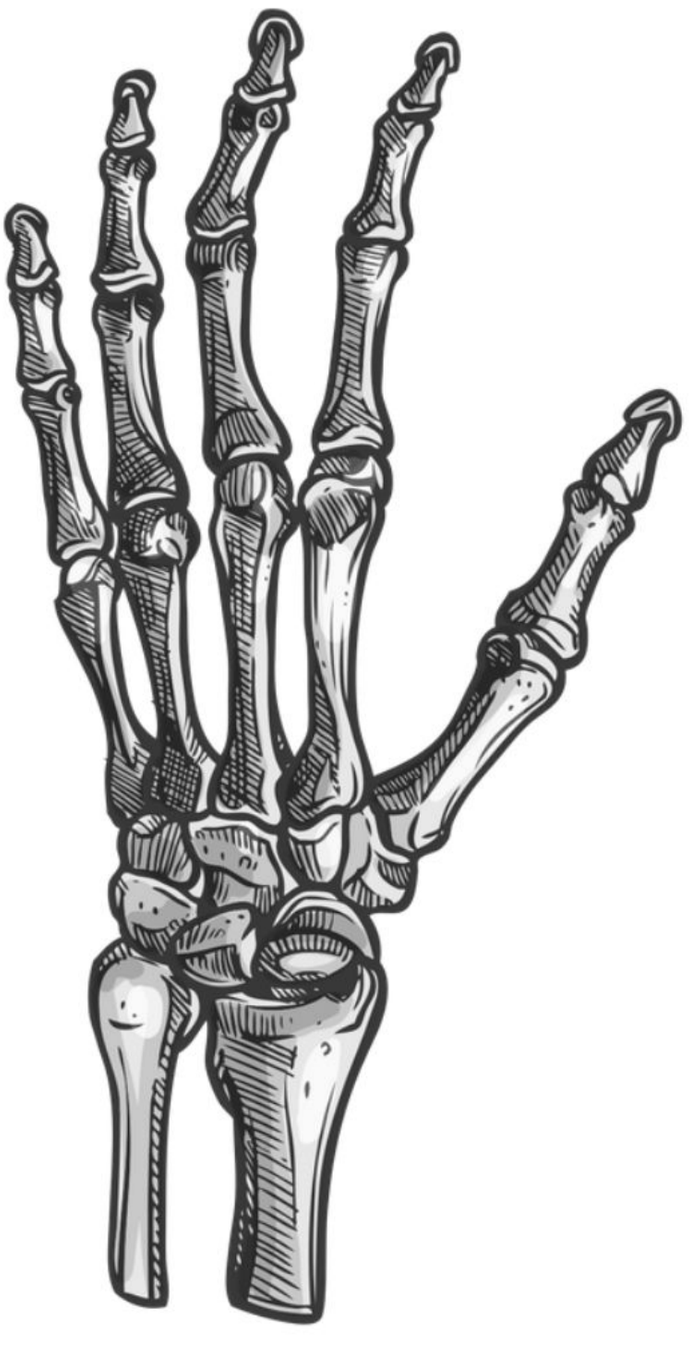
PLABABLE

GEMS

VERSION 1.3

RHEUMATOLOGY



Autoantibodies

Disease	Autoantibody
SLE	• Anti-ds DNA (specific) • Antinuclear antibody (sensitive)
Drug induced lupus	Anti-histone antibody
Systemic sclerosis	Anti-scl70
CREST syndrome	Anti-centromere
Polymyositis	Anti-Jo1
Sjogren's syndrome	Anti-Ro Anti-La
Primary biliary cirrhosis	Anti-mitochondrial
Autoimmune hepatitis	Anti-smooth muscle

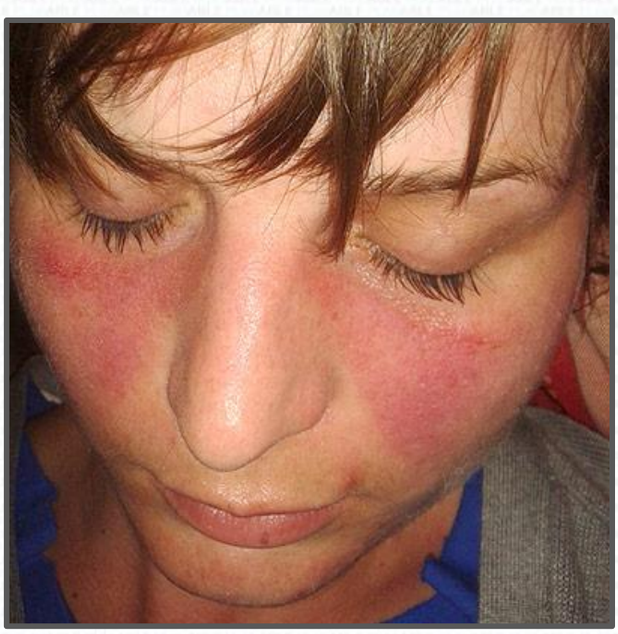
Autoantibodies

Disease	Autoantibody
Churg Strauss syndrome	p-ANCA
Wegener's granulomatosis	c-ANCA (remember as WBC)
Coeliac disease	• Anti-tissue transglutaminase • Anti-gliadin • Anti-endomysial
Graves disease	TSH-receptor antibody
Rheumatoid arthritis	Antinuclear antibody

Systemic Lupus Erythematosus

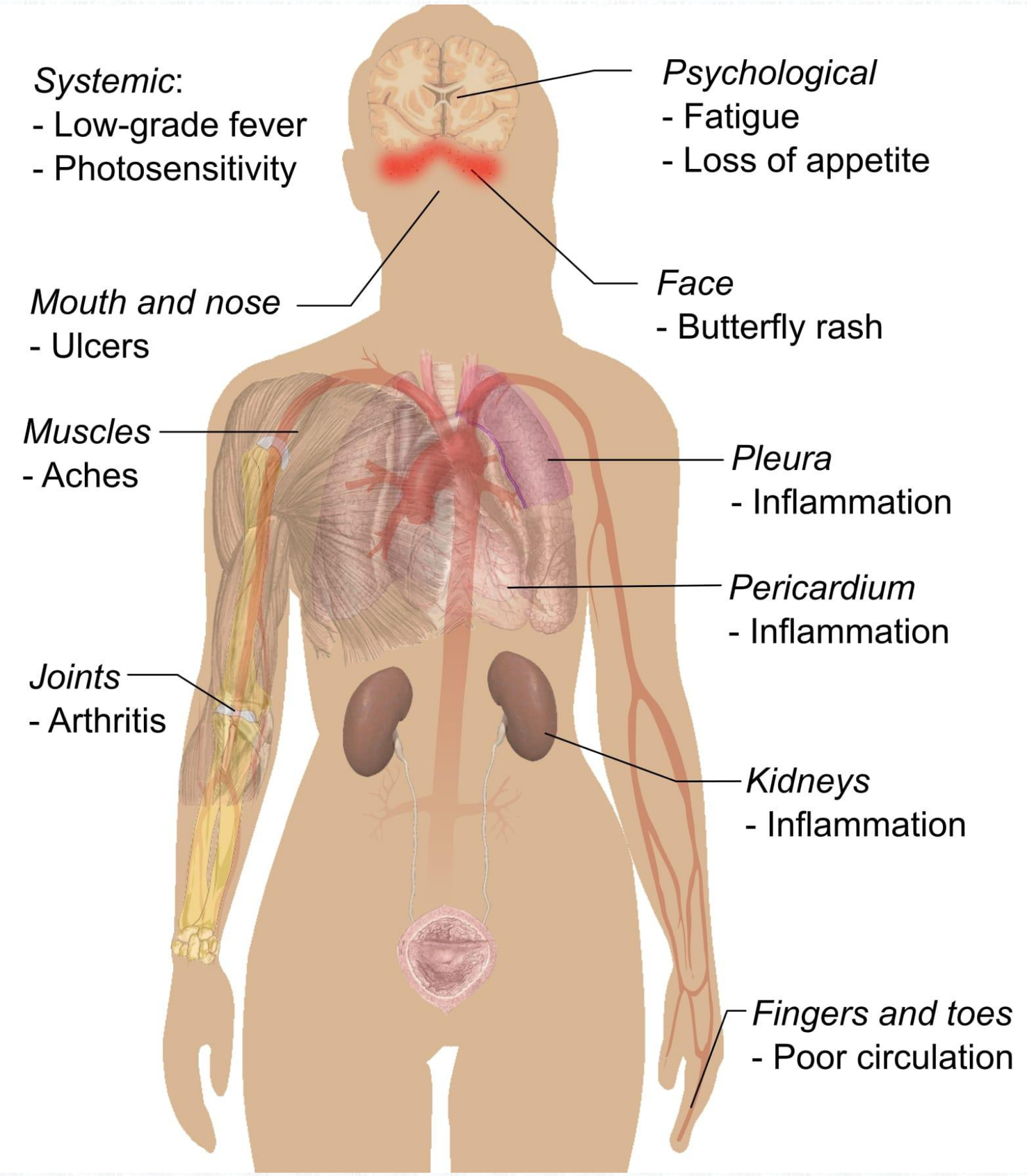
Autoimmune disorder with multi-organ involvement

Organ involved	Presentation
General	• **Fatigue** • Fever • Mouth ulcers
Skin	• **Malar (butterfly) rash** on face • Photosensitivity (discoid rash) • Raynaud's phenomenon
Joints	• **Arthralgia** • Peripheral and symmetrical polyarthritis
Cardiovascular	• Pericarditis • Libman-Sacks endocarditis
Respiratory	Pleuritis
Renal	**Glomerulonephritis** (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

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

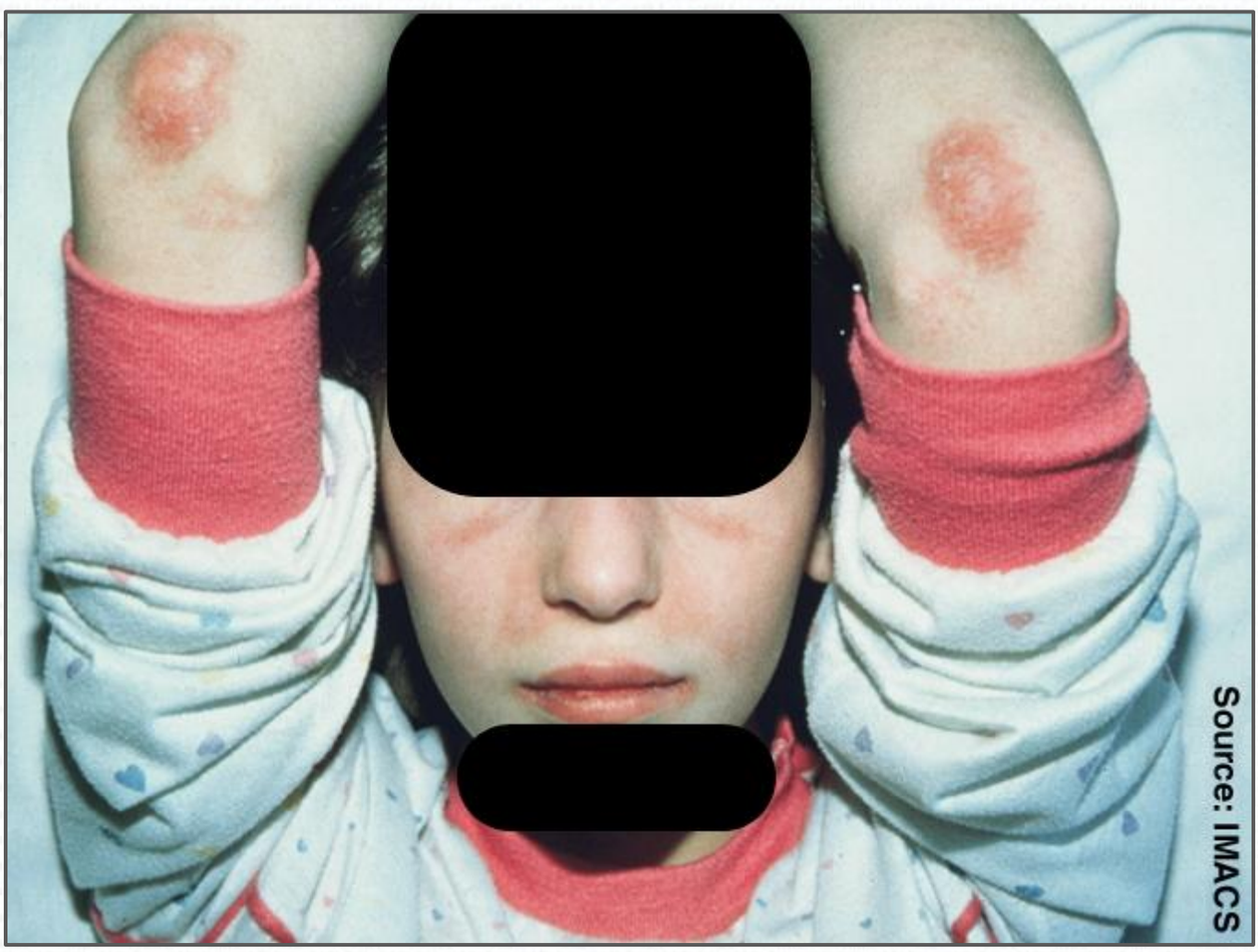
Treatment

- Steroids

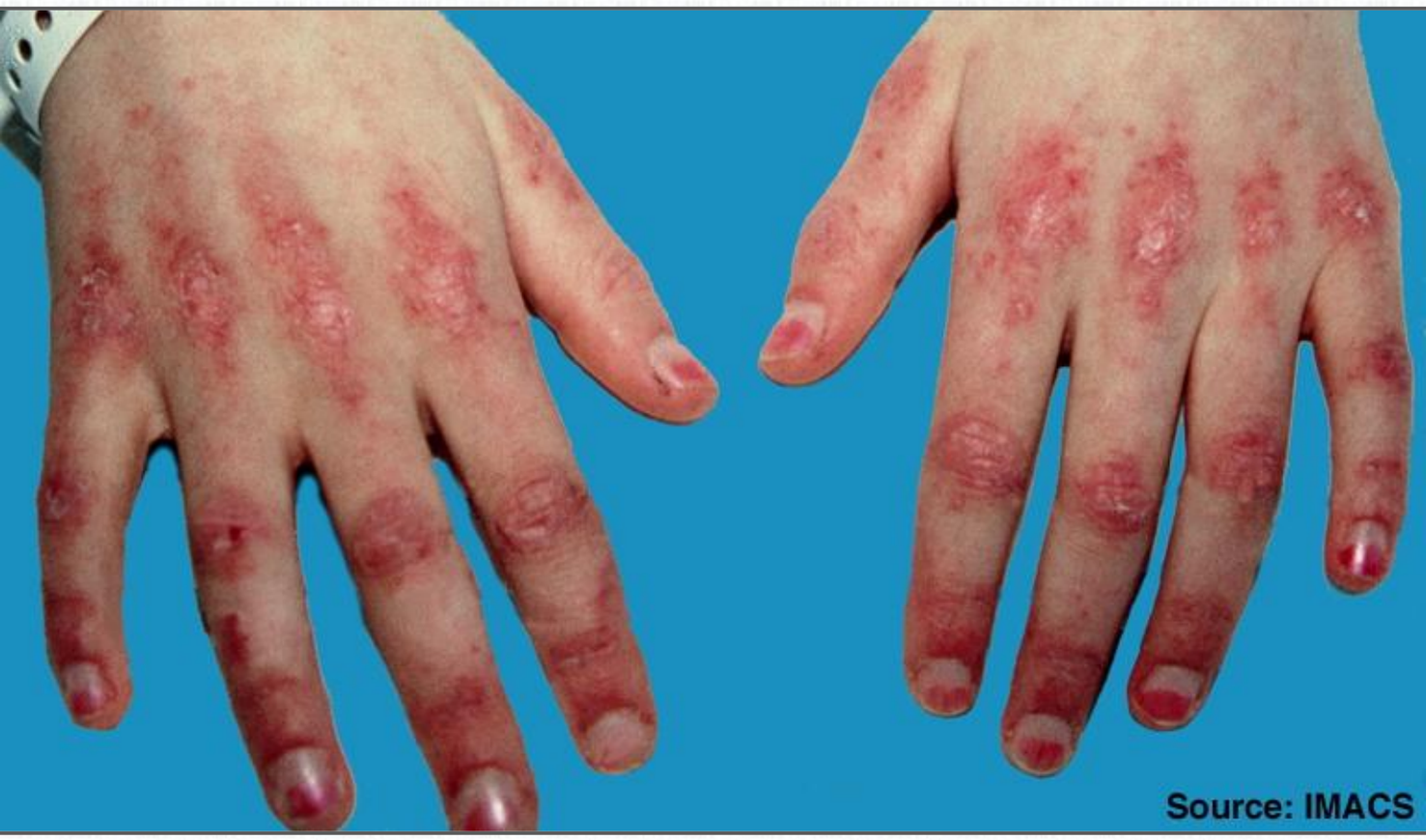
Dermatomyositis

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



Investigation

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

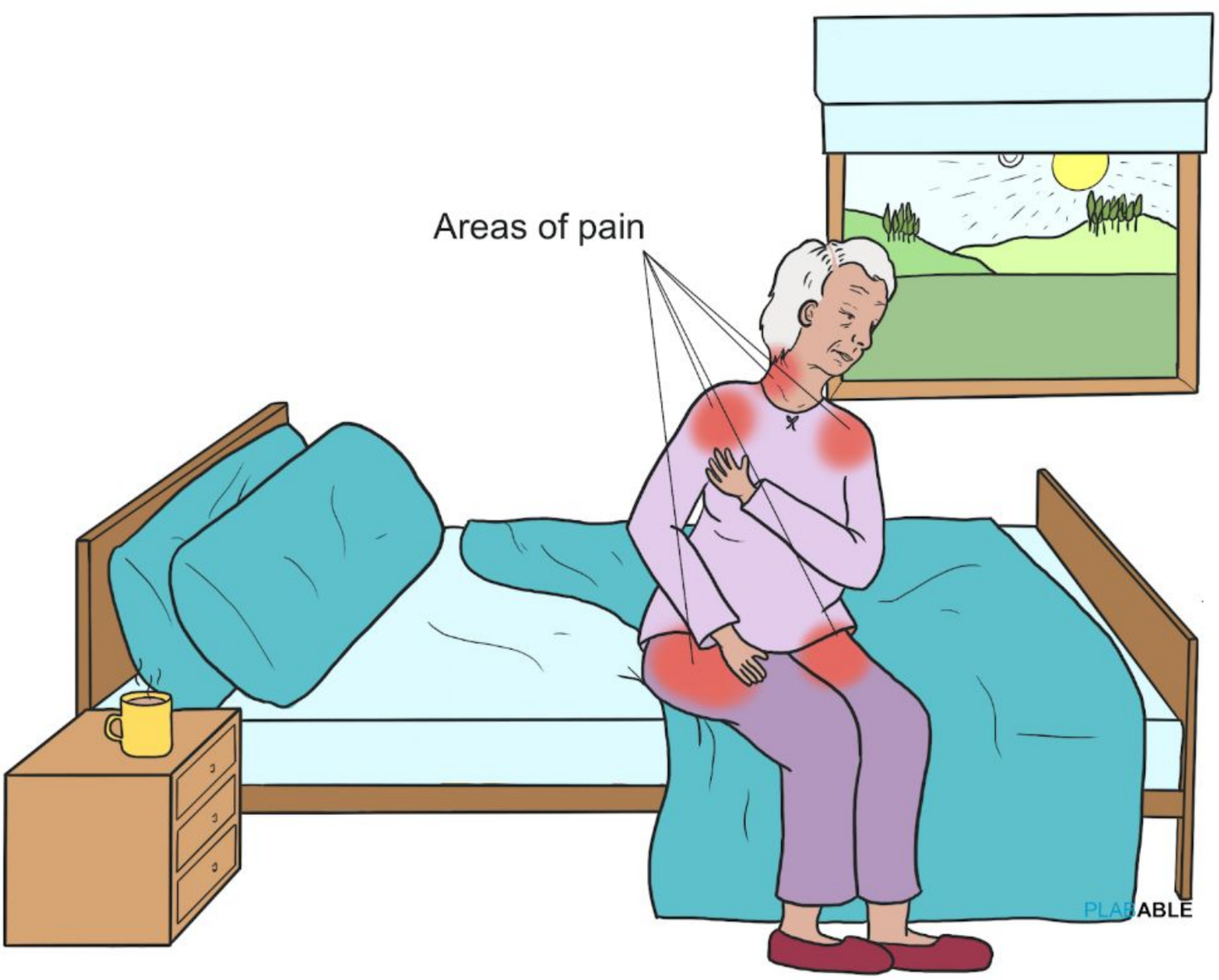
Treatment

- 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

Giant Cell or 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)

Sjogren's Syndrome

Autoimmune disorder affecting exocrine glands

Presentation:

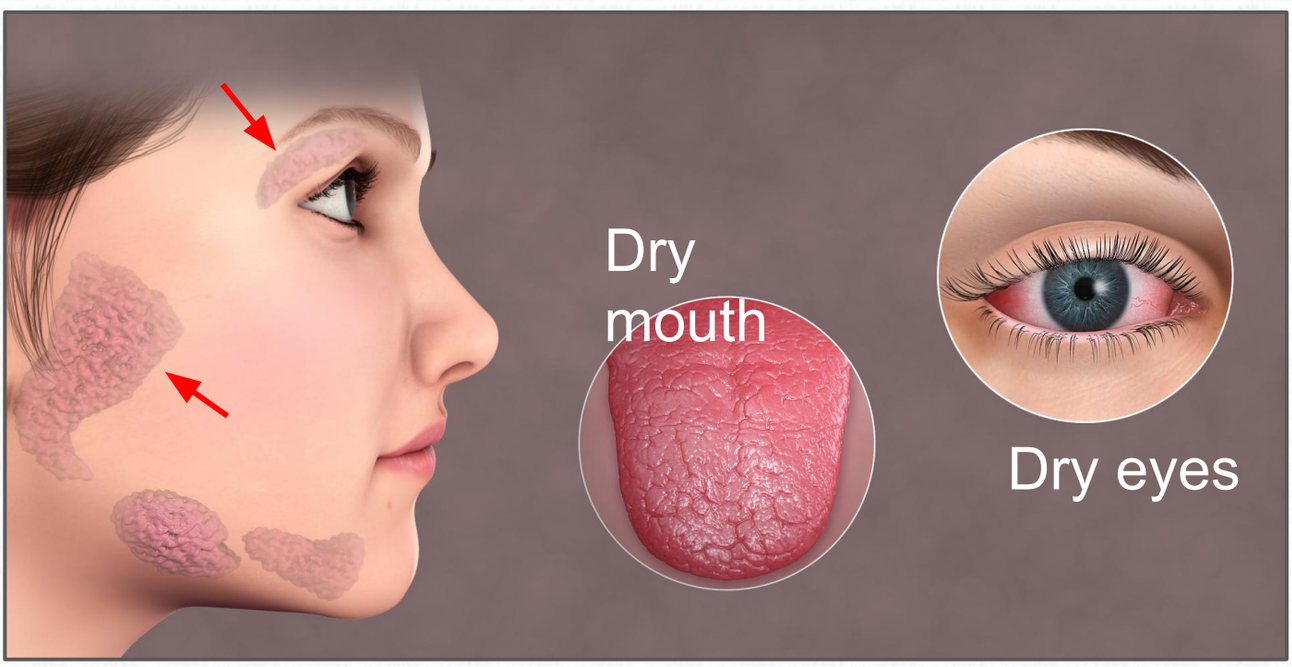
- Enlargement of parotid and lacrimal glands
- **Dry eyes** - Sandy feeling under the eyes
- **Dry mouth** - Difficulty swallowing
- Associated with: RA, SLE and scleroderma

Investigations

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

Management

- Artificial tears
- Plenty of water and artificial saliva



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 (→diffuse)
- Anti-centromere antibody (→limited)

Seronegative Spondyloarthropathies

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

Conditions

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

Behcet’s Disease

Presentation

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

Treatment

- Topical corticosteroids

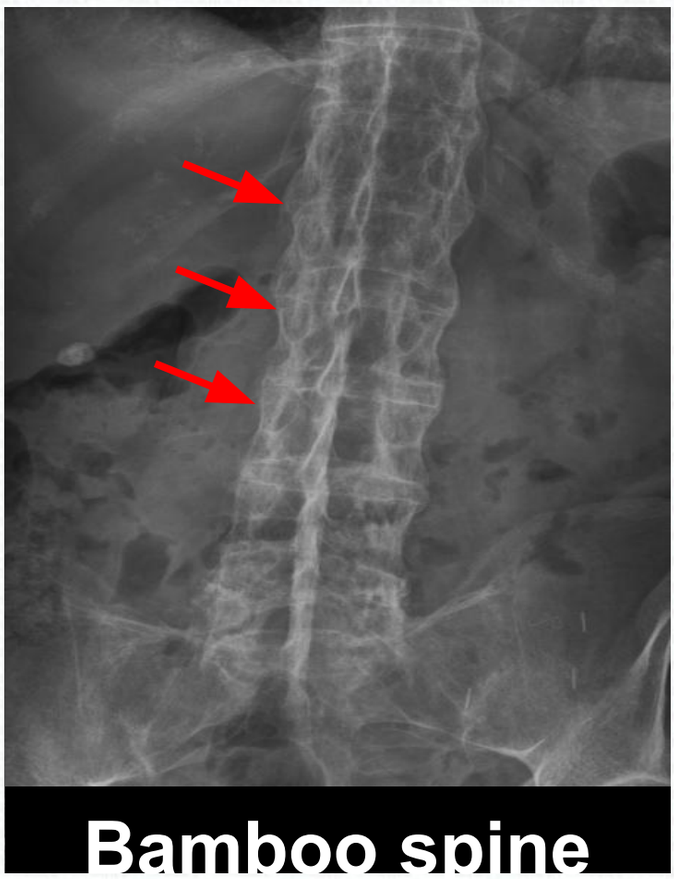
Ankylosing Spondylitis

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

Treatment

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

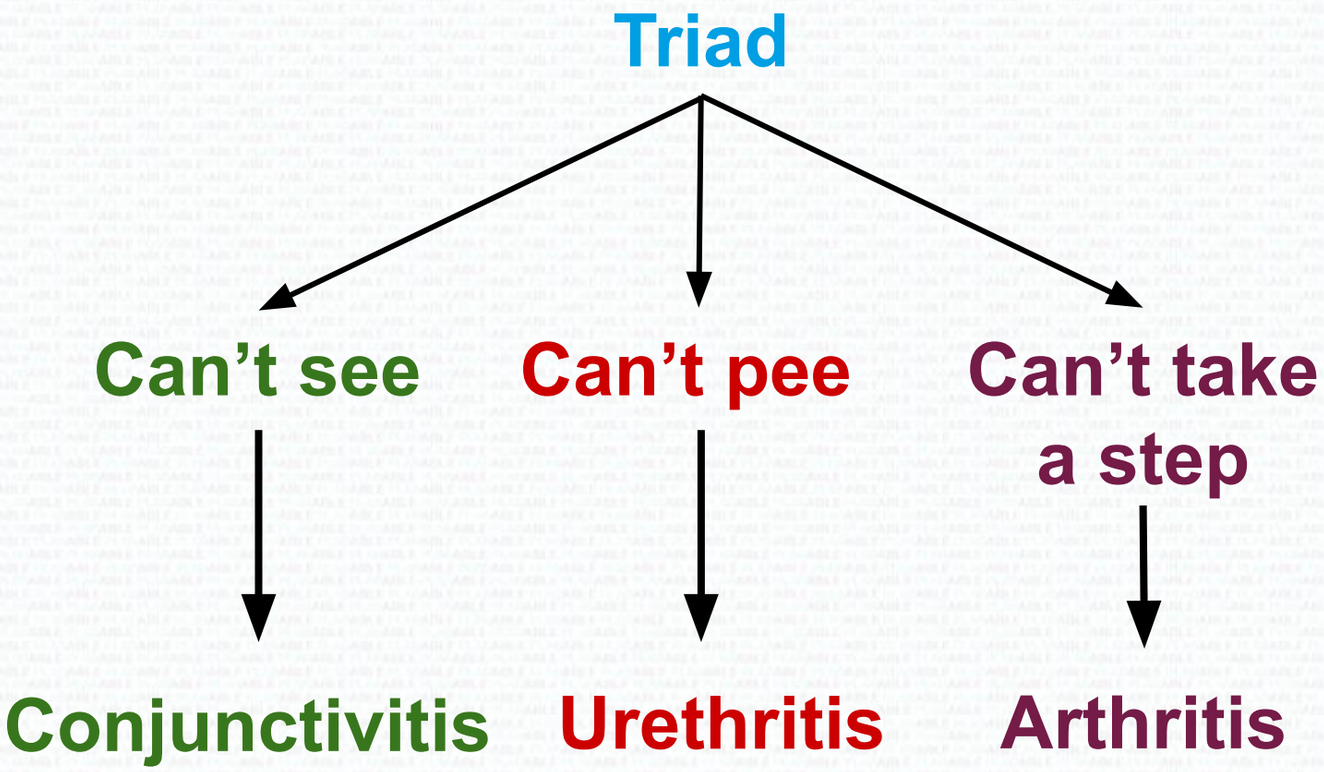
Complication: Fusion of the spine (bamboo spine)



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

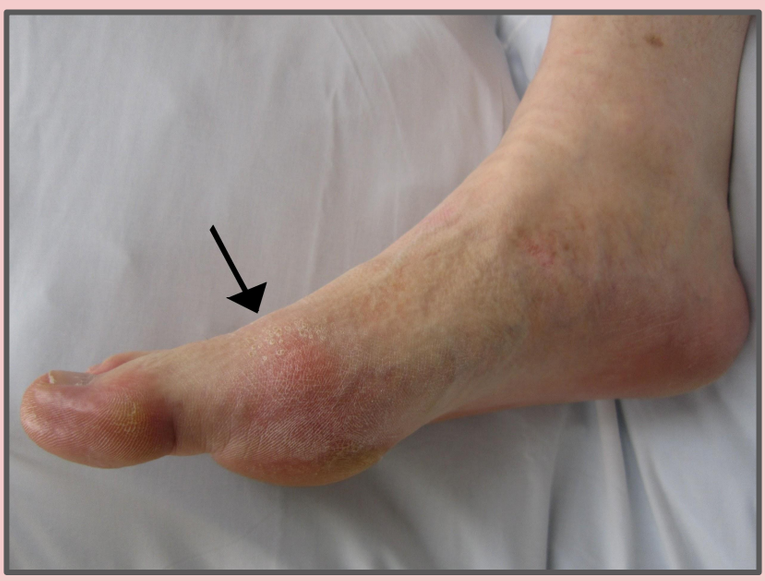
Most commonly involved joint is **first metatarsal phalangeal joint**

Diagnosis

Presence of MSU crystals in the synovial fluid

Raised uric acid level in blood

Treatment



Acute:

- NSAIDs
- Colchicine
- Corticosteroids

Prevention:

- **Allopurinol** and **febuxostat -** decreased formation of uric acid
- **Uricosurics** (Sulfinpyrazone) - increased excretion of uric acid

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

Treatment

- Steroids

Wegener's Granulomatosis

Granulomatosis with polyangiitis

Presentation

- **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

- Cyclophosphamide
- Prednisolone

Rheumatoid Arthritis

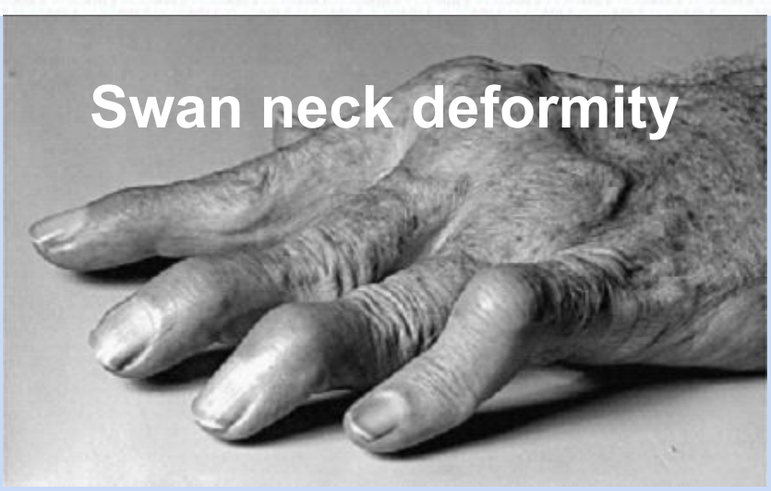
Presentation

- **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

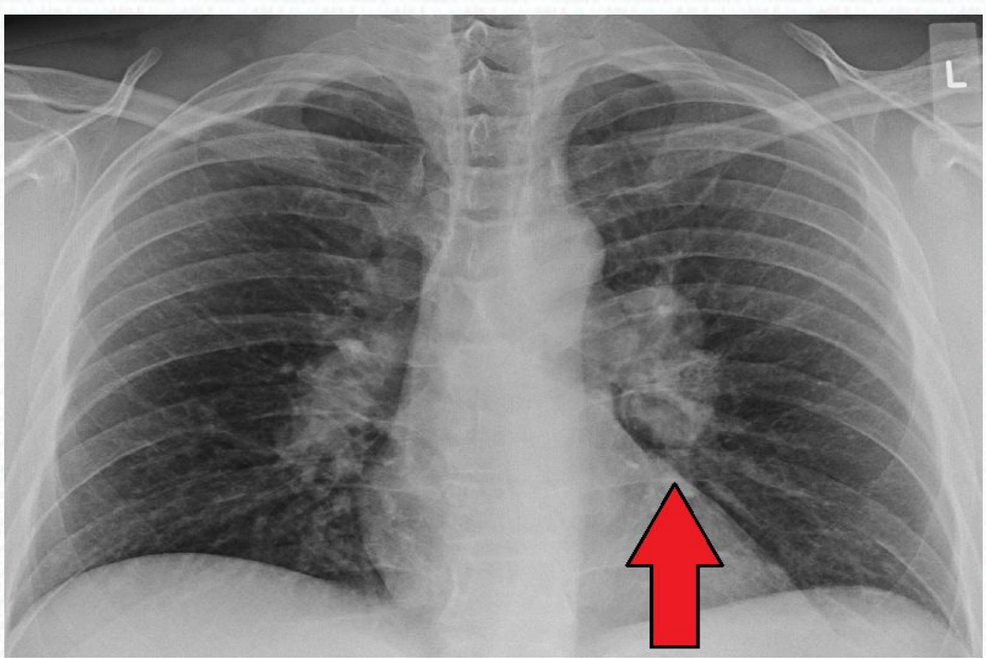
- Erythema nodosum
- Polyarthralgia
- Fever and night sweats
- Lupus pernio

Investigation

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

Treatment

- Oral corticosteroids



Sarcoidosis - Hilar adenopathy

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

Image Attributions

https://commons.wikimedia.org/wiki/File:Symptoms_of_SLE.png
Mikael Häggström. CC: Public domain

https://commons.wikimedia.org/wiki/File:Lupusfoto.jpg
Doktorinternet CC BY-SA 4.0

https://commons.wikimedia.org/wiki/File:Dermatomyositis4.jpg
Elizabeth M. Dugan, Adam M. Huber, Frederick W. Miller, Lisa G. Rider CC BY-SA 3.0

https://en.m.wikipedia.org/wiki/File:Dermatomyositis2.jpg
Elizabeth M. Dugan, Adam M. Huber, Frederick W. Miller, Lisa G. Rider CC BY-SA 3.0

https://commons.wikimedia.org/wiki/File:Sjogrens_Syndrome.jpg
Scientific Animations CC BY-SA 4.0

https://commons.wikimedia.org/wiki/File:Bamboo_spine_ankylosing_spondylitis.jpg
Stevenfruitsmaak CC BY-SA 3.0

https://commons.wikimedia.org/wiki/File:SWAN_NECK_SS.png
Abdulaziz Alkanderi CC BY-SA 3.0

https://commons.wikimedia.org/wiki/File:HilarAdenopathy.png
James Heilman, MD CC BY-SA 4.0