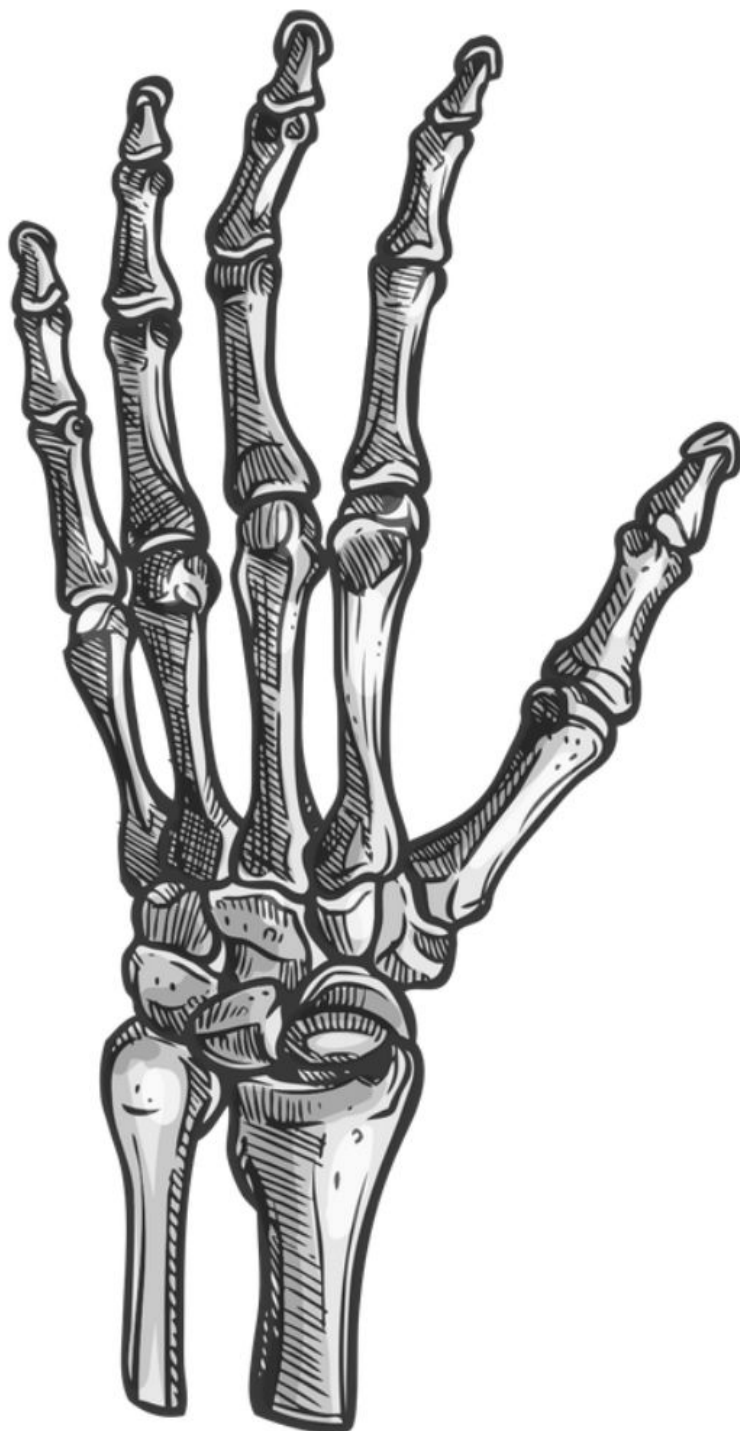


# PLABABLE

## GEMS

VERSION 3.3

# RHEUMATOLOGY



# Autoantibodies

| Disease                              | Autoantibody  |
|--------------------------------------|---|
| SLE                                  | <ul style="list-style-type: none"><li>● Anti-ds DNA (specific)</li><li>● Antinuclear antibody (sensitive)</li></ul> |
| Drug induced lupus                   | Anti-histone antibody   |
| Diffuse scleroderma                  | Anti-scl70  |
| Limited scleroderma (CREST syndrome) | Anti-centromere   |
| Polymyositis                         | Anti-Jo1  |
| Sjogren's syndrome                   | Anti-Ro<br>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<br>(remember as WBC)   |
| Coeliac disease          | <ul style="list-style-type: none"><li>• Anti-tissue transglutaminase</li><li>• Anti-gliadin</li><li>• Anti-endomysial</li></ul> |
| Graves disease           | TSH-receptor antibody   |
| Rheumatoid arthritis     | <ul style="list-style-type: none"><li>• Rheumatoid factor (initial)</li><li>• Anti-CCP (most specific)</li></ul>                |

# Systemic Lupus Erythematosus

Autoimmune disorder with multi-organ involvement

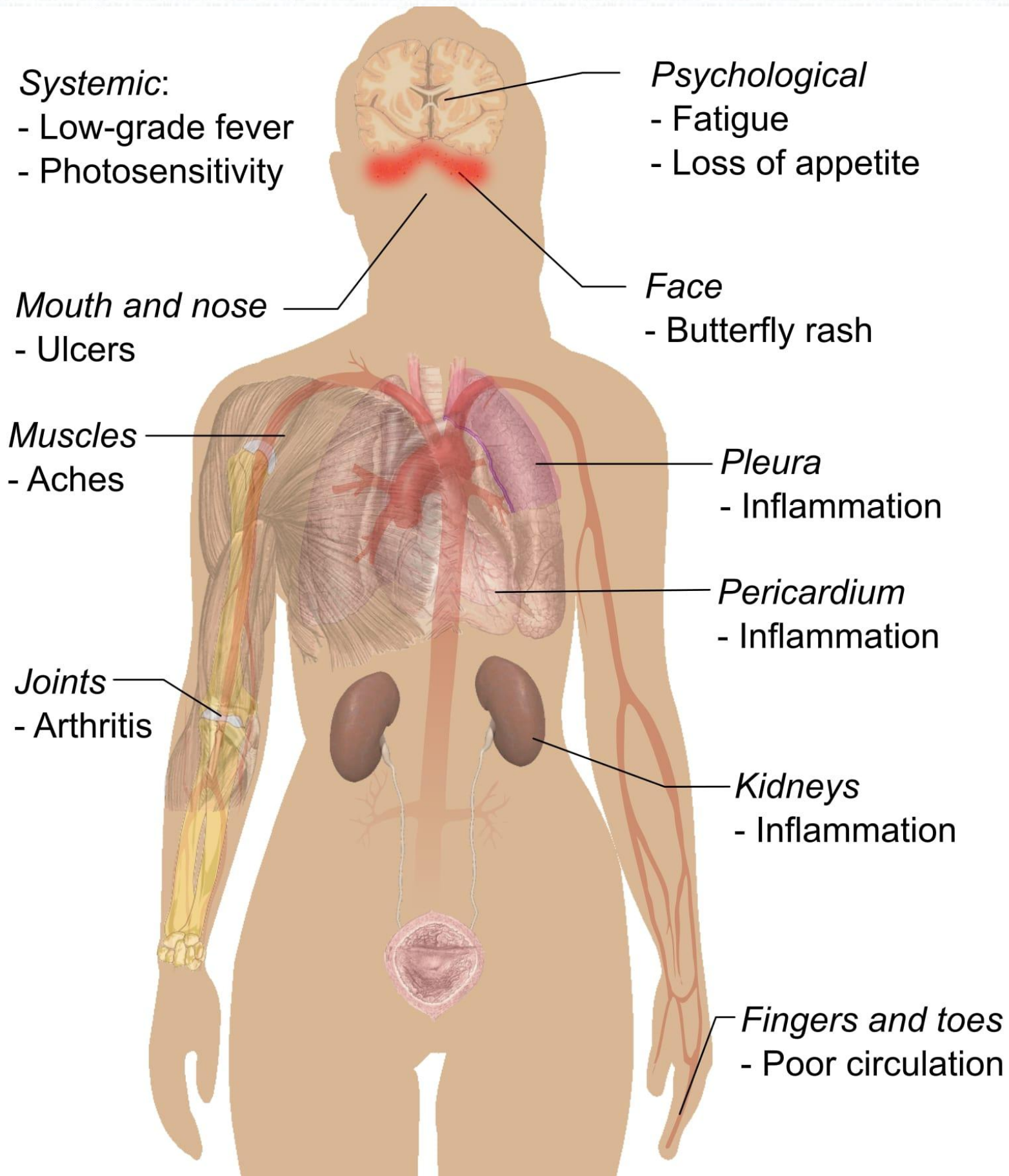
| Organ involved | Presentation   |
|----------------|--|
| General        | <ul style="list-style-type: none"><li>● <b>Fatigue</b></li><li>● Fever</li><li>● Mouth ulcers</li></ul>  |
| Skin           | <ul style="list-style-type: none"><li>● <b>Malar (butterfly) rash</b> on face</li><li>● Photosensitivity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



Butterfly rash



# Systemic Lupus Erythematosus



# 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





# Dermatomyositis



Source: IMACS

## Investigation

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

## Treatment

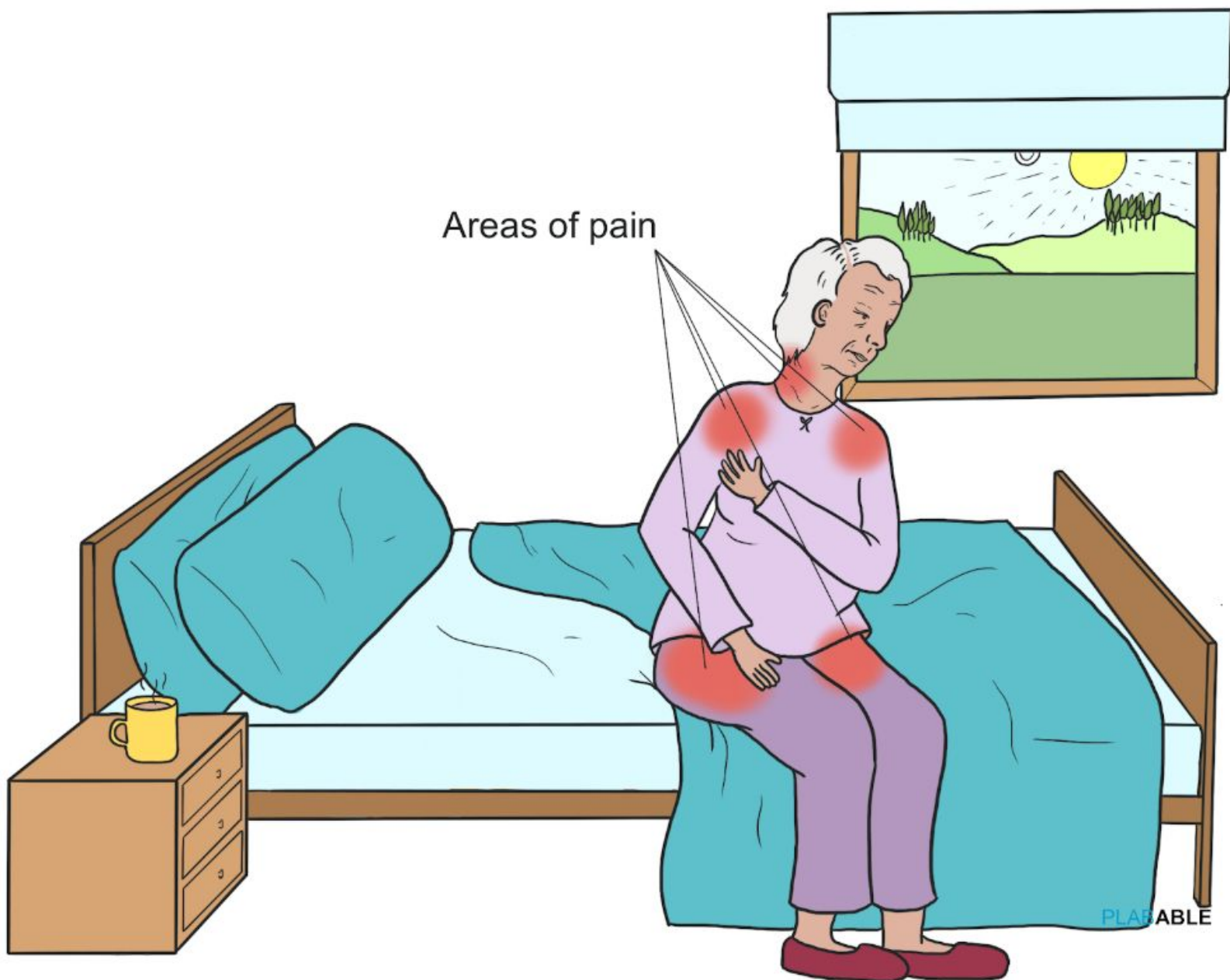
- Steroids
- Sun-blocking agents



# 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

- $\uparrow$  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 +  $\uparrow$ CK

**Polymyalgia** - pain > weakness + normal CK

# Inflammatory Conditions Involving Muscles

This card is to recap what is already learnt!

## Polymyositis

## Dermatomyositis

Weakness of muscles

Skin manifestations

- Gottron's papules
- Heliotrope rash
- Shawl sign

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



PLABABLE



# Giant Cell (Temporal) Arteritis

## Presentation

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

## Investigation

- ↑ 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



## Limited scleroderma

**C** Calcinosis  
**R** Raynaud's phenomenon  
**E** Esophageal dysmotility  
**S** Sclerodactyly  
**T** Telangiectasia

## Diffuse scleroderma

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

## 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

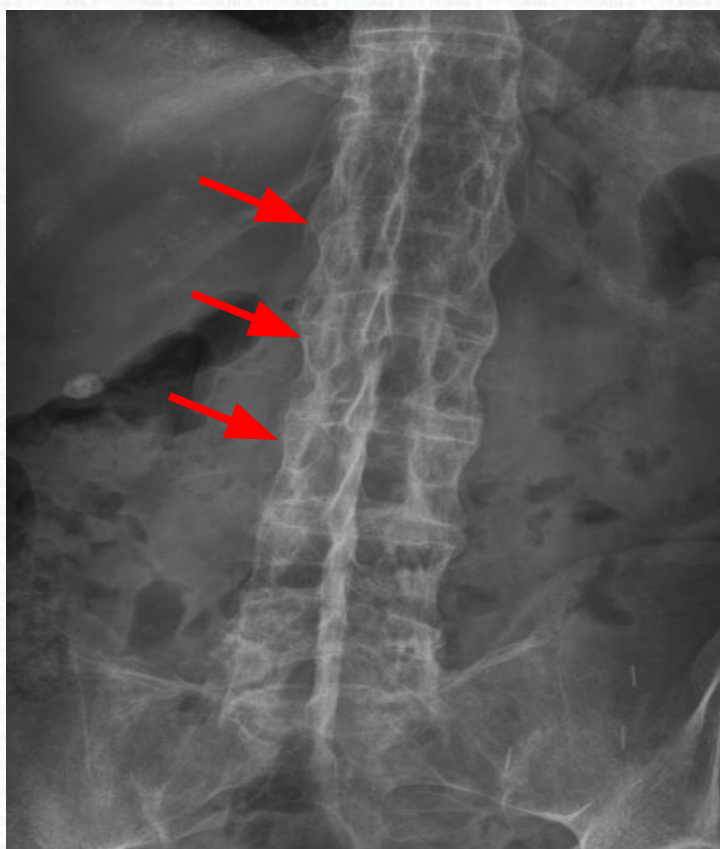
# Ankylosing Spondylitis

- 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)

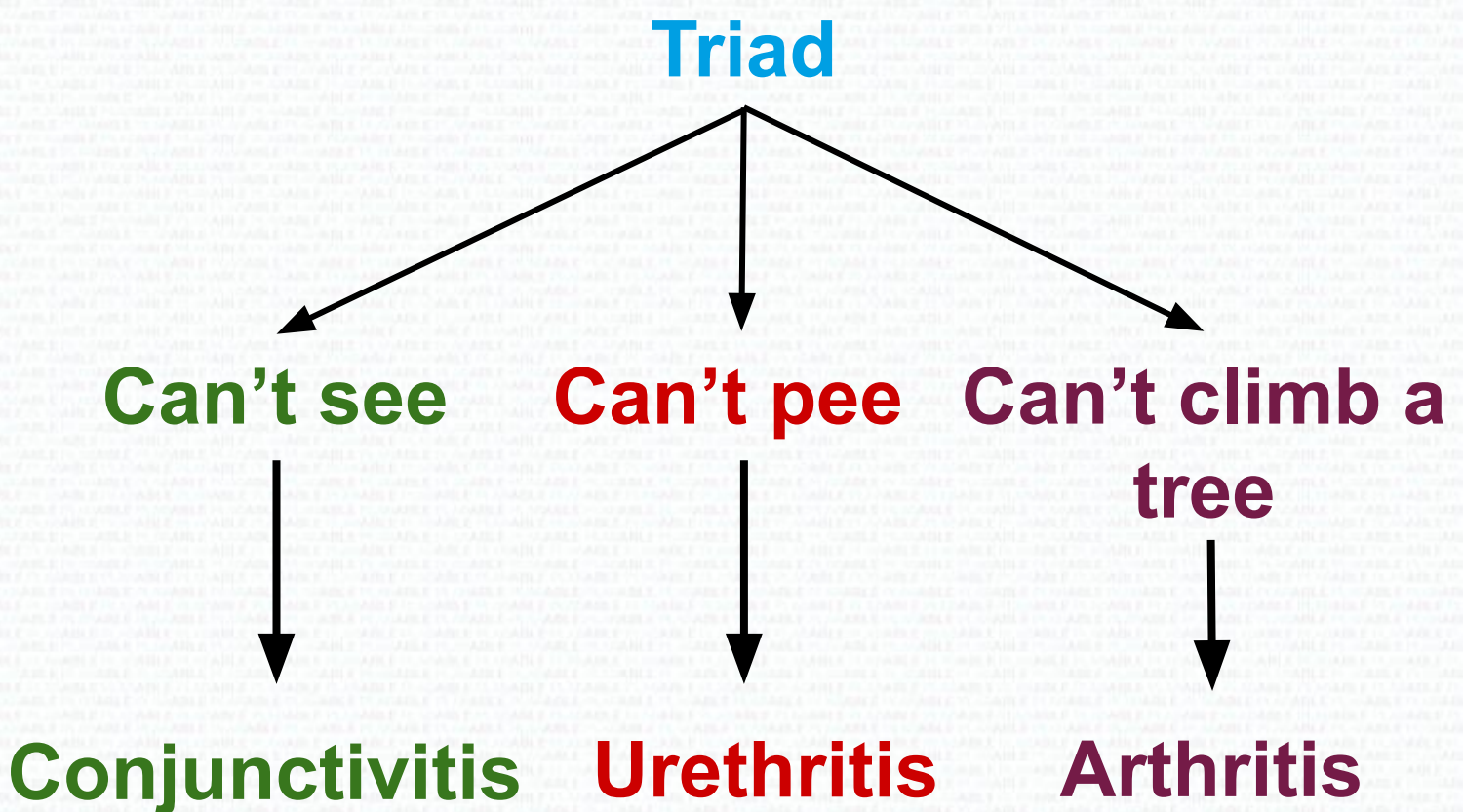
**Complication:** Fusion of the spine (bamboo spine)



**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
- **A**sthma
- **V**asculitis
- **E**osinophilia

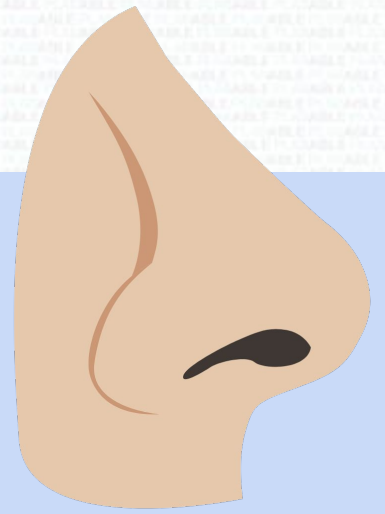
## Treatment

- Steroids

# Granulomatosis With Polyangiitis (Wegener's Granulomatosis)

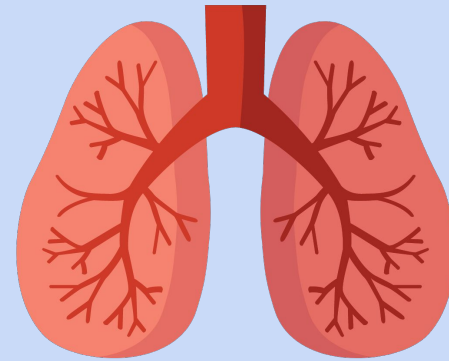
- **Upper respiratory tract**

- Nasal perforation
- Chronic sinusitis
- Epistaxis



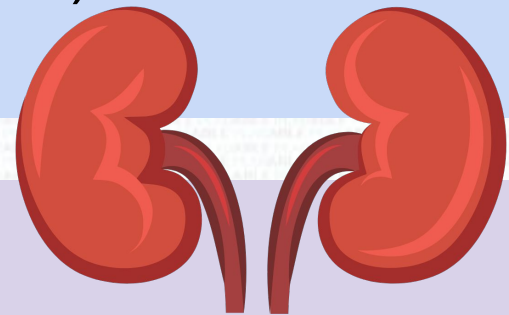
- **Lower respiratory tract**

- 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

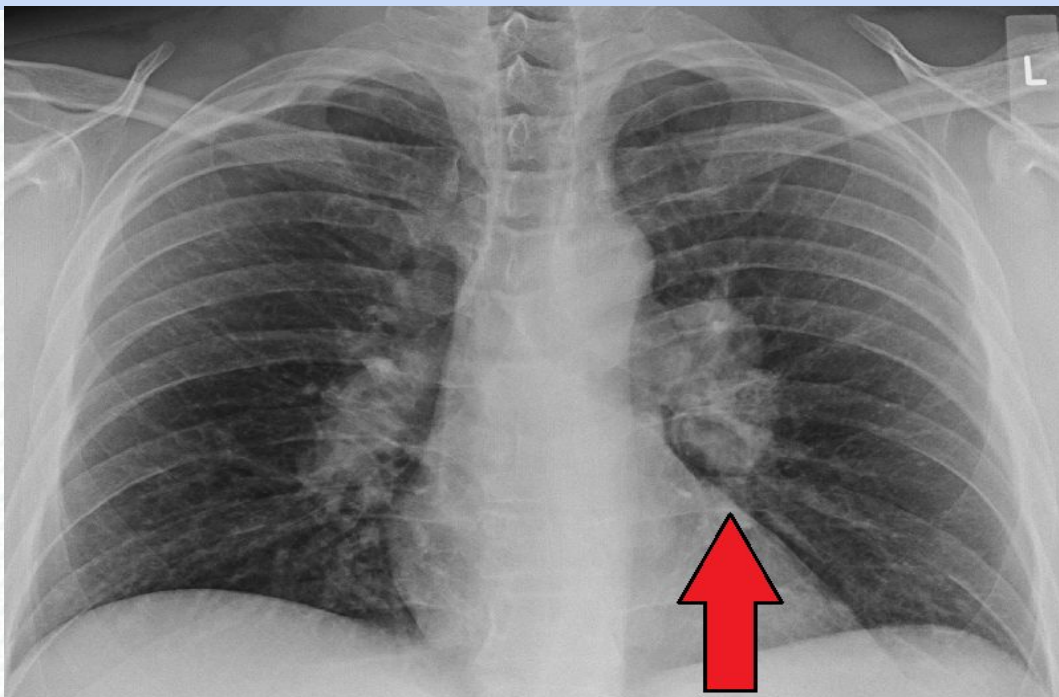
- Erythema nodosum
- Polyarthralgia
- Fever and night sweats
- Lupus pernio
- Uveitis

## Investigation

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

## Treatment

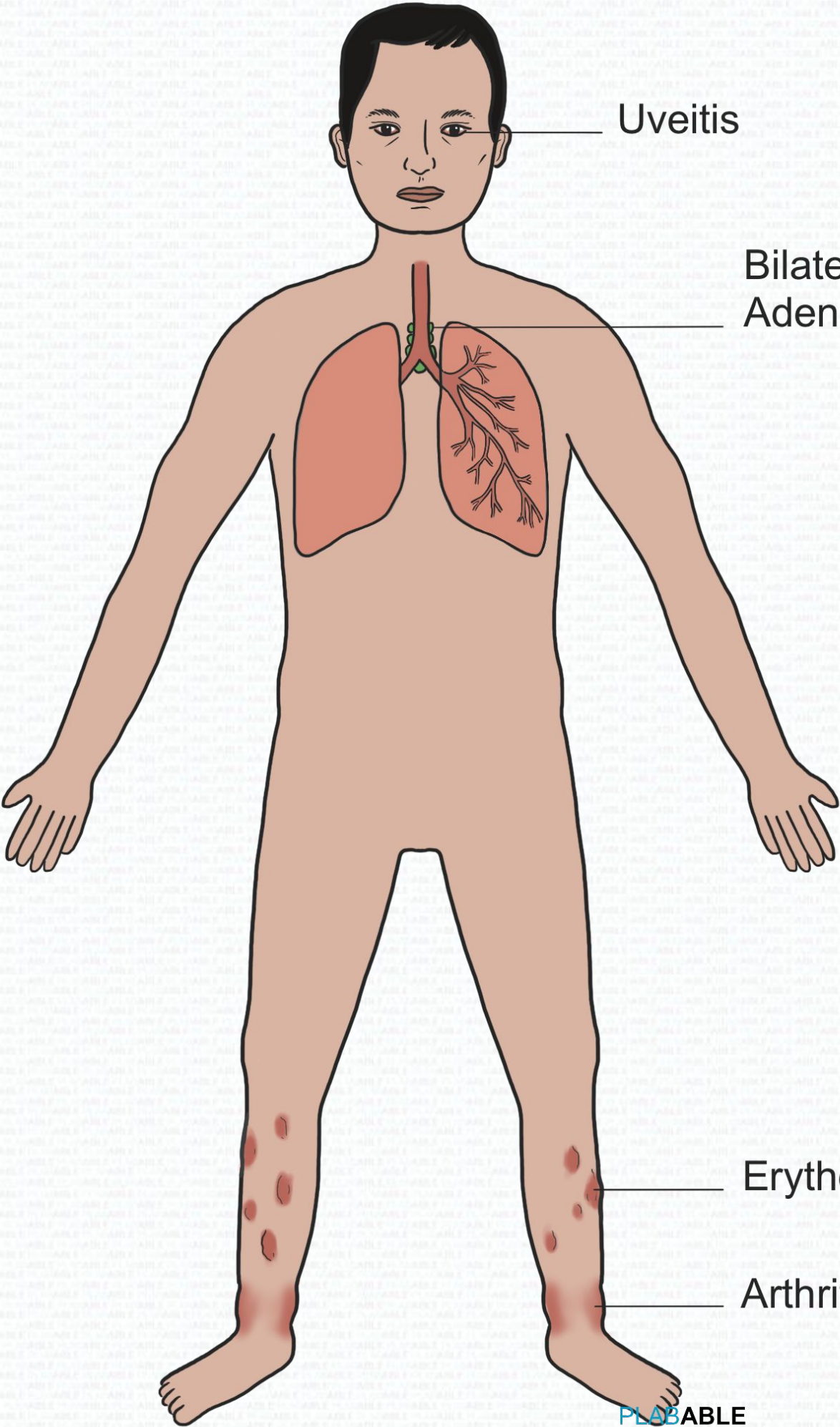
- Oral corticosteroids



**Sarcoidosis - Hilar adenopathy**



# Sarcoidosis



Uveitis

Bilateral Hilar  
Adenopathy

Erythema Nodosum

Arthritis

PLABABLE

Sarcoidosis

# 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 questions 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**

- Polyarthrititis
- 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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