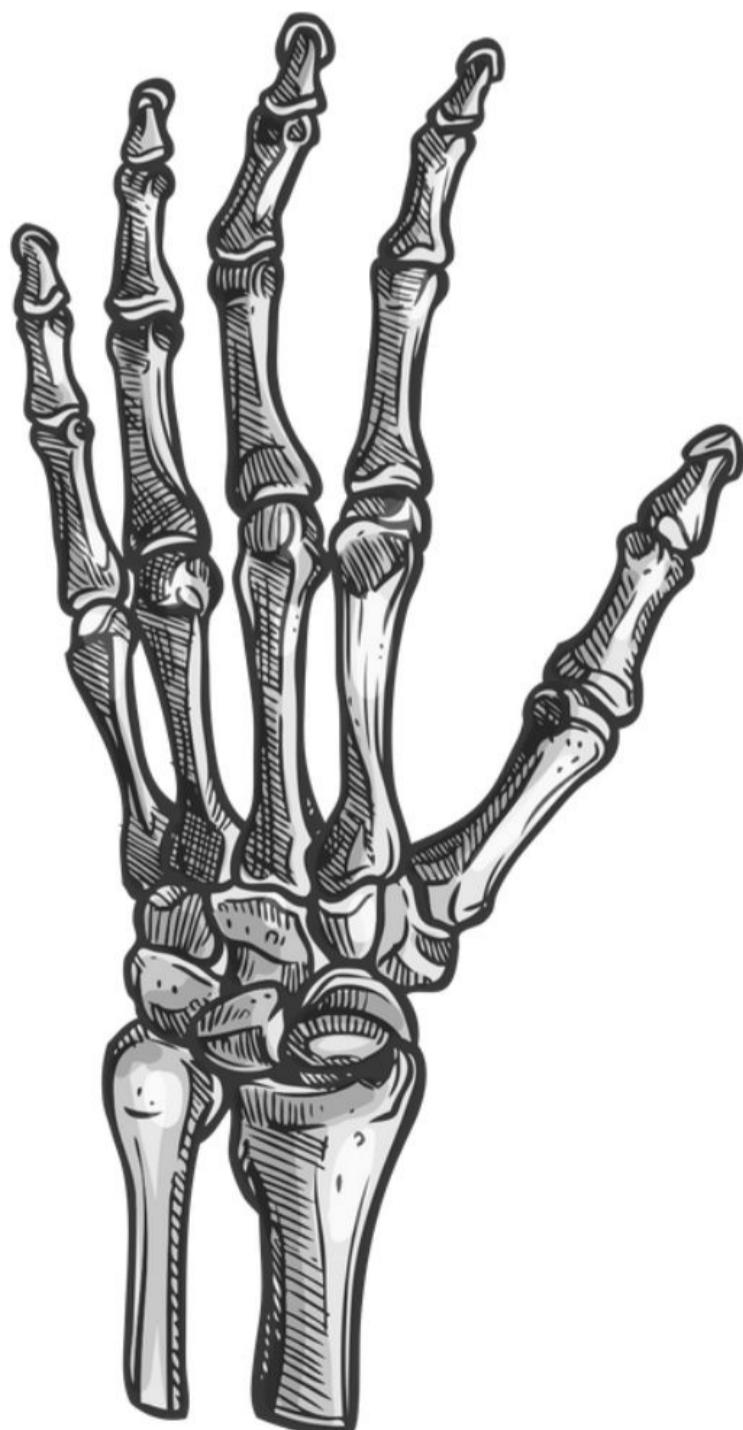


PLABABLE

GEMS

VERSION 3.3

RHEUMATOLOGY



Autoantibodies

Disease	Autoantibody
SLE	<ul style="list-style-type: none">• Anti-ds DNA (specific)• Antinuclear antibody (sensitive)
Drug induced lupus	Anti-histone antibody
Diffuse scleroderma	Anti-scl70
Limited scleroderma (CREST syndrome)	Anti-centromere
Polymyositis	Anti-Jo1
Sjogren's syndrome	Anti-Ro Anti-La
Primary biliary cirrhosis	Anti-mitochondrial
Autoimmune hepatitis	Anti-smooth muscle

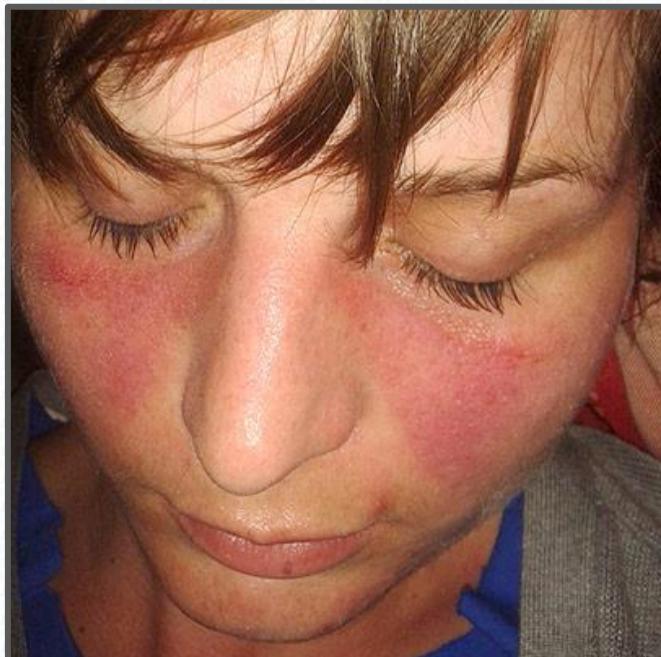
Autoantibodies

Disease	Autoantibody
Churg Strauss syndrome	p-ANCA
Wegener's granulomatosis	c-ANCA (remember as WBC)
Coeliac disease	<ul style="list-style-type: none">• Anti-tissue transglutaminase• Anti-gliadin• Anti-endomysial
Graves disease	TSH-receptor antibody
Rheumatoid arthritis	<ul style="list-style-type: none">• Rheumatoid factor (initial)• Anti-CCP (most specific)

Systemic Lupus Erythematosus

Autoimmune disorder with multi-organ involvement

Organ involved	Presentation
General	<ul style="list-style-type: none">● Fatigue● Fever● Mouth ulcers
Skin	<ul style="list-style-type: none">● Malar (butterfly) rash on face● Photosensitivity (discoid rash)● Raynaud's phenomenon
Joints	<ul style="list-style-type: none">● Arthralgia● Peripheral and symmetrical polyarthritis
Cardiovascular	<ul style="list-style-type: none">● Pericarditis● Libman-Sacks endocarditis
Respiratory	Pleuritis
Renal	Glomerulonephritis (MC: Diffuse proliferative)



Butterfly rash

Systemic Lupus Erythematosus

Systemic:

- Low-grade fever
- Photosensitivity

Mouth and nose

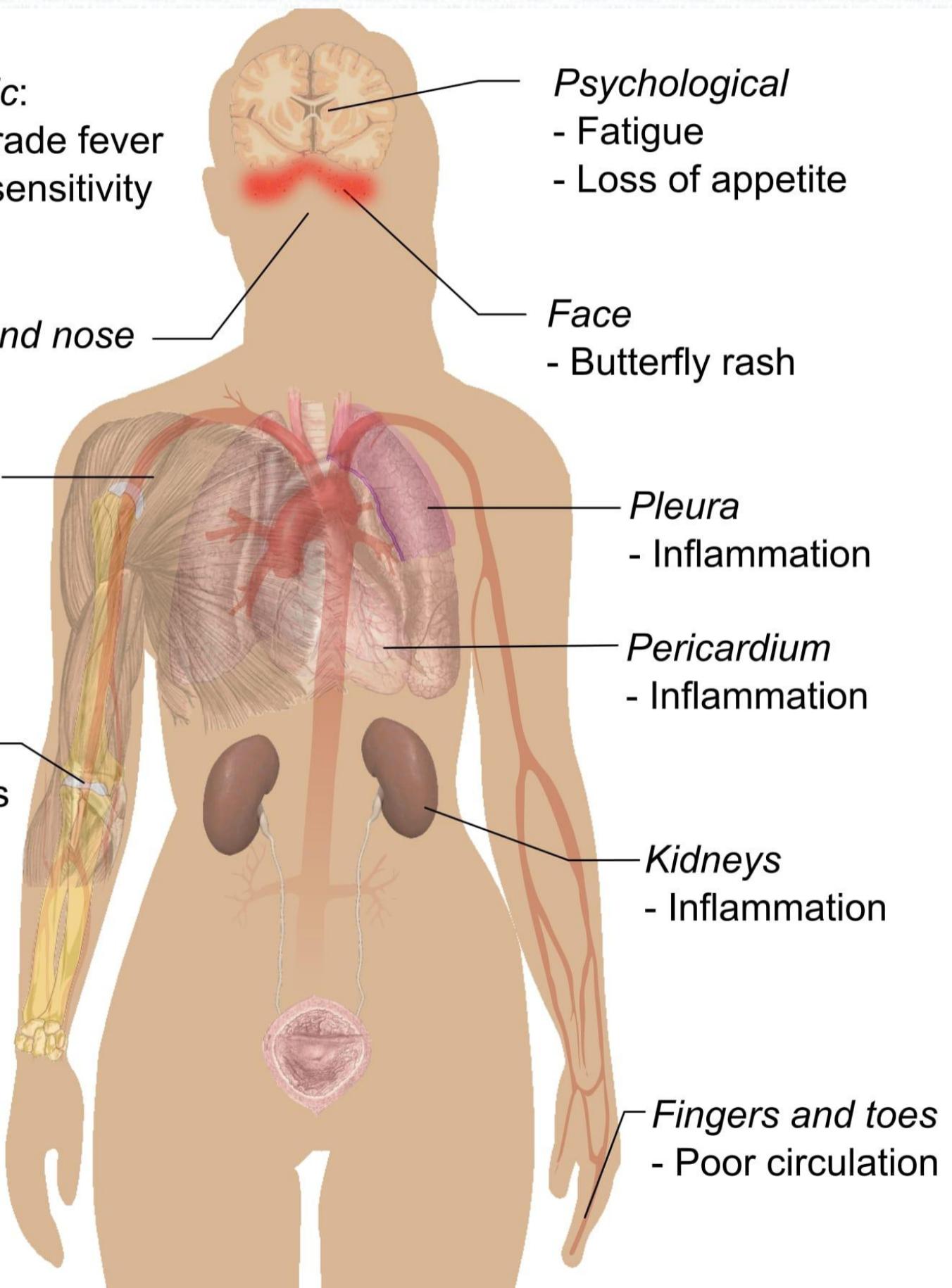
- Ulcers

Muscles

- Aches

Joints

- Arthritis



Systemic Lupus Erythematosus

Investigations

- **ANA (high sensitivity)**
- **Anti-dsDNA antibody (high specificity)**
- ↓ C3 and C4 levels
- Anti-histone (drug induced SLE)

Management

- **Hydroxychloroquine** - skin lesions, arthralgia and myalgia, oral ulcers
- **NSAIDs** - arthralgia and myalgia
- **Prednisolone** - severe cases
- **Cyclophosphamide** - life threatening lupus nephritis and vasculitis

Drug induced lupus

- Hydralazine
- Chlorpromazine
- Isoniazid



Systemic Lupus Erythematosus

A 44 year old ...*stem sounding like lupus!*

What is the **most sensitive test?**

What is the **initial test?**

What is the **best screening test?**



Antinuclear antibody (ANA)

What is the **most specific test?**

What is the **most discriminative test?**



Anti-dsDNA

Chronic Fatigue Syndrome

Also, known as **myalgic encephalomyelitis** is a chronic condition and is a diagnosis of exclusion

Presentation

- Post-exertional malaise
- Activity-induced muscle fatigue
- Cognitive dysfunction
- Sleep problems
- Generalised pain

Management

- Cognitive behavioral therapy
- Graded exercise program



Polymyositis

Presentation

- Symmetrical and diffuse muscle weakness (proximal > distal)
- Difficulty rising from a low chair, climbing steps, lifting objects and combing hair

Investigation

- ↑ Serum creatine kinase
- Anti-jo 1 antibodies
- ↑ LDH and aldolase
- Electromyography
- Muscle biopsy - definitive test

Treatment

- Steroids

Dermatomyositis

Presentation

- Muscle weakness
- Skin manifestations:
 - **Heliotrope rash:** blue-purple discolouration on the upper eyelids with periorbital oedema
 - **Gottron's papules:** Raised purple-red scaly patches over the extensor surfaces of joints and fingers
 - **Shawl sign:** rash around the neck



Source: IMACS

Dermatomyositis



Source: IMACS

Investigation

- ↑ Serum creatine kinase
- Anti-nuclear antibody
- ↑ LDH and aldolase
- Electromyography
- Muscle biopsy

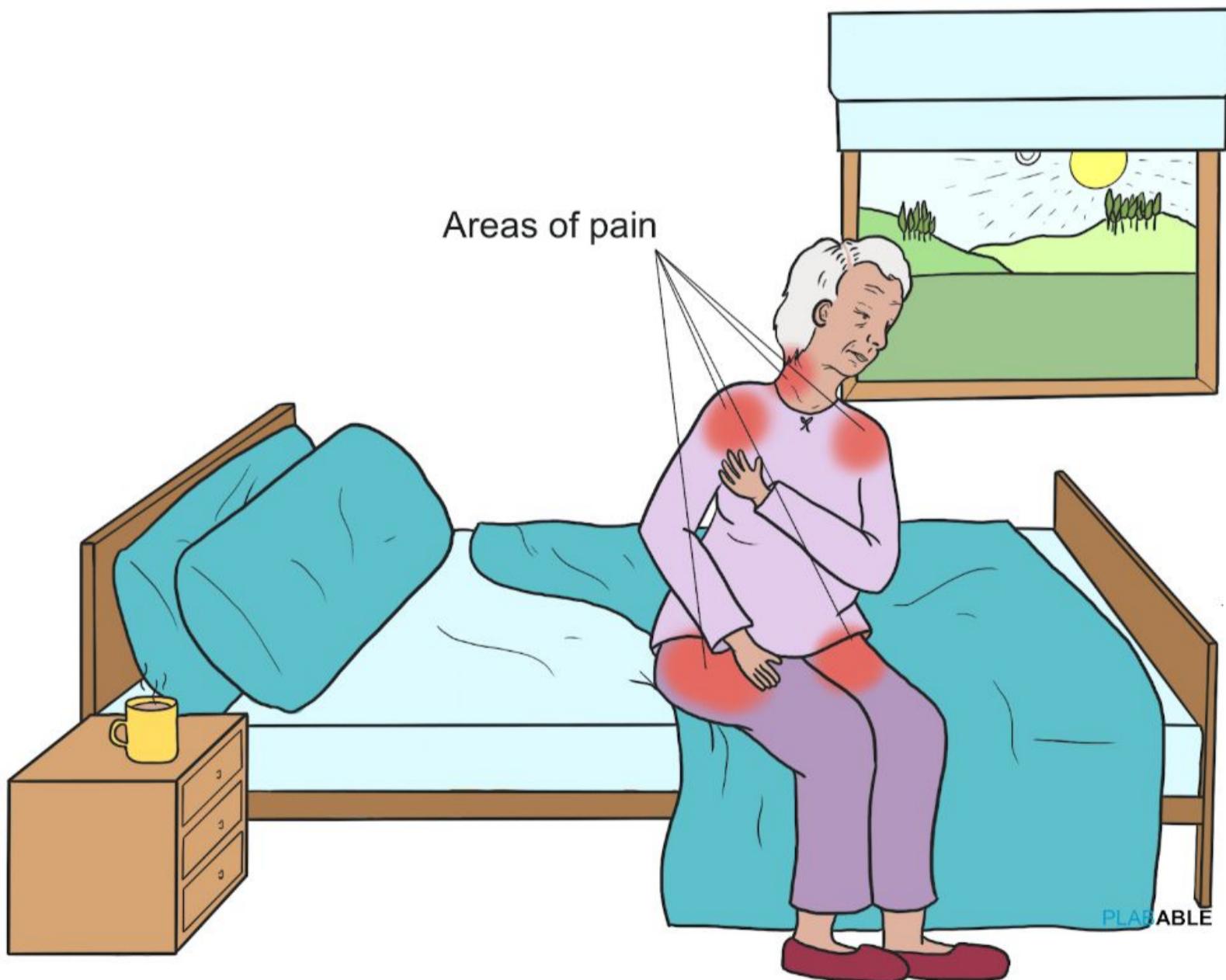
Treatment

- Steroids
- Sun-blocking agents

Polymyalgia Rheumatica

Presentation

- Bilateral pain and morning stiffness of the shoulder, neck and pelvic girdle
- Difficulty in getting out of bed, or raising the arm to brush the hair
- Associated with **giant cell arteritis**
- Mean age >50 yrs



Polymyalgia Rheumatica

Investigation

- ↑ ESR / CRP
- **Normal creatine kinase**

Treatment

- Steroids (oral prednisolone)

Plabable's Tip

Remember the letter P

Polymyalgia = Pain (not weakness)

Muscles are tender with no true weakness!

Differential Diagnosis

Myositis - weakness > pain + ↑CK

Polymyalgia - pain > weakness + normal CK

Inflammatory Conditions Involving Muscles

This card is to recap what is already learnt!

Polymyositis

Weakness of muscles

Creatine kinase high

Anti-Jo antibodies present

Polymyalgia Rheumatica

Painful, stiff muscles

ESR raised

All three of these conditions are treated with steroids

Press the icon for one of our teaching videos by Dr Jigyasa Saini



Giant Cell (Temporal) Arteritis

Presentation

- Headache
- **Scalp tenderness** (pain during combing)
- Transient visual field loss
- Age > 50
- Jaw claudication
- Associated with polymyalgia rheumatica

Investigation

- \uparrow ESR (best initial)
- Temporal artery biopsy (definitive)

Treatment

- High dose **prednisolone** (do not wait for biopsy as delay can result in permanent loss of vision)
- Low dose **aspirin** (75mg/day)

Giant Cell (Temporal) Arteritis

Brain trainer:

A patient with a headache, scalp tenderness and jaw pain presents to the Emergency Department. Blood test have been sent. What is the most appropriate management?

→ **Start prednisolone**

Starting prednisolone is the most appropriate medication to decrease the risk of her losing her vision. Low-dose aspirin can be given in addition to prednisolone as it helps to prevent myocardial infarctions and strokes.

Key learning point here is if you are given both prednisolone and aspirin as options to choose, remember prednisolone is the more important medication to start acutely.

Sjogren's Syndrome

Autoimmune disorder affecting exocrine glands

Presentation:

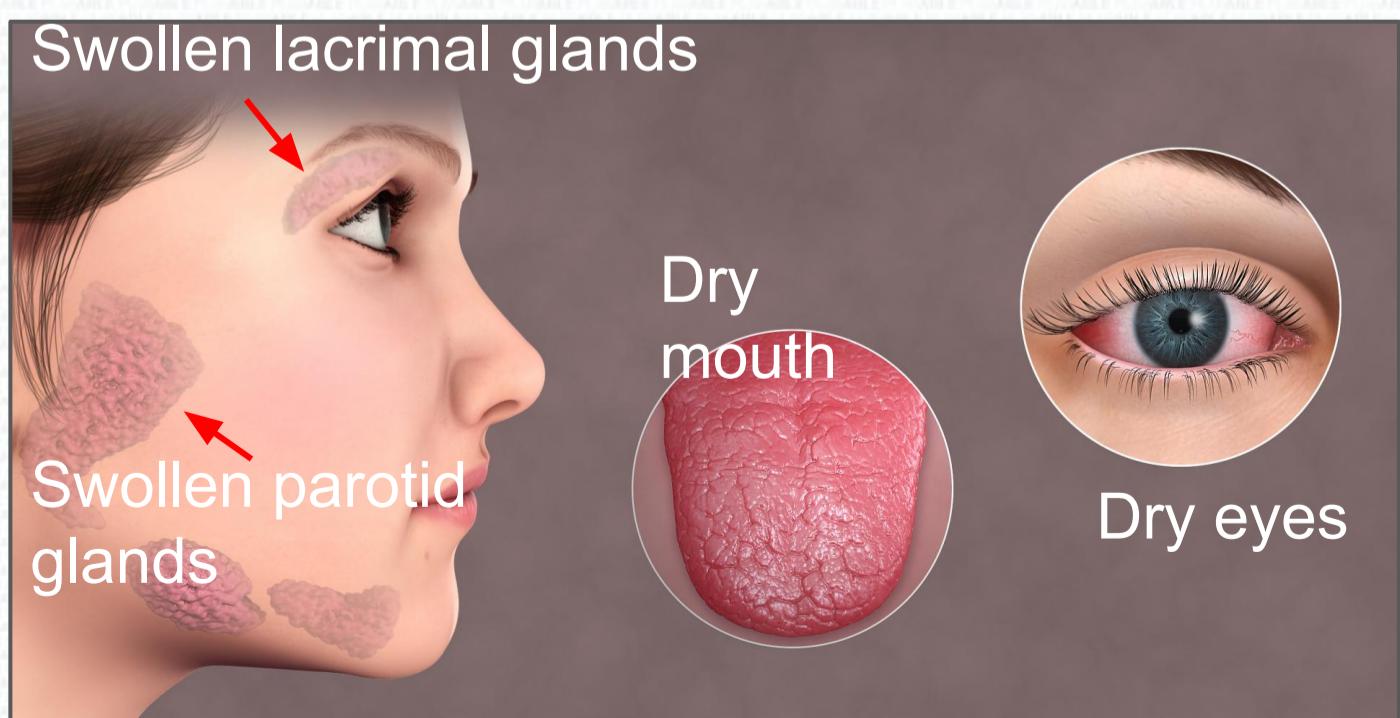
- Enlargement of parotid and lacrimal glands
- **Dry eyes** - Sandy feeling under the eyes
- **Dry mouth** - Difficulty swallowing
- Associated with: RA, SLE and scleroderma

Investigations

- **Schirmer's test:** Decreased tear production
- Anti Ro and Anti La antibody +
- RF +
- ANA +

Management

- Artificial tears
- Plenty of water and artificial saliva



What is Hypromellose

Hypromellose is a type of eye drop to help treat dry eyes.



Dry eyes can be experienced by lots of patients.

Examples include a patient who suffers with Sjögren's syndrome where Keratoconjunctivitis sicca (*dry eyes*) is seen

Systemic Sclerosis

Autoimmune disease with increased fibroblast activity → abnormal growth of connective tissues



C **Calcinosis**

- Rapid onset
- Involves trunk and limbs
- Thickening of skin
- Raynaud's phenomenon
- Scleroderma renal crisis

R **Raynaud's phenomenon**

E **Esophageal dysmotility**

S **Sclerodactyly**

T **Telangiectasia**

Investigations (only valid in diffuse scleroderma)

- Anti-Scl 70 (→diffuse)
- ANA (both)
- Anti-centromere antibody (→limited)

Seronegative Spondyloarthropathies

- Inflammatory rheumatic conditions with predominant involvement of axial and peripheral joints
- Associated with HLA-B27

Conditions

Remember the mnemonic “PAIR-B”

- Psoriatic arthritis
- Ankylosing spondylitis
- IBD associated arthritis
- Reiter's syndrome
- Behcet's disease

Behcet's Disease

Presentation

- Recurrent oral ulcers
- Recurrent genital ulcers
- Anterior or posterior uveitis
- Pathergy - Exaggerated skin injury after minor trauma

Treatment

- Topical corticosteroids

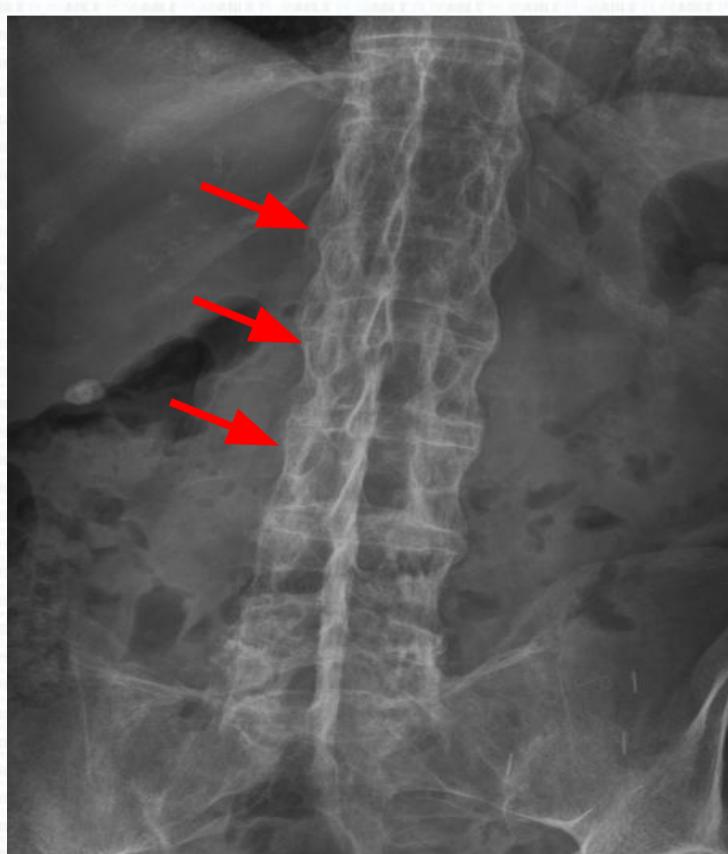
Ankylosing Spondylitis

- Back pain and morning stiffness
- Pain improves with physical activity, not with rest
- Tenderness of sacroiliac joint
- Anterior uveitis
- **X-ray** - Sacroiliitis
- Common in males < 30 years of age

Treatment

- Physiotherapy
- NSAIDs
- Oral corticosteroids
- Etanercept and adalimumab (severe cases)

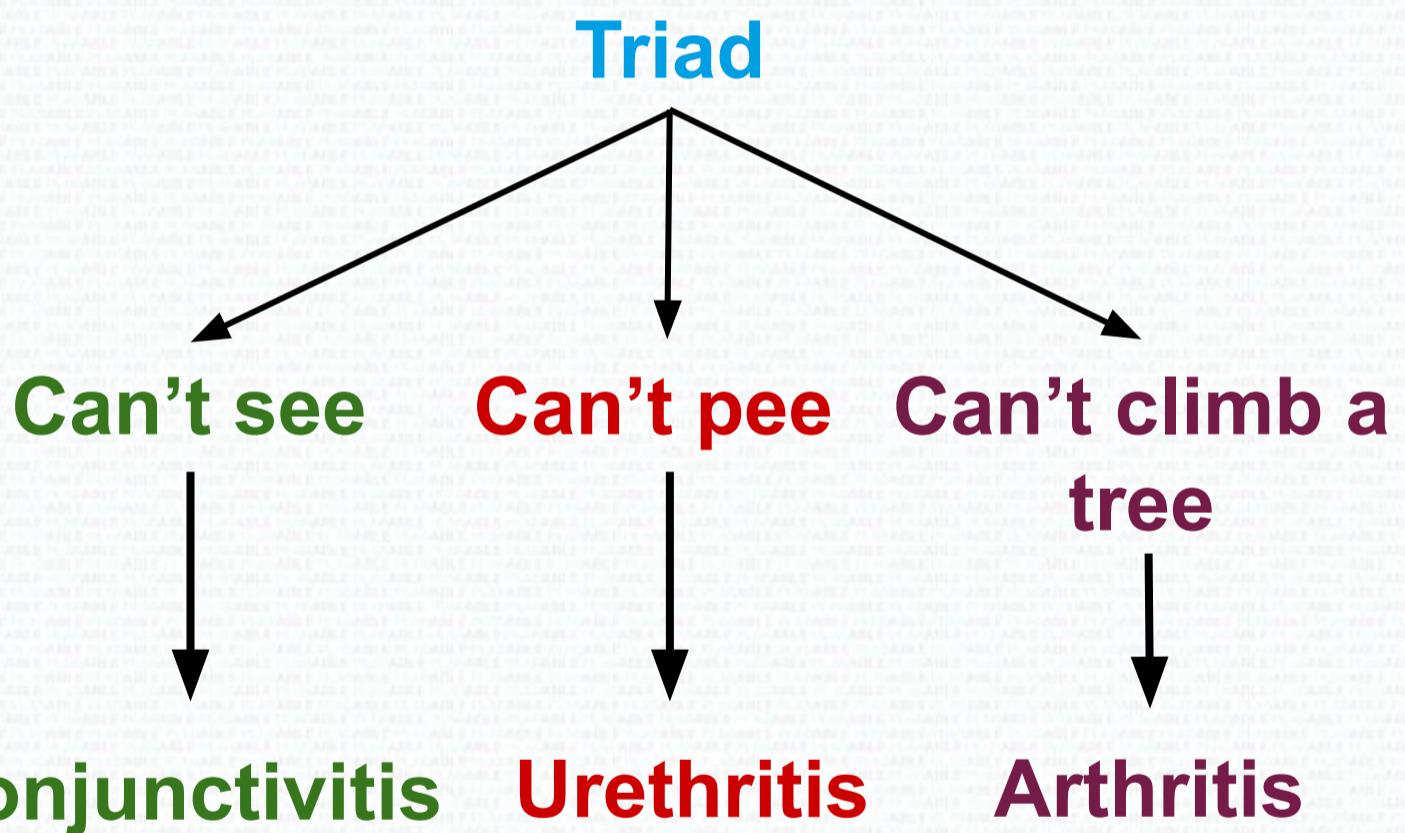
Complication: Fusion of the spine (bamboo spine)



Bamboo spine

Reiter's Syndrome

- Presents 2-4 weeks after a genitourinary or gastrointestinal infection
- C. trachomatis and C. pneumoniae are most common cause
- Also, known as **reactive arthritis**



Treatment

- Physiotherapy
- NSAIDs
- Aspiration of synovial effusions
- Oral corticosteroids

Gout

Risk factors:

- **Diet:** Meat and alcohol
- Tumour cell lysis (chemotherapy)
- Chronic kidney disease
- Diuretics

Most commonly involved joint is **first metatarsal phalangeal joint**

Diagnosis

Presence of MSU crystals in the synovial fluid

↑ Uric acid level in serum/urine (not definitive)

Treatment

Acute:

- NSAIDs
- Colchicine
- Corticosteroids



Prevention:

- **Allopurinol** and **febuxostat** - decreased formation of uric acid
 - Allopurinol → First line urate lowering agent
 - Febuxostat → Second-line urate lowering agent

The Inflamed DIP Joint



If stiff and painful, could it be gout or osteoarthritis?

Gout

Osteoarthritis

Acute course

Chronic course

History of thiazide use

Yellow substance under skin

Compare RH 1022 and RH 1101

Rheumatoid arthritis rarely affects the DIP joints

Osteomalacia

Brain trainer:

A patient has bone pain. Lab results show low serum calcium level, low serum phosphate level and raised alkaline phosphatase. What is the likely diagnosis?

→ **Osteomalacia**

De Quervain's Disease

Tenosynovitis of extensor pollicis brevis and abductor pollicis longus involved in the movement of the thumb

Presentation

- Pain and swelling at the radial side of the wrist

Risk factors

- Pregnancy
- Repetitive movements
- Rheumatic disease

Treatment

- Rest and finger splinting
- NSAIDs
- Local steroid injection

Note: Not to be confused with **De Quervain's thyroiditis** or subacute thyroiditis caused by viral infection causing hyperthyroidism and tenderness of thyroid gland

Churg Strauss Syndrome

Eosinophilic granulomatosis with polyangiitis

Presentation

- Asthma
- Nasal polyps
- Purpura
- Glomerulonephritis

Investigations

- Eosinophilia
- P- ANCA
- ↑ IgE
- Biopsy - Vasculitis with extravascular eosinophils

Remember the mnemonic “PAVE”

- P-ANCA
- Asthma
- Vasculitis
- Eosinophilia

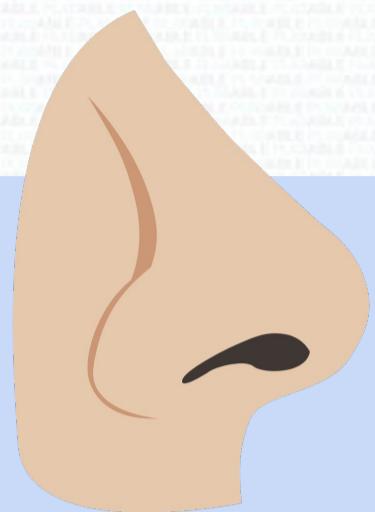
Treatment

- Steroids

Granulomatosis With Polyangiitis (Wegener's Granulomatosis)

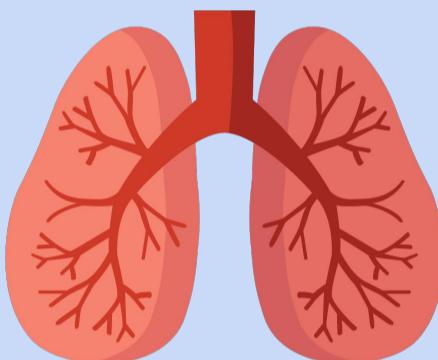
- **Upper respiratory tract**

- Nasal perforation
- Chronic sinusitis
- Epistaxis



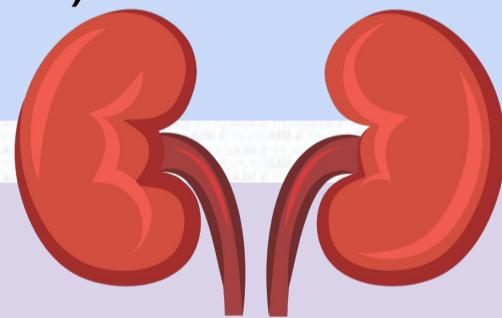
- **Lower respiratory tract**

- Haemoptysis
- Dyspnoea



- **Renal**

- Haematuria (glomerulonephritis)



Investigations

- C-ANCA
- RBC casts in urine
- Biopsy of affected site

Treatment

- Unlikely to be asked

Granulomatosis With Polyangiitis (Wegener's Granulomatosis)

Look for keywords in a patient who has
haemoptysis

Some key words

Saddle-shaped nose

Crusting around nose

Subcutaneous nodules
on the elbow

Mnemonic for investigation → **World Cup**

Wegener's → C-ANCA



Rheumatoid Arthritis

Presentation

- **Bilateral and symmetrical polyarthritis**
- Joint stiffness in the morning
- Involvement of proximal interphalangeal, metacarpophalangeal, wrist, knee and cervical spine joints
- Swan neck and Boutonnière's deformity
- Rheumatoid nodules

Investigations

- ↑ ESR / CRP
- Normocytic normochromic anaemia
- + Rheumatoid factor
- + Anti-CCP (more specific)

Management

- NSAIDs (Acute)
- Corticosteroids (flare)
- DMARDs:
 - Methotrexate
 - Sulfasalazine

DMARDs slow down disease progression and prevent deformities



Sarcoidosis

Chronic inflammatory condition characterised by the formation of non-caseating granuloma

Presentation

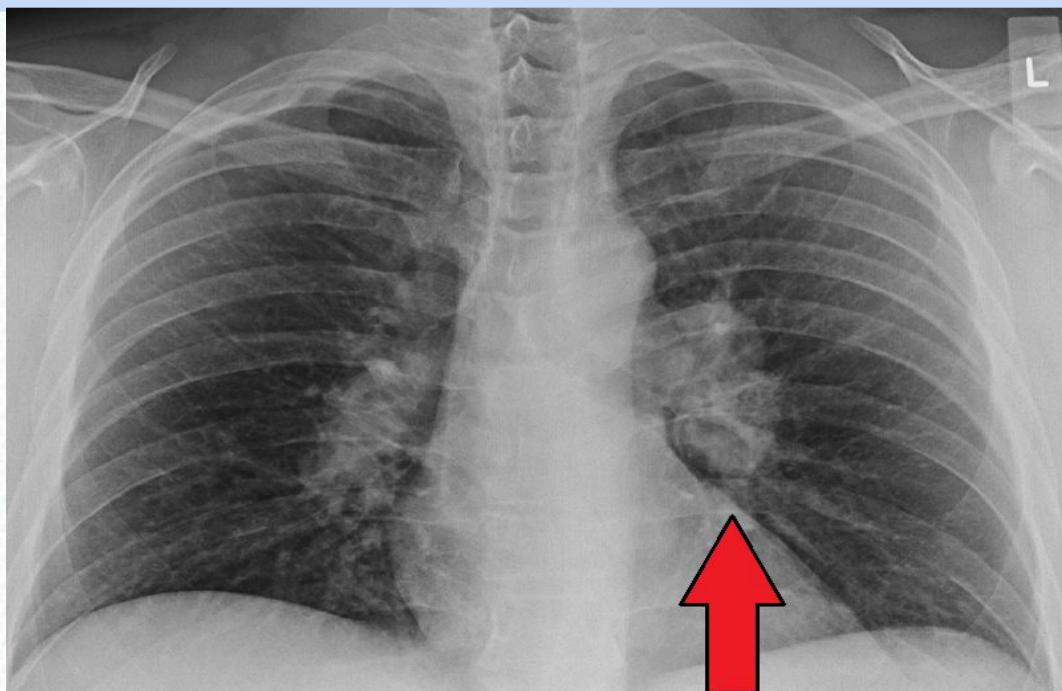
- Erythema nodosum
- Polyarthralgia
- Fever and night sweats
- Lupus pernio
- Uveitis

Investigation

- Hypercalcaemia
- ↑ ESR
- Chest X-ray - bilateral hilar lymphadenopathy

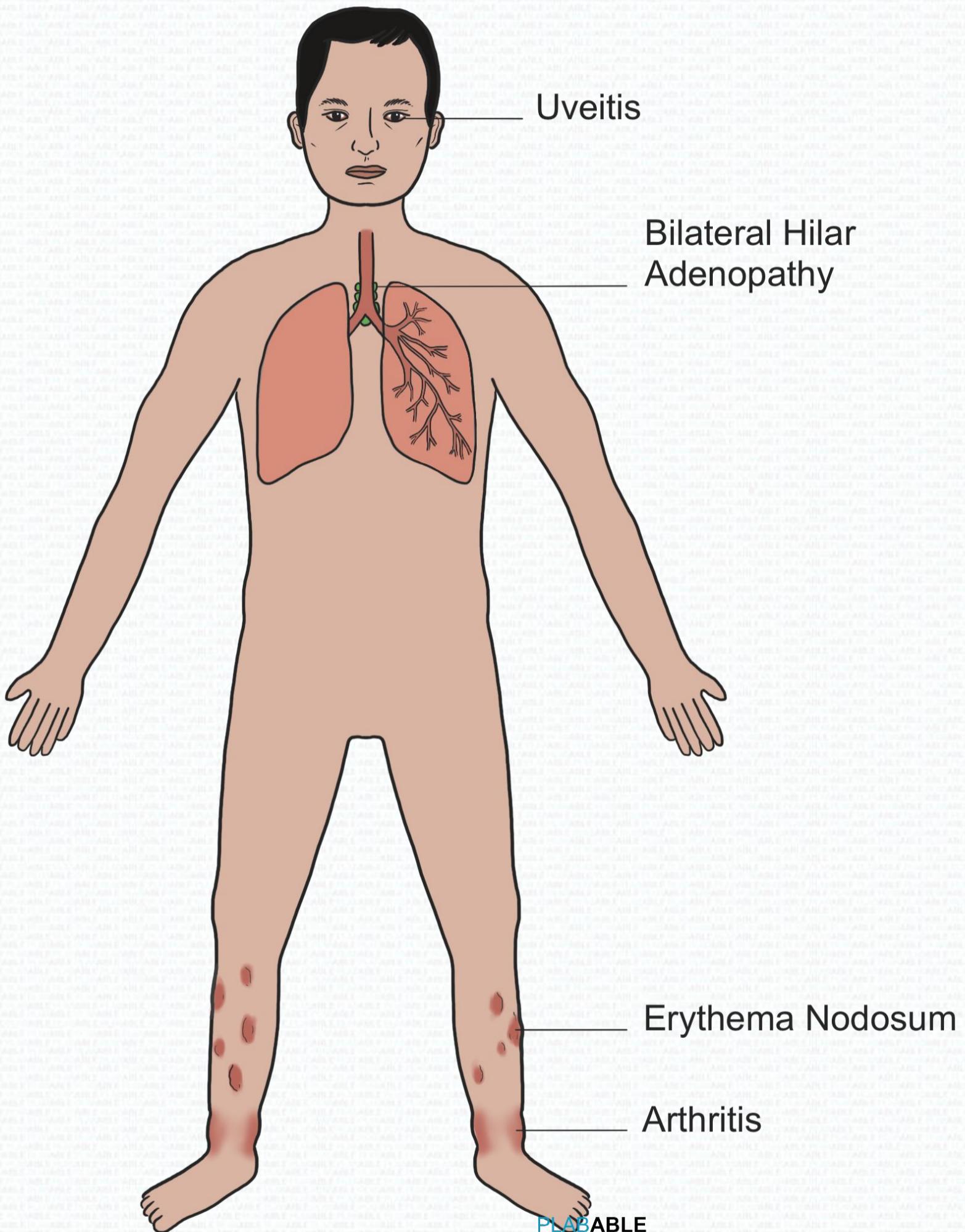
Treatment

- Oral corticosteroids



Sarcoidosis - Hilar adenopathy

Sarcoidosis



Sarcoidosis

PLABABLE

Syndromes With Sarcoidosis

Lofgren's syndrome

- Bilateral hilar adenopathy
- Erythema nodosum
- Polyarthralgia

Mikulicz syndrome

- Enlargement of parotid and lacrimal glands
- Dry mouth
- Due to sarcoidosis, tuberculosis or lymphoma

Note: Not to be confused with **Löffler's syndrome** which is a disease in which eosinophils accumulate in the lung in response to a parasitic infection

Sarcoidosis

A multisystem disorder of unknown aetiology characterised by **non-caseating granulomas**.

Common in young adult and africans

Features

- Erythema nodosum
→ Tender, red nodule over shins
- **Bilateral hilar lymphadenopathy**
- Polyarthralgia
- Hypercalcaemia
- Fever
- Elevated ACE levels

*If the question presents with **any of the 2 features** above, consider sarcoidosis.*



Associated
with



Lofgren's syndrome: (Acute form):

- Bilateral hilar lymphadenopathy
- Erythema nodules
- Fever
- Polyarthralgia

Heerfordt's syndrome

(Uveoparotid fever):

- Parotid enlargement
- Fever
- Uveitis secondary to sarcoidosis

Sarcoidosis

Brain trainer:

A previously healthy woman develops pain and swelling on both knees and ankles with a nodular rash over her shins. A chest-xray was ordered showing bilateral hilar lymphadenopathy. Which investigation will provide a definitive diagnosis?

→ Biopsy of suspected tissue

Sarcoidosis is definitively diagnosed by the presence of noncaseating granulomas on biopsy.

Raynaud's Phenomenon

Raynaud's phenomenon is episodic vasospasm of the arteries or arterioles in the extremities which leads to colour change.

Fingers or toes which are exposed to cold usually turn pale, followed by cyanosis and/or rubor.

Treatment → Nifedipine

Rheumatic Fever

Inflammation post strep throat

Can result in:

- **Arthritis** → Large joints like knees, ankles, wrist, elbows
- **Rheumatic heart disease** → Commonly mitral valve stenosis
- **Chorea** → Sydenham's chorea
- **Skin manifestation** → Subcutaneous nodules and erythema marginatum

Investigations

- ESR and CRP are high
- ASO high
- Throat cultures for group A strep
- Echocardiogram

(Note: ESR more sensitive than ASO but ASO more specific than ESR)

This means that if a question ask you which is more likely to be abnormal → ESR would be a better pick compared to ASO

Treatment

- Penicillin

Revised Jones Criteria

This is used to diagnose acute rheumatic fever

Diagnosis requires:

Evidence of recent strep infection AND either

- 2 major criteria or
- 1 major + 2 minor criteria

Major criteria

- Polyarthritis
- Carditis
- Chorea
- Erythema marginatum
- Subcutaneous nodules

Minor criteria

- Fever
- Arthralgia
- Previous rheumatic fever or rheumatic heart disease
- Raised erythrocyte sedimentation rate or C reactive protein
- Prolonged PR interval on ECG

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