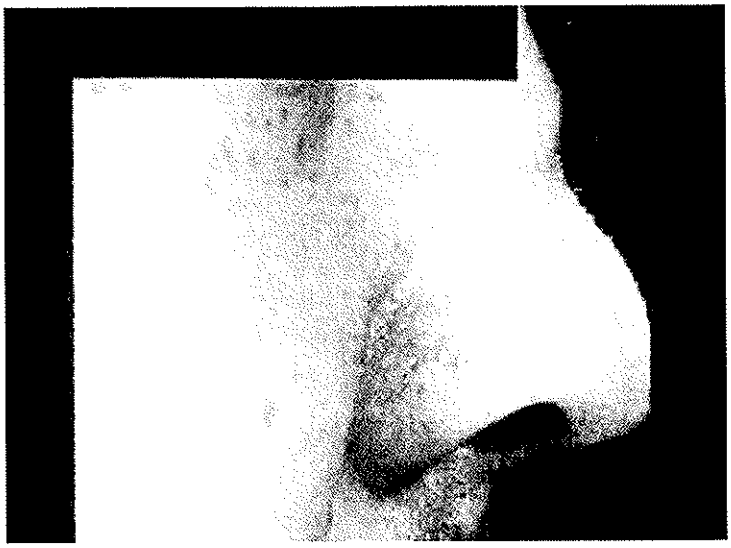
A 35-year-old man is referred to you by an otolaryngologist for evaluation of a nasal deformity. The patient, a software engineer, describes being in good health until one week ago. While playing football with some friends, he caught the ball in his face. The following morning he noted a nasal deformity (see figure).

The referring otolaryngologist performed an endoscopic exam of the upper airways and no abnormalities were identified. Plain radiographs of the sinuses reveal mild mucosal thickening and no masses or bony erosion. Chest radiograph is normal. The patient has a history of asthma for two years treated with inhaled corticosteroids but has otherwise been in good health.

Physical examination is notable only for the nasal deformity. Cardiorespiratory exam is normal. There are no swollen or tender joints.

Urinalysis is normal with no protein or blood by dipstick.

Other laboratory studies are pending.



Courtesy of Jerome H Herman, MD.

The most likely diagnosis is?

A. Wegener's granulomatosis
B. Relapsing polychondritis
C. Lymphoma
D. Lymphomatoid granulomatosis
E. Polyarteritis nodosa

Question 41

A 27-year-old woman presents with a two-week history of pain, stiffness, and swelling of the hands, wrists, knees, and ankles. She noted redness over her face and arms for two days during the first week of symptoms, but this has resolved. She denies a prior history of sore throat, fever, diarrhea, pelvic pain, or ocular pain or redness.

Physical examination reveals mild to moderate tenderness and swelling of the MCP and PIP joints, wrists and knees bilaterally. No rash, oral ulcers, adenopathy, or splenomegaly were noted.

Which of the following tests would be most helpful in establishing a diagnosis in this patient?

A. ANA
B. IgM antibodies to human parvovirus B-19
C. Lyme antibody
D. ESR
E. IgG antibodies to human parvovirus B-19

Question 42

A 17-year-old woman is referred for evaluation of new onset of photosensitivity. Upon further questioning, she also describes patchy hair loss.

Examination reveals a fine, erythematous rash over the face, neck, and trunk. There is slight alopecia and four, small mouth ulcers. The PIP joints are swollen and tender in both hands. There is minimal splenomegaly, but no hepatomegaly.

Laboratory tests:

ANA 1 = 5,120 homogeneous pattern

Rheumatoid factor: negative
C3 complement = 22 mg/dL (normal = 55 to 120 mg/dL)
C4 complement = 3 mg/dL (normal = 20 to 50 mg/dL)

Which of the following statements regarding complement deficiency states is correct?

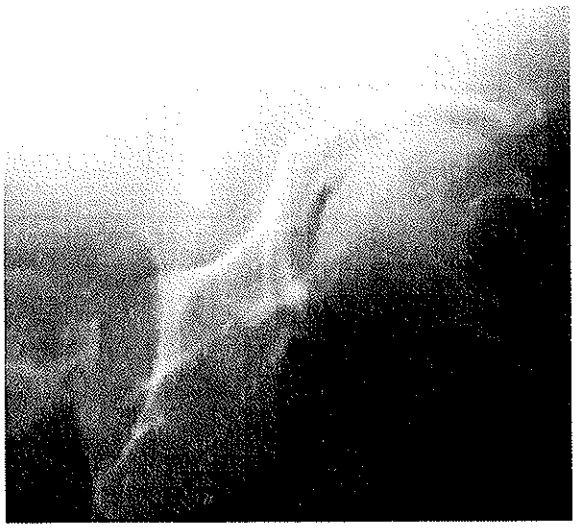
A. Complement component deficiencies are most often inherited as X-linked traits.
B. Deficiencies often result from "null" (Q0) alleles.
C. Total C4 complement deficiency is the most common complement deficiency state.
D. Unlike C1s deficiency, C1r deficiency is not associated with the development of SLE.
E. Deficiencies of components of the membrane attack complex are rarely associated with infections by Neisseria species.

Question 43

A 37-year-old woman with a 10-year history of systemic lupus erythematosus (SLE) is seen for evaluation of right groin pain. The pain began six weeks ago and has persisted. It is present with weight bearing activities, but is relieved by rest.

Physical examination reveals some pain with full internal rotation of the right hip. The patient walks with a slight limp favoring the right leg.

A plain radiograph of the hip was obtained (see figure).



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Risk factors for the development of this lesion in patients with SLE include all of the following EXCEPT?

A. Raynaud's phenomenon
B. Hyperlipidemia
C. Antiphospholipid antibodies
D. Chronic corticosteroid use
E. Male sex

Question 44

A 37-year-old librarian with systemic sclerosis comes with her husband to your office for a second opinion regarding disease prognosis. She has been told that her remaining life span may be limited to a few months or, at best, a few years.

She was diagnosed as having systemic sclerosis with limited cutaneous disease five years earlier. She had done well on no therapy apart from treatment of esophageal reflux with a proton pump inhibitor. Six months ago, she began to experience exertional dyspnea with progressively decreasing exercise tolerance.

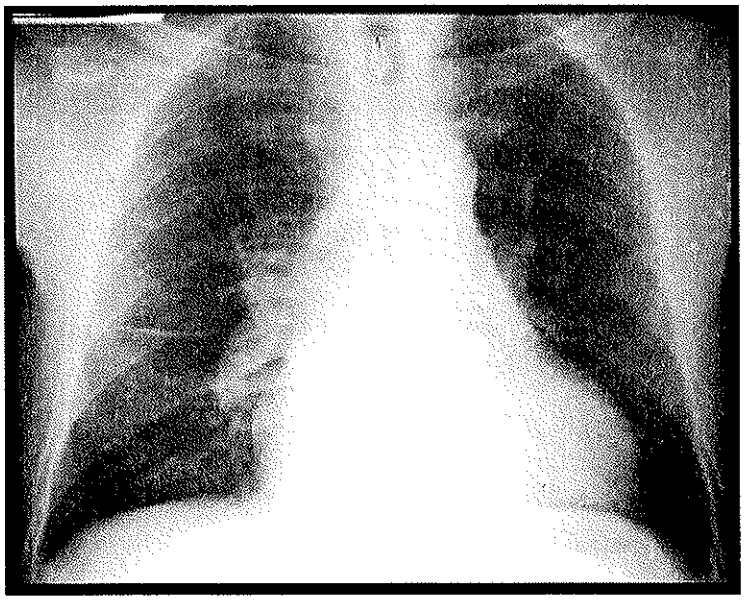
On examination, she was afebrile, pulse rate was 96 beats per minute, respiratory rate was 20 per minute, and blood pressure was 116/70 mmHg. Physical findings included skin tightening confined to the digits and face and facial telangetasia. Auscultation of the lungs was normal. On cardiac examination there was no right ventricular heave; S1 was normal, P2 was slightly increased. There was no S3 gallop and no murmur. Abdomen was soft, non-distended, and without organomegaly. Trace pretibial edema was present. Contractures were present at the PIP joints, but there was no synovitis.

She brought the report of a recent pulmonary function test. The diffusion capacity (DLCO) was reduced to 55 percent of predicted value. No other abnormalities were noted.

Findings on chest radiography are shown (see figure).

Which of the following conditions best explains her problem?

A. Pulmonary hypertension
B. Worsening interstitial lung disease
C. Superimposed opportunistic infection
D. Recurrent pulmonary emboli
E. Pulmonary sarcoidosis



Courtesy of Carol Black, MD.

Question 45

A 55-year-old man has noted the development of slowly progressive weakness over the past six years. He has difficulty getting out of a chair and climbing stairs as well as opening jars and lifting pots and pans. More recently, he has some difficulty swallowing. He denied Raynaud's phenomenon, skin rash, fevers, or joint pains. Past medical history is unremarkable. He uses no medications and denies tobacco or alcohol use. There is no family history of weakness or muscle disease.

Physical exam reveals 4/5 weakness of the psoas muscles bilaterally and the right anterior tibialis muscle. Weakness neither increases nor decreases with repetitive voluntary muscle contractions. No ptosis is noted on repeated upward gaze. Deep tendon reflexes were 1+ and symmetric. The remainder of the exam is unremarkable.

Laboratory values reveal:

CPK = 856 IU/L [14.3 µkat/L] (normal = 25 to 90 IU/L [0.42 to 1.50 µkat/L])
CBC: normal
Electrolytes: normal
ANA: negative

Nerve conduction and electromyography are normal. Muscle action potentials are unchanged with repetitive electrical stimulation.

Which of the following statements regarding this patient's condition is correct?

A. Improvement is likely after a thymectomy is performed.
B. A periodic paralysis syndrome should be considered.
C. A forearm ischemic muscle test would be abnormal.
D. A quadriceps muscle biopsy would reveal endomysial inflammation and basophilic-rimmed vacuoles with adjacent eosinophilic inclusions.
E. Dramatic improvement in strength and normalization of the CPK should occur following initiation of prednisone 1 mg/kg.

Question 46

A 70-year-old woman is referred to you for evaluation of low back pain. The pain developed insidiously two months ago and is described as a dull ache at the base of the lumbar spine without radiation to the buttocks or legs. The patient denies any prior trauma. She also denies a history of prior episodes of low back pain.

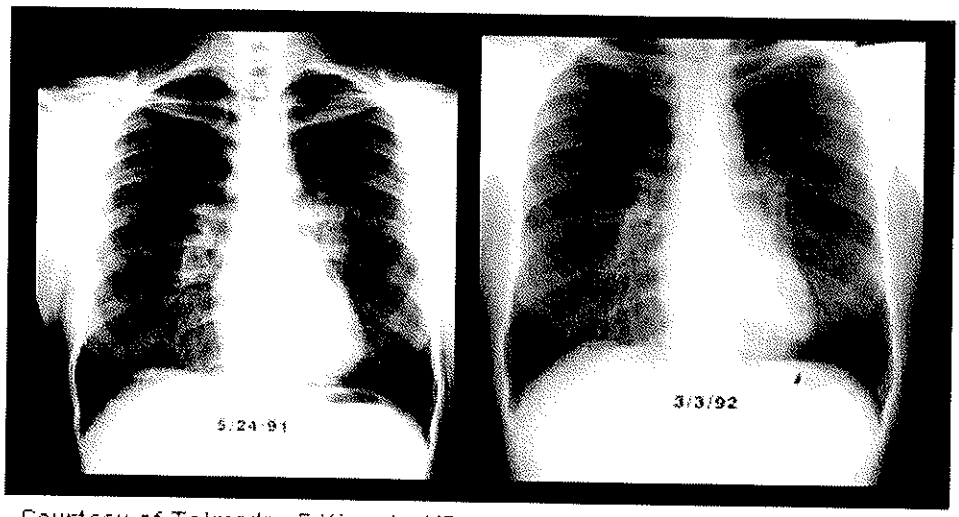
Past medical history is notable for breast cancer diagnosed six years ago and treated with lumpectomy. There has been no evidence of disease recurrence. She recalls being told ten years ago that she had a "slippage of the lower spine." You decide to obtain plain radiographs of the lumbar spine.

Which of the following statements regarding radiographs in the evaluation of low back pain in this patient is correct?

A. Oblique views should be obtained to assess for spondylolisthesis.
B. There is no utility in obtaining plain radiographs; a magnetic resonance imaging study (MRI) should be obtained.
C. A normal film would rule out the possibility of malignancy.
D. Flexion-extension views are helpful in the evaluation of spondylolisthesis.

Question 47

A 44-year-old man is referred to you for evaluation of persistent fatigue. He was in good health until one year ago when he developed an acute illness characterized by fever, dyspnea, and ankle pain. A chest radiograph revealed bilateral hilar adenopathy (see figure). He was treated with a tapering dose of prednisone over six weeks and felt better. About one month later, he noted the onset of fatigue, which has persisted. He denies any fever, chills, weight loss, shortness of breath, skin rash, or joint pain.



Courtesy of Talmadge E King Jr, MD.

He is an accountant. He denies recent travel or exposure to sick children. A trial of naproxen 500 mg bid for two weeks was not helpful.

His physical examination is totally normal.

Laboratory results:

Hemoglobin = 13.6 g/dL [8.4 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 7,700 cells/mm3 (normal = 4,300 to 10,800/mm3)
ESR = 12 mm/hr (normal = 0 to 15 mm/hr)
CK = 102 IU/L [1.7 µkat/L] (normal = 60 to 400 IU/L [1.0 to 6.7 µkat/L])

Chest radiograph: Hilar adenopathy, unchanged compared to nine months ago. No parenchymal disease.

Which of the following options would you recommend doing next?

A. Explain to the patient that he suffers from a post-sarcoid chronic fatigue syndrome
B. Obtain biopsy of hilar node
C. Order a high resolution computed tomography of the lungs
D. Measure the serum angiotensin converting enzyme level
E. Obtain an MRI of the head

Question 48

A 48-year-old construction worker presents with bilateral knee pain. It has been present for three years but has gradually worsened. There is no swelling or warmth. Pain increases with walking and is relieved with rest. He denies other joint pain. He uses acetaminophen, 1000 mg twice daily with adequate control of pain.

His only other medical problem is hypertension, which is well-controlled with lisinopril 10 mg per day.

He recalls several knee injuries obtained while playing football in high school.

Physical exam reveals a blood pressure of 140/86 mmHg, a height of 74 inches (188 cm), and a weight of 275 pounds (124 kg). Knees are cool and without effusions, with full range of motion and crepitus with flexion-extension. There is mild tenderness over the medial compartments.

Plain radiographs confirm grade II osteoarthritis involving both knees.

Which of the following statements is most accurate regarding recommendations for his treatment?

A. Weight loss would not be helpful.
B. The patient should change jobs to allow more complete rest of joints in order to slow progression of his osteoarthritis.
C. The patient should use a cane consistently since it reduces joint forces at the knee by over 50 percent.
D. The patient should use padded, protective shoes in order to reduce joint load on impact while walking.
E. Walking would be beneficial from a cardiovascular standpoint but would not have an impact on the arthritis.

Question 49

A 72-year-old woman was found to have Paget's disease involving the T12 vertebral body, posterior rib cage, and pelvis bilaterally. She presented with bone pain in these areas and was treated with alendronate for six months. Her symptoms improved and she is seen today for a follow-up visit. Her only complaint is some hearing loss over the past year.

She currently takes atenolol 50 mg qd and ASA 81 mg qd as her only medications.

Laboratory results reveal:

Serum alkaline phosphatase = 298 IU/L [3.3 nkat/L] (normal = 30 to 120 IU/L [0.5 to 2.0 nkat/L])
Serum calcium = 11.0 mg/dL [2.75 mmol/L] (normal = 9 to 10.5 mg/dL [2.2 to 2.6 mmol/L])
Serum phosphorus = 2.0 mg/dL [0.65 mmol/L] (normal = 3.0 to 4.5 mg/dL [1.0 to 1.4 mmol/L])
Creatinine = 1.0 mg/dL [88.4 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])

CBC and liver function studies: normal

Which of the following statements regarding her condition is CORRECT?

A. Her laboratory findings are consistent with Paget's disease, but one cannot ascertain the level of disease activity.
B. The hypercalcemia and elevated serum alkaline phosphatase suggest active Paget's disease.
C. Hearing loss is unlikely to be due to her Paget's disease.
D. The hypercalcemia and elevated serum alkaline phosphatase suggest primary hyperparathyroidism.
E. Hand radiographs are likely to show osteitis fibrosa cystica and brown tumors.

Question 50

A 33-year-old woman with diffuse scleroderma seeks your advice regarding treatment options for her condition. She was diagnosed three years ago following the onset of Raynaud's phenomenon, cutaneous involvement, muscle weakness, and dyspnea. Her serum creatinine concentration is currently 1.5 mg/dL (133 mmol/L) (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L]) and she is hypertensive.

All of the following statements regarding treatment options for this patient are true EXCEPT?

A. Interferon-gamma has been effective in halting the progression of renal disease.
B. Corticosteroids can be effective for the treatment of myositis.
C. Treatment with interferon-alpha has been associated with a greater deterioration in lung function.
D. High-dose corticosteroid use has been associated with scleroderma renal crisis.

Question 51

In August, a 28-year-old camp counselor from Massachusetts presents with generalized musculoskeletal aches, headache, fever, and fatigue for the last five days. She has not noted a recent tick bite. She recalls a history of Lyme disease five years ago treated with a one-month course of antibiotics. She has not traveled outside North America.

She has been vaccinated against hepatitis B, meningococcus, measles, mumps, and rubella.

On examination, temperature is 103.2°F (39.6°C). There is a faint petechial rash over the legs. Neck is supple. The spleen tip is palpable. The joint exam is normal.

Laboratory results:

Hemoglobin = 12.0 g/dL [7.4 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 2,300 cells/mm3, 70 percent polymorphonuclear cells, 10 percent lymphocytes, 37 percent atypical lymphocytes (normal = 4,300 to 10,800/mm3)
Platelets = 62,000/mm3 (normal = 130,000 to 400,000/mm3)
Creatinine = 1.0 mg/dL [88 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
Lyme antibody = IgG positive, IgM negative
AST = 66 IU/L (normal = 35 IU/L)
ALT = 75 IU/L (normal = 35 IU/L)
LDH = 498 U/L [8.3 µkat/L] (normal = 100 to 190 U/L [1.7 to 3.2 µkat/L])
Urinalysis is normal

Examination of the Wright-Giemsa stained blood smear reveals intraerythrocytic, round to oval rings with pale blue cytoplasm and red-staining nuclei.

What is the treatment of choice?

A. Amoxicillin 500 mg po tid for three weeks
B. Ceftriaxone 2 g/d for three weeks
C. Replenish fluids, acetaminophen for fever, and observe
D. Clindamycin 600 mg tid and quinine sulfate 650 mg tid for 10 days
E. Doxycycline 200 mg per day

Question 52

A 32-year-old man is referred to you for evaluation and treatment of a three-week history of painful joints in his lower extremities. He notes pain and swelling of several toes in both feet and a pain in the right groin whenever he attempts to walk. He denies any previous episodes of arthritis except for one episode of Achilles tendonitis 10-years ago while playing basketball in college.

He has had intermittent left low back pain for several years. Recently, he has started taking ibuprofen 400 mg TID without any benefit noted. He denies any antecedent infections or fever, but has noted slight discomfort upon initiating urination.

The general physical examination is remarkable for mild tenderness of the prostate to palpation. The joint exam reveals an antalgic gait favoring his right leg. There is pain and limitation of motion of the right hip with both internal and external rotation. There is diffuse swelling and tenderness of the left third toe (sausage digit) and swelling and tenderness of the right Achilles tendon.

Laboratory results reveal an ESR of 53 mm/hr (normal = 0 to 15 mm/hr).
Urine analysis is positive for leukocyte esterase and the urine sediment has 8 to 10 WBC per high power field. There is no proteinuria or hematuria. Culture of the urine for routine bacterial pathogens is negative. A urine sample tested for Chlamydia trachomatis by polymerase chain reaction (PCR) is positive.

A radiograph of the pelvis and right hip is normal.

Which of the following statements regarding this patient's condition is correct?

A. The likelihood of chronic arthritis is not increased in patients who are HLA-B27 positive.
B. The prognosis is worse than if he had developed symptoms following an episode of dysentery.
C. Following the acute episode, he will most likely continue to have a waxing and waning course.
D. The risk for the development of a chronic arthritis is greatest following infections with Yersinia species.
E. His age is associated with a poorer outcome than that of adolescents with this type of arthritis.

Question 53

A 16-year-old boy is seen for evaluation of a two-week history of fever, polyarthralgias, and skin nodules. There are several firm, painless nodules present over both olecranon surfaces.

You are concerned that these are rheumatic fever nodules. Which one of the following features is not observed with this condition?

A. Rheumatic fever nodules are firm and painless.
B. Rheumatic fever nodules primarily occur in patients with carditis.

C. When numerous, rheumatic fever nodules are usually symmetric.
D. Rheumatic fever nodules often persist for several months following resolution of other features of rheumatic fever.
E. Compared to rheumatoid nodules, rheumatic fever nodules are more commonly found on the olecranon surface.

Question 54

You are seeing a 65-year-old woman for a routine visit following a bone density study. Despite being on estrogen replacement therapy for the past decade, the study demonstrated scores in the range of osteopenia. She admits that she has never consistently taken supplemental calcium.

Many of her friends have offered her advice regarding calcium supplementation, and she is very confused about the optimal approach. She proceeds to read to you five "facts" that she has collected from different sources.

Which of the following "facts" is correct?

A. Her optimal daily calcium intake should total 2000 mg from all sources.
B. Calcium absorption from vegetables such as spinach is better than that from dairy products.
C. Many of the natural calcium carbonate preparations such as oyster shells or bone meal have unacceptably high lead levels.
D. Adequate calcium intake can allow her to discontinue estrogen use.
E. Calcium carbonate should be avoided because of its cost.

Question 55

A 25-year-old woman is referred to your office by an ophthalmologist for consideration of an autoimmune illness. Her initial symptoms began with abrupt onset of redness, irritation, and watering of the right eye. The eye is painful in the early morning and at night. Visual acuity is intact. The ophthalmologist diagnosed diffuse anterior scleritis.

The erythrocyte sedimentation rate (ESR) is 35 mm/hr (normal = 0 to 20 mm/hr).

This patient may have any of the following disorders EXCEPT?

A. Rheumatoid arthritis
B. Sarcoidosis
C. Wegener's granulomatosis
D. Polyarteritis nodosa
E. Giant cell arteritis (GCA)

Question 56

A 62-year-old man presents with a new onset of severe low back pain, which radiates into both legs. Over the past 24 hours, the pain has become so severe that he cannot get up from a chair without assistance. He recently completed a 10-day course of antibiotics for treatment of cellulitis due to staphylococcus aureus.

On examination his temperature is 100.4°F (38°C). He has marked low back pain with radiation down the right leg. Intense low back spasm precludes a thorough exam. There is localized tenderness on percussion over the L3-L4 vertebral bodies. His deep tendon reflexes are absent in the lower extremities. Muscle strength in the legs is difficult to assess because of pain; however, it appears that he is quite weak.

You suspect a spinal epidural abscess. All of the following statements regarding this diagnosis are true EXCEPT?

A. Many spinal epidural abscesses begin as a pyogenic infectious diskitis.
B. The most common bacterial pathogen is Staphylococcus aureus.
C. Patients often present with painless motor weakness or paralysis.
D. The back pain is typically worsened by percussion over the affected area.
E. Pain usually travels down the leg in the distribution of the affected nerve root.

Question 57

A 32-year-old man is referred because of a six-month history of recurrent episodes of pain, redness, and enlargement of several toe joints, primarily the left second interphalangeal joint. Each episode lasted two to three weeks. Enlargement of the left second interphalangeal joint has persisted since his last painful toe flare six weeks ago. He underwent a cadaveric renal transplant 12 years ago for end-stage renal failure resulting from IgA nephropathy. His renal function has been stable for the past five years. His past history is notable for longstanding hypertension.

Medications include: azathioprine 100 mg qd, cyclosporine 200 mg qd, prednisone 10 mg qd, hydrochlorothiazide 25 mg qAM, and verapamil sustained release 240 mg qd.

On examination his blood pressure is 120/70 mmHg. There is a nontender, soft tissue swelling over the left second interphalangeal joint with a whitish-yellow discoloration. A similar swelling, with the same whitish-yellow appearance, is noted at the medial aspect of the left great toe.

Laboratory studies reveal:

Creatinine = 1.7 mg/dL [150.3 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
BUN = 25 mg/dL [8.9 mmol/L] (normal = 10 to 20 mg/dL [3.6 to 7.1 mmol/L])
Uric acid level = 11.2 mg/dL [666.2 µmol/L] (normal = 2.5 to 8.0 mg/dL [150 to 480 µmol/L])

Under polarizing light microscopy, the aspirate from the toe mass reveals a solid field of needle-shaped, negatively birefringent crystals.

After conferring with the patient's nephrologist, which of the following initial treatment strategies do you recommend?

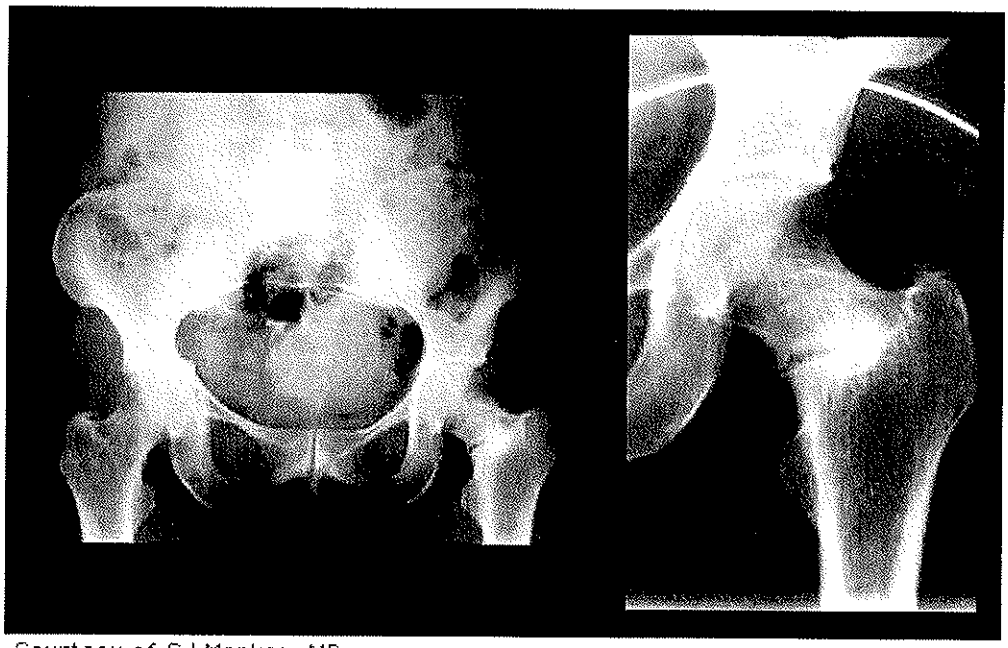
A. Start allopurinol with colchicine (0.6 mg qd)
B. Discontinue azathioprine and increase dose of cyclosporine
C. Rofecoxib (25 qd)
D. Discontinue azathioprine and replace with mycophenolate, start allopurinol with colchicine (0.6 mg qd)
E. Colchicine (0.6 mg bid)

Question 58

You are asked to see a 79-year-old woman because of the sudden onset of pain and difficulty walking. She denies any trauma but describes feeling a sensation of her left leg "giving way."

Past medical history is notable for celiac sprue. Medications include atenolol and aspirin.

A pelvic film is shown (see figure).



Courtesy of CJ Menkes, MD.

All of the following statements regarding this condition are correct EXCEPT?

A. These types of fractures often lie in apposition to arteries and may be due to erosion via arterial pulsations.
B. These may be stress fractures which have been repaired by inadequately mineralized osteoid.
C. These fractures are typically asymmetric or unilateral.
D. The upper extremity bones may be sites for these fractures.
E. An elevated serum alkaline phosphatase is observed in patients with osteomalacia due to vitamin D deficiency.

Question 59

A 61-year-old Caucasian woman is referred to your office for evaluation of fatigue and muscle pain. She describes a five-year history of muscle and joint pain over her entire body, deep and aching in quality. Pain and stiffness are especially bad in the morning, but persist all day. Onset was insidious without obvious precipitant. She states she has great difficulty with sleep, and the pain has made her somewhat depressed. Fatigue is "overwhelming." She denies fever, weight loss, night sweats, swollen joints, rash, photosensitivity, alopecia, Raynaud's symptomatology, oral ulcers, sicca, pleuritic chest pain, abdominal pain, headaches, seizures, psychosis, visual changes, or dysphagia.

She exhibits mild pain-related proximal muscle weakness. Musculoskeletal exam reveals good joint range of motion without any evidence of synovitis. Exam is remarkable for 18/18 positive fibromyalgia tender points.

You suspect a diagnosis of fibromyalgia, but wish to rule out other less likely conditions with laboratory tests.

Which of the following tests is least appropriate?

A. CBC and chemistry panel
B. ESR
C. TSH
D. CK
E. ANA

Question 60

A 54-year-old man with a 13-year history of scleroderma is referred to you for evaluation. His scleroderma is characterized by diffuse cutaneous thickening, contractures of his elbows and hands, and a myopathy with proximal muscle weakness. For the past year, he has been bothered by bouts of abdominal pain, alternating constipation and diarrhea, and a sense of bloating. His stools have been somewhat greasy. He has lost 40 pounds (18 kg) in the past 1 1/2 years. He has consciously increased his overall caloric intake but continues to lose weight. He denies dysphagia, although he previously had problems with gastroesophageal reflux for which he had been treated with omeprazole 20 mg twice daily for the past three years.

Other medications include long-acting nifedipine 60 mg qd, celecoxib 200 mg bid, and aspirin 81 mg daily.

On examination, he is a thin man with sclerodermatous facies. Weight is 162 pounds (73 kg), blood pressure is124/80 mmHg, pulse 88, and respirations 18. The oropharynx is notable for restricted inter-incisor distance, xerostomia, and numerous filled caries. The chest is clear and heart sounds are normal. The abdomen is not distended and his bowel sounds are somewhat hyperactive. Extremities are notable for sclerodactyly and joint contractures.

A plain radiograph of the abdomen reveals no evidence of obstruction, pneumatosis cystoides intestinalis, or pneumoperitoneum.

The addition of which of the following medications is most likely to provide some relief of his symptoms?

A. D-penicillamine
B. Ranitidine
C. Cyclophosphamide
D. Diphenoxylate
E. Tetracycline

Question 61

A 34-year-old woman with scleroderma presents for follow-up. Ten years ago she began to notice episodes of Raynaud's phenomenon. Six months ago, she began to notice a sense of breathlessness ascending stairs. She denies nonexertional dyspnea, cough, or chest pain. She has noted recurrent bouts of severe Raynaud's phenomenon, which has improved minimally since starting nifedipine five months ago.

On exam: She is well appearing and not dyspneic walking to the exam room. Blood pressure is 130/72 mmHg, heart rate 78, and respirations are 16 and non-labored. She has numerous facial telangiectasias. Her chest is clear to auscultation and heart rhythm is regular without a rub or a particularly prominent P2 sound. There is no S3 or S4. There is an A-wave in the jugular venous pulse distention. Her extremities are notable for sclerodactyly, nailfold capillary dilatation, and capillary drop out. There are sclerodermatous skin changes extending to her wrists. Her lower extremities are normal in appearance.

Recent labs done by her internist are notable for a normal CBC, electrolytes, renal function, and urinalysis. The ANA titer is positive at 1:1280.

Which of the following tests is most likely to best define the cause of her new symptoms?

A. CK determination with MB fractionation
B. High resolution chest computed tomography (CT) scan
C. Pulmonary function studies
D. Doppler echocardiogram
E. Ventilation-perfusion scan

Question 62

You are asked to see a 45-year-old Caucasian woman with recent onset polyarthritis. She complains of pain, swelling, and warmth of her hands, wrists, knees, and ankles. Physical examination reveals moderate-to-severe synovitis of the wrists, MCPs, PIPs, knees, and ankles.

Which of the following best applies to the diagnostic work-up of this patient?

A. Rheumatoid factor positivity would provide prognostic information
B. A normal ESR would preclude the diagnosis of inflammatory arthritis
C. Normal x-rays would preclude the diagnosis of osteoarthritis
D. Positive MRI findings would be highly specific for the diagnosis of inflammatory arthritis
E. A synovial fluid WBC count of less than 50,000/mm3 would rule out a polyarticular presentation of septic arthritis

Question 63

A 66-year-old woman with a 20-year history of rheumatoid arthritis (RA) complains of numbness of both hands. The numbness has been present for three months. It does not awaken her from sleep. In recent weeks, she has noted a "clunking" sensation in the back of her head when she looks at the floor. She has had an episodic occipital headache.

Her RA has been treated with methotrexate. She has undergone a left total knee replacement and a right wrist fusion. You are concerned about her new symptoms.

Which one of the following statements regarding the cause of her neck pain is true?

A. Vertical movement of C-1 on C-2 is the most common form of subluxation at the neck in similar patients.
B. Her sensation of the head falling forward upon flexion of the cervical spine demands immediate attention.
C. The earliest and most common symptom of cervical subluxation in such patients is numbness radiating from the occiput to the shoulders.
D. If she has reduced deep tendon reflexes in the upper extremities, it would suggest a cervical myelopathy.
E. The intervertebral joints of the cervical spine are probably not involved.

Question 64

A 54-year-old woman with a 20-year history of scleroderma is seen for a routine follow-up visit. She complains of increasing abdominal pain, bloating, and intermittent diarrhea for the past two months. She has a history of esophageal dysmotility.

An upper GI radiograph with small bowel follow-through is ordered (see figure).


Courtesy of Carol M Black, MD, FRCP.

Which of the following statements regarding this patient's bowel disease and scleroderma is CORRECT?

A. The pathophysiology of her small bowel dysfunction differs from the cause(s) of her esophageal disease.
B. The histopathology of her small bowel would likely reveal an abnormal villous structure with an inflammatory cell infiltrate.
C. The major cause of malabsorption in patients like her is bowel ischemia.
D. She is unlikely to develop anorectal dysfunction.
E. If you suspect bacterial overgrowth in this patient, a good screening test would be the glucose hydrogen breath test.

Question 65

An internist calls you regarding a patient whom he would like to refer to you. She is a previously healthy 38-year-old executive who now has had four months of hand pain and diffuse swelling, new onset Raynaud's phenomenon, and possibly some dyspnea, although she has been somewhat ambiguous regarding this complaint. Routine lab studies have been unremarkable, including a normal CBC, metabolic profile, CPK, and urinalysis.

Serological evaluation includes an ANA positive at 1:640 titer (mouse liver substrate) with a nucleolar pattern. All the following antibody titers are pending: anti-centromere, anti-Scl-70 (topoisomerase-1), dsDNA, Sm, U1-RNP, and U3-RNP.

He would like guidance regarding the proper interpretation of the laboratory results.

Which of the following statements regarding these findings is most accurate?

A. If present, antibodies to topoisomerase-1 are highly specific for limited cutaneous disease in scleroderma.
B. If present, antibodies to topoisomerase-1 would predict the likely development of pulmonary fibrosis in patients with scleroderma.
C. The presence of antibodies to U3-RNP would confer a favorable prognosis with limited major organ involvement.
D. Anticentromere antibodies are detected less frequently than antibodies to U1-RNP in patients with scleroderma.
E. A positive anti-centromere antibody titer would reduce the likelihood of this patient developing pulmonary hypertension.

Question 66

A 70-year-old woman was diagnosed with polymyalgia rheumatica three years ago and is treated with prednisone 7.5 mg daily. She is concerned about corticosteroid-related toxicities. Her daughter, a psychologist, has cautioned her about its use. Specifically, she is concerned about mood swings, skin bruising, bone disease, susceptibility to infection, and the development of cataracts. You attempt to reassure her.

All of the following statements regarding corticosteroid-related toxicities are true EXCEPT?

A. The most common toxicities attributable to prednisone are skin thinning and purpura.

B. Children are more susceptible than adults for the development of cataracts.
C. Alternate day prednisone appears to protect against the development of osteoporosis.
D. Corticosteroid-associated affective disorders may be successfully treated with lithium.
E. Corticosteroid therapy does not block the antibody response to influenza vaccine.

Question 67

A 74-year-old businessman is seen by you for the evaluation of bilateral arm pain that began abruptly three months ago. He began to notice pain in the upper arm muscles. Only the left arm was initially involved; however, within a month, both upper arms were involved. He has trouble getting dressed in the morning and sometimes awakens during the night if he sleeps on the left shoulder. He is stiff for two to three hours each morning. He was prescribed naproxen 500 mg bid for two weeks without any substantial improvement.

His past history is notable for hyperlipidemia treated with lovastatin for the past year. He denies weight loss or fever but says that he feels miserable and is considering retiring.

On physical examination, there is limitation of range of motion in both shoulders, particularly with external rotation. The wrists and hands are normal. Motor strength and reflexes are intact.

Laboratory tests reveal:

Hemoglobin = 13.0 g/dL [8.1 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 6,700 cells/mm3 (normal = 4,300 to 10,800/mm3)
ESR = 12 mm/hr (normal = 0 to 20 mm/h)
TSH = 2.4 mU/L (normal = 0.4 to 5 mU/L)

Which of the following would you recommend?

A. Check the serum CPK level
B. Begin a therapeutic trial of prednisone 10 mg per day for three weeks
C. Obtain bilateral shoulder radiographs and inject each with intraarticular corticosteroids
D. Repeat the ESR and check the serum C-reactive protein level
E. Hold the lovastatin for two weeks and reevaluate at that time

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Question 68

An 82-year-old woman with arm claudication is referred to you for further management. She notes the onset of right arm discomfort with activities. The discomfort improves with rest. She denies any headache, jaw claudication, or visual problems.

Physical examination is unremarkable except for the absence of an arterial pulse in the right arm.

Laboratory tests:

Hemoglobin = 11.8 g/dL [7.3 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
ESR = 67 mm/hr (normal = 0 to 30 mm/hr)
ANCA: pending

You suspect a diagnosis of large-vessel giant cell arteritis (GCA). Which of the following features is characteristic of such patients?

A. Similar to this patient, they tend to be older than those with cranial GCA.
B. This condition is more commonly seen in men.
C. The results of the anti-neutrophil cytoplasmic antibody test (ANCA) will help to distinguish large-vessel from cra-

nial GCA.
D. The lack of headaches in this patient is common.

Question 69

A 40-year-old woman is brought to your office by her parents because of back pain following a fall from bed. Her past medical history is significant for severe disabling mental retardation and a seizure disorder that has been well-controlled with phenytoin and phenobarbital. Physical examination reveals a cachectic woman with tenderness over the mid-thoracic spine and pelvis. She is not ambulatory and has flexion contractures of the legs.

Radiographs reveal a T-11 compression fracture.

Laboratory testing reveals:

Serum calcium = 7.1 mg/dL [1.8 mmol/L] (normal = 9 to 10.5 mg/dL [2.2 to 2.6 mmol/L])
Serum phosphorus = 1.9 mg/dL [0.6 mmol/L] (normal = 3 to 4.5 mg/dL [1.0 to 1.4 mmol/L])
Serum alkaline phosphatase = 344 IU/L [5.7 nkat/L] (normal = 30 to 120 IU/L [0.5 to 2.0 nkat/L])

Which of the following conditions is the most likely diagnosis?

A. Paget's disease
B. Osteoporosis
C. Osteomalacia
D. Primary hyperparathyroidism
E. Primary hypoparathyroidism

Question 70

A 29-year-old man presents with a three-week history of a painful, swollen right knee. He has recently returned from a trip to Central America. He describes a one-week history of diarrhea and eye discomfort.

On examination, he has red eyes and an ulcer on his soft palate. The joint exam is notable for a moderately swollen and tender right knee.

The complete blood count is normal. An erythrocyte sedimentation rate is 29 mm/hr (normal = 0 to 15 mm/hr). A synovial fluid analysis from the knee reveals 21,000 WBC/mm. There are no crystals, and the gram stain and microbiologic cultures are negative.

Which of the following statements regarding the pathogenesis of this patient's condition is true?

A. A transgenic rat bearing the HLA-B27 gene has been developed as a model for this condition.
B. Individuals who are HLA-B27 positive have a 5-fold increased risk of developing a spondyloarthropathy.
C. In affected individuals, expression of the disease-associated HLA- B27 gene is limited to antigen presenting cells.
D. The presence of HLA-B27 is not associated with a worse prognosis.
E. Determining the HLA-B27 status of an individual is helpful in predicting response to therapy.

Question 71

A 33-year-old woman is admitted to the Neurology service with an acute cerebrovascular accident. The patient was in her usual good health until she developed the sudden onset of right-sided hemiplegia five days earlier. She has never smoked nor used oral contraceptive pills. She has used over-the-counter diet pills frequently.

Physical exam reveals a blood pressure of 120/74 mmHg, a pulse of 70, and respiratory rate of 13. There is a right facial droop, tongue deviation to the right, and a right-sided hemiplegia. Deep tendon reflexes are brisk on the right with a Babinski sign. The rest of the exam is unremarkable.

Lab results include a normal complete blood count, serum electrolytes, liver function studies, and urinalysis. ESR is 42 mm/hr (normal = 0 to 20 mm/hr). ANA, RF, ANCA, and antiphospholipid antibodies are all negative.

All of the following studies are normal: PT/PTT, proteins C and S, antithrombin III, prothrombin mutation, and factor V Leyden. Transesophageal echocardiogram is normal.

A brain MRI is read as normal. Cerebrospinal fluid analysis reveals normal protein and glucose concentrations without cells. A cerebral angiogram reveals multiple areas of arterial narrowing throughout the cerebrum with evidence of vessel beading. You initiate high dose intravenous solumedrol.

What is the next most appropriate course of action?

A. Leptomeningeal biopsy
B. Continue to treat empirically with high dose intravenous solumedrol and add aspirin
C. Continue to treat empirically with high dose intravenous solumedrol and add cyclophosphamide
D. Initiate supportive measures, refer patient to rehabilitation program
E. Begin treatment with intravenous heparin and nifedipine

Question 72

A 72-year-old woman is referred to you by a colleague who follows her for type II diabetes mellitus requiring insulin therapy. She developed pain, stiffness, and swelling of the second and third metacarpal-phalangeal (MCP) joints bilaterally six-months ago. Her symptoms have exacerbated over the past two weeks. She reports feeling excessively tired over the past year, and did not work in her garden this past summer because of fatigue, listlessness, and finger joint stiffness and aching.

Examination shows trace to 1+ synovitis of bilateral second and third MCP joints, in addition to mild flexion contracture of the involved MCP joints. There is normal alignment of the wrists and MCPs.

Laboratory results include negative rheumatoid factor, Westergren sedimentation rate of 26 mm/hr (normal = 0 to 30 mm/hr), and mild elevations of AST and ALT. Transferrin saturation is 65 percent (normal = 25 to 45 percent).

All of the following statements regarding this patient are true EXCEPT?

A. Hand radiographs are likely to show hook-shaped osteophytes over the radial aspect of the metacarpal heads.
B. Removal of systemic iron by means of phlebotomy would likely improve her joint symptoms and signs.
C. She has an increased likelihood of developing calcium pyrophosphate dihydrate (CPPD) crystal deposition.
D. The joint symptoms are more severe in patients of her age.
E. Ulnar deviation is not a feature of her condition.

Question 73

A 52-year-old woman is evaluated for a three-month history of pain over the radial aspect of the wrist near the base of the thumb. The pain interferes with gripping. There has been no trauma to the area, but she does a great deal with her hands in craft activities and knitting. Her mother has osteoarthritis of the hands.

Examination reveals tenderness near the base of the thumb and over the area of the distal portion of the radial styloid. Pain is aggravated by passively stretching the thumb tendons over the radial styloid in thumb flexion (positive Finkelstein maneuver).

Which one of the following statements about this patient is true?

A. A positive Finkelstein maneuver is rarely seen in any disorder other than de Quervain's tenosynovitis.
B. Local anesthetic injection directly over the radial styloid is often necessary to differentiate de Quervain's tenosynovitis from osteoarthritis of the thumb carpometacarpal (CMC) joint.
C. Calcification of the affected tendon is seen frequently on plain radiographs in patients with this disorder.
D. Ganglia are a frequent cause of pain in this area, even in the absence of swelling on examination.
E. Local corticosteroid injection is not likely to result in substantial relief, and 60 percent of patients with this disorder need surgical therapy.

Question 74

A 45-year-old man is seen with a one-week history of pain and swelling over the right elbow. He denies trauma to the area, but often rests his elbows on his desk. He has a history of diabetes, controlled with Glucophage.

On physical examination, his temperature is 98.4°F (36.9°C). He has full motion of the elbow with a small collection of fluid over the tip of the olecranon. There is moderate surrounding warmth, erythema, and induration.

Needle aspiration of the olecranon bursa yields 6 cc of slightly opaque fluid. Cell count shows 4500 WBC/mm3 with 90 percent polys. Gram stain is negative, and no crystals are seen by polarized microscopy.

Which one of the following management approaches would you recommend for this patient?

A. Bursal fluid should be sent for culture, oral dicloxacillin started, and a follow-up visit scheduled in two days
B. The bursa should be injected with 20 mg triamcinolone acetonide
C. Oral naproxen should be started, frequent ice massage should be performed to the area, and a follow-up visit scheduled in three weeks
D. Oral dicloxacillin should be started and a follow-up visit scheduled in three weeks
E. The patient should be hospitalized and started on broad spectrum intravenous antibiotics

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Question 75

A 52-year-old woman with a three-year history of systemic sclerosis with limited scleroderma is seen in your office. She is upset because of the occasional loss of control of her bowels. She notes more frequent episodes of fecal soilage. There is no diarrhea, rectal bleeding, tenesmus, or abdominal pain.

Her disease course has been complicated by severe reflux esophagitis and stricture formation, the latter treated by repeated dilation. There has been no evidence of other internal organ involvement. She takes omeprazole 20 mg twice daily.

On examination, the abdomen is soft and nontender with normal active bowel sounds. On rectal examination, there are no masses felt and no tenderness. Anal sphincter tone is reduced. Formed brown feces are present in the rectal vault. A stool specimen is negative for occult blood.

Which one of the following do you suggest as the best diagnostic test?

A. Glucose hydrogen breath test
B. Empiric trial of erythromycin
C. Air contrast barium enema of the lower gastrointestinal tract
D. Colonoscopy
E. Anorectal manometry

Question 76

All of the following associations have been noted between specific autoantibodies and manifestations of systemic lupus erythematosus EXCEPT?

A. Anti-dsDNA with an increased risk of nephritis
B. Anti-Ro with the presence of C2 deficiency
C. Anti-La antibodies with Sjögren's syndrome
D. Anti-U1 RNP antibodies and an increased risk of diffuse proliferative glomerulonephritis

Question 77

A 44-year-old graphic designer is admitted to the medical service of your hospital for further evaluation of a progressive multisystem disease. She was in good health until eight months earlier when she developed a nonproductive cough. The cough has been persistent, and she has become short of breath with moderate activity. She also wheezes.

For the past month, she has noted the onset of numbness and tingling in both feet, extending upwards towards the knees. She feels weaker and two days prior to admission could not stand up and walk without assistance.

The patient describes excellent health previous to the onset of this illness. She travelled extensively to Europe and Asia. She does not smoke or drink alcohol. She is divorced and lives with a friend.

On physical examination the patient looks ill. Her blood pressure is 110/70 mmHg, her pulse is 78, respiratory rate is 18, and temperature is 98.5°F (36.9°C).

HEENT: No sinus tenderness, oral cavity clear, normal ear and eye exam
Chest: A few rales heard at the lung bases, scattered wheezes are auscultated.
Heart: S1, S2, S4 no murmurs
Abdomen: Soft, nontender, without organomegaly or masses felt
Neurologic: Marked diminution of sensation to light touch, pinprick and proprioception noted in lower extremities extending in a stocking distribution

Motor strength is asymmetrically reduced measuring 4/5 right foot extensors versus 3/5 left foot. Reflexes are 2+ at both knees and trace at both ankles.

Laboratory results:

Hemoglobin = 10.9 g/dL [6.8 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 25,000/mm3 with 50 percent neutrophils, 35 percent eosinophils and 15 percent lymphocytes (normal = 4,300 to 10,800/mm3)
Platelets = 375,000/mm3 (normal = 130,000 to 400,000/mm3)
Creatinine = 1.5 mg/dL [132.6 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
AST = 34 IU/L (normal = 35 IU/L)
ALT = 33 IU/L (normal = 35 IU/L)

Stools were negative for ova and parasites. A chest radiograph is normal. You suspect that the underlying disease is either the idiopathic hypereosinophilic syndrome (HES) or Churg-Strauss syndrome (CSS).

Which of the following is NOT commonly observed in both disorders?

A. Renal disease
B. Central nervous system disease
C. Skin or subcutaneous nodules
D. Congestive heart failure
E. Peripheral neuropathy

Question 78

A 58-year-old oyster fisherman is seen with a six-week history of pain and swelling in his right hand and wrist. He remembers having problems initially in the proximal IP joint of his ring finger, but this improved on its own. Shortly after this, he began having pain and swelling in the 3rd and 4th MCP joints, as well as over the dorsal aspect of his wrist. He has had no previous musculoskeletal problems except for a single attack of "gout" in his right great toe about two years ago. He cuts his hands occasionally during his work, and he remembers having some drainage for a few days from a cut about two months ago. He drinks alcohol heavily on weekends but has been healthy otherwise.

Examination reveals a generally healthy, robust male. The joint exam is normal except for the right hand and wrist, where there is tenosynovial thickening and fluid over the extensor tendons over the right wrist. Range of motion is normal with minimal pain on forced extension. There is no drainage from any skin lesion. The 3rd and 4th right metacarpalphalangeal joints are swollen and minimally sore to touch.

The complete blood counts, ESR, and chest X-ray are normal. A serum urate is 8.7 mg/dL [517.5 µmol/L] (normal = 2.5 to 8.0 mg/dL [150 to 480 µmol/L]).

Following aspiration of tenosynovial fluid, which of the following studies would most likely lead to the correct diagnosis in this patient?

A. Inspection of the fluid for monosodium urate crystals
B. Synovial fluid gram stain and culture for bacteria
C. Synovial fluid culture for acid fast organisms
D. Synovial fluid culture for fungi

Question 79

A 40-year-old woman with a 20-year history of primary Raynaud's phenomenon presents with an exquisitely painful right index finger. This digit has been persistently cold and painful for the past two weeks. She denies any other symptoms.

Current medications include sulindac 200 mg daily and long-acting nifedipine 30 mg daily.

On exam, the patient is in severe pain. The right index finger is cold, dusky in appearance, and exquisitely tender to touch distal to the MCP joint. The most distal 4 to 5 mm of the fingertip is black.

What is the best course of action?

A. Admit the patient to the hospital for pain control and an emergency cervical sympathectomy
B. Admit the patient to the hospital for pain control and initiate aggressive vasodilator plus antiplatelet drug therapy
C. Admit the patient to the hospital for pain control and initiate anti-coagulation with heparin
D. Give the patient codeine for pain control, begin aspirin and nitroglycerine and ask her to return to your office in one week
E. Admit the patient to the hospital for pain control and IV solumedrol

Question 80

A 35-year-old white woman with a history of SLE for 13 years presents to the emergency room with severe chest pain. The patient describes the chest pain as being similar to the pain that she has experienced before except now it is more constant and intense. She feels nauseated.