Scleroderma

• What is it?
  – “hard skin”
  – affects skin, joints, blood vessels, internal organs

• Epidemiology
  – 😄📞😊 (1:3)

• Pathogenesis
  – overproduction of collagen?
Pathogenesis of Scleroderma

- Over production of Collagen
  - Vascular disease
  - Autoimmune reaction
  - Fibrotic reaction
  - Genetics?
Diagnosis of Scleroderma

• May overlap with CTD
• History
• Physical Exam
• Lab Tests
  – ANA
    • anticentromere
    • topoisomerase I
Scleroderma Types

• Systemic Scleroderma (SSc) (Generalized)
  – CREST
    – Limited: Hands, face, & neck
    – Diffuse: Wrists & Elbows

• Localized
  – linear
  – morphea
  – generalized morphea
  – en coup de sabre
Diffuse vs Limited

- 1st 2-4 years
- Skin above the wrists
- Scleroderma *sans*
  Scleroderma
  - internal organs fibrosis
  - no skin tightening
- Milder form
- Pitting digital ulcers
- Raynauds
- Esophageal problems
- Slow onset
- Good prognosis
Diffuse vs Limited
Localized Scleroderma

• Morphea
  – hard, oval shaped patches w/ purplish ring
  – improves over time

• Generalized Morphea
  – larger patches

• Linear
  – line of thickened skin and deeper layers
  – can effect bones, muscles & joints
  – younger age

• en coup de sabre : “stroke of a sword”
Morphea vs Linear
Morphea
CREST Syndrome

- **C**alcinosis
- **R**aynaund’s phenomenon
- **E**sophageal dysfunction
- **S**clerodactyly
- **T**elangiectasia
Crest Types

- Pure CREST
  - 2 or more symptoms
  - Not limited or diffuse form

- Plus CREST
  - Crest symptoms with other form of Scleroderma
Calcinosis

- Calcium deposits in skin
- Elbows, knees, fingers
- Potential infection
Raynaud’s Phenomenon

- 90% affected
- induced by cold
- bilateral
- white to blue to red
- ↓ perfusion pressure
- ↓ capillary radius
- ↑ blood viscosity
Raynaud’s Phenomenon
Esophageal Dysfunction

- Hard to swallow due to internal scarring
- 50% of patients
- lower 2/3 of esophagus
- Wide esophagus
  - heartburn
  - scarring
  - inflammation
Sclerodactyly

- Tightening of skin on digits
- ↑ collagen production
- Shiny appearance
- Difficult to bend
Sclerodactyly
Telangiectasia

- Dilation of small blood vessels
- Form red spots
- Not harmful
- On face, hands, & mouth behind lips
Treatment

• No cure, treat symptoms
• PT & ROM exercises
• Medications
  – Nsaids
  – Skin moisturizers
  – Abx
  – Vasodilators
• Treat the individual patient
Summary

- Connective Tissue & Autoimmune disease
- 2 main types
  - Systemic (Adults)
  - Localized (Children)
- CREST is Systemic limited form
- More common in Females
- No genetic etiology
- No cure right now