Practice file

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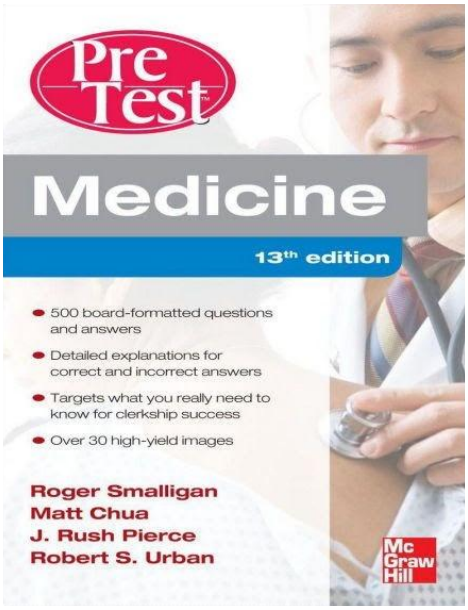

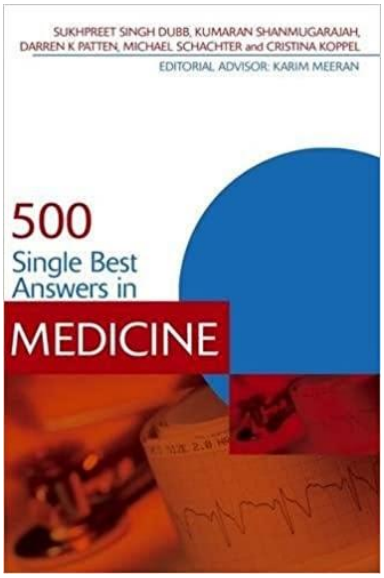

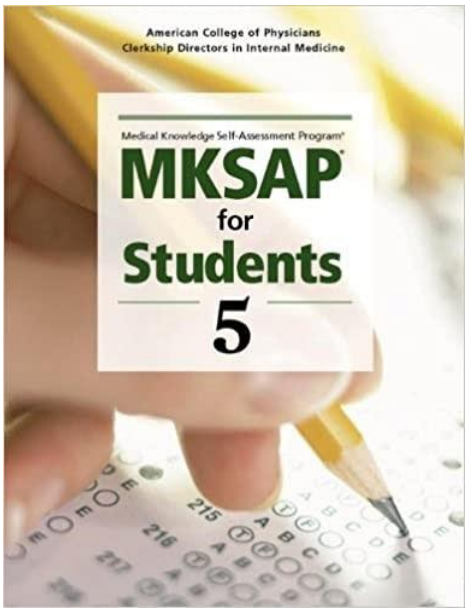

AMBOSS
medical knowledge distilled

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Osteoarthritis

Q1 (Pretest): A 60-year-old man complains of pain in both knees coming on gradually over the past 2 years. The pain is relieved by rest and worsened by movement. The patient is 5 ft 9 in tall and weighs 210 lb. There is bony enlargement of the knees with mild warmth and small effusions. Crepitation is noted on motion of the knee joint bilaterally. There are no other findings except for bony enlargement at the distal interphalangeal joint. Which of the following is the best way to prevent disease progression?

A. Weight reduction
B. Calcium supplementation
C. Total knee replacement
D. Long-term nonsteroidal anti-inflammatory drug (NSAID) administration
E. Oral prednisone

Explanation: The clinical picture of pain in weight-bearing joints made worse by activity is suggestive of degenerative joint disease, also called osteoarthritis. Osteoarthritis may have a mild to moderate inflammatory component. Crepitation in the involved joints is characteristic, as is bony enlargement of the DIP joints. In this overweight patient, weight reduction is the best method to decrease the risk of further degenerative changes. Aspirin, other NSAIDs, or acetaminophen can be used as symptomatic treatment, but these agents do not affect the course of the disease. The long-term use of NSAIDs is limited by potential side effects, including renal insufficiency and gastrointestinal bleeding. Calcium supplementation is relevant for osteoporosis, but does not treat osteoarthritis. Oral prednisone would not be indicated. Intra-articular corticosteroid injections may be given two to three times per year for symptom reduction. Knee replacement is the treatment of last resort, usually when symptoms are not controlled by medical regimens and/or activities are severely limited.

Q2 (500Best): A 79-year-old woman presents to her GP with pain in the left knee. This is particularly bad in the evenings and is stopping her from sleeping. The GP explains that her discomfort is most likely due to osteoarthritis and arranges for her to have an x-ray of the knee. Which of the following descriptions are most likely to describe the x-ray?

A. Reduced joint space, subchondral sclerosis, bone cysts and osteophytes
B. Increased joint space, subchondral sclerosis, bone cysts and osteophytes
C. Reduced joint space, soft tissue swelling and peri-articular osteopenia
D. Increased joint space, soft tissue swelling and peri-articular osteopenia
E. Normal x-ray

Explanation: The main radiographical features of osteoarthritis are reduced joint space, subchondral sclerosis, bone cysts and osteophytes (A). Knowledge of the radiograhical features of osteoarthritis is commonly examined in both written and clinical exams. Joint space is reduced radiographically in osteoarthritis, making answers (B) and (D) incorrect. Reduced joint space, soft tissue swelling and peri-articular osteopenia (C) are radiographical features of rheumatoid arthritis. Additional features include bony erosions and joint subluxation. X-ray findings are likely to be present in this patient, making normal x-ray (E) the incorrect option. However, it is important to note that the extent of radiographical abnormality may not correlate well with the extent of symptoms, and treatment options should be based on the patient and the level of disability rather than the extent of joint damage seen on the x-ray.

Q3 (500Best): A 76-year-old man presents to accident and emergency with pain in his knees. It is worse in the right knee. He describes the pain as being worse in the evening and after exertion. On examination, bony nodules are palpable on the distal interphalangeal joints of both his hands. The right knee is swollen and there is a reduced range of active movement. X-rays show reduction in the joint space, subchondral sclerosis and osteophyte formation. What is the most appropriate treatment?

A. Anti-TNF therapy
B. NSAIDs and urgent orthopaedic follow up
C. NSAIDs and GP follow up
D. NSAID and intramuscular depot injection of methylprednisolone with GP follow up
E. Admit the patient for orthopaedic assessment

Explanation: The case in this question is describing a patient with osteoarthritis. Appropriate analgesia and GP follow up (C) is the most appropriate treatment plan in this question. Analgesia should follow the WHO analgesic ladder with simple analgesia as first-line therapy. NSAID analgesia is likely to be effective for pain relief. Given that the patient has had ongoing symptoms for some months and is able to mobilize, follow up from the GP is the best option. Anti-TNF therapy (A) and steroid therapy (D) are not used in the treatment of osteoarthritis. Orthopaedic follow up (B) to consider surgical options would be appropriate if the pain is uncontrollable and particularly if the patient is getting pain at rest or during the night. Admission (E) should be considered in some patients where the pain is too severe for discharge or social circumstances or co-morbidities mean that management of symptoms at home will not be possible.

Answers: Q1: A | Q2: A | Q3: C

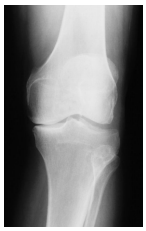
Osteoarthritis

Q4 (500Best): A 32-year-old man presents to accident and emergency with a 1-day history of pain in the right knee. He also mentions that he has had a fever and is feeling generally unwell. On examination, the right knee is swollen, warm and extremely painful to move. What is the most appropriate next step?

A. Empirical intravenous antibiotic treatment
B. X-rays of the right knee
C. Aspiration of the joint and blood cultures
D. Referral for physiotherapy
E. Immobilize the joint

Explanation: The case presented in this question should raise the suspicion of septic arthritis. This is a medical emergency and if left untreated can result in rapid destruction of the joint. Staphlococcus aureus is the most common organism that causes septic arthritis, although gonococcus is also common in young people. Septic arthritis presents as a hot, red, swollen joint that is extremely painful. The systemic features and fever in the case should also point to septic arthritis, but patients may also have no systemic features. The management of patients with suspected septic arthritis should be swift. Aspiration of the joint and blood cultures (C) should be taken first and sent to the microbiology laboratory for culture. Empirical intravenous antibiotic treatment (A) must then be commenced without delay. An example of an empirical regime for septic arthritis would include flucloxacillin, benzylpenicillin and gentamicin. These can then be altered when sensitivites are known. X-rays of the right knee (B) should be performed but are unlikely to aid the diagnosis. Other blood tests including full blood count and CRP should also be sent. The joint should be initially immobilized (E), but this is not the most appropriate next step. Physiotherapy (D) should be started early, but after an initial period of immobilization, making this the wrong answer.

Q5 (MKSAP): A 70-year-old male dairy farmer is evaluated for a 1-year history of pain in the left knee that worsens with activity and is relieved with rest. On physical examination, vital signs are normal. A small effusion is present on the left knee, but there is no erythema or warmth. Range of motion of the left knee elicits pain and is slightly limited Extension of this joint is limited to approximately 10 degrees, but flexion is nearly full. The remainder of the musculoskeletal examination is normal The erythrocyte sedimentation rate is 15 mm/h. A standing radiograph of the left knee is shown Which of the following is the most likely diagnosis ?



A. Avascular necrosis
B. Osteoarthritis
C. Rheumatoid arthritis
D. Torn medial meniscus

Explanation: This patient most likely has osteoarthritis of the knee. He has two risk factors for this condition, advanced age and an occupation involving repetitive bending and physical labor. Osteoarthritis commonly affects weight-bearing joints such as the knees and is characterized by pain on activity that is relieved with rest. Swelling in patients with this condition is usually minimal, and range of motion may be limited. According to the American College of Rheumatology, osteoarthritis of the knee can be diagnosed if knee pain is accompanied by at least three of the following features: age greater than 50 years, morning stiffness lasting less than 30 minutes, crepitus, bony tenderness, bony enlargement, and an absence of palpable warmth. This patient's radiographic findings of osteophytes, joint-space narrowing, sclerosis, and cyst formation are typical of this condition. Arthrocentesis is not necessary to establish a diagnosis of osteoarthritis.
Patients with vascular necrosis of the knee typically experience pain on weight bearing and may have a painful, limited range of motion. However, this condition also is associated with pain on rest and most commonly occurs in patients who use corticosteroids, have systemic lupus erythematosus, or consume excessive amounts of alcoholic beverages. Radiographs in patients with vascular necrosis usually reveal density changes; subchondral radiolucency; cysts; sclerosis; and, eventually, joint-space narrowing. Rheumatoid arthritis may be associated with a limited range of motion and joint-space narrowing visible on radiography.
Patients with rheumatoid arthritis is usually have symmetric arthritis that affects at least three joints as well as an elevated erythrocyte sedimentation rate and is associated with morning stiffness that persists for more than 30 minutes. In addition, rheumatoid arthritis also would not explain the presence of subchondral sclerosis and osteophytes on radiography.
A torn medial meniscus would cause pain in the knee and can occur in the elderly in association with osteoarthritis. Patients with acute meniscal damage often describe a twisting injury with the foot in a weight-bearing position in which a popping or tearing sensation is felt, followed by severe pain; in addition, this condition is characterized by the sensation that the knee "locks" or "gives out".

Answers: Q4: C | Q5: B

Osteoarthritis

Q6 (MKSAP): A 72-year-old woman is evaluated for a 1-year history of progressive pain in the right knee. The pain is most acute along the medial aspect of the joint, worsens with activity, and is relieved with rest. She has no stiffness in the morning and has had no swelling. She also has not experienced locking or giving away of this joint. On physical examination, vital signs are normal. There is bony enlargement of the proximal and distal interphalangeal joints. There is no evidence of a right knee effusion. Passive flexion and extension of the right knee are painful. Laboratory studies, including complete blood count, erythrocyte sedimentation rate, and C-reactive protein, are normal. Radiograph of the right knee also is normal.
In addition to acetaminophen as needed, which of the following is the most appropriate next step in this patient's management?

A. Arthroscopy
B. Aspiration of the knee
C. MRI of the knee
D. Physical therapy

Explanation: This patient has osteoarthritis of the knee. The most appropriate next step in her management is referral for physical therapy, which is an appropriate first-line management option for patients with this condition. Quadriceps muscle training in particular has been shown to reduce pain in this population group. Use of over-the-counter acetaminophen or an NSAID on an as-needed basis also may benefit this patient.
Arthroscopy and MRI of the knee would most likely reveal abnormalities of the articular cartilage not visible on plain radiography but are not needed to establish the diagnosis of osteoarthritis. Similarly, aspiration of the knee joint would be warranted in patients with an effusion to obtain a synovial fluid leukocyte count but is not needed to establish a diagnosis; furthermore, this patient does not have an effusion.

Q7 (MKSAP): A 60-year-old woman is evaluated for wrist pain of 6 months' duration. The pain is located at the base of the right thumb at the wrist. She is a watercolor artist and a graphic designer and is right handed. The pain is described as a persistent ache for which she takes acetaminophen, which provides moderate relief. She has 20 minutes of morning stiffness in the right thumb that improves with a hot shower. On physical examination, vital signs are normal. Examination of her hands shows no soft tissue swelling, warmth, or redness of any hand or wrist joints. Bilateral boney hypertrophy of the proximal interphalangeal joints is present bilaterally. Range of motion of the right thumb is limited by pain. Point tenderness is elicited at the base of the right thumb at the first carpometacarpal joint. Circular movement of her right thumb exacerbates the pain. Tapping the flexor retinaculum does not reproduce or aggravate the pain nor does passively stretching the tendons over the radial styloid.
Which of the following is the most likely diagnosis?

A. Carpal tunnel syndrome
B. Crystal-induced arthritis
C. de Quervain tenosynovitis
D. Osteoarthritis
E. Rheumatoid arthritis

Explanation: This patient most likely has osteoarthritis of the first carpometacarpal joint. Osteoarthritis in this location often presents with well localized tenderness to palpation. Movement of the thumb in a circular motion ("grind test") will often elicit the pain. Predisposing factors include repetitive use of the wrist or thumb. Patients may present with pain, swelling, or enlargement of the carpometacarpal joint recognized as squaring or boxing at the base of the thumb. Associated findings of osteoarthritis are common and may include boney enlargement of the distal interphalangeal joints (Heberden nodes) or the proximal interphalangeal joints (Bouchard nodes).
Rheumatoid arthrit is is an inflammatory arthritis most often affecting the small joints of the hands (wrist, metacarpophalangeal and proximal interphalangeal joints) and feet (met at arsophalangeal joints) in a symmetric pattern. Stiffness in rheumatoid arthritis usually lasts more than 1 hour, and synovial swelling, tenderness, and warmth are apparent on examination.
de Quervain tenosynovitis is an inflammation of the abductor pollicis longus and extensor pollicis brevis tendons. Pain is present on palpation of the distal aspect of the radial styloid. This point is more proximal than the first carpomet acarpal joint. Pain elicited by flexing the thumb into the palm, closing the fingers over the thumb, and then bending the wrist in the ulnar direction (Finkelstein test) is confirmatory.
Carpal tunnel syndrome will often present with pain, numbness, and tingling in the thumb and first two fingers of the hand. Tapping the flexor retinaculum (Tinel sign) or flexing the wrists against each other (Phalen sign) may exacerbate symptoms. Symptoms are often worse at night or with repetitive movements or continuous pressure on the wrist such as typing on a keyboard.
Crystal-induced arthritis in the hands is usually associated with evidence of joint inflammation such as redness and warmth. Additionally, the chronicity of this patient's symptoms argues against crystal-induced arthropathy. Secondary osteoarthritis, however, can develop in the setting of chronic inflammation from crystal-induced arthritis

Answers: Q6: D | Q7: D

Osteoarthritis

Q8 (MKSAP): A 67-year-old man is evaluated in the office for acute right knee pain that developed 6 days ago after working on his car. The pain is worse when he walks or climbs stairs and improves when he rests. He has no fever and denies knee stiffness. He has minimal pain relief with maximal dosage of acetaminophen and the application of ice. He has no other medical illness and takes no medications. On examination, his temperature is 36.8°C (98.2°F), blood pressure is 132/75 mm Hg, heart rate is 92/min and respiratory rate is 14/min. BMI is 27. Moderate valgus deformity of both knees is present. The right knee has a moderate effusion without warmth or erythema. Pain is elicited with passive and active range of motion. The remainder of his examination is normal. Plain radiographs of his left knee show severe medial joint space narrowing with subchondral sclerosis and osteophyte formation. Analysis of the synovial fluid reveals 1100 leukocytes/uL (1.1 × 10°/L) (79% lymphocytes, 10 % macrophages)
Which of the following is the best management for this patient's knee pain?

A. Begin a nonsteroidal anti-inflammatory drug
B. Begin empiric antibiotics
C. Inject intra-articular corticosteroids
D. Obtain right knee magnetic resonance imaging (MRI)
E. Refer for total knee arthroplasty

Explanation: The best management for this patient is to begin a nonsteroidal anti-inflammatory drug (NSAID). This patient most likely has acute osteoarthritis of the knee. Classic findings of osteoarthritis include pain with activity that is relieved with rest. The patient's radiographic findings of joint space narrowing, subchondral sclerosis and osteophyte formation are consistent with osteoarthritis, and his valgus deformity predisposes him to medial compartment osteoarthritis due to uneven loading forces when ambulating. He has failed treatment with acetaminophen and therefore an oral NSAID such as ibuprofen is the appropriate next management step. Physical therapy, temporary use of a cane and bracing or taping are also reasonable initial interventions. NSAIDs are associated with an increased risk of gastrointestinal bleeding and cardiovascular disease. The American College of Rheumatology guidelines recommend that physicians and patients weigh the potential risks and benefits of treatment with NSAIDs, but no evidence-based guidelines yet exist to indicate which patients can safely use these agents.
Intra-articular corticosteroid or hyaluronan injections may be considered in patients with mono- or pauciarticular osteoarthritis in whom NSAIDs are either contraindicated or do not provide adequate pain relief. Referral to an orthopedic surgeon for consideration of total joint arthroplasty (replacement) of the knee is warranted only when no further medical therapy is available and the patient decides that the impairment caused by his or her condition warrants this intervention. This patient has not had a sufficient trial of more conservative measures to warrant either of these therapies.
Magnetic resonance imaging would be indicated if there were features on history or physical exam that suggested another etiology for the pain. Meniscal tears are seen almost universally in patients with osteoarthritis of the knee and are not necessarily a cause of increased symptoms. Removal of menisci should be avoided unless symptoms of knee locking or the inability to extend the knee are present.
Antibiotic treatment is not indicated because arthrocentesis does not suggest septic arthritis. In noninflammatory arthritis, the synovial fluid leukocyte count is usually less than 2000/uL (2.0 x 10'/L). Septic arthritis usually has leukocyte counts greater than 50,000/uL (50 x 10'/L) and a predominance of polymorphonuclear cells.
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Q9 (AMBOSS): A 67-year-old woman comes to the physician for the evaluation of bilateral knee pain for the past year. She reports that the pain is worse with movement and is relieved with rest. She has type 2 diabetes mellitus. The patient says her mother takes leflunomide for a "joint condition." The patient's medications include metformin and a multivitamin. She is 165 cm (5 ft 5 in) tall and weighs 85 kg (187 lb); BMI is 31 kg/m2. Vital signs are within normal limits. Physical examination shows pain both in complete flexion and extension, crepitus on joint movement, and joint stiffness and restricted range of motion of both knees. X-ray of the knee joints shows irregular joint space narrowing, subchondral sclerosis, osteophytes, and several subchondral cysts. There is no reddening or swelling. Which of the following is the most appropriate pharmacotherapy?

A. Intra-articular glucocorticoid injections
B. Administration of ibuprofen
C. Administration of infliximab
D. Administration of methotrexate
E. Administration of oral prednisolone

Explanation: This patient presents with symptoms of osteoarthritis. Management is focused on treating pain, as well as maintaining and improving joint mobility and function. While nonpharmacological treatment approaches include physical therapy, exercise, and patient education (e.g., maintaining a healthy weight), NSAIDs such as ibuprofen are recommended as first-line pharmacological treatment for pain relief.

Answers: Q8: A | Q9:B

Osteoarthritis

Q10 (AMBOSS): A 52-year-old woman comes to the physician because of a 4-month history of progressive pain and stiffness of the fingers of her right hand that is worse at the end of the day. She works as a hairdresser and has to take frequent breaks to rest her hand. She has hypertension for which she takes hydrochlorothiazide. Two weeks ago, she completed a course of oral antibiotics for a urinary tract infection. Her sister has systemic lupus erythematosus. She drinks one to two beers daily and occasionally more on weekends. Over the past 2 weeks, she has been taking ibuprofen as needed for the joint pain. Her vital signs are within normal limits. Physical examination shows swelling, joint-line tenderness, and decreased range of motion of the right first metacarpophalangeal joint as well as the 2nd and 4th distal interphalangeal joints of the right hand. Discrete, hard, mildly tender swellings are palpated over the 2nd and 4th distal interphalangeal joints of the right hand. Which of the following is the most likely underlying mechanism for these findings?

A. Bacterial infection of the joint space
B. Monosodium urate crystal precipitation in the joints
C. Autoimmune-mediated cartilage erosion
D. Calcium pyrophosphate dihydrate crystal precipitation in the joints
E. Degenerative disease of the joints

Explanation: Osteoarthritis (OA) is a degenerative arthritis that is characterized by pain and stiffness that worsens with activity. It can affect any joint, but frequently affects the knees and hand joints (most commonly DIP and PIP). The Heberden nodes seen in this patient are a characteristic sign of OA.
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Q11 (AMBOSS): A 61-year-old man with progressive left hip pain comes to the physician for a follow-up examination. One year ago, he was diagnosed with osteoarthritis of the left hip. Since then, he has had an 8-kg (18-lb) weight loss after changing to a vegetarian diet, regular swimming, and physical therapy. The pain worsens when he climbs stairs, which makes it increasingly difficult for him to reach his apartment located on the second floor. Over the last few weeks, he gradually increased the frequency of diclofenac intake but says that even a daily intake does not provide complete pain relief. He asks for a treatment that will lead to a long-term improvement of his symptoms. He has no history of major medical illness. His only other medication is pantoprazole. He does not smoke or drink alcohol. He is 179 cm (5 ft 10 in) tall and weighs 80 kg (176 lb); BMI is 25 kg/m2. Physical examination of the left hip shows crepitus, a limited internal rotation, and pain with full flexion and extension. An x-ray of the left hip shows joint space narrowing, osteophytes, and subchondral sclerosis and cysts. Which of the following is the most appropriate recommendation for this patient?

A. Arthroscopic hip debridement
B. Intraarticular glucocorticoid injections
C. Walking aids
D. Total hip arthroplasty
E. Fentanyl patch

Explanation: Total hip arthroplasty is recommended in patients with hip OA who have experienced inadequate relief using conservative measures (physical therapy, exercise, weight loss, pain medication), there is evidence of advanced OA (e.g., severe pain, radiological signs of osteoarthritis), and symptoms impact the patient's daily life significantly, as seen here. In addition, this patient has no evidence of contraindications to surgery (e.g., active infection, major medical illness).
Intraarticular glucocorticoid injections can be used as second-line therapy for short-term pain relief (e.g., while waiting for surgery).

Answers: Q10: E | Q11:D

Gout

Q1 (Pretest): A 40-year-old man complains of acute onset of exquisite pain and tenderness in the left ankle. There is no history of trauma. The patient is taking hydrochlorothiazide for hypertension. On examination, the ankle is very swollen and tender. There are no other physical examination abnormalities. Which of the following is the best next step in management?

A. Begin colchicine and broad-spectrum antibiotics.
B. Perform arthrocentesis.
C. Begin allopurinol if uric acid level is elevated.
D. Obtain ankle x-ray to rule out fracture.
E. Apply a splint or removable cast.

Explanation: The sudden onset and severity of this monoarticular arthritis suggests acute gouty arthritis, especially in a patient on diuretic therapy. However, an arthrocentesis is indicated in the first episode to document gout by demonstrating needle-shaped, negatively birefringent crystals and to rule out other diagnoses such as infection. The level of serum uric acid during an episode of acute gouty arthritis may actually fall. Therefore, a normal serum uric acid does not exclude a diagnosis of gout. For most patients with acute gout, NSAIDs are the treatment of choice. Colchicine is also effective but causes nausea and diarrhea. Systemic corticosteroids can be used if NSAIDs are contraindicated. Antibiotics should not be started for suspected septic arthritis before an arthrocentesis is performed. Treatment for hyperuricemia should not be initiated in the setting of an acute attack of gouty arthritis. Long-term goals of management are to control hyperuricemia, prevent further attacks, and prevent joint damage. Long-term prophylaxis with allopurinol is considered for repeated attacks of acute arthritis, urolithiasis, or formation of tophaceous deposits. X-ray of the ankle would likely be inconclusive in this patient with no trauma history. In addition, the x-ray changes of tophaceous gout take years to develop. In the absence of trauma, there is no indication for immobilization.

Q2 (Pretest): A 65-year-old man develops the onset of severe knee pain over 24 hours. The knee is red, swollen, and tender. The patient does not have fever or systemic symptoms. He has a history of diabetes mellitus and cardiomyopathy. Definitive diagnosis is best made by which of the following?

A. Serum uric acid
B. Serum calcium
C. Arthrocentesis and identification of positively birefringent rhomboid crystals
D. Rheumatoid factor
E. ANA

Explanation: Acute monoarticular arthritis in association with linear calcification of the cartilage of the knee (chondrocalcinosis) suggests the diagnosis of pseudogout, a form of calcium pyrophosphate dihydrate deposition disease (CPPDD). In its acute manifestation, the disease resembles gout. Positively birefringent crystals (looking blue when parallel to the axis of the red compensator on a polarizing microscope) can be demonstrated in joint fluid, although careful search is sometimes necessary. Serum uric acid and calcium levels are normal, as are rheumatoid factor and antinuclear antibodies. Pseudogout is about half as common as gout, but becomes more common after age 65. Calcium pyrophosphate dihydrate deposition disease is diagnosed in symptomatic patients by characteristic x-ray findings and crystals in synovial fluid. Pseudogout is treated with NSAIDs, colchicine, or steroids. Arthrocentesis and drainage with intraarticular steroid administration is also an effective treatment. Linear calcifications or chondrocalcinosis are often found in the joints of elderly patients who do not have symptomatic joint problems; such patients do not require treatment.

Q3 (500Best): A 70-year-old woman presents to accident and emergency with sudden onset pain and swelling in the right knee. Her past medical history includes hypertension and hypercholesterolaemia. She is currently taking aspirin, ramipril and simvastatin. On examination, she is apyrexial and the right knee is swollen. There is reduced range of movement in the knee due to swelling and pain. X-ray of the right knee shows chondrocalcinosis. What is the most likely diagnosis?

A. Gout
B. Pseudo-gout
C. Septic arthritis
D. Reactive arthritis
E. Osteoarthritis

Explanation: Pseudo-gout (B) is caused by the presence of calcium pyrophosphate crystals in the joint, causing an acute synovitis. Pseudo-gout most commonly affects **elderly women and usually involves the knee or wrists.** It may also be seen in younger patients with underlying conditions causing the deposition of calcium pyrophosphate crystals such as hypothyroidism, hyperparathyroidism, acromegaly Wilson's disease or haemochromatosis. X-ray of the affected joint may show **chondrocalcinosis** (calcification of the hyaline cartilage). Treatment of pseudo-gout is with aspiration of the joint and NSAIDs. Intra-articular steroid injection can be used if pain is not controlled. The acute synovitis of pseudogout resembles gout (A). While acute gout most commonly affects the first metatarsophalangeal joint, other joints may be affected. However, the finding of chondrocalcinosis makes pseudo-gout more likely than gout. Septic arthritis is a differential diagnosis of pseudo-gout and should be considered, despite the apyrexia. Therefore, the joint aspirate should be sent for culture. The chondrocalcinosis on x-ray makes the diagnosis of pseudo-gout more likely than septic arthritis (C) in this question. Reactive arthritis (D) presents as an asymmetrical polyarthritis of the lower limbs, making this answer incorrect. Osteoarthritis (E) may affect the knee. However, the chondrocalcinosis again makes pseudo-gout the more likely answer. In addition, a history of pain in the knee would be expected.

Answers: Q1: B | Q2: C | Q3: B

Gout

Q4 (500Best): A 74-year-old woman presents to accident and emergency with sudden onset pain and swelling in the left knee. On examination, she is apyrexial and the left knee is swollen. There is reduced range of movement in the knee due to swelling and pain. X-ray of the right knee shows chondrocalcinosis. Microscopy of the fluid aspirated from the joint is most likely to show:

A. Rhomboidal, weakly positively birefringent crystals under polarized light microscopy
B. Needle-shaped negatively birefringent crystals under polarized light microscopy
C. Atypical mononuclear cells
D. ReedSternberg cells
E. Tophi

Explanation: The presence of rhomboidal, weakly positively birefringent crystals (A) under polarized light microscopy in joint fluid is diagnostic of pseudogout. Needle-shaped negatively birefringent crystals (B) are seen in gout. Atypical mononuclear cells (C) are found on microscopy of blood samples in patients with infectious mononucleosis. Microscopic analysis of lymph node biopsy specimens in patients with Hodgkin's lymphoma may show Reed–Sternberg cells (D). Tophi (E) are the white deposits seen in skin and soft tissue in some patients with gout. They are composed of sodium urate and the presence of tophi in a patient with long-standing gout is called 'chronic tophaceous gout'.

Q5 (500Best): A 59-year-old man presents to his GP with sudden onset severe pain, tenderness and swelling of the first metatarsophalangeal joint. He is known to suffer from acute gout and has had several previous similar episodes. What is the most appropriate treatment?

A. Allopurinol
B. NSAIDs
C. Conservative measures including reduced alcohol intake and weight loss
D. Intra-articular steroid injection
E. Methotrexate

Explanation: The most appropriate treatment of acute episodes is with a strong NSAID (B) such as indomethacin. If NSAIDs are contraindicated, for example in peptic ulcer disease, colchicine can be used. For patients with recurrent attacks of gout, such as this case, serum urate should be reduced with conservative measures and long-term allopurinol (A). Conservative measures (C) include weight loss and avoiding excess alcohol, purine-rich food and low-dose aspirin. However, the treatment of the acute episode is the immediate priority here. Allopurinol may exacerbate an acute episode and must only be started after the attack has resolved. Steroids (D) by oral, intramuscular, intra-articular routes can be given in an acute episode when NSAIDs and colchicine are contraindicated, for example in renal failure. Methotrexate (E) is a DMARD that is not used in the management of gout.

Q6 (MKSAP): A 36-year-old man is evaluated for the acute onset of a warm swollen right ankle of 3 days' duration. He had a similar episode 2 years ago involving his left great toe that resolved in 5 days. He is otherwise healthy and takes no medications.
On physical examination, temperature is 36.7°C (98.0°F), blood pressure is 140/90 mm Hg, pulse rate is 80/min, and respiration rate is 12/min. Abnormal findings are limited to a warm swollen right ankle with painful range of motion.
An Arthrocentesis Is performed. Synovial fluid cell count is 30,000/µL (30 × 109/L) with 95% polymorphonuclear cells and 5% lymphocytes. Gram stain is negative for bacteria.
Polarized microscopy demonstrates intracellular monosodium urate crystals.
Which of the following is the most appropriate treatment?

A- Allopurinol
B- Colchicine
C- Febuxostat
D- Indomethacin

Explanation: The most appropriate treatment for this patient is administ rat ion of an NSAID such as indomethacin. Definitive diagnosis of gout requires the identification of monosodium urate crystals on arthrocentesis or aspiration of a tous. During an at t ack of gout , needle-shaped monosodium urate crystals that typically appear engulfed by the neutrophils are visible on compensated polarized light microscopy. NSAIDs, corticosteroids, and colchicine are options in the treatment of an acute attack of gout . NSAIDs are highly effective when administered during an acute attack, but they should be used with caution in patients at risk for renal impairment , bleeding, or ulcer disorders, especially in the elderly. Oral, intra-articular, or intravenous corticosteroid therapy is also effective in acute gouty attacks. However, oral and intravenous therapy may be problematic in patients with diabet es mellitus. Colchicine is most effective in patients with monoarticular involvement and, when used within the first 24 hours of symptoms, can abort a severe attack. At the first sign of an attack in patients with normal renal function, this agent is usually administered two or three times daily until the patient experiences symptomatic relief, develops gastrointestinal toxicity, or reaches a maximum dose of 6 mg per attack. Allopurinol and febuxostat are xanthine oxidase inhibitors useful in reducing uric acid levels in patients with recurrent attacks of acute gout and patients with uric acid tophi or renal stones. Rapid control of serum uric acid levels generally is not necessary during an acute attack, and acute increases and decreases in the uric acid level alter the steady state and may prolong the current attack or precipitated new attacks. Prophylactic colchicine, low-dose corticosteroids, or NSAIDs initiated at least 1 week before beginning or adjusting the dose of uric acid-lowering therapy help t o prevent disease flares associated with changes in uric acid levels and may need t o be continued until therapeutic serum uric acid levels have been achieved (<6 mg/dL [0.4 mmol/L]). Prolonged use of these agents may be indicated in patients with chronic tophaceous gout until the disease is controlled.

Answers: Q4: A | Q5: B | Q6: D

Gout

Q7 (MKSAP): A 68-year-old man is evaluated for hyperuricemia. He has had multiple attacks of acute gout typically affecting the great toe, but 2 weeks ago he had an attack involving his right great toe and both ankles. Arthrocentesis confirmed the presence of monosodium urate crystals. He was successfully treated with ibuprofen. Physical examination is normal except for a tophaceous deposit on the right forefoot. Laboratory studies are normal except for a serum uric acid level of 11.2 mg/dL (0.7 mmol/L).
Which of the following is the most appropriate therapy for this patient?

A- Allopurinol
B- Low-dose colchicine
C- Low-dose colchicine and allopurinol
D- Low-dose indomethacin

Explanation: The most appropriate therapy for this patient is low-dose colchicine and allopurinol. Criteria for initiating treatment of hyperuricemia in patients with symptomatic gout include the presence of tophi or renal stones, multiple attacks of acute gout, or a history of a decreasing period between attacks. Uric acid-lowering therapy typically is not initiated until a patient experiences two documented acute attacks. Dietary purine restriction, weight loss, and discontinuation of alcohol may help to decrease uric acid levels in patients with mild hyperuricemia and symptomatic gout. Medications that raise serum uric acid levels, such as thiazide diuretics and low-dose salicylates, should be discontinued if alternative therapy is available. However, most patients with recurrent gouty attacks, particularly those with tophaceous deposits, require pharmacologic therapy to lower serum levels of uric acid. The goal in uric acid-lowering therapy is to achieve a serum uric acid level less than 6.0 mg/dL (0.4 mmol/L), not just levels within the normal range. When the uric acid levels are below 6.0 mg/dL (0.4 mmol/L), monosodium urate crystals from within the joint and from soft-tissue tophaceous deposits are reabsorbed. Prophylactic colchicine, low-dose corticosteroids (10 mg/d or less), or nonsteroidal anti-inflammatory drugs (NSAIDS) initiated at least 1 week before beginning or adjusting the dose of uric acid-lowering therapy help to prevent disease flares associated with changes in uric acid levels and may need to be continued until therapeutic serum uric acid levels have been achieved. Low-dose NSAIDS, such as indomethacin, or low-dose colchicine may prevent attacks of gout but do not lower uric acid levels and, therefore, cannot prevent the continued accumulation of uric acid in soft tissues (tophi), uric acid kidney stones, or destructive arthritis.

Q8 (AMBOSS): A 56-year-old man comes to the physician for a follow-up examination. Two weeks ago, he was treated for an acute gout attack of the metatarsophalangeal joints of his right big toe. His symptoms improved with naproxen. He has had three other similar episodes of joint pain in his toes and ankles during the last year that improved with over-the-counter analgesics. He does not currently take any medications. He used to drink 3–5 beers daily but has recently cut down. He is a chef at a steakhouse. His temperature is 37.0°C (98.6°F), pulse is 76/min, and blood pressure is 147/83 mm Hg. Examination of his right big toe shows minimal tenderness; there is no warmth or apparent deformity. The remainder of the examination shows no abnormalities. His serum creatinine concentration is 0.9 mg/dL. Long-term treatment with which of the following drugs is most appropriate to prevent future gout attacks?

A- Allopurinol
B- Colchicine
C- Aspirin
D- Prednisolone

Explanation: This patient with a history of chronic gout has been adequately treated with NSAIDs for his acute gouty attacks. Given his repeated exacerbations (≥ 2 gout attacks during the past year), he should be started on urate-lowering therapy, which is also indicated in patients with evidence of tophaceous gout. Urate-lowering therapy should also be considered in patients with a first episode of acute gout who have hyperuricemia (> 9 mg/dL), advanced chronic kidney disease, or a history of urolithiasis. Allopurinol is the preferred first-line urate-lowering drug. Despite its therapeutic effect, it may also trigger an acute gout attack; to prevent this, antiinflammatory prophylaxis (colchicine, NSAIDs, or glucocorticoids) should be given to all patients during initiation of treatment. Other side effects of allopurinol include nausea and diarrhea.

Answers: Q7: C | Q8: A

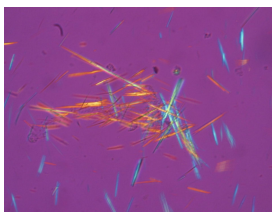
Gout

Q9 (AMBOSS): A 53-year-old man comes to the emergency department for severe left knee pain for the past 8 hours. He describes it as an unbearable, burning pain that woke him up from his sleep. He has been unable to walk since. He has not had any trauma to the knee. Ten months ago, he had an episode of acute pain and swelling of the right great toe that subsided after treatment with indomethacin. He has hypertension, type 2 diabetes mellitus, psoriasis, and hyperlipidemia. Current medications include topical betamethasone, metformin, glipizide, losartan, and simvastatin. Two weeks ago, hydrochlorothiazide was added to his medication regimen to improve blood pressure control. He drinks 1–2 beers daily. He is 170 cm (5 ft 7 in) tall and weighs 110 kg (242 lb); BMI is 38 kg/m2. His temperature is 38.4°C (101.1°F). Examination shows multiple scaly plaques over his palms and soles. The left knee is erythematous, swollen, and tender; range of motion is limited by pain. Which of the following is the most appropriate next step in management?

A- Serum uric acid level
B- X-ray of the knee
C- Arthrocentesis
D- Oral methotrexate

Explanation: This patient's presentation (acute, severe knee pain, swelling, and fever) could be caused by gout, pseudogout, or septic arthritis. While acute gouty arthritis is the most likely diagnosis in this patient, arthrocentesis and synovial fluid analysis are indicated to establish a definitive diagnosis in all new-onset acute gout attacks and/or if past suspected gout attacks were not confirmed via arthrocentesis. Synovial fluid analysis allows alternative diagnoses to be ruled out and ensures the underlying condition is treated appropriately. In gout and pseudogout, fluid samples taken during arthrocentesis are examined with polarized light microscopy. Needle-shaped, negatively birefringent urate crystals are characteristic of gout, while rhomboid-shaped, positively birefringent crystals indicate pseudogout. Arthrocentesis in septic arthritis shows yellowish-green synovial fluid with an elevated WBC count (typically > 50,000/µL).

Q10 (AMBOSS): A 57-year-old man is brought to the emergency department for worsening pain and swelling of his left ankle for the past 2 hours. The pain is severe and awakened him from sleep. He has poorly-controlled hypertension and hyperlipidemia. Current medications include hydrochlorothiazide and pravastatin. His temperature is 37.8°C (100.1°F), pulse is 105/min, and blood pressure is 148/96 mm Hg. Examination shows exquisite tenderness, erythema, and edema of the left ankle; active and passive range of motion is limited by pain. Arthrocentesis of the ankle joint yields cloudy fluid with a leukocyte count of 19,500/mm3 (80% segmented neutrophils). Gram stain is negative. A photomicrograph of the joint fluid aspirate under polarized light is shown. Which of the following is the most appropriate acute pharmacotherapy?



A- Colchicine
B- Allopurinol
C- Aspirin
D- Colchicine and allopurinol

Explanation: This patient's presentation is consistent with an acute gout attack, for which his history of hypercholesterolemia, hypertension, and thiazide diuretic use are risk factors. First-line therapy for acute gout exacerbations includes nonsteroidal anti-inflammatory drugs (e.g., indomethacin), colchicine, or corticosteroids. The choice of treatment is typically based on the patient's age, comorbidities, and drug availability; because this patient has poorly-controlled hypertension, NSAIDs should be avoided. Colchicine, a microtubule polymerization inhibitor that blocks leukocyte phagocytosis of urate crystals, is contraindicated in patients with advanced liver disease and severe renal dysfunction, neither of which this patient has a history of. Colchicine is therefore an acceptable treatment option for this patient. Combination therapy with colchicine and allopurinol is not indicated for the acute management of gout exacerbations. Allopurinol is indicated for maintenance therapy in chronic gout.

Answers: Q9: C | Q10: A

Scleroderma spectrum disease

Q1 (500Best): A 60-year-old woman presents to her GP with a two-month history of lethargy and weakness. She mentions that she is finding it increasingly difficult to climb the stairs and do the housework. On examination, there is wasting and weakness of the proximal muscles in the upper and lower limbs. What is the most likely diagnosis?

A. Dermatomyositis
B. Polymyositis
C. Polymyalgia rheumatica
D. Kawasaki's disease
E. Polyarteritis nodosa

Explanation: Polymyositis (B) occurs due to inflammation of striated muscle, resulting in proximal muscle weakness. It affects women more than men. The onset usually occurs over a period of months and may include systemic features such as lethargy and weight loss. The proximal muscles of the shoulder and pelvis girdle become weak and wasted. The disease may then progress to involve the pharyngeal, laryngeal or respiratory muscles. When features of polymyositis are accompanied with overlying skin changes, it is termed 'dermatomyositis' (A). **These include a heliotrope (lilac) rash over the eyelids, scaly red papules over the knuckles, elbows or knees (Gottron's papules), a macular rash over the back and shoulders (shawl sign) or painful cracking over the tips of the fingers (mechanic's hands). The absence of these features makes dermatomyositis the incorrect answer.** Polymyalgia rheumatica (C) may affect the proximal muscles. However, it is associated with pain, which is an uncommon feature of polymyositis. Although investigations have not been mentioned in this question, it is worth noting that polymyalgia rheumatica is associated with a raised ESR while in polymyositis, the ESR is usually not raised. Kawasaki's disease (D) is a medium-vessel vasculitis that affects mainly children and does not cause proximal muscle weakness, making this answer incorrect. Polyarteritis nodosa (E) is also a medium vessel vasculitis, which affects multiple organs. Proximal muscle weakness is not a feature, making this answer incorrect.

Q2 (500Best): A 45-year-old woman presents to the rheumatology clinic with a three-month history of itchy, dry eyes and a persistently dry mouth. She also mentions that her fingers have been extremely cold, occasionally turning blue after going outside in the morning. Shirmer's test is positive. What is the most likely diagnosis?

A. Systemic sclerosis
B. Raynaud's disease
C. SLE
D. Primary Sjögren's syndrome
E. Secondary Sjögren's syndrome

Explanation: Primary Sjögren's syndrome (D) occurs due to underlying fibrosis of exocrine glands. Clinical features include decreased tear production, decreased salivation and parotid gland swelling. There may be systemic disease, including arthritis, vasulitis or organ involvement (including pulmonary disease, renal tubular involvement, thyroid disease, myaesthenia gravis, primary biliary cirrhosis). Schirmer's test uses filter paper under the lower eyelid to measure tear production. Reduced tear production and salivation are not features of systemic sclerosis (A), making this answer incorrect. While Raynaud's phenomenon is a systemic manifestation of Sjögren's syndrome, primary Raynaud's disease (B) just affects the hands and is thus the incorrect answer. Secondary Sjögren's syndrome may occur in patients with other connective tissue disorders including SLE or systemic sclerosis. **The absence of features suggestive of these other connective tissue disorders means SLE (C) and secondary Sjögren's syndrome (E) are the incorrect answers.**

Q3 (500Best): A 24-year-old woman presents to her GP complaining of cold hands and feet. This has been ongoing for the past three months and is especially bad when she goes out in the mornings and may last for hours. On further questioning, she mentions that her hands sometimes turn blue or red and that gloves are unhelpful. She has otherwise been feeling well and has no past medical history. What is the most appropriate treatment?

A. Propanolol
B. Aspirin
C. Nifedipine
D. Subcutaneous injection of low molecular weight heparin
E. Prednisolone

Explanation: The case in this question is describing Raynaud's syndrome. Vasospasm results in peripheral digital ischaemia. Precipitating factors include exposure to cold. Patients may describe their hands becoming pale, then blue and then red. Raynaud's disease is when Raynaud's syndrome occurs without an underlying cause, as in this case. However, Raynaud's syndrome is associated with a wide range of underlying connective tissue diseases. In these cases, it is termed Raynaud's phenomenon. Treatment of Raynaud's disease is with conservative measures such as warm gloves. In addition, nifedipine (C) can be used, making this the correct answer. Propanolol (A) is contraindicated in patients with Raynaud's syndrome as it may worsen the digital ischaemia. Aspirin (B), subcutaneous injection (D) and prednisolone (E) are not treatments for Raynaud's syndrome.

Answers: Q1: B | Q2: D | Q3: C

Scleroderma spectrum disease

Q4 (500Best): A 42-year-old woman presents to accident and emergency with retrosternal discomfort. She was diagnosed with systemic sclerosis a year ago. Which of the following statements is true about systemic sclerosis?

A. Microstomia is only seen in diffuse cutaneous systemic sclerosis
B. Skin involvement is limited to face, hands and feet in limited cutaneous systemic sclerosis
C. Oesophageal dysmotility is only seen in limited cutaneous systemic sclerosis
D. Anti-double stranded DNA antibodies are normally detected in patients with systemic sclerosis
E. Raynaud's phenomenon occurs as a result of skin fibrosis (scleroderma)

Explanation: Systemic sclerosis is a multisystem disease of unknown cause. It may be limited to skin and soft tissue in limited cutaneous systemic sclerosis or also involve the organs in diffuse cutaneous systemic sclerosis. Skin involvement of the face may be seen in both forms and may produce the characteristic beak-like nose and small mouth, termed microstomia (A). In limited cutaneous systemic sclerosis, skin involvement is limited to the hands, face and feet, making this the correct answer (B). In diffuse cutaneous systemic sclerosis, skin across most of the body can be affected in the worst cases. Oesophageal dysmotility (C) may occur in both forms of the disease, making this option incorrect. Raised levels of anti-nuclear antibody, anti-centromere antibody (limited cutaneous systemic sclerosis), anti-Ro and anti-topoisomerase antibodies are associated with systemic sclerosis. Anti-double stranded DNA antibodies (D) are normally associated with SLE not systemic sclerosis, making this answer wrong. Raynaud's phenomenon (E) results from digital ischaemia, not skin fibrosis as mentioned here.

Q5 (Pretest): A 60-year-old woman complains of dry mouth and a gritty sensation in her eyes. She states it is sometimes difficult to speak for more than a few minutes. There is no history of diabetes mellitus or neurologic disease. The patient is on no medications. On examination, the buccal mucosa appears dry and the salivary glands are enlarged bilaterally. Which of the following is the best next step in evaluation?

A. Lip biopsy
B. Schirmer test and measurement of autoantibodies
C. IgG antibody to mumps virus
D. A therapeutic trial of prednisone for 1 month
E. Administration of a benzodiazepine

Explanation: The complaints described are characteristic of Sjögren syndrome, an autoimmune disease with presenting symptoms of dry eyes and dry mouth. The disease is caused by lymphocytic infiltration and destruction of lacrimal and salivary glands. The Schirmer test, which assesses tear production by measuring the amount of wetness on a piece of filter paper placed in the lower eyelid for 5 minutes, is the appropriate screening test. Most patients with Sjögren syndrome produce autoantibodies, particularly anti-Ro (SSA). Lip biopsy is needed only to evaluate uncertain cases, such as when dry mouth occurs without dry eye symptoms. Mumps can cause bilateral parotitis, but would not explain the patient's complaint of a gritty sensation, which is the most typical symptom of dry eye syndrome. Corticosteroids are reserved for severe vasculitis or other serious complications. Although anxiety (for which a benzodiazepine could be administered) can cause a dry mouth, it would not cause either parotid swelling or dry eyes.

Q6 (Pretest): A 45-year-old woman has pain in her fingers on exposure to cold, arthralgias, and difficulty swallowing solid food. She has a few telangiectasias over the chest but no erythema of the face or extensor surfaces. There is slight thickening of the skin over the hands, arms, and torso. What is the best diagnostic test?

A. Rheumatoid factor
B. Antinuclear, anti-topoisomerase I, and anticentromere antibodies
C. ECG
D. BUN and creatinine
E. Reproduction of symptoms and findings by immersion of hands in cold water

Explanation: The symptoms of Raynaud phenomenon, arthralgia, and dysphagia point toward the diagnosis of scleroderma. Scleroderma, or systemic sclerosis, is characterized by a systemic vasculopathy of small- and medium-sized vessels, excessive collagen deposition in tissues, and an abnormal immune system. It is an uncommon multisystem disease affecting women more often than men. There are two variants of scleroderma—a limited type (previously known as CREST syndrome) and a more severe, diffuse disease. Antinuclear antibodies are almost universal. Topoisomerase-I antibody occurs in only 30% of patients with diffuse disease, but a positive test is highly specific. Anti-centromere antibodies are more often positive in limited disease. Cardiac involvement may occur, and an ECG could show heart block but is not at all specific. Renal failure can develop insidiously, but BUN and creatinine levels would not be diagnostically specific. Rheumatoid factor is nonspecific and present in 20% of patients with scleroderma. Reproduction of Raynaud phenomena is nonspecific and is not recommended as an office test.

Answers: Q4: B | Q5: B | Q6: B

Scleroderma spectrum disease

Q7 (MKSAP): A 63-year-old woman with dermatomyositis is evaluated for cough and dyspnea. She was diagnosed with polymyositis 6 months ago and has been treated with prednisone and methotrexate. She was doing very well until 6 weeks ago when she developed a dry cough and progressive dyspnea. She has no history of pulmonary disease and does not smoke. On physical examination, temperature is 37.0°C (98.6°F), blood pressure is 110/60 mm Hg, pulse rate is 88/min, and respiration rate is 20/min. The cardiac examination is normal. No jugular venous distention or peripheral edema is evident. On pulmonary auscultation, bibasilar crackles are heard. Muscle strength is normal, and no skin rash is evident. Laboratory evaluation shows a normal complete blood count, comprehensive chemistry panel, and serum creatine kinase level. Chest x-ray shows increased interstitial markings in both lung bases.
Which of the following is the most likely diagnosis?

A. Community-acquired pneumonia
B. Heart failure
C. Interstitial lung disease
D. Pneumocystis pneumonia

Explanation: The most likely diagnosis is interstitial lung disease (ILD). ILD with progressive pulmonary fibrosis and secondary pulmonary arterial hypertension is one of the leading causes of death in patients with polymyositis and dermatomyositis is. ILD may be prominent at the onset of myopathy or develop over the course of the disease. The presence of anti-Jo-1 antibodies is associated with an increased risk for ILD. Patients with ILD have progressive dyspnea, basilar crackles, bibasilar infiltrates on chest radiographs, and restrictive changes on pulmonary function studies, including a decreased forced vital capacity, total lung capacity, and diffusing capacity of the lungs for carbon monoxide. Chest radiographs demonstrate an interstitial pattern, and high-resolution CT scans of the chest most commonly suggest a diagnosis of nonspecific interstitial pneumonia. Lung biopsy is generally not needed for diagnosis, but bronchoscopy may be needed to exclude infection.
Typical community-acquired pneumonia is characterized by rapid onset of high fever, productive cough, and pleuritic chest pain, all of which are absent in this patient. Cardiac involvement in patients with an inflammatory myopathy is rare and includes arrhythmias and cardiomyopathy. Furthermore, this patient has no findings to support heart failure, including an S,, jugular venous distention, or peripheral edema. Approximately 1% to 2% of patients with rheumatic diseases can develop pneumocystis pneumonia, usually in patients taking combination immunosuppressant therapy that includes corticosteroids. The risk may be higher in patients with dermatomyositis or polymyositis compared with other rheumatic diseases. Most patients with rheumatic disease and pneumocystis pneumonia have an abrupt onset of acute respiratory failure and fever. This patient's 6-week course of progressive dyspnea and absence of fever make pneumocystis pneumonia an unlikely diagnosis.

Q8 (MKSAP): A 41-year-old woman is evaluated for intermittent pain and cyanosis of the fingers that is usually associated with exposure to cold temperatures or stress. She does not smoke, and her efforts to keep room temperatures warm and to wear gloves and layers of clothing to maintain her core temperature have not been successful in managing her symptoms. She was diagnosed with limited cutaneous systemic sclerosis 1 year ago. She also has gastroesophageal reflux disease. Her only medication is omeprazole.
On physical examination, temperature is 37.0°C (98.6°F), blood pressure is 128/72 mm Hg, and pulse rate is 88/min. Cutaneous examination of the hands shows sclerodactyly. Radial and ulnar pulses are 2+ and equal bilaterally.
Which of the following is the most appropriate additional treatment for this patient?

A. Amlodipine
B. Isosorbide dinitrate
C. Prednisone
D. Propranolol

Explanation: This patient has Raynaud phenomenon, which is present in more than 95% of patients with systemic sclerosis and is particularly likely to develop in patients with limited cutaneous disease. The most appropriate treatment for this patient is amlodipine. Systemic sclerosis is classified according to the degree of skin involvement. Systemic sclerosis with limited cutaneous involvement, or CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia), manifests as skin thickening distal to the elbows and knees. Conversely, systemic sclerosis with diffuse cutaneous involvement is associated with skin thickening proximal to the elbows and knees. Diffuse and limited cutaneous systemic sclerosis may affect the face. Episodes of Raynaud phenomenon are often precipitated by cold exposure or stress and usually involve the extremities. In patients with Raynaud phenomenon, cigarette smoking is contraindicated and avoidance of cold is recommended; pharmacologic therapy is warranted for patients in whom these interventions do not provide sufficient relief.
Dihydropyridine calcium channel blockers such as amlodipine have been shown to reduce the frequency and severity of attacks in patients with both primary and secondary Raynaud phenomenon, and these agents are frequently used as first-line treatment in this condition. Other agents used to manage Raynaud phenomenon include peripherally acting a-1 blockers, phosphodiesterase inhibitors, and endothelin receptor antagonists.
Topical nitrates applied to the finger webs are often used in the treatment of Raynaud phenomenon but are usually used as second-line therapy. Oral therapy with nitroglycerin is less effective and less well tolerated than amlodipine and is not indicated as a first-line drug for this condition.
Raynaud phenomenon is caused by microvascular involvement in patients with systemic sclerosis and is characterized by intimal proliferation and progressive luminal obliteration, as well as digit al spasm. This process does not respond to anti-inflammatory agents; therefore, prednisone is not indicated in the treatment of Raynaud phenomenon.
ß-Blockers such as propranolol are not indicated in the treatment of Raynaud phenomenon and may actually worsen symptoms by preventing ß-adrenergic-mediated vasodilation

Answers: Q7: C | Q8: A

Scleroderma spectrum disease

Q9 (MKSAP): A 68-year-old woman is evaluated because of recent soft tissue swelling posterior to the mandible, and dryness of both eyes, dry mouth. Her medical history is otherwise unremarkable. Physical examination reveals normal vital signs, bilateral parotid gland swelling, diffuse lymphadenopathy, and trace joint effusion in both knees. Schirmer's test shows 8mm, right eye; 5 mm, left eye (normal, greater than 15 mm/5 min), consistent with decreased tear production.
Which of the following is the most likely diagnosis?

A. Hodgkin disease
B. Sarcoidosis
C. Sjogren syndrome
D. Systemic lupus erythematosus

Explanation: This patient has Sjogren's syndrome. Sjogren's syndrome is an autoimmune disease characterized by keratoconjunctivitis sicca, xerostomia, and the presence of multiple autoantibodies. This condition may occur as a primary disease process or may be associated with another autoimmune disease. Primary Sjogren's syndrome usually is diagnosed in patients between 40 and 60 years of age, and this condition has a 9:1 female predominance. The characteristic manifestations of Sjogren's syndrome are symptomatic oral and ocular dryness. Lymphocytic inflammation of the lacrimal glands causes an aqueous tear deficiency with resultant keratoconjunctivitis sicca, whereas lymphocytic inflammation of the major and minor salivary glands is associated with salivary gland enlargement and xerostomia. A cardinal feature of Sjogren's syndrome is the presence of autoantibodies, which may include antibodies to Ro/SSA and La/SSB. These autoantibodies are not specific for Sjogren's syndrome; they may also occur in subsets of patients with systemic lupus erythematosus and in asymptomatic women. Antinuclear antibodies and rheumatoid factor also frequently are present in patients with this condition, as is hypergammaglobulinemia.
Hodgkin disease is an aggressive lymphoid malignancy that typically presents with rapidly progressive, symptomatic disease, often initially localized to one organ or compartment (for example, bone marrow)
Sarcoidosis is a multisystem, granulomatous inflammatory disease of unknown cause. It occurs most commonly in young and middle-aged adults, with a peak incidence in the third decade. The most common presenting manifestations involve the lymphatic and pulmonary systems, along with the eyes and skin
Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease of unknown cause. Manifestations of this heterogeneous syndrome range from mild to severe and life threatening and most commonly involve the skin and joints. Other manifestations include cytopenias, hemolytic anemia, serositis, aphthous ulcers, and kidney
disease.
The syndrome of dry eyes, parotid gland enlargement and arthritis is not found in systemic lupus erythematosus, lymphoma, or sarcoidosis.

Q10 (AMBOSS): A 40-year-old woman comes to the physician for a 2-month history of chest pain and heartburn after meals. The patient reports that the pain is worse at night and especially when lying down. She has a history of Raynaud disease treated with nifedipine. There is no family history of serious illness. She emigrated to the US from Nigeria 5 years ago. She does not smoke or drink alcohol. Vital signs are within normal limits. Cardiopulmonary examination shows no abnormalities. Thickening and hardening of the skin is seen on the hands and face. There are several firm, white nodules on the elbows and fingertips. Further evaluation of this patient is most likely to show which of the following findings?

A. Anti-RNA polymerase III antibodies
B. Anticentromere antibodies
C. Anti-Scl-70 antibodies
D. Anti-dsDNA antibodies

Explanation: Anticentromere antibodies are diagnostic for CREST syndrome, a form of lcSSc that can be found in about half of individuals with this condition. Other typical laboratory findings include increased γ-globulins in serum protein electrophoresis and nonspecific inflammatory markers..
Anti-Scl-70 antibodies are most commonly associated with diffuse cutaneous systemic sclerosis (dcSSc) and indicate a poor prognosis. The patient's symptoms and history are suggestive of CREST syndrome, a form of lcSSc. While anti-Scl-70 antibodies may also be seen in patients with CREST syndrome, another finding is more likely.

Answers: Q9: C | Q10: B

Scleroderma spectrum disease

Q11 (AMBOSS): A 35-year-old woman comes to the physician because of a 2-year history of progressive fatigue and joint pain. She has a 1-year history of skin problems and a 4-month history of episodic pallor of her fingers. She reports that the skin of her face, neck, and hands is always dry and itchy; there are also numerous "red spots" on her face. She has become more "clumsy" and often drops objects. She has gastroesophageal reflux disease treated with lansoprazole. She does not smoke. She occasionally drinks a beer or a glass of wine. Her temperature is 36.5°C (97.7°F), blood pressure is 154/98 mm Hg, and pulse is 75/min. Examination shows hardening and thickening of the skin of face, neck, and hands. There are small dilated blood vessels around her mouth and on her oral mucosa. Mouth opening is reduced. Active and passive range of motion of the proximal and distal interphalangeal joints is limited. Cardiopulmonary examination shows no abnormalities. Her serum creatinine concentration is 1.4 mg/dL. The patient is at increased risk for which of the following complications?

A. Swan neck deformity
B. Dactylitis
C. Antiphospholipid syndrome
D. Interstitial lung disease
E. Urolithiasis

Explanation: Patients with systemic sclerosis (SSc) are at an increased risk of lung disease with more than 50% having some form of lung involvement in the course of their lifetime. The most common forms of lung involvement include interstitial lung disease (ILD) and pulmonary hypertension, which can occur separately or in combination. ILD can be seen in both types of systemic sclerosis but is far more common in patients with diffuse systemic sclerosis. Other complications of SSc include renal disease (with scleroderma renal crisis being the most severe form of renal involvement), cardiac disease (fibrosis, myocarditis, and pericarditis), GI tract involvement (esophageal dysmotility and small bowel dysmotility), and thromboembolic events.

Q12 (AMBOSS): A 55-year-old woman with gastroesophageal reflux disease comes to the physician because of progressively worsening shortness of breath. She appears younger than her stated age. Skin examination shows small ulcerations and pitting in the fingertips. A serum anti-DNA topoisomerase antibody test is positive. Pulmonary function testing is most likely to show which of the following findings?

A. Decreased ratio of forced expiratory volume in 1 second to forced vital capacity
B. Increased residual volume
C. Increased total lung capacity
D. Decreased diffusing capacity of the lung for carbon monoxide
E. Increased forced vital capacity

Explanation: This patient with systemic sclerosis is likely to have restrictive lung disease as a result of aberrant interactions between immune cells and native fibroblasts, which leads to fibroblast proliferation and excess extracellular matrix (collagen) deposition that disrupts the surfaces between capillaries and alveoli. Restrictive lung disease is characterized by stiff lung parenchyma, decreased lung compliance, decreased total lung capacity, decreased residual volume, and impaired gas exchange, which results in a decreased diffusing capacity of the lung for carbon monoxide (DLCO)
A decreased FEV1/FVC ratio is seen in patients with obstructive lung diseases, such as asthma and chronic obstructive pulmonary disease. This patient with systemic sclerosis is likely to have restrictive lung disease, which results in stiff lung parenchyma. As a result, FEV1 and FVC are likely to be decreased, with FVC slightly more decreased than FEV1. Therefore, she is likely to have an increased FEV1/FVC ratio.

Answers: Q11: D | Q12: D

Rheumatoid arthritis

Q1 (500Best): A 45-year-old woman presents to the rheumatology outpatient clinic with a three- month history of stiff hands and wrists. She mentions that the pain is particularly bad first thing in the morning. On examination, the wrists, metacarpophalangeal joints and proximal interphalyngeal joints are swollen and warm. A diagnosis of rheumatoid arthritis is suspected. Which of the following investigations is most specific for confirming the diagnosis?

A. X-rays
B. Rheumatoid factor levels
C. Anti-citrullinated peptide antibody (anti-CCP) levels
D. C-reactive protein
E. Erythrocyte sedimentation rate

Explanation: Anti-citrullinated peptide (anti-CCP) antibodies (C) levels are the most specific investigation for rheumatoid arthritis. **X-rays (A) early in the disease course will demonstrate soft tissue swelling but are unlikely to show much else. However, x-rays of the hands, feet and any other affected joint should be performed early in the disease to establish a baseline.** Rheumatoid factor (B) is positive in approximately 70 per cent of patients with rheumatoid arthritis. However, it is not specific to rheumatoid arthritis and may be raised in a number of other conditions. Rheumatoid factor should be tested for in patients with suspected rheumatoid arthritis. Anti-CCP antibodies can be subsequently sent if rheumatoid factor is negative or to inform decision making about starting therapy. It is important to note that anti-CCP antibodies are not presently a routine test for all patients with suspected rheumatoid arthritis. C-reactive protein (D) is raised in many infective and inflammatory conditions. It is therefore not the most specific test for rheumatoid arthritis. However, it is raised in rheumatoid arthritis and can be used to monitor the disease and guide treatment decisions. Erythrocyte sedimentation rate (E) is raised in rheumatoid arthritis and a range of other conditions. It is, therefore, not the correct answer.

Q2 (500Best): A 40-year-old woman presents to the rheumatology outpatient clinic with a three- month history of stiff hands and wrists. She mentions that the pain is particularly bad first thing in the morning. On examination, the wrists, metacarpophalangeal joints and proximal interphalangeal joints are swollen and warm. A diagnosis of rheumatoid arthritis is suspected. Blood tests for rheumatoid factor return as positive. What is the most appropriate management?

A. Non-steroidal anti-inflammatory drugs (NSAIDs)
B. Intramuscular depot injection of methylprednisolone plus NSAIDs
C. Anti-TNF therapy
D. Intramuscular depot injection of methylprednisolone plus NSAIDs and methotrexate and sulfasalazine
E. Physiotherapy

Explanation: Most patients will be offered NSAIDs (A) first by their GP, or over the counter, but if these fail, referral to hospital rheumatology is required. If NSAIDs do not resolve symptoms, therapy would be escalated to include methotrexate and one other disease-modifying anti-rheumatic drug (DMARD), such as sulfasalazine plus a steroid (D). DMARDs should be offered early within the treatment of the disease to limit joint destruction. Short-term steroid therapy is useful for controlling disease flares and can be given by oral, intramuscular and intra-articular routes. Intramuscular depot injection of methylprednisolone plus NSAIDs (B) will be very useful in managing symptoms of the flare, but will not alter the disease course. Anti-TNF therapy (C) is reserved for patients who have active disease despite DMARDs (usually two trials of DMARDs should be tried before considering anti-TNF). Physiotherapy (E) is very useful and should be encouraged, but alone cannot control symptoms or disease course, making this answer incorrect.

Q3 (500Best): A 50-year-old woman, who has received a recent diagnosis of rheumatoid arthritis, presents to her GP with ongoing pain and stiffness in her hands and feet. Which joints are usually spared at onset of rheumatoid arthritis?

A. Proximal interphalangeal joints
B. Distal interphalangeal joints
C. Metacarpophalangeal joints
D. Wrists
E. Metatarsophalangeal joints

Explanation: The hand changes associated with rheumatoid arthritis are frequently tested in clinical and written exams. The most common presenting features are pain and stiffness of the small joints of the hands and feet, which is worse in the mornings. The most common joints at onset of disease are proximal interphalangeal joints (A), metacarpophalangeal joints (C), wrists (D) and metatarsophalangeal joints (E). The distal interphalangeal joints (B) are usually spared at onset, making this the correct answer here. It is important to note, however, that this is a variable disease and some patients may present with other joint involvement including elbows, shoulders, knees or ankles. As the disease progresses and joint damage occurs in the hands, a variety of deformities may be seen. These include ulnar deviation and palmar subluxation of the metacarpophalangeal joints, Boutonniere deformity (flexion of PIPs, hyperextension of DIPs), swan-neck deformity (hyperextension of PIP, flexion of DIPs) or dorsal subluxation of the ulnar styloid. Inflammation of the flexor tendon sheath may result in carpal tunnel syndrome and inflammation of the extensor tendon sheath can cause tendon rupture. Therefore, it is worth looking out for scars of carpal tunnel decompression and tendon repair during clinical examination.

Answers: Q1: C | Q2: A | Q3: B

Rheumatoid arthritis

Q4 (500Best): A 55-year-old woman presents to her GP with shortness of breath and dry cough. The symptoms began a few months ago and have progressed. She has a past medical history of rheumatoid arthritis, diagnosed ten years earlier. On respiratory examination, there are bibasal fine inspiratory crackles on auscultation. What is the most likely cause of her symptoms?

A. Pulmonary oedema
B. Consolidation
C. Pleural effusions
D. Pulmonary fibrosis
E. Intrapulmonary nodules

Explanation: The collection of shortness of breath, dry cough and bibasal fine inspiratory crackles should point to pulmonary fibrosis (D) as the answer in this question. In patients with rheumatoid arthritis, this may occur as a result of extra-articular manifestation of the disease. Pulmonary fibrosis is also a side effect of methotrexate, a disease-modifiying anti-rheumatic drug, which is commonly used in rheumatoid arthritis. Pulmonary oedema (A) will cause shortness of breath and be heard as fine inspiratory crackles during respiratory examination. However, it would characteristically produce a productive cough of frothy sputum. In addition, pulmonary oedema is **not an extra-articular manifestation** of rheumatoid arthritis or a side effect of the drugs used to treat it, making this answer incorrect. Consolidation (B) occurs as a result of an infective process and can present in a number of ways including shortness of breath, productive cough or pleurisy. On respiratory examination, there is likely to be bronchial breathing and end expiratory crackles. Pleural effusions (C) are an extra- articular manifestation in rheumatoid arthritis. Clinical features would include shortness of breath and, on examination, t**here would be reduced air entry, stony dullness to percussion and reduced vocal resonance**. Therefore, this is the incorrect answer. Intrapulmonary nodules (E) are also an extra-articular manifestation of rheumatoid arthritis, but do not cause the clinical features outlined in this question, making this answer wrong.

Q5 (Pretest):A 40-year-old woman complains of 7 weeks of pain and swelling in both wrists and knees. She has several months of fatigue. After a period of rest, resistance to movement is more striking. On examination, the meta-carpophalangeal joints and wrists are warm and tender. There are no other joint abnormalities. There is no alopecia, photosensitivity, kidney disease, or rash. Which of the following is correct?

A. The clinical picture suggests early rheumatoid arthritis, and a rheumatoid factor and anti-CCP (anti- cyclic citrullinated peptide) should be obtained.
B. The prodrome of lethargy suggests chronic fatigue syndrome.
C. Lack of systemic symptoms suggests osteoarthritis.
D. X-rays of the hand are likely to show joint space narrowing and erosion. e. An aggressive search for occult malignancy is indicated.

Explanation: The clinical picture of symmetrical swelling and tenderness of the metacarpophalangeal (MCP) and wrist joints lasting longer than 6 weeks strongly suggests rheumatoid arthritis (RA). Rheumatoid factor, an immunoglobulin directed against the Fc portion of IgG, is positive in about two-thirds of cases and may be present early in the disease. The history of lethargy or fatigue is a common prodrome of RA. The inflammatory joint changes on examination are not consistent with chronic fatigue syndrome; furthermore, patients with CFS typically report fatigue existing for many years. The MCP-wrist distribution of joint symptoms makes osteoarthritis very unlikely. The x-ray changes described are characteristic of RA, but would occur later in the course of the disease. Although arthritis can occasionally be a manifestation of hematologic malignancies and, rarely, other malignancies, the only indicated screening would be a complete history and physical examination along with a CBC.

Q6 (Pretest):A 48-year-old woman complains of joint pain and morning stiffness for 4 months. Examination reveals swelling of the wrists and MCPs as well as tenderness and joint effusion in both knees. The rheumatoid factor is positive, antibodies to cyclic citrullinated protein are present, and subcutaneous nodules are noted on the extensor surfaces of the forearm. Which of the following statements is correct?

A. Prednisone 60 mg per day should be started.
B. The patient should be evaluated for disease-modifying antirheumatic therapy.
C. A nonsteroidal anti-inflammatory drug should be added to aspirin.
D. The patient's prognosis is highly favorable.
E. The patient should receive a 3-month trial of full-dose nonsteroidal anti-inflammatory agent before determining whether and/or what additional therapy is indicated.

Explanation: b The patient has more than four of the required signs or symptoms of RA, including morning stiffness, swelling of the wrist or MCP, simultaneous swelling of joints on both sides of body, subcutaneous nodules, and positive rheumatoid factor. Subcutaneous nodules and anti-CCP antibodies are poor prognostic signs for the activity of the disease, and disease-modifying antirheumatic drugs (DMARDs) such as methotrexate, antimalarials, sulfasalazine, leflunomide, anti-TNF agents, or a combination of these drugs should be instituted. Methotrexate has emerged as a cornerstone of most disease-modifying regimens, to which other agents are often added. Low-dose corticosteroids (eg, prednisone 7.5 mg a day or less) have recently been shown to reduce the progression of bony erosions and, although controversial, are useful additions to DMARD therapy. High-dose steroids, however, should be avoided. Use of anti-inflammatory doses of both aspirin and nonsteroidals together is not desirable because it will increase the risk of side effects. Given the aggressive nature of this woman's rheumatoid arthritis and negative prognostic signs, use of DMARDs is indicated. Significant joint damage has been shown by MRI to occur quite early in the course of disease.

Answers: Q4: D | Q5: A | Q6: B

Rheumatoid arthritis

Q7 (500Best): A 30-year-old woman presents to accident and emergency with worsening stiffness in the hands, wrists and feet. She mentions that the pain has been particularly bad in the mornings. On examination, there is a palpable spleen. Initial blood tests reveal a low neutrophil count and a raised C-reactive protein. The most likely diagnosis is:

A. Felty's syndrome
B. Reactive arthritis
C. Still's disease
D. Infectious mononucleosis
E. Serum sickness

Explanation: **Felty's syndrome (A) is splenomegaly and neutropenia in a patient with rheumatoid arthritis**. Reactive arthritis (B) is an asymmetrical lower limb arthritis occurring 1–4 weeks following an infection. A range of other clinical features may also be present (see question 8). Still's disease (C) is systemic juvenile idiopathic arthritis and is characterized by swinging pyrexia, rash and arthritis. Juvenile idiopathic arthritis is the most common form of persistent arthritis in those under 16 years of age. There is a variable pattern of arthritis including oligoarthritis, polyarthritis and systemic arthritis. Infection mononucleosis (D) may cause splenomegaly. However, arthritis and neutropenia are not features of infectious mononucleosis, making this answer incorrect. Serum sickness (E) is caused by a hypersensitivity reaction to antibodies derived from animals. The clinical features include fever, rashes, arthralgia, malaise, splenomegaly and lymphadenopathy. Arthritis affecting and hands and feet and neutropenia are not common features, making this option incorrect.

Q8 (MKSAP): A 45-year-old woman is evaluated in the office for a 3-month history of pain, stiffness, and swelling of the small joints of the hands and feet . She also has increasing fatigue that has caused her to miss work at least 1 day per week. She has no other medical problems.
On physical examination, the vital signs and general physical examination, including skin examination, are normal. A photograph of one of her hands is shown (P lat e 29).
Complete blood count , serum chemistries, and urinalysis are all normal. Erythrocyte sedimentation rate is 44 mm/h.
Which of the following is the most likely diagnosis ?

A- Osteoarthritis
B- Psoriatic arthritis
C- Rheumatoid arthritis
D- Systemic lupus erythematosus

Explanation: This patient has symptoms and signs consistent with rheumatoid arthritis. Different joints are variably affected by different disorders. Rheumatoid arthritis and osteoarthritis can both involve the proximal interphalangeal joint s of the hands, but metacarpophalangeal joint involvement occurs in rheumatoid arthritis but not typically in osteoarthritis is. Distal interphalangeal joint involvement is charact erist ic of osteoarthritis but not rheumatoid arthritis. Unless a secondary condition, such as trauma, metabolic disorder, or inflammatory arthritis, has already affected the joint , osteoarthritis is does not occur in the metacarpophalangeal, wrist , elbow, shoulder, and ankle joint s. This patient has erythema and swelling of the metacarpophalangeal joint s and loss of function leading to absenteeism from work; these findings are most consistent with rheumatoid arthritis.
Psoriasis is associated with an underlying inflammatory arthritis in up t o 30% of patients with skin disease; nail pitting suggest s psoriatic arthritis, even in the absence of psoriatic skin lesions. These changes are not present in this patient .
More than 90% of patients with SLE develop joint involvement that can manifest as arthralgia or true arthritis. Joint pain is often migratory and can be oligo articular or polyarticular and asymmetric or symmetric. Pain typically involves the large and small joints; the wrist s and metacarpophalangeal and proximal interphalangeal joints in particular are most commonly affected. The absence of other manifest at ions of SLE (serositis, cytopenias, kidney disease, rash, photosensitivity) make this diagnosis unlikely.

Q9 (MKSAP): A 62-year-old woman is evaluated for a 3-day history of fever and left knee pain and swelling. She has a 30-year history of rheumatoid arthritis treated with methotrexate and non steroidal anti-inflammatory drugs. She has no other medical problems.
On physical examination, temperature is 37.8°C (100.0°F), blood pressure is 140/78 mm Hg, pulse rate is 86/min, and respiration rate is 14/min. T he left knee is swollen, red, warm, and tender to palpation. Range of motion is limited because of pain.
Arthrocentesis is performed, and the following results are reported: synovial fluid leukocyte count 75,000/µL (75 × 109/L) wit h 69% neutrophils. Gram stain is positive for gram-positive cocci.
Which would be the most appropriate initial antibiotic therapy pending culture results ?

Answers: Q7: A | Q8: C

A- Cefazolin
B- Ceftriaxone
C- Nafcillin
D- Vancomycin

Explanation: The most appropriate initial antibiotic selection for this patient is vancomycin. Septic arthritis is a medical emergency. Hematogenous spread is the most common mechanism of joint infection, because the synovium has no basement membrane and is, therefore, particularly vulnerable t o infection. Staphylococcus is the most common gram-positive organism affecting nat ive and prosthetic joints, and infection with the methicillin-resistant strain is becoming increasingly common. Joints that have been previously damaged are more likely t o become infected than structurally normal joint s. In particular, patient s with rheumatoid arthritis have usually used intra-articular corticosteroids or immunosuppressive agent s at some point in their disease course and are, therefore, particularly susceptible t o infection. Septic arthritis also is more likely t o have a polyarticular present at ion in patients with pre-existing rheumatoid arthritis than in other patients. In general, vancomycin is the empiric therapy of choice for community-acquired septic arthritis and synovial fluid positive for gram-positive cocci or in patient s at low risk for gram-negative infection and with a negative synovial fluid Gram stain. Because this patient 's synovial fluid is positive for gram-positive cocci and because of increasing concern about methicillin-resistant Staphylococcus aureus (MRSA) infection in the community, vancomycin is the initial treatment of choice pending culture result s. Modifications of the initial antibiotic regimen can be made t o narrow coverage with cefazolin or nafcillin when culture results are available but are not generally recommended as initial empiric therapy because of the prevalence of MRSA in the community.
Ceft riaxone is t he init ial ant ibiot ic of choice in pat ient s at risk for gonococcal infect ion. Gonococcal art hrit is is t he most common form of bact erial art hrit is in young, sexually act ive persons and should be considered in pat ient s who present wit h migrat ory t enosynovit is and art hralgia.

Q10 (MKSAP): A 32-year-old woman is evaluated in the emergency department for a 4-day history of pain and swelling of the right wrist and low-grade fever. She has a 7-year history of severe rheumatoid arthritis. She does not recall any specific trauma involving the wrist but has recently been very physically active. Medications are methotrexate, a folic acid supplement , etanercept , prednisone, and ibuprofen.
On physical examination, temperature is 37.8°C (100.0°F), blood pressure is 118/68 mmHg, pulse rate is 90/min, and respiration rate is 18/min. BMI is 22. Cardiopulmonary examination is normal. There is no rash. The right wrist is swollen and tender and has a decreased range of motion. There are a subcutaneous nodule and small flexion deformity on the left elbow but no active synovitis. Mild synovitis is present on the second metacarpophalangeal joint s bilaterally. T he hips, knees, and feet are not tender or swollen and have full range of motion.
Which of the following diagnostic studies of the wrist will be most helpful in establishing this patient's diagnosis ?

A- Arthrocentesis
B- Arthroscopy
C- Bone scan
D- MRI
E- Radiography

Explanation: This patient most likely has septic arthritis, which usually manifests as acute monoarthritis is and is characterized by pain on passive range of motion in the absence of known trauma. Arthrocentesis of the wrist will most likely help t o establish a diagnosis in this patient .
Septic arthritis should particularly be suspected in patients with underlying hematologic disorders such as rheumatoid arthritis who presented with a sudden single joint flare that is not accompanied by other features of the pre-existing disorder. However, all patients who present with acute monoarthritis is should be presumed t o have septic arthritis unt il synovial fluid analysis via arthrocentesis excludes this condition. Synovial fluid analysis is the only definitive way t o diagnose septic arthritis and is critical to guide antibiotic treatment . Patients with suspicion for this condition should begin empiric systemic antibiotic therapy until culture results are available. Surgical drainage or debridement via arthroscopy may be warranted in patients with septic arthritis who do not respond to repeated percutaneous drainage and appropriate antibiotic therapy but would not be an appropriate initial intervention.
Joint and bone damage due t o infection are relatively lat e radiographic findings. In acute septic arthritis, nonspecific soft -t issue fullness and joint effusions are often the only initial radiographic findings and do not establish the diagnosis of infection. Bone scans are more sensitive in detecting inflammatory lesions in bones and joint s but also are not specific for infection.
MRI of the affected joint is especially useful in detecting avascular necrosis, soft -t issue masses, and collections of fluid not visualized by other imaging modalities but would not establish the diagnosis of infection.

Rheumatoid arthritis

Q11 (MKSAP): A 19-year-old woman with a 4-year history of rheumatoid arthritis is evaluated because of a 3-month history of worsening symptoms. She had previously good disease control with methotrexate. She is otherwise well and denies fevers, night sweats and weight loss. On physical examination, vital signs are normal. There is active synovitis involving the left first and second metacarpophalangeal joints and right wrist. The remainder of the physical examination is normal Hand x-rays reveal a new erosion on the right ulnar styloid and the left second MCP joint. Initiation of the tumor necrosis factor (TNF)-a inhibitor, adalimumab, is recommended.
Which of the following tests should be performed prior to initiating adalimumab therapy?

A- Brain MRI
B- Chest CT
C- Thyroid stimulating hormone measurement
D- Tuberculin skin test

Explanation: A tuberculin skin test should be performed prior to initiating therapy with adalimumab. When adequate disease control is not achieved with oral disease modifying antirheumatic drugs (DMARDs) such as methotrexate, biologic therapy should be initiated. The initial biologic therapy should be a TNF-a inhibitor. This agent generally should be added to the baseline methotrexate therapy, because the rate of radiographic progression has been shown to decrease with combination therapy. Rarely, serious infections have occurred in patients treated with these agents; among these infectious complications, reactivation tuberculosis is the most common. Tuberculin skin testing is indicated before beginning treatment with these agents, and positive results on this test warrant treatment for latent tuberculosis. Furthermore, periodic tuberculin skin testing for tuberculosis is now recommended during treatment with a TNF-a inhibitor. A chest CT scan is not necessary prior to initiating therapy with a TNF-a inhibitor. If the patient has a positive tuberculin skin test (>5 mm induration), a chest x-ray will be required to exclude active pulmonary tuberculosis but neither this test nor a chest CT scan is indicated in an asymptomatic person with a negative tuberculin skin test.
Rare cases of multiple sclerosis or demyelinating conditions such as optic neuritis have been reported as a potential complication of TNF-a inhibitor therapy but usually remit upon discontinuation of therapy. There is no value in screening asymptomatic patients for multiple sclerosis with a brain MRI. Numerous other conditions, such as migraine, cerebrovascular disease, hypertension, smoking, diabetes mellitus, hyperlipidemia, and head trauma, are also associated with white matter abnormalities on brain MRI. Misinterpretation of white matter abnormalities discovered incidentally in a patient with nonspecific symptoms is a leading cause of multiple sclerosis misdiagnosis Many drugs can interfere with thyroid hormone production, release, transport and activity; however drugs to treat rheumatoid arthritis do not usually fall into this category. Corticosteroids, for example, can interfere with TSH release and decrease thyroid binding globulin, but TNF-a inhibitors do not interfere with thyroid function and there is no need to obtain a TSH measurement in this patient prior to initiating therapy.

Q12 (MKSAP): A 26-year-old woman is evaluated for a 2-month history of pain and swelling in the hands and daily morning stiffness that lasts for 3 to 4 hours. She is 4 months postpartum and her pregnancy was without complications. She has no history of rash and is otherwise well. Her only medication is ibuprofen, which has not sufficiently relieved her symptoms. On physical examination, temperature is normal, blood pressure is 110/68 mm Hg, pulse rate is 82/min, and respiration rate is 16/min. The second and third proximal interphalangeal and metacarpophalangeal joints and the wrists are tender and swollen bilaterally. Laboratory studies show an erythrocyte sedimentation rate of 67 mm/h, and titers of IgM antibodies against parvovirus B19 are negative.
Which of the following is the most likely diagnosis?

A- Gout
B- Osteoarthritis
C- Parvovirus B19 infection
D- Rheumatoid arthritis

Explanation: This patient most likely has rheumatoid arthritis, which is the most common cause of chronic, inflammatory polyarthritis in premenopausal women. Rheumatoid arthritis commonly affects the metacarpophalangeal, proximal interphalangeal, and wrist joints. This patient's swelling, prolonged morning stiffness, and elevated erythrocyte sedimentation rate are consistent with this diagnosis. Furthermore, women are three times more likely to develop rheumatoid arthritis than men and have a slightly increased risk of developing this condition during the first 3 months postpartum.
Gout may involve the hand and wrist and is associated with inflammatory features. However, gout usually has an asymmetric presentation and is unlikely to develop in a premenopausal woman.
Osteoarthritis may manifest as chronic arthritis involving the proximal interphalangeal joints but would not affect the metacarpophalangeal joints or the wrists. Secondary osteoarthritis related to trauma or a metabolic condition such as hemochromatosis may explain this patient's pattern of joint involvement, but this condition would be unlikely in a 26-year-old woman. Osteoarthritis also would not have an inflammatory presentation.
Viral arthritis usually is self-limited except when associated with hepatitis B and C virus infection. Parvovirus B19 infection in adults may induce an acute rheumatoid factor- positive oligo- or polyarthritis. Most adult patients with parvovirus B19 infection also develop rash, but only rarely in adults does rash manifest as the classic rash seen in childhood erythema infectiosum, the "slapped cheek" rash. Diagnosis of acute parvovirus B19 infection may be established by detecting circulating IgM antibodies against parvovirus B19
Viral arthritis usually resolves within 3 weeks, although a minority of patients may develop persistent arthritis. The arthritis associated with acute parvovirus B19 infection does not cause joint destruction, and supportive analgesic therapy with NSAIDs is appropriate as tolerated. Parvovirus B1 9 infection is unlikely in this patient considering the duration of her symptoms, absence of rash, and negative titers of IgM antibodies against parvovirus B19

Answers: Q11: D | Q12: D

Spondyloarthritis

Q1 (500Best): A 30-year-old man presents to his GP with a 1-week history of painful, swollen knees and a painful right heel. Further history reveals that he has been experiencing burning pains while urinating for the past 2 weeks and that his eyes have become red and itchy. What is the most likely diagnosis?

A. Septic arthritis
B. Gout
C. Ankylosing spondylitis
D. Enteropathic arthritis
E. Reactive arthritis

Explanation: Reactive arthritis (E) is a sterile arthritis, which follows an attack of dysentery (caused by Campylobacter, Salmonella, Shigella or Yersinia spp.) or urethritis (caused by Chlamydia or Ureaplasma spp.). **Clinical features of reactive arthritis are an acute, asymmetrical lower limb arthritis occurring 1–4 weeks following an infection.** Other features of reactive arthritis include conjunctivitis (as described by this case), enthesitis (which may result in plantar fasciitis or Archilles tendonitis), circinate balanitis (painless superficial ulceration of glans penis), keratoderma blenorrhagica (painless, red plaques on the soles and palmes), nail dystrophy, mouth ulcers and, rarely, aortic incompetence. The triad of urethritis, arthritis and conjunctivitis is known as Reiter's disease. Treatment of reactive arthritis is with NSAIDs and local steroid injection for symptomatic control. Any underlying infection should be treated but is unlikely to influence the course of the arthritis. Individuals who develop recurrent attacks of arthritis can be considered for therapy with sulfasalazine or methotrexate. Septic arthritis (A) is a monoarthritis, presenting as a hot, swollen, tender joint. Therefore this is the incorrect answer. Gout (B) also usually presents as an acute monoarthritis and does not cause conjunctivitis or urethritis, making this answer incorrect. Ankylosing spondylosis (C) does not affect the knees or cause urethritis, although it can cause conjunctivitis. Enteropathic arthritis (D) is an asymmetrical lower limb arthritis, associated with inflammatory bowel disease. The absence of bowel symptoms in this patient means this is the incorrect answer.

Q2 (500Best): A 23-year-old man presents to the rheumatology clinic with lower back and hip pain. These have been occurring every day for the past two months. Pain and stiffness are worse in the mornings. He also mentions that his right heel has been hurting. He is previously fit and well, but had occasions of lower back pain when he was a teenager. His symptoms have stopped him from playing tennis. Recent blood tests organized by his GP have shown a raised C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR). What is the most appropriate treatment?

A. NSAID and spinal exercises
B. NSAID and bed rest
C. Oral prednisolone
D. Methotrexate plus sulfasalazine
E. Bed rest

Explanation: This is a difficult question as it requires knowledge of the diagnosis and then knowledge of the treatment. The case presented is of a patient with ankylosing spondylosis. The diagnosis is clinical, with involvement of the sacroiliac joint as the earliest manifestation. The disease course is variable and may progress to a marked kyphosis of the spine. Other features include enthesitis (such as the Archilles tendon enthesitis in this case), costochondritis, peripheral joint involvement (usually asymmetrical and involving the large joints), aortic regurgitation, apical pulmonary fibrosis and amyloidosis. The ESR and CRP are usually raised. Initial x-rays may be unremarkable. However, later in the disease, syndesmophytes (bony spurs due to inflammatory enthesitis) may be seen between vertebrae resulting in the characteristic bamboo spine appearance. Ankylosing spondylosis is managed with exercises, not bed rest. NSAIDs are given, unless there are contraindications, for the management of pain. Therefore, NSAID and spinal exercises (A) is the correct answer and NSAID and bed rest (B) and bed rest (E) are incorrect. Local steroid injections may be used for pain relief, particularly for peripheral arthritis and enthesitis. However, oral prednisolone (C) is not normally used. Methotrexate and sulfasalazine (D) may be given to patients with peripheral arthritis, but do not help the back pain, making this answer incorrect.

Q3 (Pretest): A 22-year-old man develops the insidious onset of low back pain improved with exercise and worsened by rest. There is no history of diarrhea, conjunctivitis, urethritis, rash, or nail changes. On examination, the patient has loss of mobility with respect to lumbar flexion and extension. He has a kyphotic posture. A plain film of the spine shows sclerosis of the sacroiliac joints. Calcification is noted in the anterior spinal ligament. Which of the following best characterizes this patient's disease process?

A. He is most likely to have acute lumbosacral back strain and requires bed rest.
B. The patient has a spondyloarthropathy, most likely ankylosing spondylitis.
C. The patient is likely to die from pulmonary fibrosis and extrathoracic restrictive lung disease.
D. Rheumatoid factor is likely to be positive.
E. A colonoscopy is likely to show Crohn disease.

Explanation: Insidious back pain occurring in a young male and improving with exercise suggests one of the spondyloarthropathies—ankylosing spondylitis, reactive arthritis (including Reiter syndrome), psoriatic arthritis, or enteropathic arthritis. In the absence of symptoms or findings to suggest one of the other conditions and in the presence of symmetrical sacroiliitis on x-ray, ankylosing spondylitis is the most likely diagnosis. Acute lumbosacral strain would not be relieved by exercise or worsened by rest. The prognosis in ankylosing spondylitis is generally good, with only 6% dying of the disease itself. While pulmonary fibrosis and restrictive lung disease can occur, they are rarely a cause of death (cervical fracture, heart block, and amyloidosis are leading causes of death as a result of ankylosing spondylitis). Rheumatoid factor is negative in all the spondyloarthropathies. Crohn disease can cause an enteropathic arthritis, which may precede the gastrointestinal manifestations, but this diagnosis is far less likely in this case than ankylosing spondylitis.

Answers: Q1: E | Q2: A | Q3: B

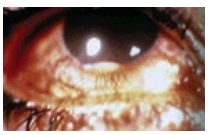
Spondyloarthritis

Q4 (500Best): A 20-year-old man presents to accident and emergency with sudden onset pain in the right eye, with associated blurred vision and discomfort when gazing at the lights. He has a history of back pain and has recently been diagnosed with ankylosing spondylosis. What is the most likely cause of his eye pain?

A. Conjunctivitis
B. Retinal detachment
C. Anterior uveitis
D. Corneal ulceration
E. Acute glaucoma

Explanation: Anterior uveitis (C) is associated with ankylosing spondylosis and may occur in up to one-third of patients. The symptoms described in this patient with ankylosing spondylosis means anterior uveitis is the correct answer. Conjunctivitis (A) is associated with ankylosing spondylosis but would present with red, itchy eyes without blurred vision or photophobia, making this answer incorrect. Retinal detachment (B) is not associated with ankylosing spondylosis and usually presents with flashes of light in the affected eye and increase in the number of floaters followed by visual loss. Therefore, this answer is incorrect. Corneal ulceration (D) is not associated with ankylosing spondylosis and presents with redness and eye pain without visual loss, making it the wrong answer. Acute glaucoma (E) may present with eye pain but is not associated with ankylosing spondylosis, making this answer incorrect. Patients would present with a red eye, sudden onset eye pain, decreased visual acuity and nausea or vomiting.

Q5 (MKSAP): A 23-year-old man is evaluated in the emergency department for a 5-day history of headache, blurred vision, and right eye pain. His eye pain increases when he attempts to read or when exposed to light. He also has a 3-year history of back stiffness that is worse in the morning and tends to improve as he becomes more active. He does not have arthralgia, arthritis, or rash. He takes no medications and is monogamous. On physical examination, temperature is 36.8°C (98.2°F), blood pressure is 130/76 mm Hg, pulse rate is 85/min, and respiration rate is 14/min. There are no skin lesions. The appearance of the right eye is shown. Photophobia is present during the penlight examination of the pupil. Both pupils react to light. An emergency referral is made to an ophthalmologist.
Following resolution of the eye problem, this patient should be evaluated for which of the following systemic diseases?



A- Ankylosing spondylitis
B- Sarcoidosis
C- Sjogren syndrome
D- Systemic lupus erythematosus

Explanation: The patient has anterior uveitis with a hypopyon, and the associated systemic disease is most likely ankylosing spondylitis. The classic triad for acute anterior uveitis is pain, sensitivity to light, and blurred vision; headache, tenderness, and tearing may also occur. Photophobia during penlight examination has a positive predictive value of 60% for severe eye disease and a negative predictive value of 90%.
Prospective studies have documented systemic illness in 53% of patients with anterior uveitis. Patients with uveitis associated with systemic disease usually have a history or physical examination findings that suggest an underlying disorder. The most commonly diagnosed systemic illnesses in this setting are reactive arthritis, ankylosing spondylitis, and sarcoidosis.
Acute anterior uveitis, particularly unilateral presentations that fluctuate between both eyes over time, is strongly associated with the HLA-B27-related arthropathies, including ankylosing spondylitis. In addition, this patient's chronic back stiffness is highly suggestive of ankylosing spondylitis. Furthermore, in up to 65% of patients with uveitis, spondyloarthropathy remains undiagnosed until these patients present with uveitis.
Posterior uveitis may be related to sarcoidosis or vasculitis but is not typically associated with pain or redness of the eye. Patients with posterior uveitis also often have decreased visual acuity and floaters, which is not consistent with this patient's presentation. Furthermore, sarcoidosis is an unlikely cause of this patient's chronic low back pain.
Sicca syndrome manifests as dryness of the mouth, eves, and vagina and variable enlargement of the parotid glands in association with concomitant redness and gritty irritation of the eyes. This condition is suggestive of primary or secondary Sjogren's syndrome. However, Sjogren syndrome would not cause anterior uveitis and also would not explain the presence of chronic low back pain in a young man.
Anterior uveitis is associated with psoriasis and, in rare cases, Whipple disease, systemic lupus erythematosus, and the systemic vasculitides. However, the patient's long history of back pain in the absence of cutaneous and other manifestations of systemic lupus erythematosus makes this diagnosis unlikely.:

Answers: Q4: C | Q5: A

Spondyloarthritis

Q6 (MKSAP): A 26-year-old female electrical engineer is evaluated for a 2-year history of persistent pain and stiffness involving the low back. These symptoms are worse in the morning and are alleviated with exercise and hot showers. There are no radicular symptoms. Her only medication is ibuprofen, which has helped to relieve her symptoms. She has no other medical problems and takes no additional medications. On physical examination, vital signs are normal. Cutaneous examination is normal. Palpation of the pelvis and low back elicits pain. There is loss of normal lumbar lordosis, and forward flexion of the lumbar spine is decreased. Reflexes and strength are intact. Radiographs of the lumbar spine and pelvis are normal. Which of the following studies is most likely to establish the diagnosis in this patient?

A- Anti-cyclic citrullinated peptide antibodies
B- Erythrocyte sedimentation rate
C- HLA-B27
D- MRI of the sacroiliac joints

Explanation: This patient most likely has ankylosing spondylitis, and MRI of the sacroiliac joints is most likely to establish a diagnosis. Radiographic evidence of sacroiliitis is required for definitive diagnosis and is the most consistent finding associated with this condition. Onset of ankylosing spondylitis usually occurs in the teenage years or 20s and manifests as persistent pain and morning stiffness involving the low back that are alleviated with activity. This condition also may be associated with tenderness of the pelvis. Typically, the earliest radiographic changes in affected patients involve the sacroiliac joints, but these changes may not be visible for several years; therefore, this patient's normal radiographs of the pelvis do not exclude sacroiliitis. MRI, especially with gadolinium enhancement, is considered a sensitive method for detecting early erosive inflammatory changes in the sacroiliac joints and spine and can assess sites of active disease and response to effective therapy.
Anti-cyclic citrullinated peptide antibodies are highly specific for rheumatoid arthritis. However, rheumatoid arthritis does not involve the sacroiliac joints or lumbar spine, and testing for this condition in this patient is therefore not indicated.
An elevated erythrocyte sedimentation rate would raise suspicion for an inflammatory process but would not help to establish a specific diagnosis. In addition, the erythrocyte sedimentation rate does not correlate with disease activity in patients with ankylosing spondylitis, and measurement of this value is therefore not useful in diagnosing or monitoring patients with this condition.
HLA-B27 positivity is a strong risk factor for ankylosing spondylitis. However, less than 5% of patients who have this allele develop this condition. In addition, not all patients who have ankylosing spondylitis have this allele. Therefore, it is neither 100% sensitive nor 100% specific for the diagnosis of ankylosing spondylitis.

Q7 (AMBOSS): A 34-year-old man comes to the physician because of increasing lower back and neck pain for the past 7 months. The pain is worse in the morning and improves when he plays basketball. He has noticed shortness of breath while playing for the past 2 months. He is sexually active with two female partners and uses condoms inconsistently. He appears lethargic. His vital signs are within normal limits. Examination of the back shows tenderness over the sacroiliac joints. Range of motion is limited by pain. The lungs are clear to auscultation. Chest expansion is decreased on full inspiration. His leukocyte count is 14,000/mm3 and erythrocyte sedimentation rate is 84 mm/h. An x-ray of the spine shows erosion and sclerosis of the sacroiliac joints and loss of spinal lordosis. Further evaluation of this patient is most likely to show which of the following?

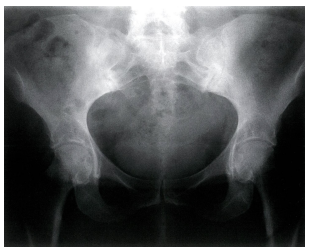
A- Paresthesia over the anterolateral part of the thigh
B- Hyperextension of the proximal interphalangeal joints with flexion of the distal interphalangeal joints
C- Nail pitting and separation of the nail from the nailbed
D- Tenderness at the Achilles tendon insertion site
E. Erythema and inflammation of the conjunctiva

Explanation: Tenderness at the Achilles tendon insertion site indicates heel enthesitis, which is a common finding in individuals with AS. About half of the patients with AS will have enthesitis at some point in the course of the disease. Enthesitis is generally a clinical diagnosis, but ultrasonography can also be used for the detection and assessment of the condition. Individuals with AS commonly develop restrictive pulmonary disease as a result of the decreased mobility of their spine and thorax. This is likely the cause of this patient's shortness of breath and decreased chest expansion on inspiration. Erythema and inflammation of the conjunctiva (conjunctivitis) are seen in reactive arthritis.

Answers: Q6: D | Q7: D

Spondyloarthritis

Q8 (AMBOSS): A 32-year-old woman comes to her physician because of increasing back pain for the past 10 months. The pain is worse in the morning when she wakes up and improves with activity. She used to practice yoga, but stopped 5 months ago as bending forward became increasingly difficult. She has also had bilateral hip pain for the past 4 months. She has not had any change in urination. She has celiac disease and eats a gluten-free diet. Her temperature is 37.1°C (98.8°F), pulse is 65/min, respirations are 13/min, and blood pressure is 116/72 mmHg. Examination shows the range of spinal flexion is limited. Flexion, abduction, and external rotation of bilateral hips produces pain. An x-ray of her pelvis is shown. Further evaluation of this patient is likely to show which of the following?



A- High levels of rheumatoid factor
B- HLA-B27 positive genotype
C- Presence of anti-dsDNA antibodies
D- HLA-DR3 positive genotype
E. High levels of creatine phosphokinase

Explanation: More than 90% of patients with AS are HLA-B27 positive. HLA-B27 testing is indicated to diagnose AS in patients with inconclusive findings on physical examination and/or pelvic x-ray. Although most patients with AS are HLA-B27 positive, less than 5% of people who are HLA-B27 positive develop the disease. Isolated HLA-B27 positivity has little clinical significance. Gene positivity only supports the diagnosis in patients with clinical suspicion of an associated disease (e.g., chronic low back pain and morning stiffness).

Q9 (AMBOSS): A 33-year-old man comes to the physician 1 hour after he slipped in the shower and fell on his back. Since the event, he has had severe neck pain. He rates the pain as an 8–9 out of 10. On questioning, he has had lower back pain for the past 2 years that radiates to the buttocks bilaterally. He reports that the pain sometimes awakens him at night and that it is worse in the morning or when he has been resting for a while. His back is very stiff in the morning and he is able to move normally only after taking a hot shower. His temperature is 36.3°C (97.3°F), pulse is 94/min, and blood pressure is 145/98 mm Hg. Range of motion of the neck is limited due to pain; the lumbar spine has a decreased range of motion. There is tenderness over the sacroiliac joints. Neurologic examination shows no abnormalities. An x-ray of the cervical spine shows decreased bone density of the vertebrae. An MRI shows a C2 vertebral fracture as well as erosions and sclerosis of the sacroiliac joints bilaterally. The patient's condition is most likely associated with which of the following findings?

A- Stiffness and deformity of the fingers and hands
B- Recent episode of urethritis
C- Recurring eye redness and pain
D- Back swelling and erythema
E. Foot drop and difficulty heel walking

Explanation: Ankylosing spondylitis is a seronegative spondyloarthropathy and is associated with extraarticular manifestations, the most common of which is acute unilateral anterior uveitis (~ 25% of cases). Further findings in this patient would, therefore, most likely include eye redness and pain. Other associated, though rarer, extraarticular findings of ankylosing spondylitis include gastrointestinal (inflammatory bowel disease), cardiac (aortic insufficiency, AV block), and renal (IgA nephropathy).

Answers: Q8: B| Q9: C

Spondyloarthritis

Q10 (AMBOSS): A 36-year-old man comes to the physician because of increasing back pain for the past 6 months. The pain is worse when he wakes up and improves throughout the day. He has difficulty bending forward. He has taken ibuprofen and celecoxib for the past 3 months, which provided limited relief. His only current medication is a topical corticosteroid for an itchy skin condition. His mother has rheumatoid arthritis. Temperature is 37.1°C (98.8°F), pulse is 75/min, respirations are 14/min, and blood pressure is 126/82 mm Hg. Examination shows a limited spinal flexion. He has two patches with erythematous papules on his right forearm. There is tenderness on percussion of the sacroiliac joints. An x-ray of the patient's spine is shown. Which of the following is most likely to improve this patient's mobility?

A- Methotrexate
B- Etanercept
C- Prednisolone
D- Rituximab

Explanation: Additional treatment with a TNF-α inhibitor, such as etanercept, is indicated for improving spine mobility and reducing pain in patients with AS who have persistent symptoms despite NSAID use, which is the first-line treatment for ankylosing spondylitis, for > 2 months. Some studies suggest that these agents may also slow down radiographic disease progression after several years of treatment. In addition, every patient should receive regular physical therapy to maintain range of motion and posture. However, there is no curative treatment for AS. Although treatment with DMARDs such as methotrexate (MTX) may be beneficial for patients with AS, especially when accompanied by peripheral arthritis, it is not a common treatment option to improve mobility. Moreover, among the DMARDs used in the treatment of AS, sulfasalazine is preferred over MTX. MTX is used as first-line treatment of rheumatoid arthritis and psoriatic arthritis.

Answers: Q10: B

SLE

Q1 (Pretest): A 20-year-old woman has developed low-grade fever, a malar rash, and arthralgias of the hands over several months. High titers of anti-DNA antibodies are noted, and complement levels are low. The patient's white blood cell count is 3000/µL, and platelet count is 90,000/µL. The patient is on no medications and has no signs of active infection. Which of the following statements is correct?

A. If glomerulonephritis, severe thrombocytopenia, or hemolytic anemia develops, high-dose glucocorticoid therapy would be indicated.
B. Central nervous system symptoms will occur within 10 years.
C. The patient can be expected to develop Raynaud phenomenon when exposed to cold.
D. Joint deformities will likely occur.
E. The disease process described is an absolute contraindication to pregnancy.

Explanation: The combination of fever, malar rash, and arthritis suggests systemic lupus erythematosus (SLE), and the patient's thrombocytopenia, leukopenia, and positive antibody to native DNA provide more than four criteria for a definitive diagnosis. Other criteria for the diagnosis of lupus include discoid rash, photosensitivity, oral ulcers, serositis, renal disorders (proteinuria or cellular casts), and neurologic disorder (seizures). High-dose corticosteroids would be indicated for severe or life-threatening complications of lupus such as described in item a. The arthritis in SLE is nondeforming. Patients with SLE have an unpredictable course. Few patients develop all signs or symptoms. Neuropsychiatric disease occurs at some time in about half of all SLE patients and Raynaud phenomenon in about 25%. Pregnancy is relatively safe in women with SLE who have controlled disease and are on less than 10 mg of prednisone.

Q2 (Pretest): A 28-year-old woman presents to her primary care physician with a 3-month history of fatigue. Her past medical history includes severe acne. She has had 3 uncomplicated vaginal deliveries and has healthy children aged 5, 3, and 2 years. Questioning reveals that she develops an erythematous rash upon minimal sun exposure, and has heavy menstrual periods despite being on oral contraceptives for the past 2 years. For the past 6 months, she has taken minocycline for acne. Physical examination reveals small joint effusions and tenderness to palpation of the knees bilaterally. Lab testing reveals a normocytic anemia, thrombocytopenia, mild hyper-bilirubinemia, and a marked elevation in her ANA titer. Which of the following statements best characterizes this patient's illness?

A. Her anemia is due to bone marrow suppression from chronic disease.
B. Her anemia is due to iron deficiency.
C. Minocycline should be discontinued.
D. Anti-histone antibodies are likely to be negative.
E. The likelihood of this patient developing venous thromboembolism is comparable to the general population.

Explanation: This patient likely has drug-induced lupus erythematosus. Minocycline is one of many medications implicated. Other common offenders include procainamide, hydralazine, propylthiouracil, carbamazepine, phenytoin, and isoniazid. Stopping the offending agent is essential and will lead to resolution of the disease in weeks to months. Renal and CNS disease are uncommon in drug-induced lupus; usually skin and joint manifestations predominate. In lupus, immune- mediated hemolysis is the usual cause of the anemia. Although depressed erythropoiesis from anemia of chronic disease (answer a) can contribute to the patient's low hemoglobin, the elevated bilirubin suggests hemolysis. Likewise, despite the heavy periods from her thrombocytopenia, long-standing iron deficiency anemia (answer b) will cause a low MCV (microcytic anemia). Anti-histone antibodies (answer d) are very common in drug-induced lupus. Answer e is incorrect as patients with lupus (drug-induced or otherwise) have a higher rate of clot formation and may suffer from antiphospholipid antibody syndrome. Although not directly related to venous thromboembolism, long-standing inflammation with lupus accelerates the rate of atherosclerosis, predisposing to arterial occlusive disease over time.

Q3 (500Best): A 34-year-old Afro-Carribean woman has been admitted for management and investigation of increasing shortness of breath. On further questioning, she mentions that her hands have been painful and stiff over the past few months and she has been having recurrent mouth ulcers. Chest x-ray confirms bilateral pleural effusions and blood tests reveal a raised ESR and a normal CRP. A diagnosis of systemic lupus erythematosus (SLE) is suspected and a full autoantibody screen is sent to the laboratory. Which of the following auto-antibodies is most specific to the suspected diagnosis?

A. Anti-nuclear antibody
B. Rheumatoid factor
C. Anti-double stranded DNA antibody
D. Anti-centromere antibody
E. Anti-mitochondrial antibody

Explanation: Anti-double stranded DNA antibody (C) is the most specific antibody for SLE and is raised in 60 per cent of cases. Levels of other autoantibodies may also be raised in SLE but these are less specific. They include anti- nuclear antibody (A) and rheumatoid factor (B). Anti-centromere antibody (D) levels may be raised in patients with limited systemic sclerosis but not in patients with SLE. Raised anti-mitochondrial antibody (E) levels are associated with primary biliary cirrhosis.

Answers: Q1: A | Q2: C | Q3: C

SLE

Q4 (500Best): A 27-year-old woman presents to accident and emergency complaining of sudden onset shortness of breath and right-sided pleuritic chest pain. She has a past medical history of three miscarriages and a deep venous thrombosis in the right leg. On examination, pulse is 110bpm, respiratory rate is 24bpm, oxygen saturation is 88 per cent on room air. An arterial blood gas shows pH 7.40, PO2 8.0, PCO2 3.1. What is the diagnostic investigation of choice?

A. Full blood count
B. Chest x-ray
C. D-dimer
D. CT pulmonary angiogram (CTPA)
E. ECG

Explanation: The clinical features outlined in this question of sudden onset shortness of breath and pleuritic chest pain with tachycardia and tachypnoea with a low oxygen saturation on room air, should raise the suspicion of pulmonary embolism. The blood gas demonstrates a low P02 and PCO2. This indicates that the patient is hypoxic despite hyperventilation. The diagnostic investigation of choice in patients with pulmonary embolism is CTPA (D). Full blood count (A) is not useful in the diagnosis of pulmonary embolism. Chest x-ray (B) is often normal in patients with pulmonary embolism but may show a wedge-shaped infarct. D-dimer (C) is a fibrin degradation product that is commonly used in the diagnosis of deep venous thrombosis or pulmonary oedema. When negative, it indicates that venous thrombosis is unlikely. D-dimer may be positive for a number of reasons. Therefore, it is often used in patients where the clinical picture is unclear. In the case outlined in the question, there should be a high degree of suspicion for pulmonary embolism and a CTPA should be arranged without delay, making D-dimer the incorrect answer. ECG (E) may be useful and may commonly demonstrate sinus tachycardia. In addition, ECG might show signs of right heart strain, such as right bundle branch block or right axis deviation. Rarely, the S1Q3T3 pattern is seen (large S wave in lead I, Q wave in lead III and inverted T wave in lead III). While ECG is useful, it is not the diagnostic investigation of choice, making this the incorrect answer.

Q5 (500Best): A 27-year-old woman presents to accident and emergency complaining of sudden onset shortness of breath, right-sided pleuritic chest pain and haemoptysis. She has a past medical history of three miscarriages and a deep venous thrombosis in the right leg. CTPA confirms a large pulmonary embolism. What is the most likely underlying diagnosis?

A. SLE
B. Primary anti-phospholipid syndrome
C. Raynaud's disease
D. Systemic sclerosis
E. Beçhet's disease

Explanation: Recurrent venous or arterial thromosis with a history of miscarriages should point to primary anti-phospholipid syndrome (B) as the correct answer in this question. Anti-phospholipid syndrome is associated with SLE (A) and a proportion of patients with SLE may develop secondary anti-phospholipid syndrome. The absence of any clinical features of SLE in this case means that this answer is the incorrect option. Raynaud's disease (C) is digital ischaemia due to vasospasm, often precipitated by the cold in the absence of an underlying cause. When an underlying cause, for example SLE, is present, it is termed Raynaud's phenomenon. Recurrent venous thrombosis and miscarriage are not features of systemic sclerosis (D) or Beçhet's disease (E), making these answers incorrect.

Answers: Q4: D | Q5: B

SLE

Q6 (500Best): A 27-year-old woman presents to accident and emergency complaining of sudden onset shortness of breath, right-sided pleuritic chest pain and haemoptysis. She has a past medical history of three miscarriages and a deep venous thrombosis in the right leg. CTPA confirms a large pulmonary embolism. A diagnosis of anti-phospholipid syndrome is suspected and a full autoantibody screen is sent. Which of the following auto-antibodies would confirm the diagnosis if detected?

A. Anti-cardiolipin antibody
B. Anti-centromere antibody
C. Anti-nuclear antibody
D. Anti-mitochondrial antibody
E. Anti-histone antibody

Explanation: The presence of anti-cardiolipin antibodies (A) would confirm the diagnosis. Both anti-cardiolipin antibody and lupus anticoagulant antibodies should be sent. Raised levels of anti-centromere (B) antibodies are associated with limited systemic sclerosis, not anti-phospholipid syndrome, making this answer incorrect. Anti-nuclear antibody (C) levels may be raised in a number of conditions including SLE, systemic sclerosis and rheumatoid arthritis, but are usually negative in anti-phospholipid syndrome. Raised anti-mitochondrial antibody (D) levels are associated with primary biliary cirrhosis, but not anti-phospholipid syndrome, making this answer incorrect. Similarly, raised anti-histone (E) antibody levels are associated with drug-induced SLE, but not anti-phospholipid syndrome.

Q7 (MKSAP): A 36-year-old woman is evaluated for a 5-month history of fever, joint swelling, and pleuritic chest pain. Her symptoms have not responded to daily naproxen. On physical examination, temperature is 36.6°C (97.8°F), blood pressure is 140/85 mm Hg, pulse rate is 102/min, and respiration rate is 14/min. She has a shallow, nontender hard palate ulcer and central facial redness sparing the nasolabial folds. She has bilateral synovitis of her wrist and metacarpophalangeal joints.
Hemoglobin 10.7 mg/dL, (107 g/L)
Leukocyte count 3000/uL (3 × 10%L)
Antinuclear antibody (ANA) Positive. 1:640
Urine protein 1.5 g/24 hours
Which of the following serologic tests is most likely to confirm the diagnosis?

A. Anti-double-stranded DNA antibody
B. Anti Ribonucleoprotein antibody
C. Anti-SS-A (Ro) and ant i-SS-B (La) antibodies
D. Anti-topoisomerase I (anti-Scl-70) antibody
E. Rheumatoid factor

Explanation: The next, most helpful serologic test for this patient is the anti-double-stranded DNA antibody. This patient with pleuritic chest pain, symmetric synovitis of the hand and wrist joints, leukopenia, proteinuria, and a positive ANA likely has systemic lupus erythematosus (SLE). Patients with a high pretest probability of SLE and ANAs (titer ≥1:160) should undergo confirmatory testing, such as measurement of complement levels C3, C4, and total hemolytic complement (CH50) and more specific autoantibody testing, such as anti-double-stranded DNA antibody testing (specificity, 75%-100%)
The anti ribonucleoprotein antibody is strongly associated with mixed connective tissue disease but also can be seen in patients with SLE and myositis, but is neither very sensitive nor specific for SLE. Anti-SS-A and anti-SS-B antibodies (sometimes referred to as anti-Ro and anti-La, respectively) are neither sensitive nor specific for SLE; they are seen in Sjogren syndrome. In patients with SLE, a positive anti-SS-A antibody is often associated with subacute cutaneous lupus erythematosus. Two antibodies are associated with systemic sclerosis: anti-topoisomerase I (anti-Scl-70) and anticentromere. The anti-Scl-70 antibody is seen in approximately half of the patients with diffuse systemic sclerosis and is associated with the development of interstitial lung disease. The anticentromere antibody is associated with limited cutaneous systemic sclerosis. Approximately 75% of patients with rheumatoid arthritis are rheumatoid factor positive, but the prevalence rate of rheumatoid arthritis may be as low as 50% in early disease. Rheumatoid factor positivity is not specific for rheumatoid arthritis and frequently occurs in other autoimmune disorders, including SLE, and chronic infections, most notably chronic active hepatitis C virus infection.

Answers: Q6: A | Q7: A

SLE

Q8 (MKSPA): A 25-year-old woman is evaluated during a routine follow-up visit. Four months ago, she was diagnosed with systemic lupus erythematosus that manifested as fatigue, malar rash, oral ulcers, pleuritis, and arthralgia. At that time, she began treatment with hydroxychloroquine and a 1-month course of low-dose prednisone. On physical examination today, she states that her symptoms have resolved somewhat but that she still has slight fatigue and mild arthralgia in her hands, feet, and knees. Temperature is 36.4°C (97.6°F), blood pressure is 130/92 mm Hg, pulse rate is 84/min, and respiration rate is 18/min. She has a mild malar flush, a painless ulcer on the hard palate, and trace bilateral ankle edema. The remainder of the examination is normal
Hemoglobin 10 g/dL. (100 g/L)
Leukocyte count 2300/uL. (2.3 × 10°/L)
Platelet count 132,000/uL. (132 × 10°/L)
Erythrocyte sedimentation rate 45 mm/h
Serum creatinine 1.0 mg/dL. (88.4 umol/L)
Albumin 3.1 g/dL (31 g/L)
Serum complement (C3 and C4) Decreased
Urinalysis 2+ protein; 3+ blood; 5-10 leukocytes, 15-20 erythrocytes, and 1 erythrocyte cast/hpf
Which of the following is the next best step in this patient's treatment?

A. Amlodipine
B. High-dose prednisone
C. Ibuprofen
D. Low-dose prednisone

Explanation: This patient's hypertension, ankle edema, hematuria, proteinuria, hypoalbuminemia, and erythrocyte casts on urinalysis are highly suggestive of lupus nephritis despite the absence of renal insufficiency. To prevent irreversible renal damage, early treatment with a high-dose corticosteroid such as prednisone is indicated for patients whose condition raises strong suspicion for lupus nephritis. Whether renal biopsy is necessary in this clinical situation in order to establish a diagnosis remains uncertain, and treatment with high-dose corticosteroids would not significantly alter subsequent biopsy results.
Initiation of antihypertensive therapy would benefit this patient but is not the most appropriate next step in the management of her condition; treatment of her nephritis takes precedence and may itself help to control her hypertension. Instead of a calcium channel blocker such as amlodipine, angiotensin-converting enzyme inhibitors are the antihypertensive drugs of choice in patients with lupus nephritis because these agents help to control proteinuria.
Ibuprofen may help to control this patient's arthralgia. However, NSAIDs can significantly worsen renal function in patients with lupus nephritis and are therefore contraindicated in this patient population.
Low-dose prednisone may help to alleviate this patient's arthralgia and rash but would not treat her lupus nephritis.

Q9 (MKSAP): A 22-year-old woman is evaluated because of a 3-week history of pain in her joints and a rash. Both the skin rash and arthralgia began after a 2-week sailing vacation in July on Lake Michigan. The joint pain involves primarily the wrists and hands bilaterally, and tends to be worse in the morning and improve as the day progresses. She originally thought that the rash was the result of sunburn, but it has not gone away, and is shown (Plate 31). It is not painful and does not itch.
Which of the following conditions is most likely responsible for this patient's rash?

A. Dermatomyositis
B. Rosacea
C. Seborrheic dermatitis
D. Systemic lupus erythematosus

Explanation: This woman most likely has systemic lupus erythematosus (SLE). Patients with SLE have sun sensitivity, and their disease is triggered or exacerbated by light in the ultraviolet A and ultraviolet B spectrums. The facial rash, seen in the figure, is a classic presentation, involving the bridge of the nose, malar areas, and forehead (not seen), with erythematous plaques and a fine scale. The nasolabial folds are relatively protected from the sun, and the absence of the rash in this area helps to distinguish it from other common rashes of the face, including rosacea and seborrheic dermatitis. The rash may last for hours or days and has a tendency to recur.
Rosacea is a chronic inflammatory skin disorder that begins in early to middle adulthood and is characterized by central telangiectasis, flushing, and acneiform papules and pustules. Many patients with rosacea are misdiagnosed as having SLE. However, the nasolabial folds are not typically spared in rosacea, acne-like pustules are more
prominent, and rosacea is not associated with other systemic symptoms, such as arthritis.
Patients with dermatomyositis have pronounced proximal muscle weakness and elevated serum concentrations of muscle enzymes such as creatine kinase. Patients with dermatomyositis may have a facial rash that extends up to the eyelids, giving them a purplish (heliotrope) hue. Another characteristic finding is red to purplish plaques on the dorsal hands, more prominent over the joints, and known as Gottron papules.
The lesions of seborrheic dermatitis are ill defined (lack a distinct border), yellowish-red, and of varying size, and are usually associated with a greasy or dandruff-like scale. It occurs most commonly on the scalp, central face, upper mid-chest, and other oily areas of the body, and is not related to sun exposure or associated with systemic symptoms.

Answers: Q8: B | Q9: D

SLE

Q10 (AMBOSS): A 14-day-old male newborn is brought to the emergency department by his mother because of a rash on his face for the past 5 days. His mother says that the rash became more prominent after sun exposure. The mother did not receive prenatal care and the newborn was delivered at home. She has had episodes of arthralgia and a rash on her face and neck that often worsened during summer, but she has never been to a physician. The newborn is at the 50th percentile for length and 25th percentile for weight. His temperature is 37.1°C (98.8°F), pulse is 79/min, respirations are 45/min, and blood pressure is 82/51 mm Hg. Examination shows erythematous, annular lesions with raised margins in the periorbital area and the scalp. An ECG shows a P-wave rate of 130/min and an R-wave rate of 75/min with no apparent relation between the two. Which of the following laboratory studies for the newborn is most likely to confirm the diagnosis?

A. Serology for anti-Ro/SSA antibodies
B. Polymerase chain reaction for trypanosomal DNA
C. Serology for antitreponemal antibodies
D. Serology for anti-dsDNA antibodies

Explanation: The finding of anti-Ro/SSA antibodies in a newborn is specific for neonatal lupus syndrome, which is a passively acquired autoimmune disease caused by the transfer of maternal anti-Ro/SSA or anti-La/SSB antibodies across the placenta and is the most common cause of complete heart block in a fetus or newborn. As well as manifesting with heart block or a characteristic rash, neonatal lupus syndrome can also affect the hepatic (e.g., hepatosplenomegaly, hepatitis) and hematologic systems (e.g., anemia, neutropenia, thrombocytopenia). The combination of positive antibodies (in mothers or newborns) and either congenital heart block, rash, or hepatic and hematologic findings is required for a diagnosis of neonatal lupus syndrome. The presence of anti-dsDNA antibodies is specific for systemic lupus erythematosus (SLE), a condition that commonly manifests with a malar rash, arthralgias, and photosensitivity; the mother's arthralgias and rash are consistent with SLE. However, anti-dsDNA antibodies do not cross the placenta; therefore, they would not be seen in a newborn with suspected neonatal lupus.

Q11 (AMBOSS): A 32-year-old woman comes to the physician because of fatigue and joint pain for the past 4 months. Examination shows erythema with scaling on both cheeks that spares the nasolabial folds and two 1-cm ulcers in the oral cavity. Further evaluation of this patient is most likely to show which of the following findings?

A. Increased platelet count
B. Decreased lymphocyte count
C. Increased complement component 3
D. Increased prothrombin time
E. Decreased gamma globulin

Explanation:Lymphocytopenia is a typical finding in patients with systemic lupus erythematosus, particularly during active disease, and is presumably caused by autoimmune-mediated cell destruction. Both at diagnosis and throughout the disease, decreased cell counts of all lineages may be present, with anemia being the most common hematological abnormality.

Answers: Q10: A | Q11: B

General Rheumatology

Q1 (500Best): A 36-year-old woman presents to the rheumatology outpatient clinic with a two-month history of stiff hands and wrists. She mentions that the pain is particularly bad for the first few hours after waking up and is affecting her work as a dentist. On examination, the wrists, metacarpophalangeal joints and proximal interphalangeal joints are swollen and warm. What is the most likely diagnosis?

A. Rheumatoid arthritis
B. Osteoarthritis
C. Septic arthritis
D. Polymyalgia rheumatica
E. Reactive arthritis

Explanation: Rheumatoid arthritis (A) is a chronic, systemic inflammatory disease, which produces a symmetrical, deforming polyarthritis. The typical presentation of rheumatoid arthritis is with symmetrical pain and stiffness of the small joints of the hands and feet. Pain is characteristically worse in the morning and improves with exercise. As the disease progresses, it may cause deformity of the affected joints. Less commonly, rheumatoid arthritis may present with a monoarthritis. Diagnosis is made from the presence of articular and extra-articular features characteristic of the disease. The most recent criteria for diagnosis is the 2010 American College of Rheumatology and the European League Against Rheumatism criteria. This criteria set can be applied to patients that have synovitis in at least one joint and the absence of an alternative diagnosis. A total score of 6 or greater is diagnostic.
Primary nodal osteoarthritis (B) affects the hands. However, this answer is incorrect as the distal interphalangeal joints (DIPs) are usually more involved than the proximal interphalangeal joints (PIPs). In addition, pain is characteristically worse at the end of the day. Septic arthritis (C) usually presents as a monoarthritis, making this answer incorrect. Polymyalgia rheumatica (D) is incorrect as this disorder is rare in those under the age of 60 and is characterized by pain and stiffness in the shoulders, neck, hips and lumbar spine, which is worse in the morning. Reactive arthritis (E) is usually an asymmetrical, lower-limb arthritis. Therefore this answer is also wrong.

Q2 (500Best): A 55-year-old man presents to his GP with a 2-week history of pain in his hands. The pain is particularly bad in his right hand. On examination, brown discoloration of the nails with onycholysis is noted and the distal interphalangeal joints are tender on palpation. What is the most likely diagnosis?

A. Rheumatoid arthritis
B. Dermatomyositis
C. Reactive arthritis
D. Osteoarthritis
E. Psoriatic arthritis

Explanation: Approximately 5 per cent of patients with psoriasis develop arthritis. The pattern of arthritis is variable but **most commonly affects the distal interphalangeal joints and is asymmetrical. Nail or skin changes of psoriasis are usually present, but may develop after the arthritis.** It is also important to note that psoriatic arthritis may present as a symmetrical polyarthritis, resembling rheumatoid arthritis. A small number of patients with psoriatic arthritis may develop arthritis mutilans, where peri-articular osteolysis and bone shortening occur, producing marked deformity. The pattern of asymmetrical arthritis affecting the distal interphalangeal joints with nail changes should indicate that psoriatic arthritis (E) is the correct answer. The distal interphalangeal joints are usually spared at onset of rheumatoid arthritis (A) and the asymmetrical pattern described with nail changes mean that this is the incorrect answer. Dermatomyositis (B) presents with symmetrical proximal muscle weakness with characteristic skin changes. It does not cause a polyarthritis of the hands, thus making this answer incorrect. Reactive arthritis (C) is an asymmetrical lower limb arthritis, making this answer wrong. Osteoarthritis (D) may affect the distal interphalangeal joints, but does not cause nail changes, making this answer incorrect as well.

Q3 (Pretest): A 66-year-old man complains of a 1-year history of low back and buttock pain that worsens with walking and is relieved by sitting or bending forward. He has hypertension and takes hydrochlorothiazide but has otherwise been healthy. There is no history of back trauma, fever, or weight loss. On examination, the patient has a slightly stooped posture, pain on lumbar extension, and has a slightly wide based gait. Pedal pulses are normal and there are no femoral bruits. Examination of peripheral joints and skin is normal. What is the most likely cause for this patient's back and buttock pain?

a. Lumbar spinal stenosis
b. Herniated nucleus pulposus
c. Atherosclerotic peripheral vascular disease
d. Facet joint arthritis
e. Prostate cancer

Explanation: Lumbar spinal stenosis is a frequent cause of back pain in the elderly. Patients typically have pain that radiates into the buttocks (and sometimes thighs) and is aggravated by walking and by lumbar extension. Decreased vibratory sensation and a wide based gait may also be seen. Narrowing of the spinal canal is usually caused by age-related degenerative changes. A recent randomized controlled trial demonstrated that surgery was more effective than medical therapy in the relief of symptoms for patients with lumbar spinal stenosis. Symptoms often recur several years after surgery.
Disc herniation and facet joint arthropathy usually cause unilateral radicular symptoms. Leg pain associated with walking can also be caused by vascular disease, but the symptoms often are unilateral and usually occur in the distal leg. Normal pedal pulses and the classic history make vascular claudication an unlikely diagnosis in this patient. The bone pain of metastatic cancer is rarely positional and is usually unremitting, causing pain both day and night.

Answers: Q1: A | Q2: E | Q3: A

General Rheumatology

Q4 (500Best): A 30-year-old Afro-Carribean woman presents to accident and emergency with a 1-week history of progressive shortness of breath and fever. On further questioning, she mentions that her hands have been painful and stiff over the past few months and she has been having recurrent mouth ulcers. Chest x-ray confirms bilateral pleural effusions and blood tests reveal a raised ESR and a normal CRP. What is the most likely diagnosis?

A. Systemic lupus erythematosus
B. Systemic sclerosis
C. Sjögren's syndrome
D. Discoid lupus
E. Beçhet's disease

Explanation: SLE is a multisystem, inflammatory disorder which is nine times more common in women than men. It is also more common in people of Afro- Carribean origin and peak age of onset is usually 20–40 years of age. Multiple genetic and aetiological factors have been associated with the development of this disease. SLE has an extremely variable presentation, with clinical features usually caused by underlying vasculitis. These include polarthritis (the most common clinical feature, often a symmetrical small joint polyarthritis similar to that seen in early RA), photosensitive rashes (characteristic erythematous rash over malar eminences, sparing the nasolabial folds), mouth ulcers, serositis (affecting pleura or pericardium) or renal disease (usually nephrotic syndrome of renal failure due to underlying glomerulonephritis). The features presented in this case are, therefore, most suggestive of SLE (A) as the diagnosis. The raised ESR and CRP should also identify SLE as the correct answer. Systemic sclerosis (B) can be localized or diffuse and does not characteristically cause mouth ulcers, pleural effusions or a raised ESR. Sjögren's (C) is a syndrome of dry eyes and dry mouth. It can be a primary syndrome or occur secondary to other autoimmune diseases. While it is associated with athralgia, the pleural effusion, mouth ulcers and blood results make this diagnosis unikely. Discoid lupus (D) is a variant of SLE where skin involvement is the only feature. Therefore, this is the incorrect answer. Beçhet's disease (E) usually manifests with oral ulceration. However, the peripheral polyarthritis, pleural effusuions and blood results (in Beçhet's both ESR and CRP may be elevated) make this the incorrect answer in this case.

Q5 (500Best): A 75-year-old woman presents to accident and emergency complaining of pain in her knees. She mentions that this has been troubling her for several months. Pain is generally worse in the evenings and after walking. On examination, there are palpable bony swellings on the distal interphalangeal joints of the fingers on both hands. In addition, there is reduced range of movement and crepitus in the knees. What is the most likely diagnosis?

A. Rheumatoid arthritis
B. Osteoarthritis
C. Reactive arthritis
D. Polymyalgia rheumatica
E. Gout

Explanation: Osteoarthritis (B) is the most common type of arthritis. It is increasingly common with age and most people over the age of 60 will have some evidence of the disease on x-rays. It is important to note that osteoarthritis occurs as a result of a complex pathological process. Osteoarthritis may be localized or generalized, with pain characterisitically worse in the evenings. Localized osteoarthritis includes nodal osteoarthritis, which usually involves the distal interphalangeal joints. However, the proximal interphalangeal joints are also affected. With time, the hands become stiff and painful and painless bony swellings develop – Heberden's nodes on the DIPs and Bouchard's nodes on the PIPs. Bony swelling of the first carpometacarpal joint may result in a squared hand appearance in nodal osteoarthritis. Localized osteoarthritis may also affect the weight-bearing joints of the hips and knees. Generalized osteoarthritis may include features of nodal disease plus widespread joint involvement including DIPs, first metatarsophalangeal joints, knees and hips. Increased pain in the evenings, nodal disease and knee involvement point to osteoarthritis being the correct answer here. Rheumatoid arthritis (A) is worse in the mornings and the DIPs are usually spared at disease onset, making this answer wrong. Reactive arthritis (C) does not involve the hands or cause nodal disease, making this answer incorrect. Polymyalgia rheumatica (D) is wrong as this does not usually involve the hands or knees and is worse in the mornings. Gout (E) normally presents as an acute monoarthritis, making this answer incorrect.

Q6 (500Best): A 53-year-old man, who works as a chef, presents to accident and emergency with sudden onset severe pain, tenderness and swelling of the first metatarsophalangeal joint. The pain is making it difficult for him to mobilize. He has had two previous similar episodes. Blood tests reveal a raised serum urate level. The most likely diagnosis is:

A. Gout
B. Pseudo-gout
C. Septic arthritis
D. Reactive arthritis
E. Osteoarthritis

Explanation: This case describes a typical presentation of acute gout (A). It is caused by a raised plasma urate level and most commonly affects the first metatarsophalangeal joint. Occassionally it is polyarticular. Acute manifestations may be precipitated by a range of causes such as trauma, surgery or the use of diuretics. Pseudo-gout (B) may present as a monoarthritis but usually affects other joints such as the knee, wrist or hip. In addition, the raised urate levels are not seen. Septic arthritis (C) should be considered in any acutely inflamed joint. The raised urate levels mean that this diagnosis is less likely. Reactive arthritis (D) presents as an asymmetrical polyarthritis of the lower limbs, making this answer incorrect. Osteoarthritis (E) is unlikely to present as an acute arthritis affecting just the first metatarsophalangeal joint. The raised urate levels and the clinical presentation mean that a diagnosis of osteoarthritis is unlikely.

Answers: Q4: A | Q5: B | Q6: A

General Rheumatology

Q7 (AMBOSS): A 58-year-old woman comes to the physician because of a 2-year history of progressively worsening pain in her knees and fingers. The knee pain is worse when she walks for longer than 30 minutes. When she wakes up in the morning, her fingers and knees are stiff for about 15 minutes. She cannot recall any trauma to the joints. She was treated with amoxicillin following a tick bite 2 years ago. She is otherwise healthy and only takes a multivitamin and occasionally acetaminophen for the pain. She drinks one to two glasses of wine daily. She is 160 cm (5 ft 3 in) tall and weighs 79 kg (174 lb); BMI is 31 kg/m2. Her temperature is 36.9°C (98.4°F), pulse is 70/min, and blood pressure is 133/78 mm Hg. Examination of the lower extremities shows mild genu varum. Range of motion of both knees is limited; there is palpable crepitus. Complete flexion and extension elicit pain. Tender nodules are present on the proximal and distal interphalangeal joints of the index, ring, and little fingers bilaterally. Which of the following is the most likely diagnosis?

A. Gout
B. Lyme arthritis
C. Rheumatoid arthritis
D. Osteoarthritis
E. Septic arthritis

Explanation: This patient presents with symptoms characteristic of osteoarthritis (OA): joint pain that worsens with ambulation/weight-bearing, joint stiffness < 30 minutes after prolonged immobility, decreased range of motion (due to osteophytes), palpable crepitus of both knees, genu varum (due to medial cartilage loss), and nodules on the DIP (Heberden's nodes) and PIP (Bouchard's nodes). Furthermore, this patient has multiple risk factors for OA, including female sex, obesity, and age > 55 years. X-ray of the involved joints would most likely show decreased joint space, osteophytes, and subchondral sclerosis (areas of increased bone density caused by unequal load-bearing

Q8 (AMBOSS): A 53-year-old woman with hypertension and gastroesophageal reflux comes to the physician because of swelling and pain in the right ankle for 3 days. She has smoked 1 pack of cigarettes daily for 7 years and drinks 1–2 glasses of wine during special occasions. She follows a vegetarian diet and exercises regularly. Current medications include atenolol, enalapril, hydrochlorothiazide, and omeprazole. Her BMI is 22 kg/m2. Physical examination shows erythema and swelling of the right ankle, with limited range of motion due to pain. Arthrocentesis of the right ankle joint is performed, and microscopic analysis of the aspirated fluid shows negatively birefringent, needle-shaped crystals. Which of the following is this patient's strongest predisposing factor for her condition?

A. Female sex
B. Medication use
C. Diet
D. Smoking history
E. Weight

Explanation: Certain medications can cause hyperuricemia, which is the strongest risk factor for acute gout. Thiazide diuretics, taken by this patient, increase uric acid reabsorption in the renal tubules. Hyperuricemia can also be caused by uric acid overproduction, for instance, due to high dietary purine intake (e.g., a high-protein diet, excessive alcohol consumption), which is unlikely in this vegetarian patient with social drinking habits. Other risk factors for gout include male sex, hypertension, obesity, diabetes, and dyslipidemia.

Answers: Q7: D | Q8:B