Answers to self-assessment questions

Rheumatology

A Clinical Handbook

SECOND EDITION

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Answers to self-assessment questions

2.1 Rheumatoid arthritis

1. How long has the pain and swelling been present? How has the pain and swelling changed over time? Are the joints stiff? If so, for how long? Is the stiffness worse in the morning? Does it change throughout the day? Is there pain anywhere else? Are any other joints affected?
2. Boutonnière deformity, swan neck deformity, Z-thumb, or ulnar deviation.
3. FBC (↑ platelet count, ↑ serum ferritin, anaemia of chronic disease); RF or anti-CCP (+), CRP/ESR (↑ during active disease); LFTs and U & Es (usually normal).
4.
   - **Eyes:** scleritis and episcleritis
   - **Skin:** rashes, vasculitis ulcers
   - **Rheumatoid nodules**
   - **Respiratory system:** pleural effusions, pulmonary nodules, interstitial lung disease
   - **Haematological:** anaemia of chronic disease, Felty's syndrome
   - **Neuropathy**
   - **Cardiovascular system:** cardiovascular disease, pericarditis
   - **Renal system:** analgesic nephropathy, amyloidosis.
5. DMARDs. Methotrexate is most commonly used. Sulfasalazine, leflunomide and hydroxychloroquine are also commonly used. Side-effects of methotrexate include bone marrow suppression, GI upset (mouth ulcers), hepatotoxicity (fibrosis/cirrhosis) and nephrotoxicity, acute pneumonitis, pulmonary fibrosis or pulmonary oedema.
6. Measuring CRP and the DAS28 score.

2.2 Osteoarthritis

1. Both hips have reduced joint space, subchondral sclerosis, subchondral cysts and osteophytes. This is indicative of osteoarthritis involving both hip joints.
2. Paracetamol. If this doesn't work, work up the pain ladder (see *Fig. 2.2.5*).
3. Exercise and weight loss, physiotherapy, walking aids, and psychosocial support. Behaviour change techniques would be useful to help implement these changes.
4. Failure of medical therapy and severe impact on the patient's life. Given the patient's age, the most appropriate surgical intervention would be a bilateral total hip replacement.

2.3 Septic arthritis

1. **Look:** signs of erythema, swelling and obvious effusion. **Feel:** tenderness, warmth and effusion. **Move:** marked limitation of movements and inability to bear weight. **Presence of systemic features:** fever, malaise, rash and tachycardia.

2. Haematogenous spread of a respiratory or urinary tract origin is the most common route.
3. Risk factors include: prosthetic joint, rheumatoid arthritis, diabetes mellitus, low socioeconomic status, extremes of age, intravenous drug use, osteomyelitis, intra-articular injection / aspiration.
4. Gram stain, WCC and culture.
5. *Staphylococcus aureus*.
6. A microbiologist should be consulted to ensure that the choice of therapy is guided by the local resistance patterns in both hospitals and the community. Flucloxacillin is typically the first-line treatment for *Staphylococcus aureus* infections, unless the patient is allergic to penicillin. In such cases, an alternative antibiotic, such as clindamycin, may be used.

2.4 Psoriatic arthritis

1. Nail changes (pitting, onycholysis and hyperkeratosis) and uveitis.
2. **Clinical features:** PsA characteristically involves the DIP joint; RA mainly affects the MCP and PIP joints of the hand (although the PsA rheumatoid pattern presents very similarly). PsA usually involves an asymmetrical large joint, whereas RA presents symmetrically and usually affects the small joints. PsA is not associated with rheumatoid nodules, whereas RA is. PsA commonly presents with dactylitis and enthesitis, RA doesn't. RA and PsA have different extra-articular manifestations.
3. **Blood tests:** PsA is RF and anti-CCP negative, whereas RA typically is RF and anti-CCP positive.
4. Soft tissue swelling may be the only radiographical finding seen in early disease. Erosion in the DIP joint and periarticular new bone formation, osteolysis and 'pencil-in-cup' deformity occur in advanced disease.
5. Oral NSAIDs (first-line) and intra-articular corticosteroid injections if NSAIDs are insufficient for pain relief. Two classes of DMARDs are cDMARDs (conventional), and bDMARDs (biologic).
6. A cDMARD, usually methotrexate. Side-effects are bone marrow suppression, GI upset (mouth ulcers), hepatotoxicity (cirrhosis) and nephrotoxicity; acute pneumonitis, pulmonary fibrosis or pulmonary oedema; hypersensitivity reactions and increased risk of infection.

2.5 Ankylosing spondylitis

1. Given the patient's age and clinical presentation of chronic lower back pain and stiffness which is particularly worse in the morning (and lasts longer than an hour), a diagnosis of ankylosing spondylitis (AS) is most likely.
2. The modified Schober's test. An inferior mark at the level of the posterior superior iliac spine is drawn and a 10 cm segment above this point is also marked on the patient's back. The increase in distance on maximal forward spinal flexion with locked knees is measured. The measured distance should increase from 10 cm to at least 13.5–15 cm in healthy adults. If it increases less than this, it is indicative of AS.
3. HLA-B27.
4. Early signs include bone erosions, widening of the SI joints and vertebral bodies appear square with shiny corners (Romanus lesions). In the long term, ossification

of longitudinal ligaments of the spine (syndesmophytes) occurs, giving it a bamboo spine appearance.

5. See *Table 2.5.2*.

2.6 Reactive arthritis

1. GU chlamydia infection, his age as well as his gender.
2. To look for any keratoderma blennorrhagica of the soles – this is a sign of reactive arthritis.
3. Circinate balanitis.
4. Raised CRP and ESR; FBC – leukocytosis and thrombocytosis acutely; ANA, RF and anti-CCP (all negative); HLA-B27 (positive in 75%).
5. Rest and splint the affected joint. Give an NSAID for pain and soft tissue inflammation. Tetracycline (follow local antimicrobial guidelines) should be administered if chlamydia infection is implicated.

2.7 Gout

1. Gout, pseudogout, septic arthritis, acute flare of osteoarthritis and cellulitis. Septic arthritis is the most important condition to rule out, since failure to treat septic arthritis promptly can result in rapid joint destruction.
2. A single peripheral joint (most commonly the MTP joint of the big toe) which acutely becomes excruciatingly painful (often nocturnal), red, hot and swollen.
3. Obese, male and middle-aged.
4. Under polarized light microscopy, the presence of negatively birefringent monosodium urate (MSU) crystals.
5. NSAIDs (first-line) e.g. naproxen or colchicine (if NSAIDs are contraindicated) or corticosteroids (if NSAIDs and colchicine are contraindicated). The main side-effects of NSAIDs are GI disturbances including discomfort, nausea, diarrhoea, and occasionally GI bleeding and ulceration. Side-effects of colchicine include nausea, vomiting, and abdominal pain. Corticosteroids have many side-effects ('**CUSHINGOID FAM**') – **C**ushing's syndrome, **C**ataracts, **U**lcers (peptic), **S**kin (striae, thinning, bruising), **H**ypertension, **I**nfections, **N**ecrosis, **G**rowth restriction (children), **O**steoporosis, **O**besity (central), **I**mmunosuppression, **D**iabetes, **F**luid retention, **A**cute pancreatitis and **M**yopathy.
6. Try to limit alcohol consumption to less than 14 units per week with two alcohol-free days (beer, champagne, red wine and port may make gout worse than other alcohol). A high intake of fluids should be recommended (at least 1.5 L of water). Regular consumption of purine-rich foods (i.e. heart, kidney, liver, anchovies, crab, sardines, prawns) should be avoided and you should have only 2 servings of protein per day. Low-fat dairy and vegetable sources of protein should be recommended.
7. Allopurinol; co-prescribe a low-dose NSAID, or low-dose colchicine, for at least 1 month to prevent acute attacks of gout. Febuxostat is second-line if allopurinol is not tolerated or is contraindicated.

2.9 Vasculitis

1. Wegener's granulomatosis.
2. C-ANCA targeting anti-proteinase 3.
3. Other signs/symptoms:

Related to the upper airway	Foul-smelling rhinorrhoea, hyposmia, epiphora, scleritis/episcleritis, sinusitis, nasal septal perforation and hoarseness of voice
Related to the pulmonary airway	Persistent cough (usually unproductive), pyrexia, haemoptysis, dyspnoea and post-obstructive infection
Related to the kidney	Nephritic syndrome
Other features	Skin rash (palpable purpura), conjunctival haemorrhages and scleritis

4. Cyclophosphamide, given in combination with high-dose corticosteroids.

2.10 Giant cell arteritis

1. Giant cell arteritis (GCA).
2. High-dose glucocorticoid: oral prednisolone (40–60 mg).
3. Due to overlapping polymyalgia rheumatica.
4. Erythrocyte sedimentation rate (ESR)/CRP.
5. Long segment biopsy and MRI (temporal/extracranial) have a similar diagnostic value.

2.11 Polymyalgia rheumatica

1. Polymyalgia rheumatica. Bilateral pain and stiffness in the shoulder and hip girdles, as well as morning stiffness which improves as the day progresses, are characteristic of PMR. Fatigue, weight loss and fever are also symptoms of PMR.
2. Giant cell arteritis.
3. Blood tests: raised ESR >40 mm/hour and CRP in PMR and GCA. Serum protein electrophoresis: measures paraprotein level to exclude multiple myeloma. Thyroid function test to exclude hypothyroidism. Radiography to exclude non-erosive joint disease. Temporal artery biopsy to rule out GCA (if clinically indicated).
4. With oral prednisolone (15–20 mg prednisolone). The dose of prednisolone is reduced slowly for 3–6 months to a low maintenance level which is sustained for a further 6–12 months and gradually reduced over the next 6 months with the view of stopping altogether. A bone protective agent (e.g. bisphosphonate) and gastroprotective agent (e.g. proton pump inhibitor (PPI)) should be used as prophylaxis against the long-term side-effects of corticosteroids.

2.12 Systemic lupus erythematosus

1. Malar and discoid rash.
2. Risk factors include: female sex, age (15–45 years), ethnicity (particularly prevalent in Afro-Caribbeans and Asians), drug use (minocycline, isoniazid, procainamide, quinidine, chlorpromazine and methyldopa), sun exposure, family history and tobacco use.
3. ANA is the most sensitive autoantibody in SLE (>95%) but may be raised in other conditions. Anti-dsDNA autoantibody is the most specific autoantibody to SLE.
4. Anti-dsDNA antibody titres, complement proteins (C3, C4) and ESR levels are the best indicators of disease activity. Other parameters such as BP, urinalysis for casts and protein, FBC, U&Es, LFTs and CRP can also be used.
5. Advice about sun exposure – if she has sun-induced rashes then she should use sunscreen regularly for about 6 months over the summer. She should be aware that sun exposure may precipitate a flare-up. Smoking cessation if she smokes. Pregnancy should be planned. Risk of problems with pregnancy is greatly reduced if disease is well controlled prior to conception. Her drug therapy should be reviewed before pregnancy. Pills that contain oestrogen may exacerbate lupus disease or thrombosis and she should be aware of this and use with caution. Suggest that barrier methods or progesterone-only contraception are a better alternative. Finally, advise that infections should be treated promptly.

2.13 Polymyositis and dermatomyositis

1. Gottron's papules, Gottron's sign and heliotrope rash.
2. Creatine kinase (CK) and other enzymes including aldolase, serum glutamic-oxaloacetic transaminase (SGOT), serum glutamic-pyruvic transaminase (SGPT), and lactate dehydrogenase (LDH) are usually raised. ESR, plasma viscosity and CRP may be raised in active disease. ANA autoantibodies as a positive ANA finding are found in approximately 60% of patients. Anti-Jo-1 antibodies should also be screened for, as these are associated with interstitial lung disease, Raynaud's phenomenon and arthritis.
3. Electromyography and muscle biopsy.
4. High-dose prednisolone should be started, i.e. 60 mg/24 hr. In resistant cases DMARDs should be used; most commonly rituximab.
5. Measure CK levels. They should decrease in response to effective treatment.

2.14 Sjögren's syndrome

1. Swelling around cheeks, vaginal dryness, dyspareunia, dry cough, dysphagia, joint pain and symptoms of Raynaud's.
2. Non-Hodgkin's B-cell lymphoma.
3. Anti-Ro (SS-A) and anti-La (SS-B) autoantibodies. They are found in up to 90% of patients with SS.
4. Schirmer's test.
5. For her eye symptoms, artificial tears are first-line. Other treatments for dry eyes include ophthalmic ciclosporin drops, spectacle eye shields and humidifiers. Also, advise patients to take regular breaks while reading. For her symptoms of dry mouth, she should be advised to drink plenty of fluids. Salivary substitutes are first-line but cholinergic drugs such as pilocarpine can be used if salivary substitutes are inadequate.

2.15 Scleroderma

1. Subcutaneous calcinosis.
2. Scleroderma.
3. The extent of skin involvement should be noted. Other signs: hand swelling, reduced range of movement (prayer sign), subcutaneous calcinosis and telangiectasia. If there is foot swelling, prompt CVS examination for signs of heart failure. Interstitial lung disease signs should also be looked for.
4. ANA, anti-topoisomerase-1 (Scl 70) antibody, anti-centromere antibody (ACA) or anti-RNA polymerase I and III antibody.
5. Interstitial lung disease. CXR, pulmonary function tests, high resolution CT scan and echocardiogram should be performed.

2.16 Fibromyalgia

1. Fibromyalgia.
2. All negative.
3. Heated pool treatment, exercise programmes (individually tailored exercise programmes which include aerobic training and muscle strengthening), cognitive behavioural therapy (CBT), physiotherapy and psychological support.
4. Paracetamol can be trialled for pain relief when symptoms are mild. For more severe symptoms, antidepressants including SSRIs (e.g. citalopram, fluoxetine, sertraline), duloxetine or amitriptyline should be considered. Neurotransmitter modulators and codeine can be trialled with caution as these may have addictive potential and have been reclassified as class C schedule drugs.

2.17 Osteoporosis

1. Hyperthyroidism, alcohol use, low BMI, testosterone deficiency, erosive bone disease or calcium deficiency.
2. Use of prophylactic bone protective agents such as bisphosphonates.
3. Bisphosphonates, e.g. alendronate. Side-effects of bisphosphonates include oesophageal reactions (oesophagitis, oesophageal ulcers, stricture and erosions), abdominal pain and distension, dyspepsia, regurgitation and osteonecrosis of the jaw (rare).
4. Osteopenia of the vertebrae. The bone density of the hip is normal.
5. FRAX tool or QFracture.
6. Smoking cessation and reduction of alcohol consumption (if indicated), weight-bearing muscle exercises, dietary (adequate calcium and vitamin D) and physiotherapy.

2.18 Paget's disease

1. Osteomalacia and bone metastasis.
2. Pelvis, spine, skull and femur.
3. Bone deformity and enlargement (particularly affecting the bones mentioned above), increased temperature over affected bones, pathological fractures, secondary osteoarthritis as well as hearing loss and tinnitus.
4. With bisphosphonates (to reduce bone turnover) and analgesics, e.g. paracetamol/NSAIDs (for bone pain).

3.1 Vitamin D deficiency

1. Rickets.
2. Inadequate vitamin D in her diet as she is exclusively breastfed.
3. Serum calcium (decreased), parathyroid hormone (increased), fasting phosphate (decreased) and ALP (increased).
4. Metaphyseal cupping and flaring, epiphyseal irregularities, widening of the epiphyseal plates.
5. Reassure them that with appropriate vitamin D replacement, her condition can be treated. This will allow her health to progress in terms of gaining weight, growing and walking properly as well as reversing the abnormalities in her legs and wrists.

3.2 Juvenile idiopathic arthritis

1. Transient synovitis of the hip; septic arthritis; JIA; osteomyelitis; slipped upper femoral epiphysis; Perthes' disease.
2. Oligoarthritis JIA.
3. Non-pharmacological: physiotherapy, hydrotherapy and occupational therapy, parent education and liaison with nursery. Pharmacological: NSAIDs. If severe, surgery is an option.