

Systemic Sclerosis (Scleroderma)

Systemic auto-immune condition resulting in excessive collagen deposition by fibroblasts.

Systemic sclerosis examination findings

- Things to ask patient
 - **CREST**
 - Raynaud's (hands change colour in cold: white→blue→red)
 - Esophageal dysfunction (heartburn)
 - Organ involvement (ask)
 - Lungs (breathlessness)
 - Cardiac problems
 - Renal (hypertension)
 - GI (heartburn)
- Hands
 - Look
 - Sclerodactyly
 - Telangiectasia
 - Tight waxy skin
 - Ulceration
 - Blanching
 - Finger-pulp atrophy
 - Feel
 - Subcutaneous calcinosis
 - Cold hands
 - Move
 - Prayer sign lost (fixed flexion deformity of fingers)
- Face
 - Mask-like face
 - Skin tethering
 - Telangiectasia
 - Microstomia
 - Beaking of nose
- Organ involvement
 - Auscultate lungs (end-inspiratory crepitations = IPF)
 - BP (hypertension)

Types

- Limited cutaneous systemic sclerosis (cutaneous sclerosis limited to below elbows)
- Diffuse cutaneous systemic sclerosis (cutaneous sclerosis extends above elbows)

Clinical Features

- **CREST** + skin involvement
 - Calcinosis
 - Raynaud's
 - Esophageal dysfunction
 - Sclerodactyly
 - Telangiectasia
- Fibrotic organ involvement
 - Lungs (fibrosis, pulmonary hypertension)
 - Cardiac (myocardial fibrosis → arrhythmias)
 - Renal (acute or chronic kidney injury, hypertension, acute renal hypertensive crises)
 - GI (reflux, dysphagia, anal incontinence)

Investigations

- Auto-antibodies
 - Limited
 - Anti-Centromere (=CREST)
 - Diffuse

- Anti-topoisomerase
- Assess extent of organ involvement (e.g. U&Es, urinalysis, CXR/high-resolution CT, hands XR, barium swallow etc)

Management

- No cure
- Symptomatic treatment
 - Raynaud's: hand warmers, Ca antagonists, IV prostacyclins
 - GI: omeprazole
- Prevention of complications
 - Lung fibrosis: immunosuppressants (cyclophosphamide)
 - Pulmonary hypertension: prostaglandins, endothelin antagonists, phosphodiesterase antagonists
 - Renal crises: low dose ACE-inhibitor