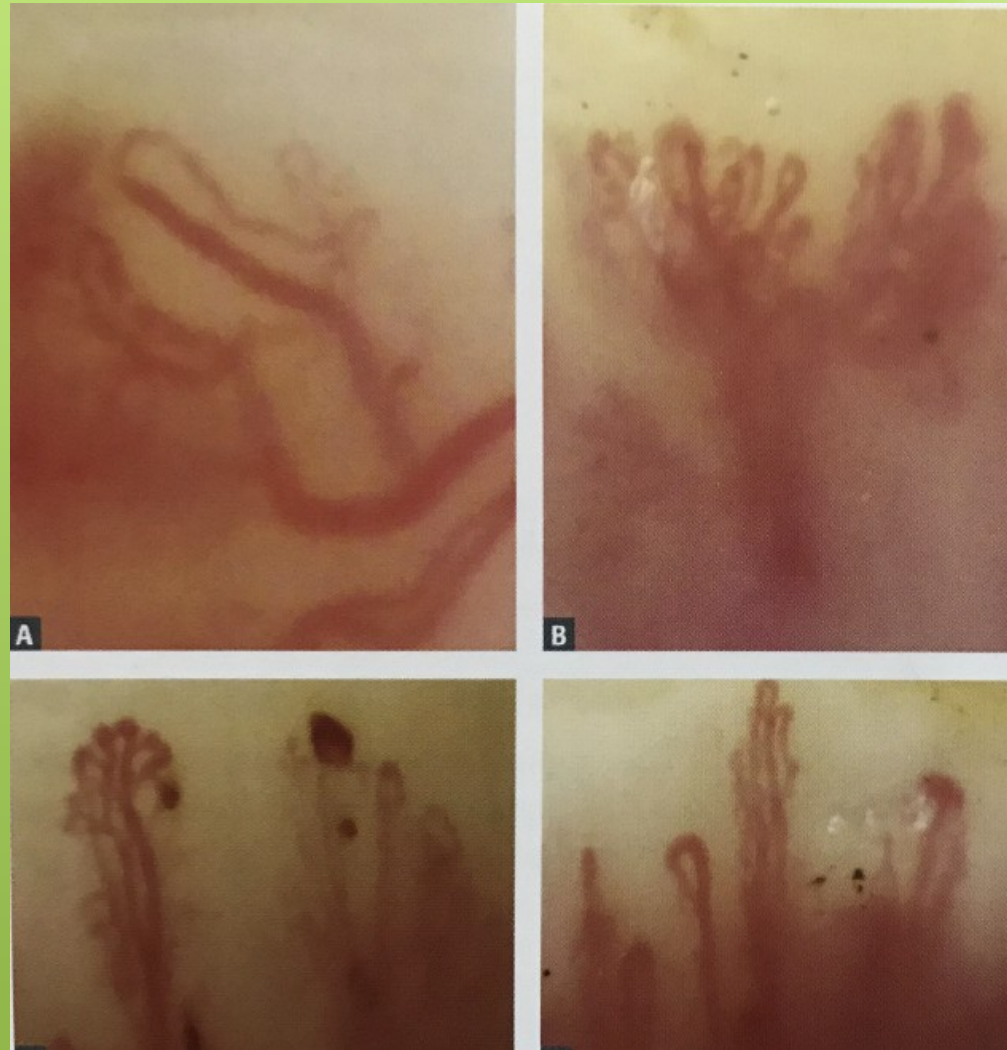


Bushing



Ramified



How to assess /score the nailfold capillary parameters?

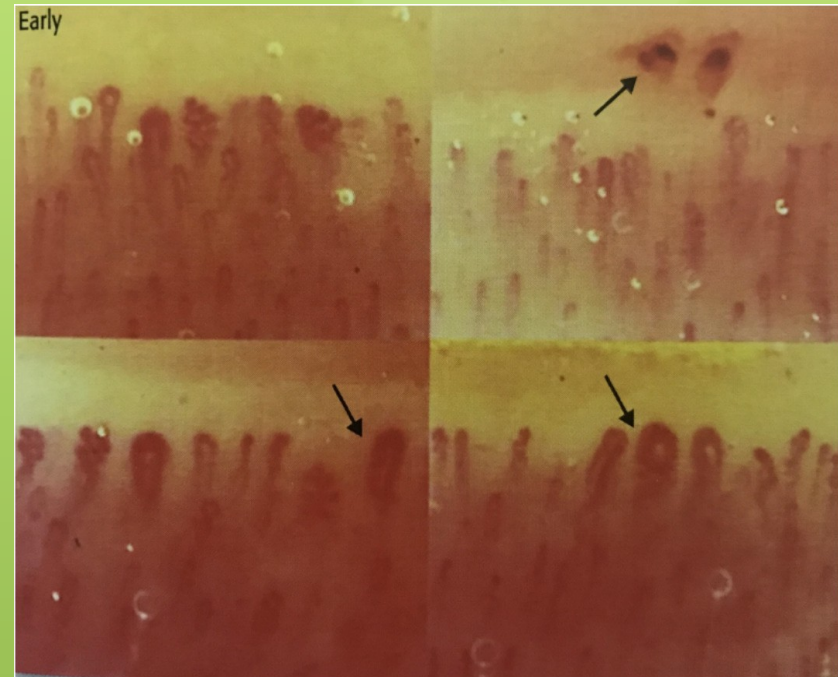
- No consensus on which scoring system to use
- Qualitative assessment
 - pattern recognition
- Semiquantitative assessment using NVC

Scleroderma pattern

- three different patterns of nailfold microvascular damage
- “Early”, “Active” and “Late”
- correlating with the evolution of the microangiopathy

Scleroderma Pattern

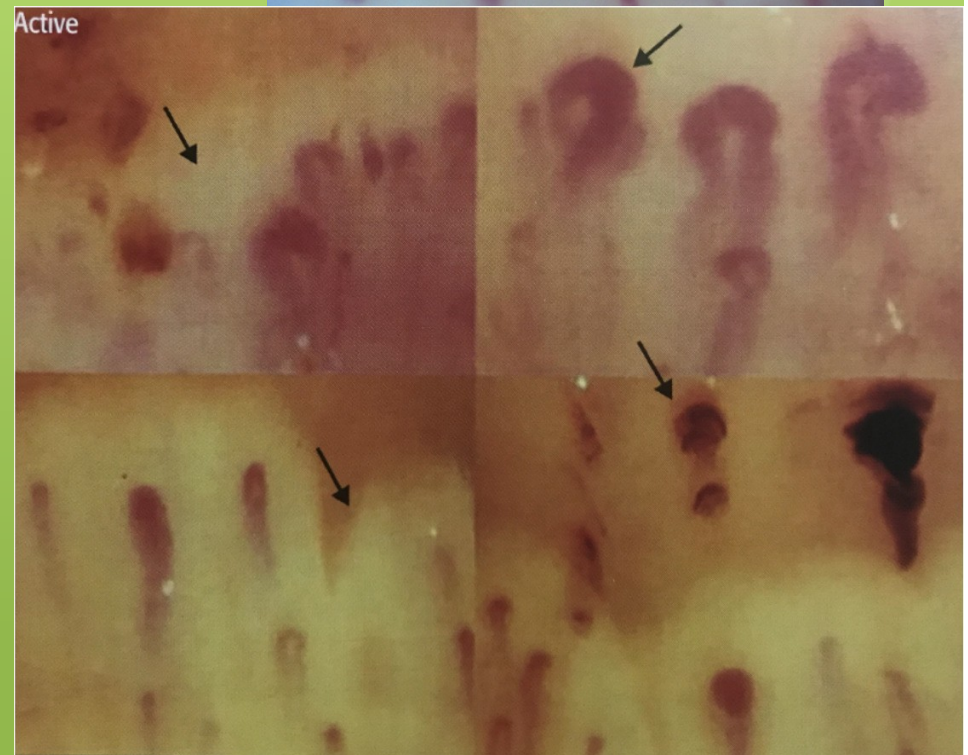
- “Early” pattern
 - Few giant capillaries (pathognomonic)
 - Microhaemorrhages
 - No loss of capillaries
 - Relatively well preserved capillaries distribution



Scleroderma Pattern

“Active” pattern

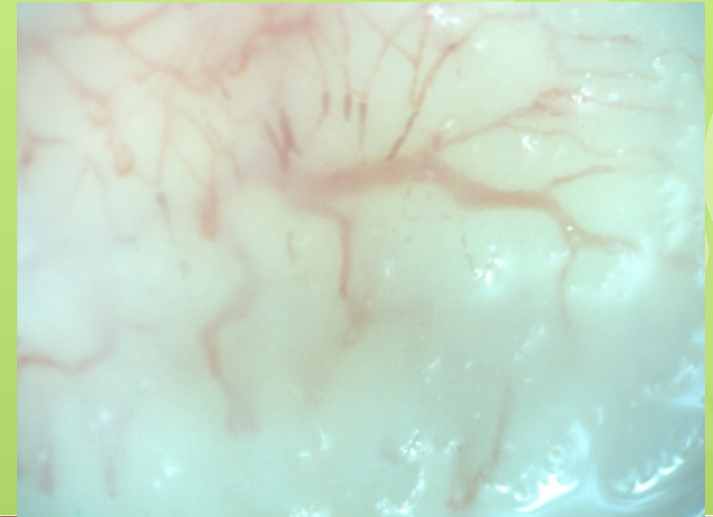
- Freq giant capillaries
- Freq microhaemorrhages
- normal-shaped capillaries progressively decrease
- Moderate capillaries loss
- Absence/mildly ramified cap
- Slight disorganisation of cap architecture



Scleroderma Pattern

Late pattern

- Only few Irregular enlarged capillaries
- Absent of giant cap and microhaemorrhage
- Severe Loss of cap/ extensive avascular areas
- Bushy/ramified cap
- Intense disorganisation of capillary array



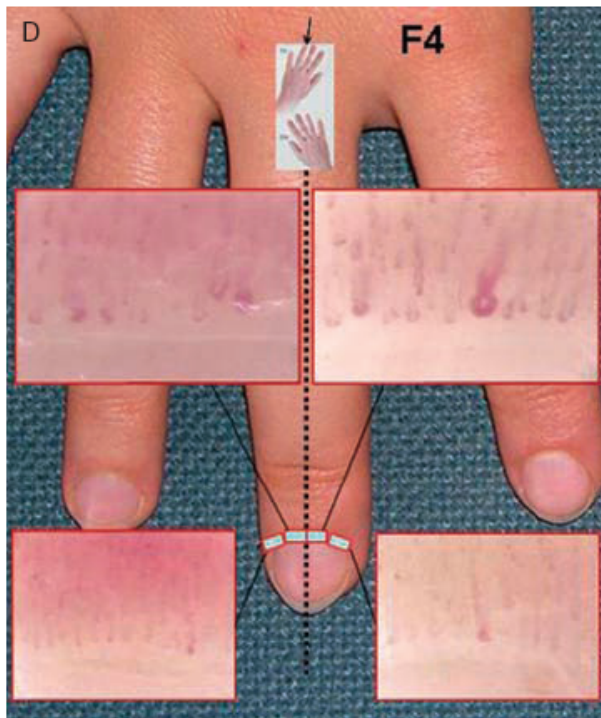
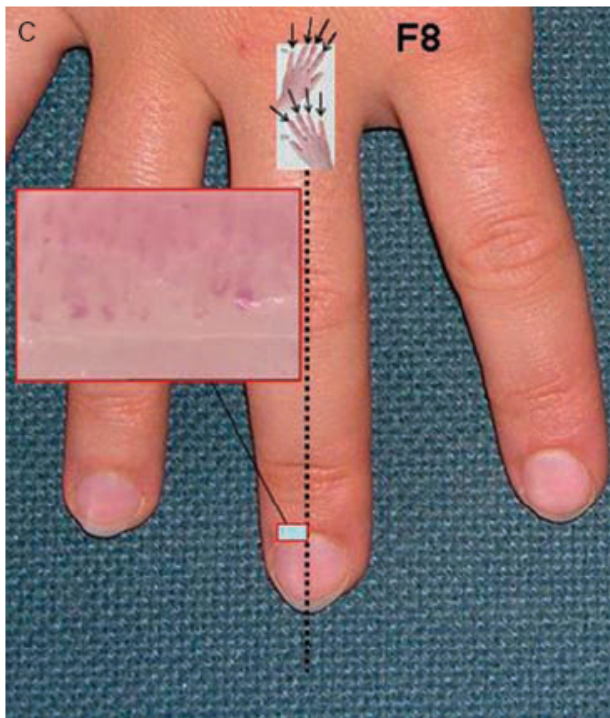
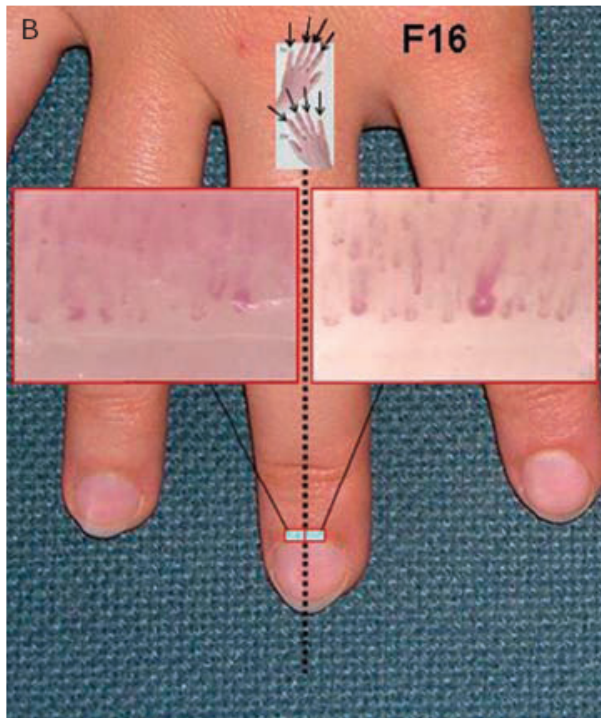
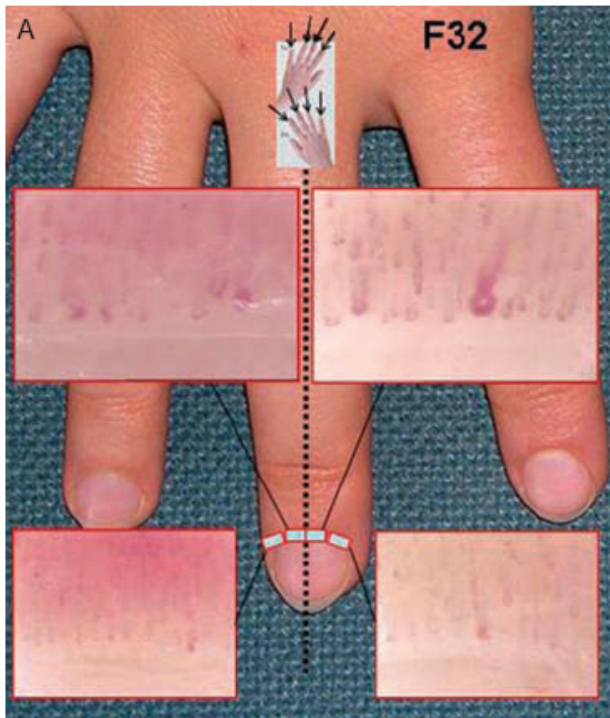
Semi-quantitative rating scale

Definition:

- Normal capillaries- > 9 per linear mm counted at the distal row of the nailfold = 0
- $< 33\%$ reduction (7-9 capillaries) = 1
- 33-66% of capillary reduction (4-6 capillaries) = 2
- $> 66\%$ reduction (1-3 capillaries) = 3

Semiquantitative assessment of capillaroscopic patterns

- Evaluate the average of the 8 fingers, fr 2nd to 5th



“Early” NVC pattern

- few (< 33%) enlarged/giant capillaries (G1)
- few (< 33%) capillary haemorrhages (H1)
- relatively well-preserved capillary distribution
- Absent or mild (<33%) loss of capillaries. (CL1)

“Active” NVC pattern

- frequent ($> 33\%$) giant capillaries (G2-3)
- frequent ($> 33\%$) capillary haemorrhages (H2-3)
- moderate ($< 66\%$) loss of capillaries (CL2-3)
- Moderate ($< 66\%$) disorganisation of the capillary architecture
- absent or few ($< 33\%$) ramified capillaries (R0-1)

“Late” NVC pattern

- irregular enlargement of the capillaries (CL3)
- few (< 33%) or absent giant capillaries and haemorrhages (G0H0)
- severe (>66%) loss of capillaries with avascular areas, disorganisation of the normal capillary array, ramified/bushy capillaries. (R3)

Scleroderma NVC pattern

	morphology	Giant	µhaemorrhage	neoangiogenesis
Early	Well preserved, absent or mild loss of capillaries (<33%)	Few < 33%	Few (<33%)	
Active	Moderate loss of capillary <66%	Frequent > 33%	Frequent >33%	Absent or few ramification <33%
Late	Severe loss of capillary >66%	absent	Few or absent	Severe >66%

NVC pattern and organ involvement

	Baseline normal	early	active	late
Future severe¹ organ disease	13%	20%	33%	63%

1. Smith V et al, Ann Rheum Dis. 2012;71:1636-9

Late SSc pattern

- Correlate with greater organ involvement and disease severity