



#### Scieroderma, the basics

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# What is scleroderma

- Sclero: fibrotic; derm: skin => fibrotic skin
- When you have scleroderma your skin tighten or harden and thicken
- But, Scleroderma is not only a skin disease, it can affect:
  - Lungs
  - Gastrointestinal track
  - Vessels
  - Kidneys
  - Etc.

# Types of scleroderma

- Localized scleroderma (skin disease only)
  - Morphea
  - Linear scleroderma
- Systemic sclerosis (SSc, but we also call it scleroderma!)
  - Diffuse scleroderma
  - Limited scleroderma
  - Scleroderma *sine* scleroderma!!

#### Localized scleroderma - Morphea



#### Localized scleroderma - En Coup De Saber



#### Localized scleroderma - linear scleroderma



## Systemic sclerosis

- >90% of Systemic sclerosis (scleroderma) patients have all 3 of following:
  - Raynaud's phenomenon
  - + ANA (lab test)
  - Sclerodactyly (skin thickening of the fingers)



# Limited and Diffuse SSc— Skin Involvement

#### Limited

Diffuse

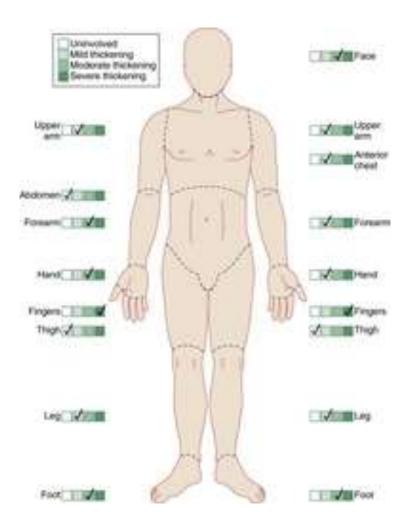
Medsger T . In Clements and Furst 2ND Edition, Systemic Sclerosis

# Diffuse vs limited Scleroderma

- Diffuse
  - Lung fibrosis 15%
  - Heart 10%
  - Kidney (15%)
  - Large joint contractures
  - Scl70 Ab

- Limited
  - Lung fibrosis less common
  - Pulmonary hypertension more common (15%)
  - Anti Centromere Ab

#### Diffuse scleroderma



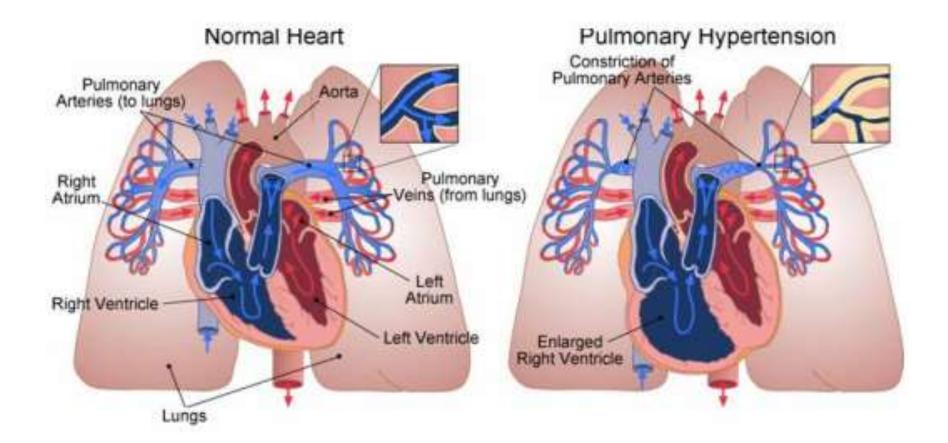
#### Diffuse Scleroderma



#### Interstitial lung disease in scleroderma



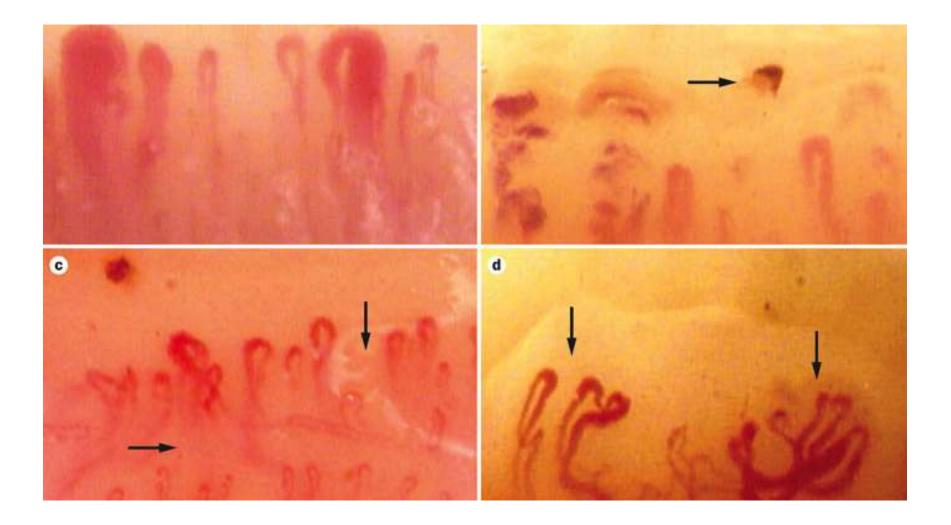
# **Pulmonary hypertension**



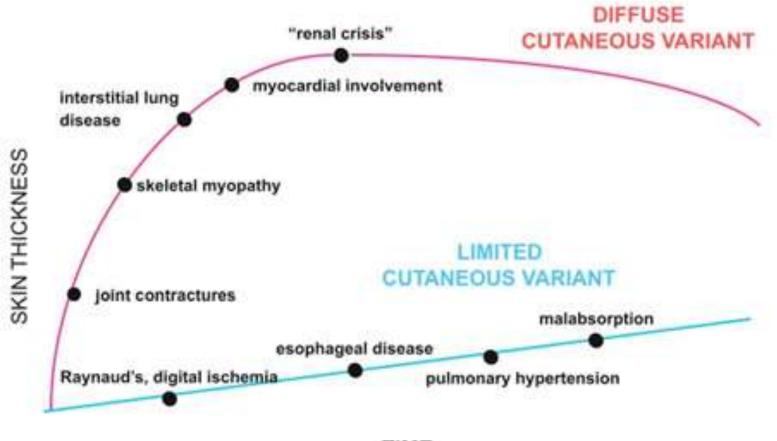
# Gastrointestinal involvement in scleroderma



#### Nailfold capillaries in scleroderma



#### Usual Timing of Problems in Patients with SSc



TIME

Thomas A. Medsger, Jr., MD

# What to expect when you visit your doctor

- Evaluation of the extent of skin fibrosis
- Listening to the lungs and heart
- Blood pressure measurement (hopefully you also do it at home twice weekly)
- Annual echocardiogram
- Annual CT scan for the first few years, then as needed
- Treatment directed to type of organ involvement

### In Scleroderma, One Size Does Not Fit All!



## Thank you

Any Questions



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