Systemic sclerosis: initial work up

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Bilan initial d’un malade sclérodermique

- Forme: diffus vs limité
- Examen clinique
- Examens complémentaires
- Atteintes viscérales
- Handicap/qualité de vie

Clinical classification of SSc

Diffuse cutaneous SSc

- Skin sclerosis proximal to elbows and knees
- Inflammatory features prominent in 1st 3 years
- Anti-Scl-70 or anti-RNA polymerase
- Increased frequency of interstitial lung disease, renal crisis, bowel & cardiac involvement

Limited cutaneous SSc

- No skin sclerosis proximal to elbows and knees
- Anti-centromere antibody (ACA)
- CREST subgroup
- Lung fibrosis, renal crisis & cardiac involvement less common than in dcSSc

Scleroderma sine Scleroderma

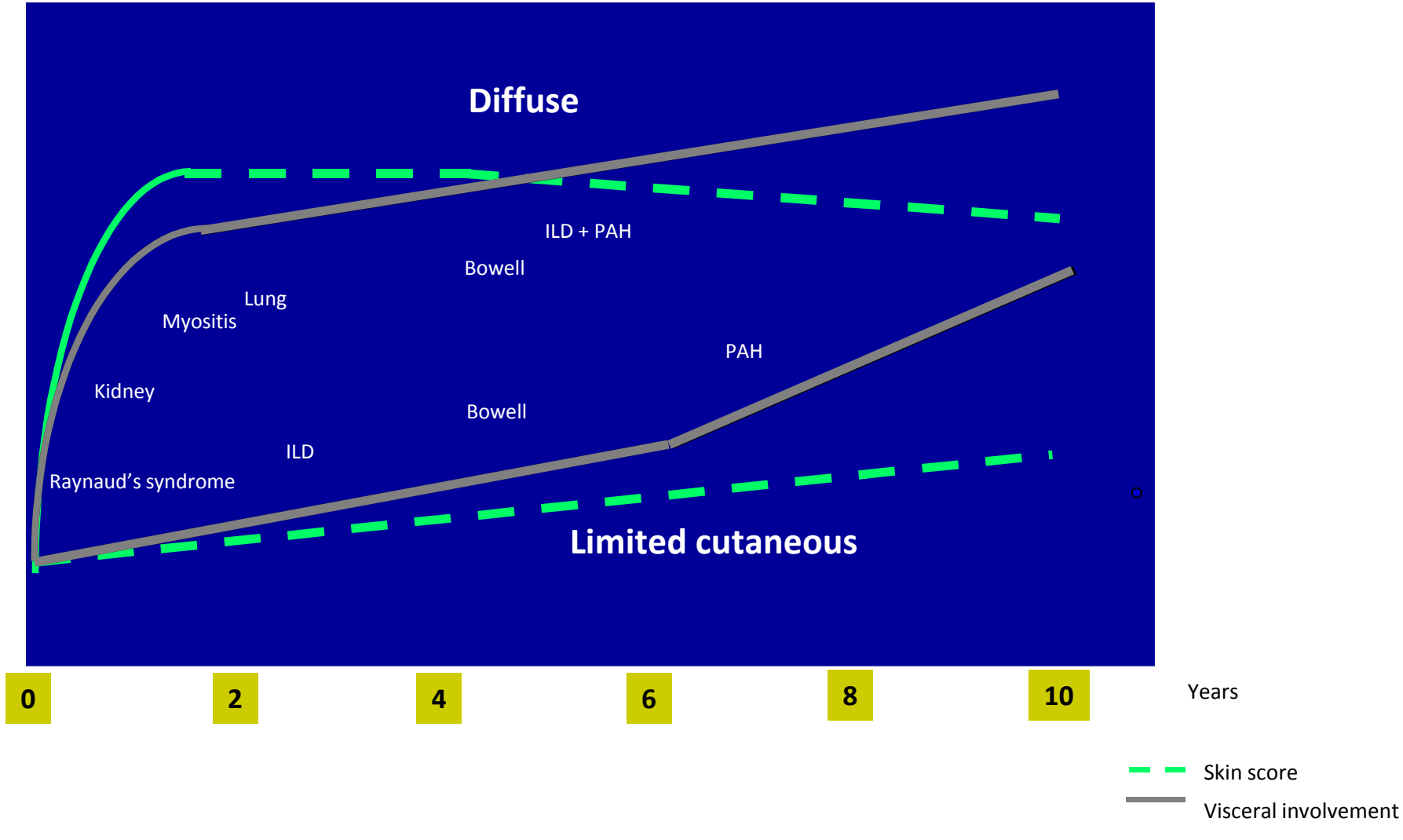
- No skin sclerosis

Overlap syndrome

- Features include those of lcSSc or dcSSc with those of other autoimmune disease(s)

Poormoghim H, *et al. Arthritis Rheum* 2000; 43:444-51,
Denton CP and Black CM, *Trends Immunol* 2005; 26:596-602.

SYSTEMIC SCLEROSIS : EVOLUTION



Evaluation clinique

Lésions cutanées
- Score de Rodnan modifié
- Télangiectasies
- Ulcérations/calcinoses
- Hyperpigmentation/dépigmentation

Pouls périphériques
- Pouls cubitaux
- Manœuvre d'Allen

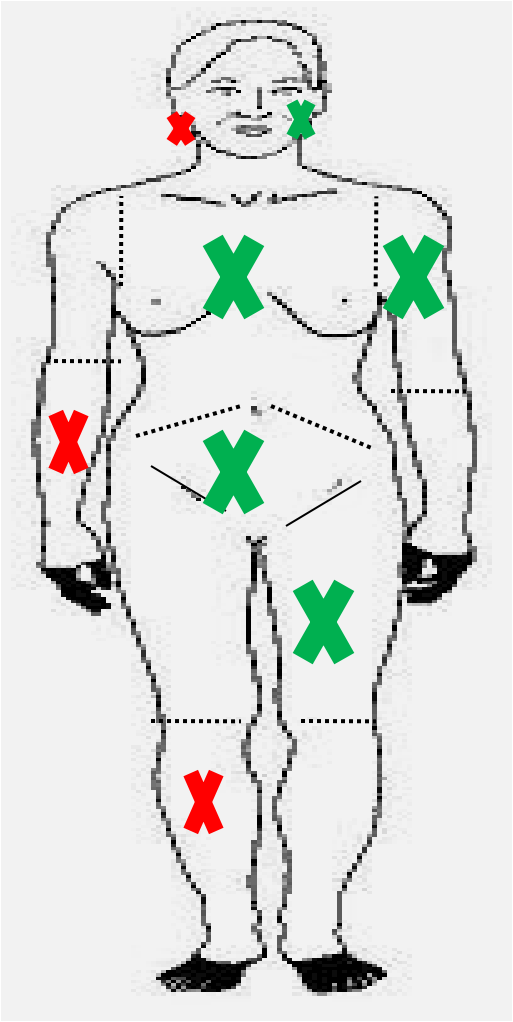
Articulations/muscles
- Douleurs
- Déformations
- Synovites
- Force musculaire
- Frictions tendineuses

Les poumons
- Dyspnée
- Crépitants velcro des bases

Le cœur
- Palpitations
- Fréquence cardiaque/bruit/souffle

Tube digestif
- reflux/dysphagie
- Vomissements
- Distention abdominale
- Constipation / diarrhée
- Incontinence anale

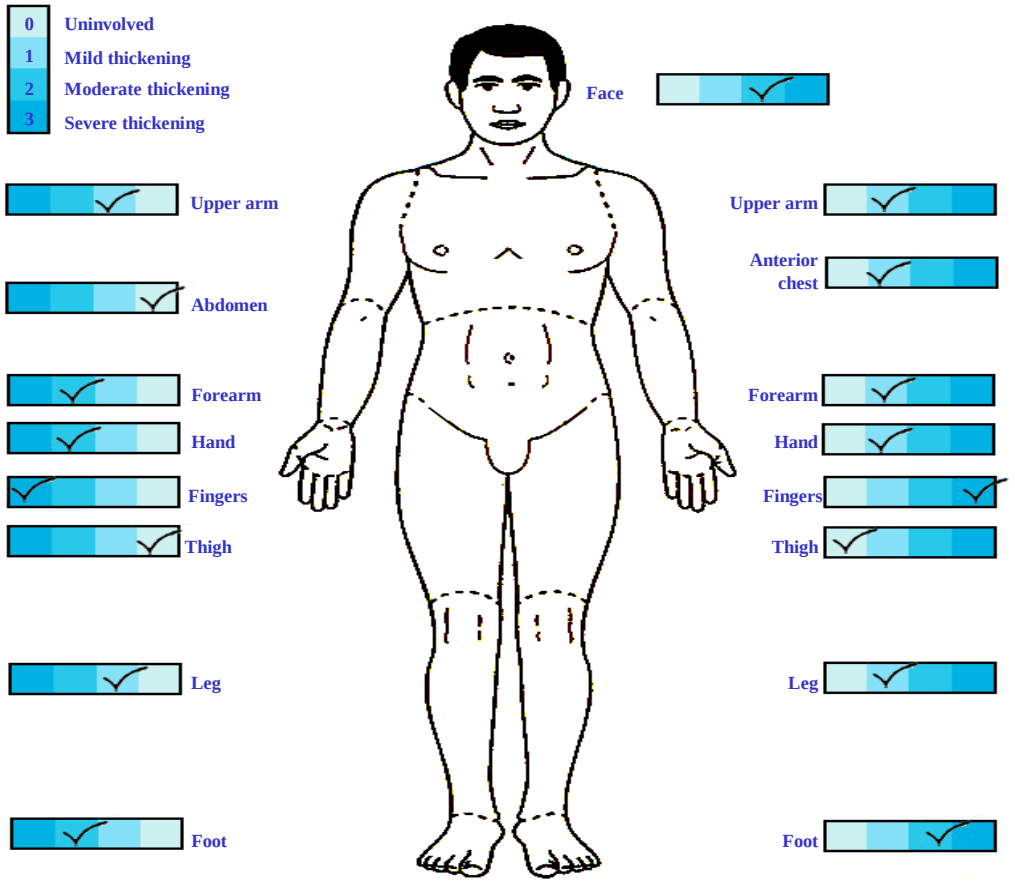
Classification en fonction de l'extension des lésions cutanées

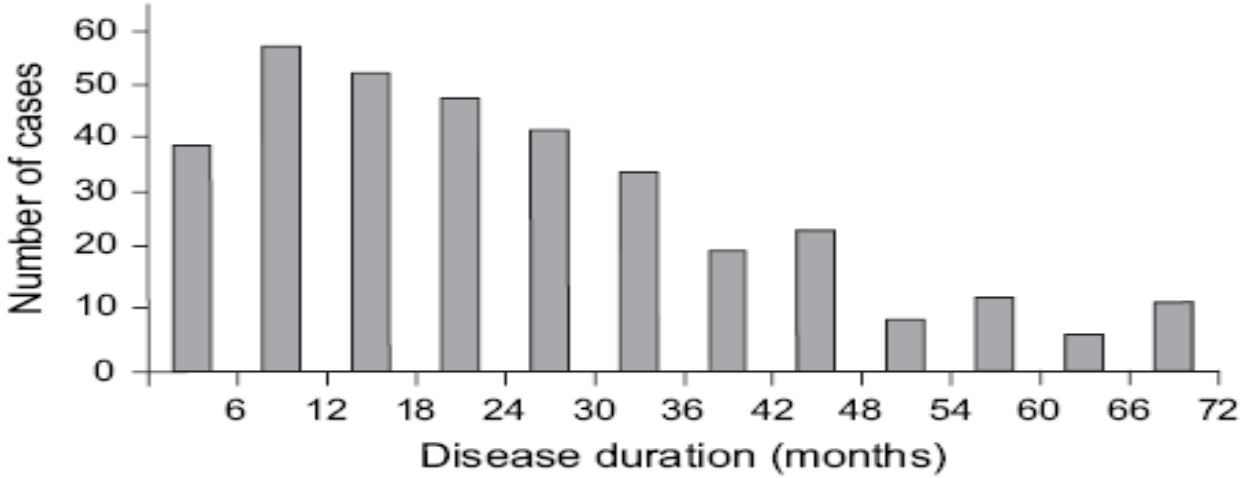


✘ Sclérodermie systémique cutanée limitée

✘ Sclérodermie systémique cutanée diffuse

The modified Rodnan skin score (MRSS)



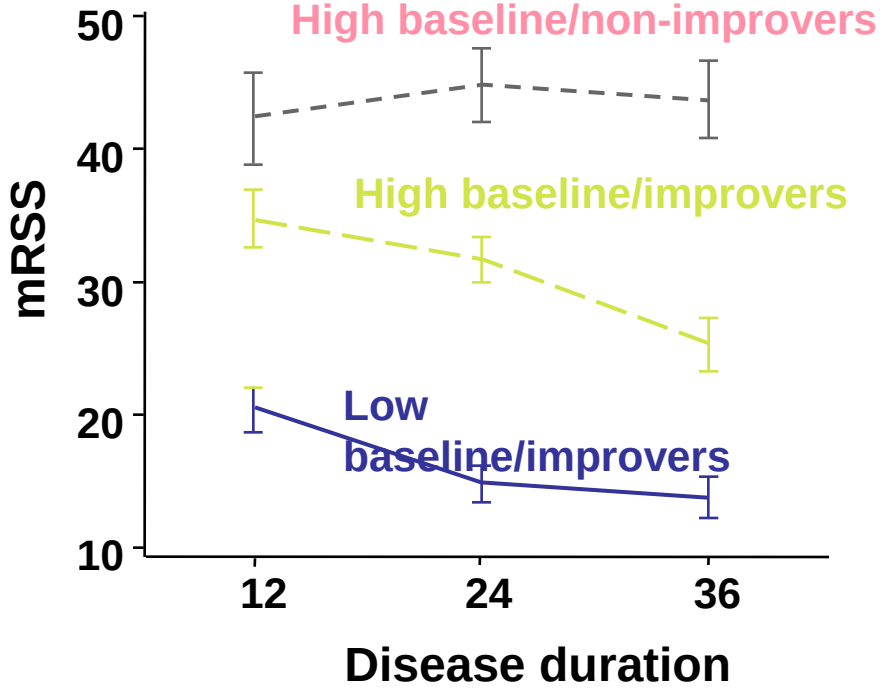


Disease duration at peak skin score of the patients who had dcSSc from the Royal Free Hospital scleroderma database.

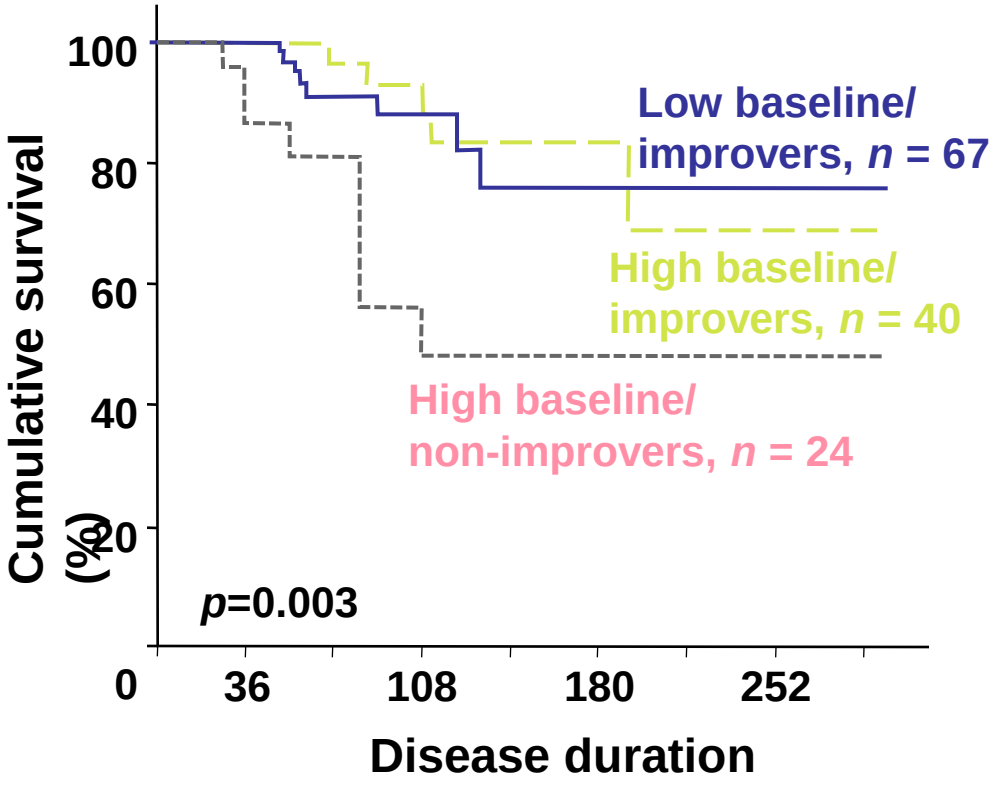
Nihtyanova SI, Denton CP. Rheum Dis Clin N Am 34 (2008) 161–179

Disease duration and skin score in dcSSc

Change in skin score over 3 years in the subgroups

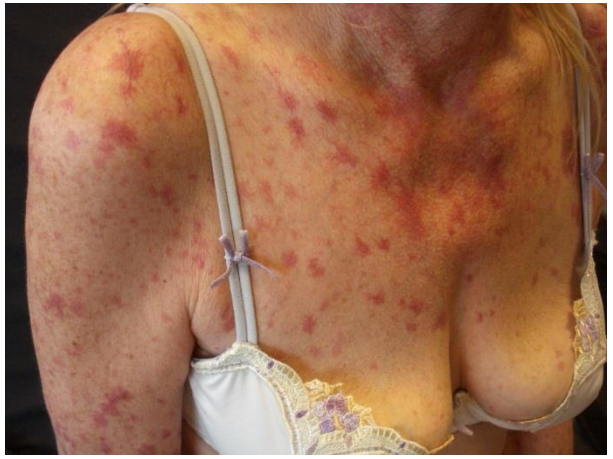


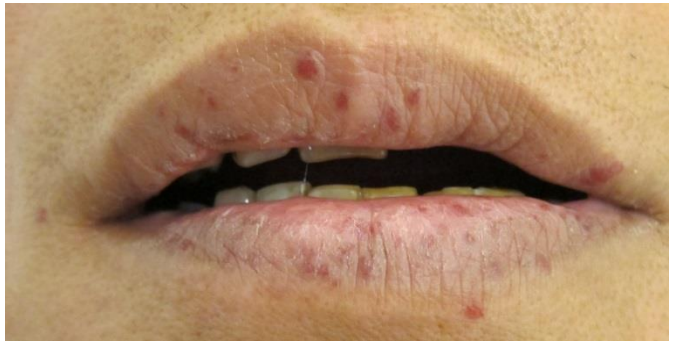
Survival in the subgroups

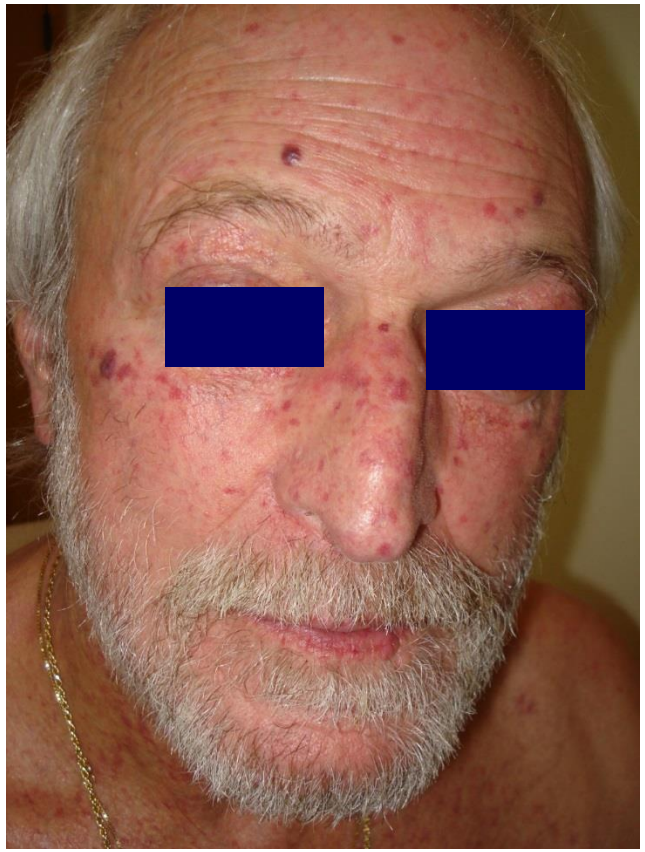


Shand L, *et al. Arthritis Rheum* 2007; 56:2422-31.

telangiectasia

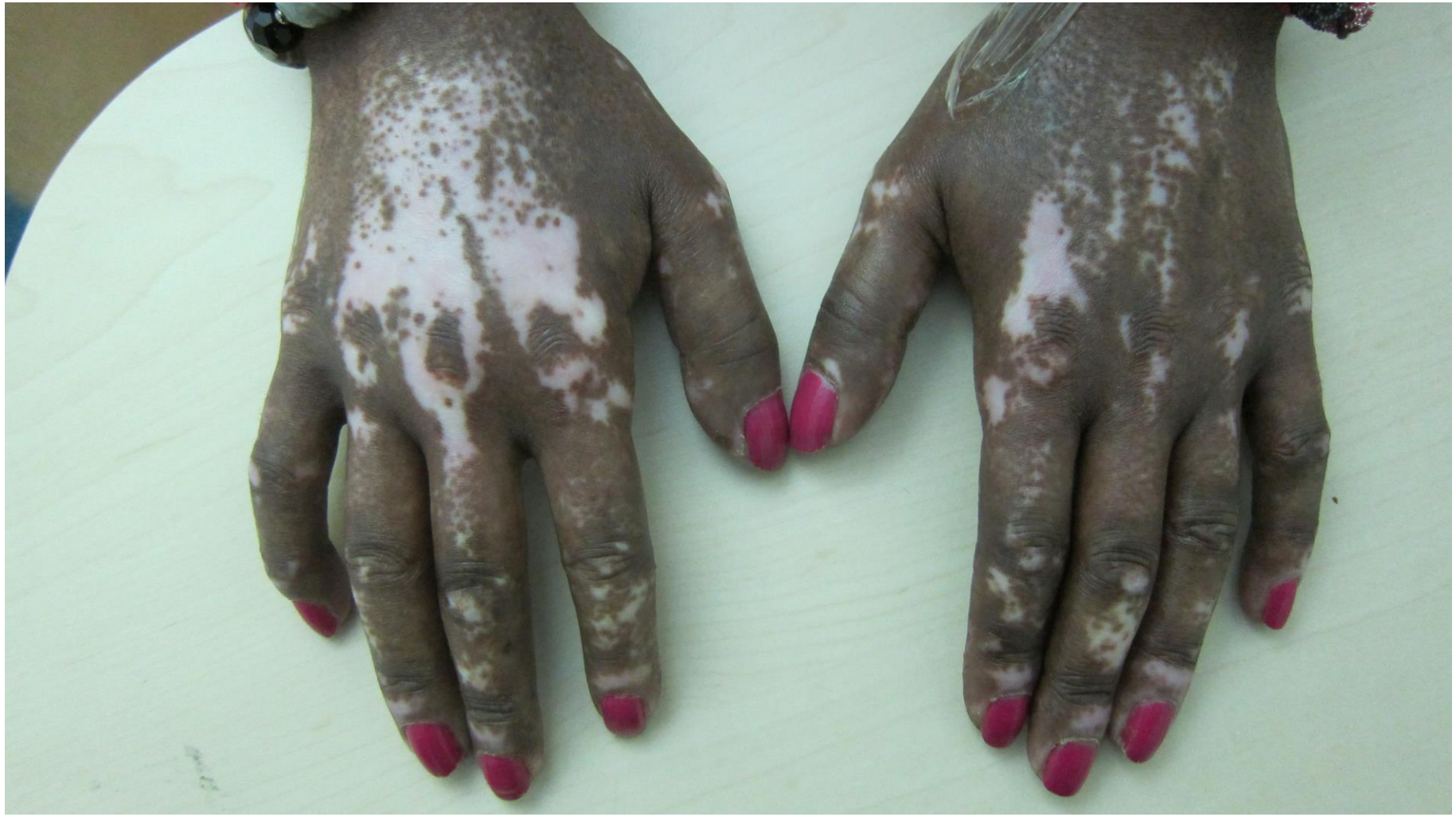






Score = 2

Systemic sclerosis: hyperpigmentation / depigmentation



Tendon friction rubs (TFR)

- Detected by physical examination
- Highly associated with dcSSc
  - 91% of patients with TFR classified as dcSSc
- Associated with poor prognosis
  - e.g. scleroderma renal crisis
- May aid early diagnosis of dcSSc and identification of patients at high risk for serious organ-based complications

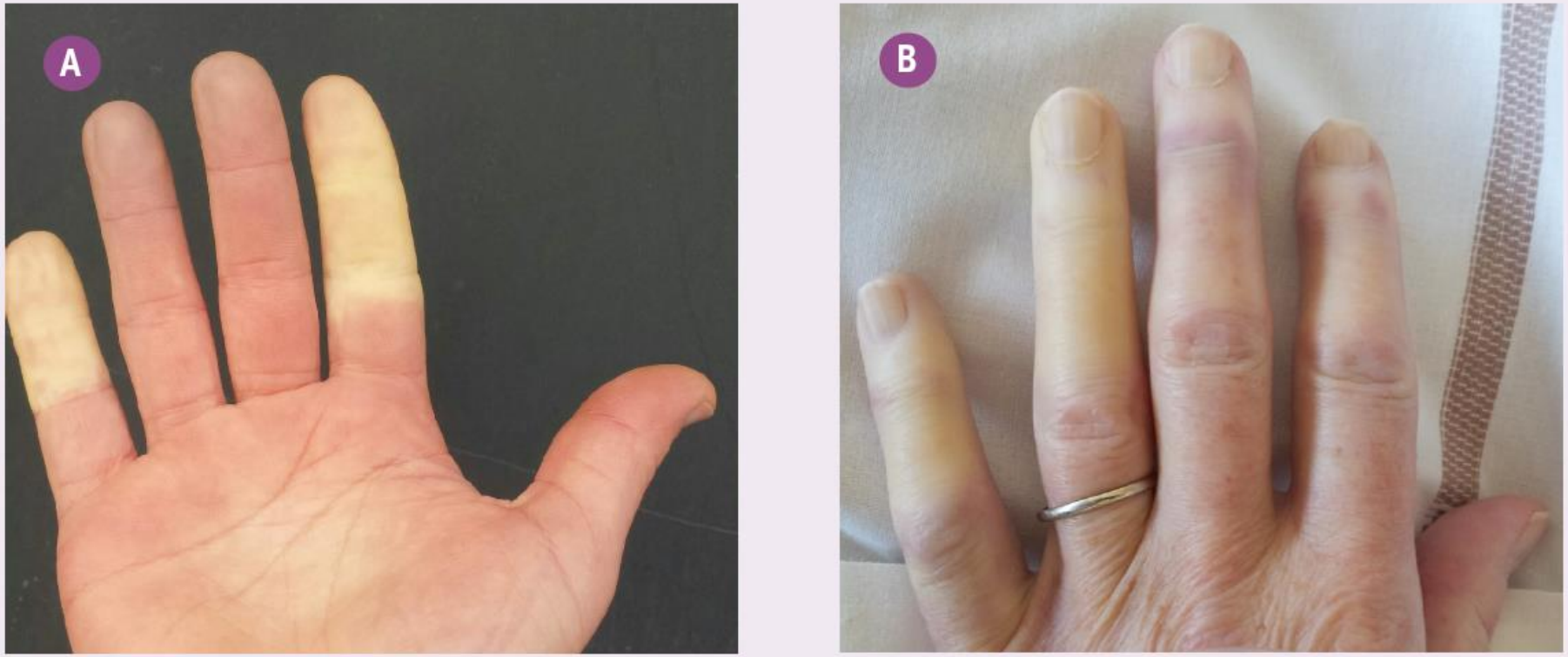
Steen VD and Medsger TA Jr, *Arthritis Rheum* 1997; 40:1146-51.

Les mains

- Phénomène de Raynaud
- Hypertrophie cuticule
- Cicatrices pulpaires
- Ulcérations digitales
- Handicap

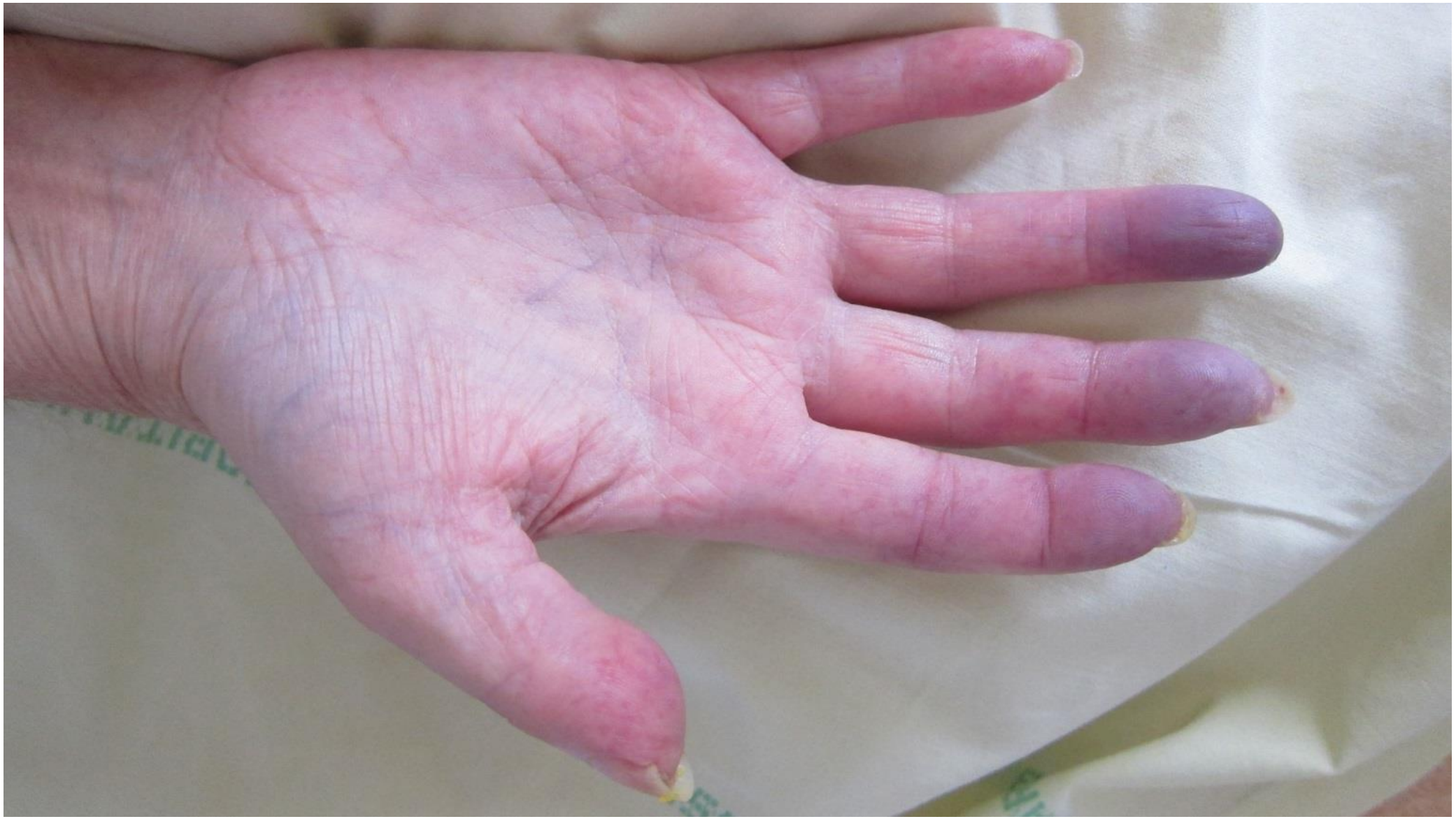
Phénomène de Raynaud

Figure 1. Phase syncopale ou « blanche » d'un phénomène de Raynaud
Remarquer les limites assez nettes. **A** : sclérodermie systémique de forme limitée
B : sclérodermie systémique de forme diffuse.

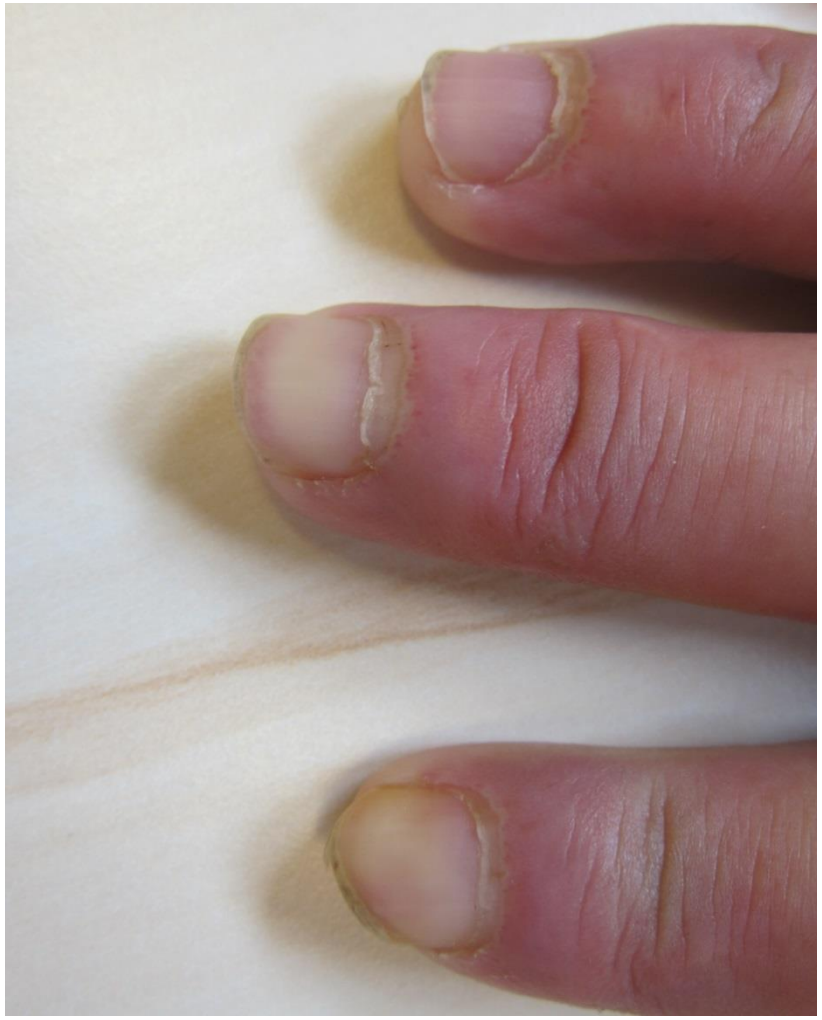


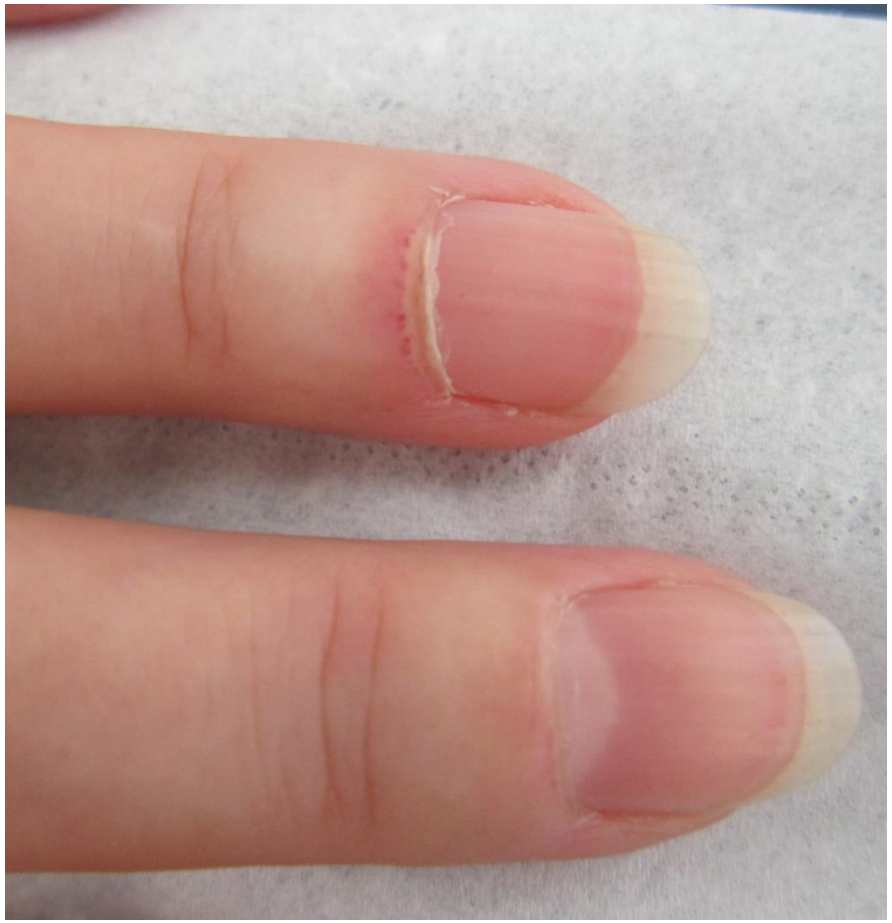
Médecine Interne CEMI, Medline 2015

Raynaud’s phenomenon

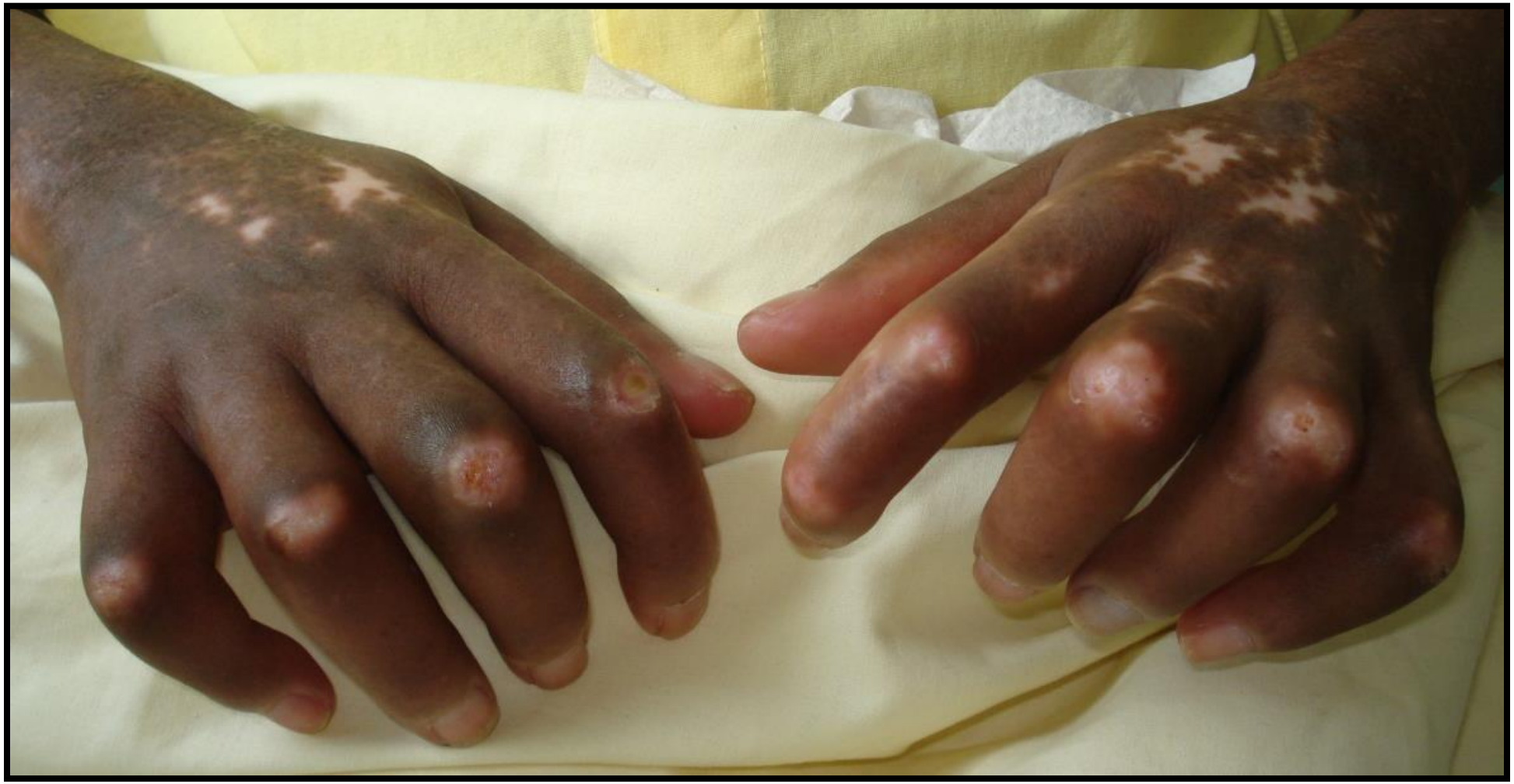


Hypertrophie de la cuticule…

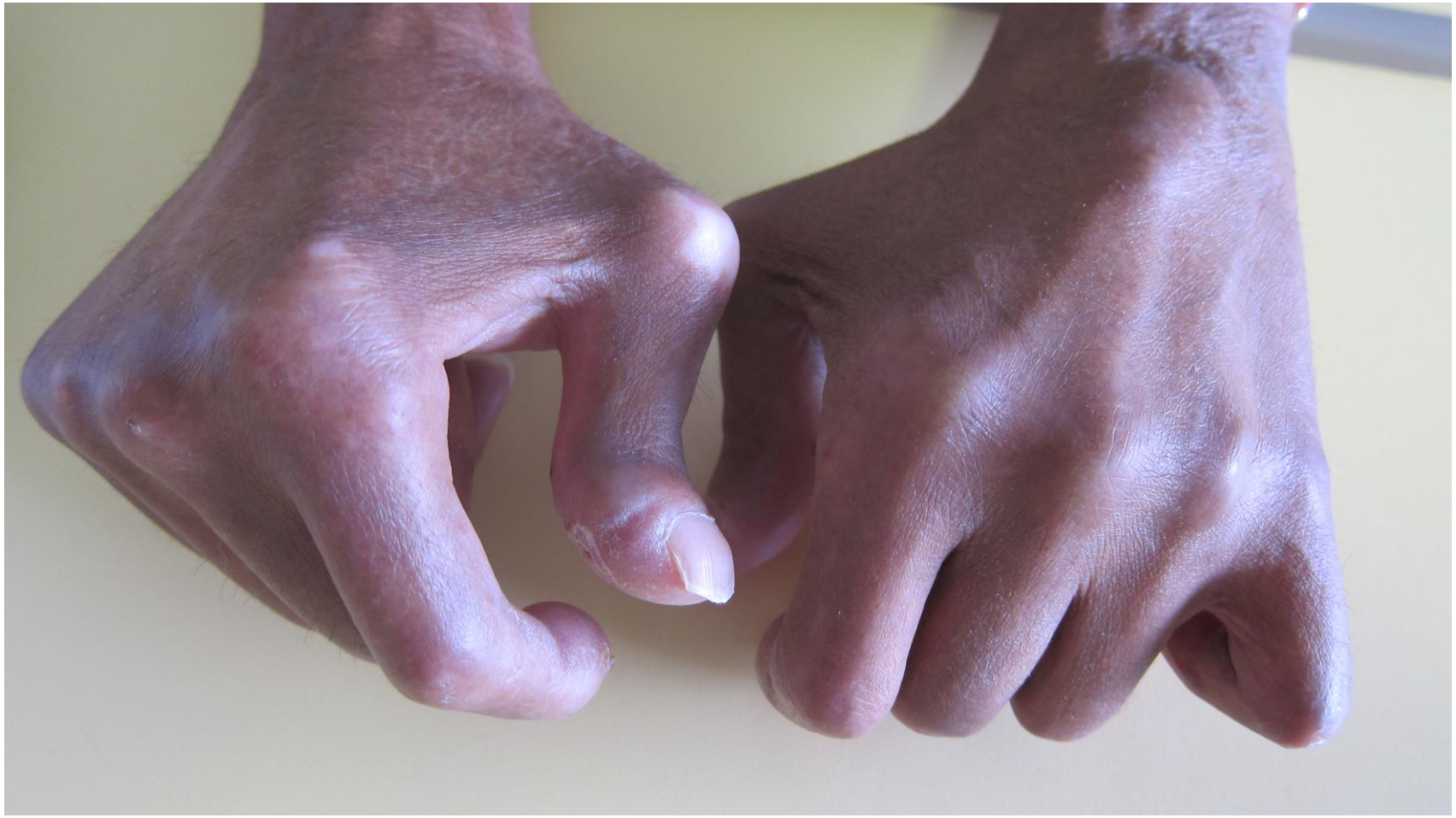




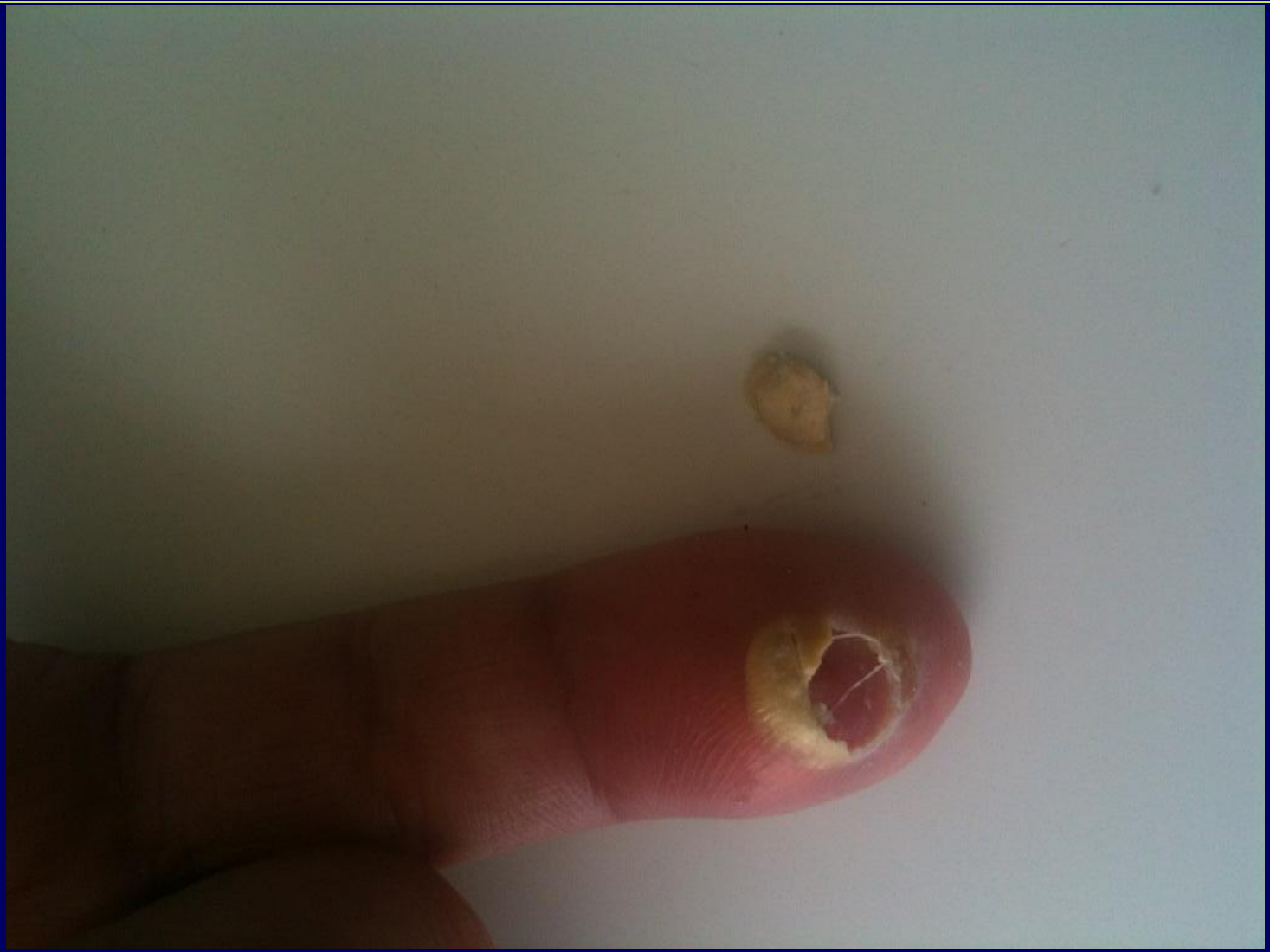
Sclerotic phase



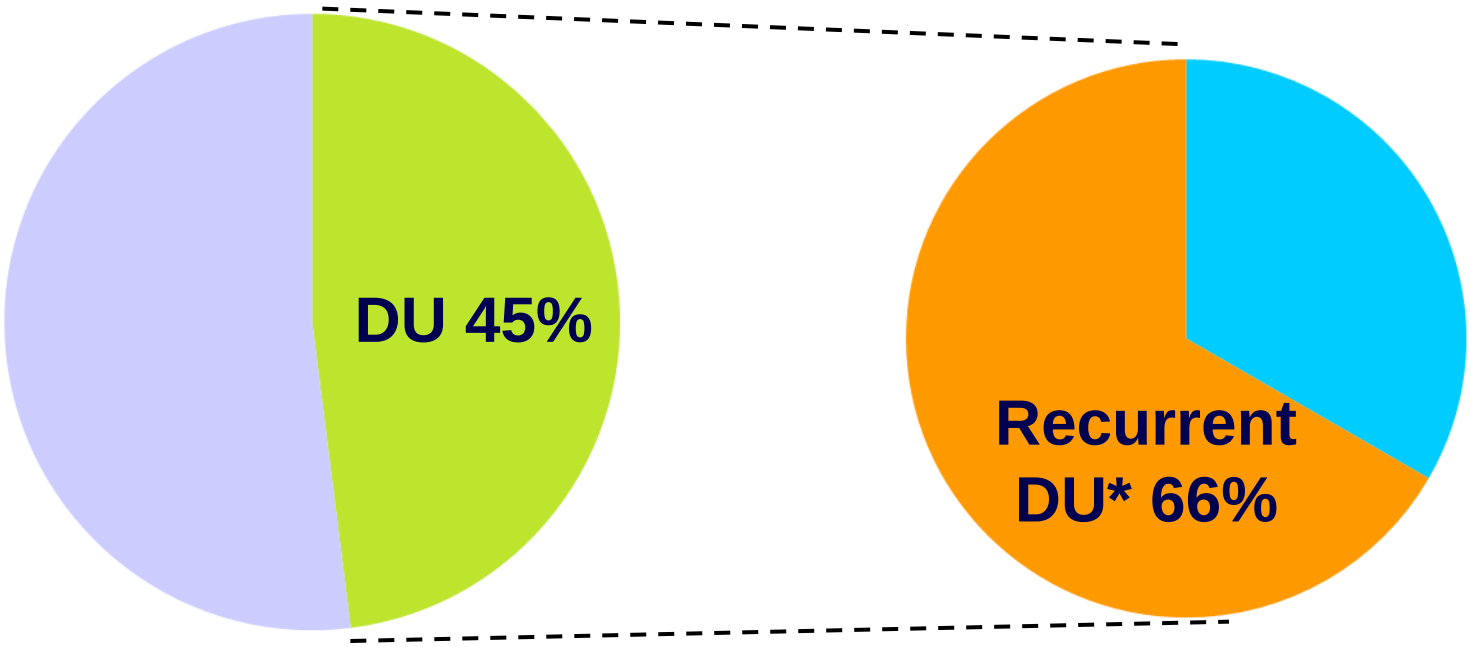
Atrophic phase



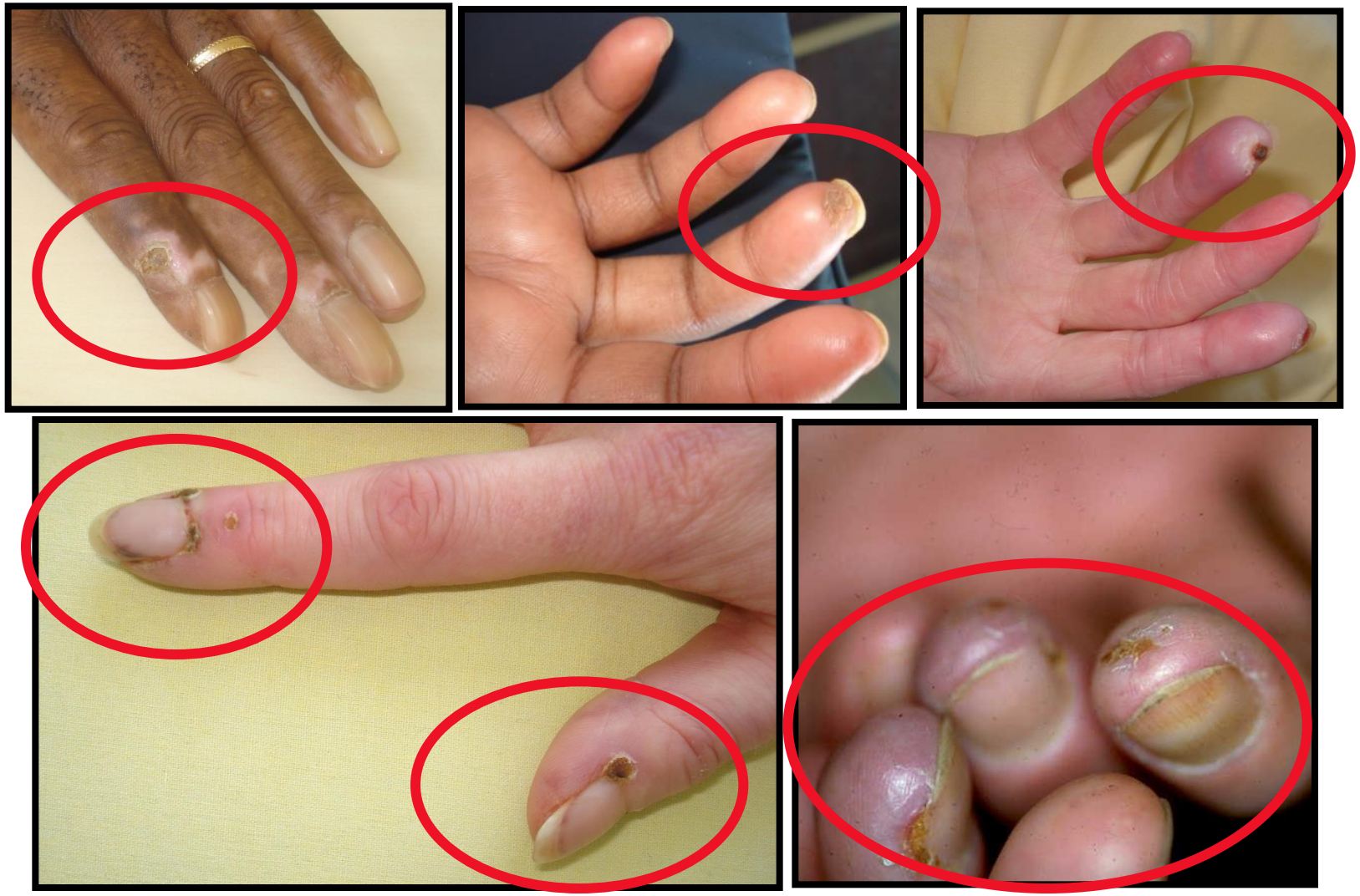
Calcinosis: spontaneous elimination



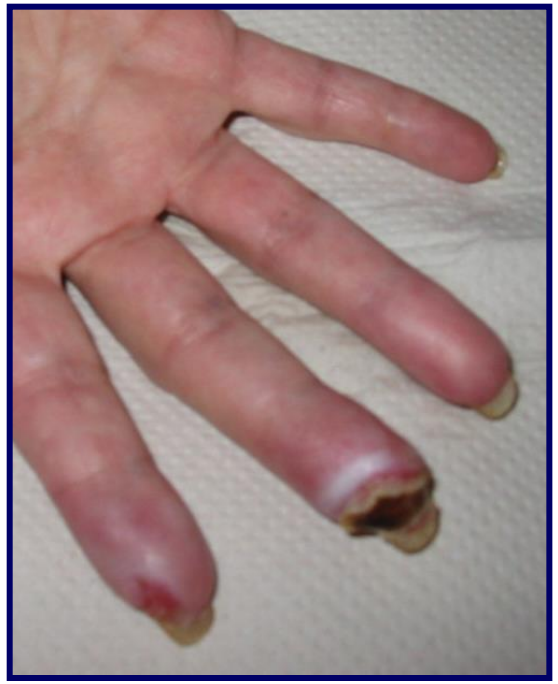
DU are a common and recurrent manifestation of SSc

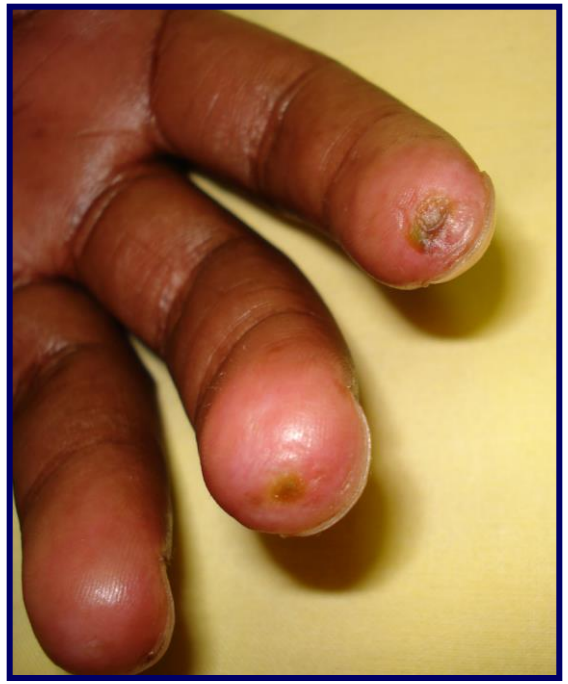


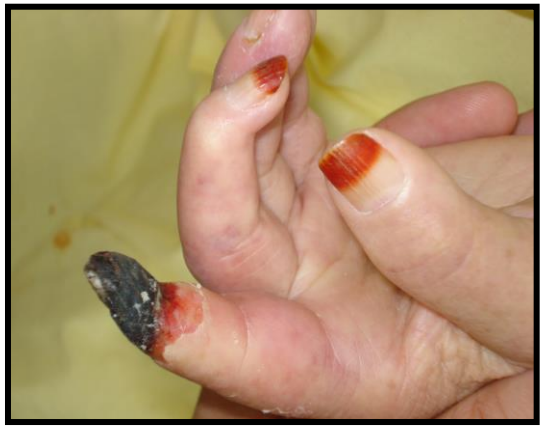
Digital ulcers:
Vascular mechanisms

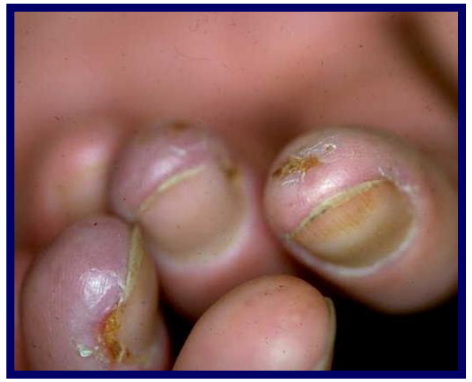


IMPACT OF DIGITAL ULCERS IN SYSTEMIC SCLEROSIS







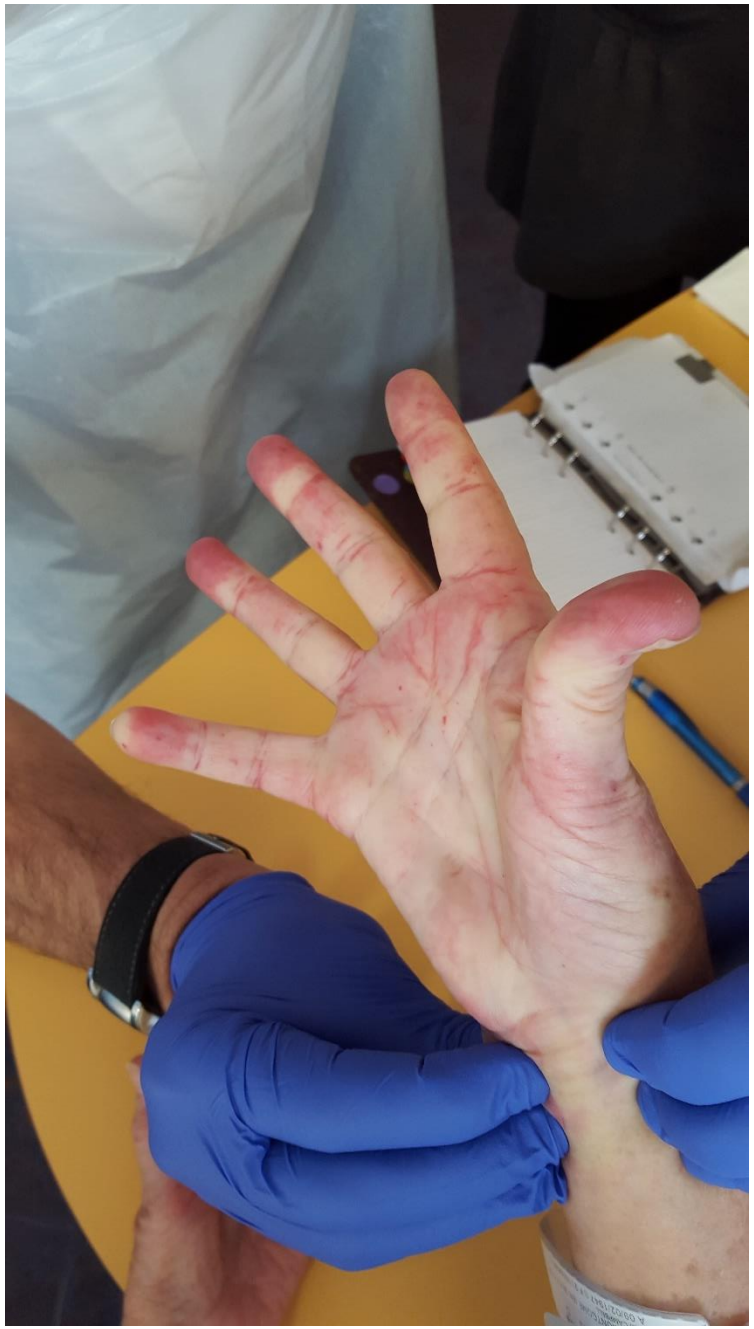


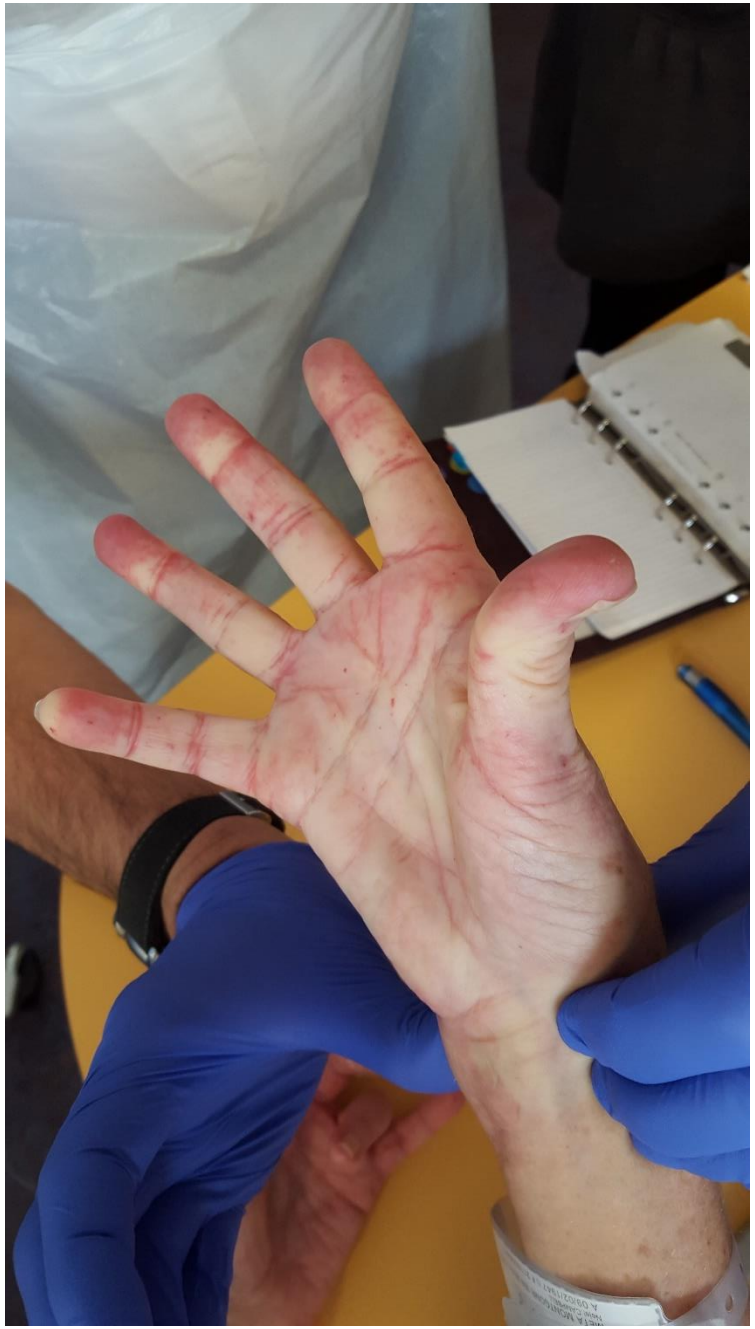
Infection
Gangrene
Amputation

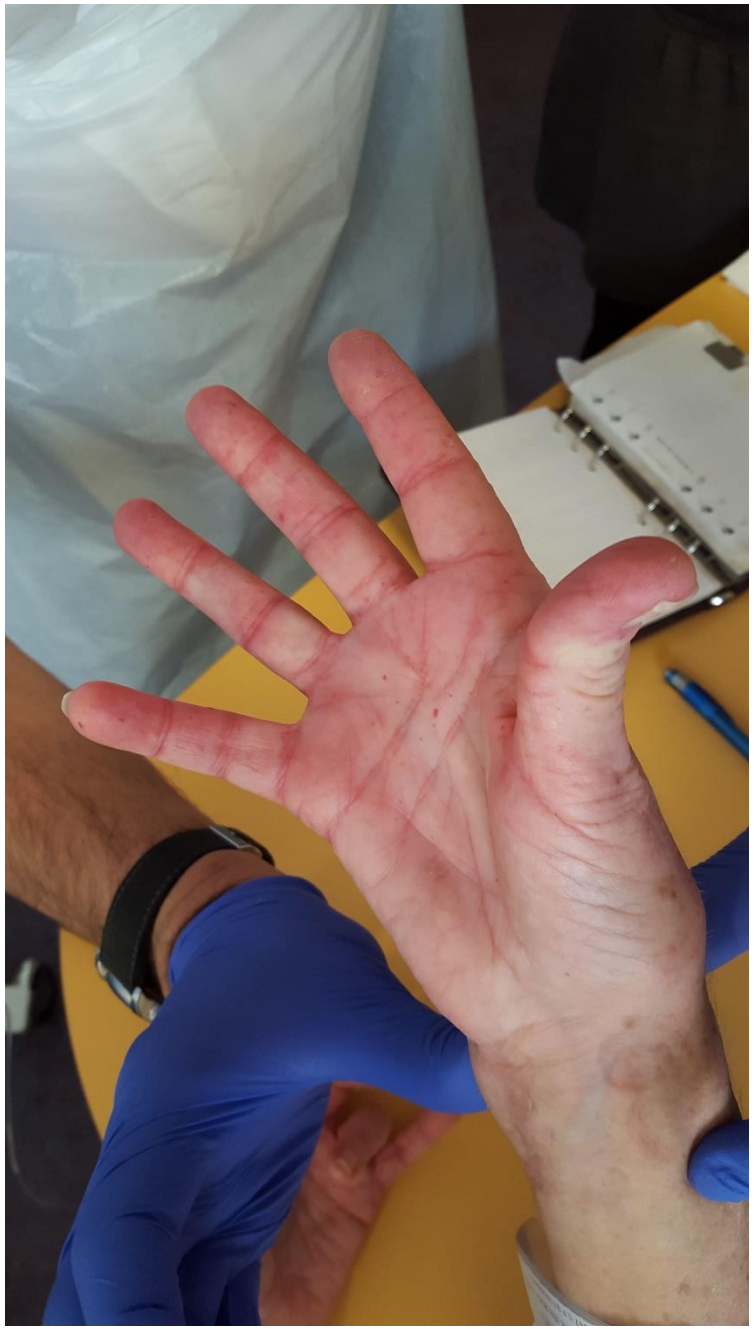
→

Disability
Pain
Loss of function

Occlusion de l’artère ulnaire: Manœuvre d’Allen

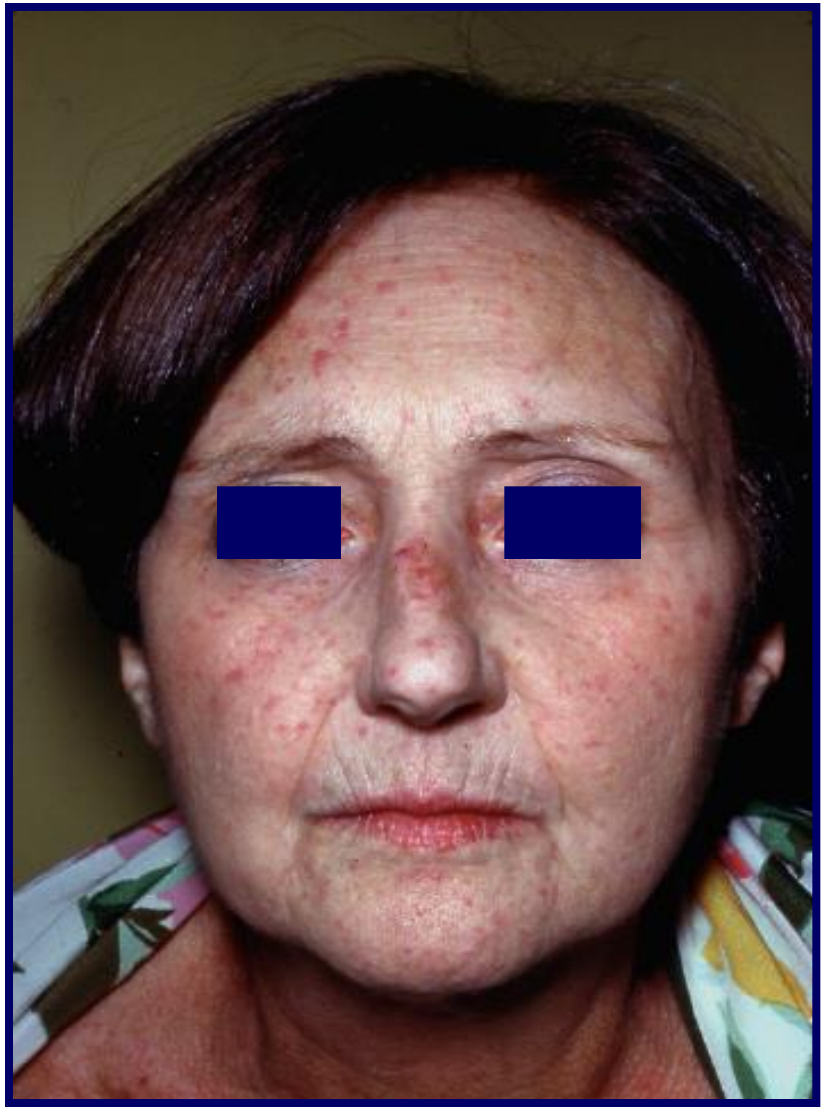


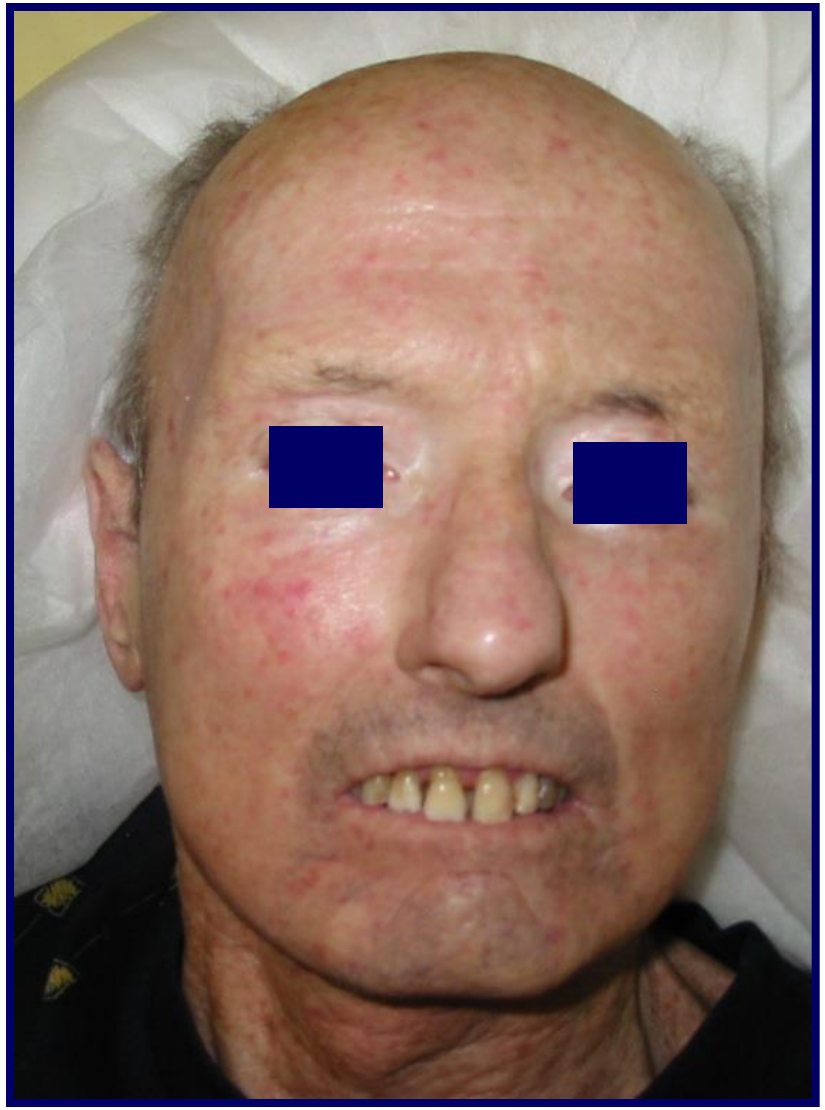




Le visage

- Télangiectasies
- Ouverture buccale
- Syndrome sec
- Etat dentaire

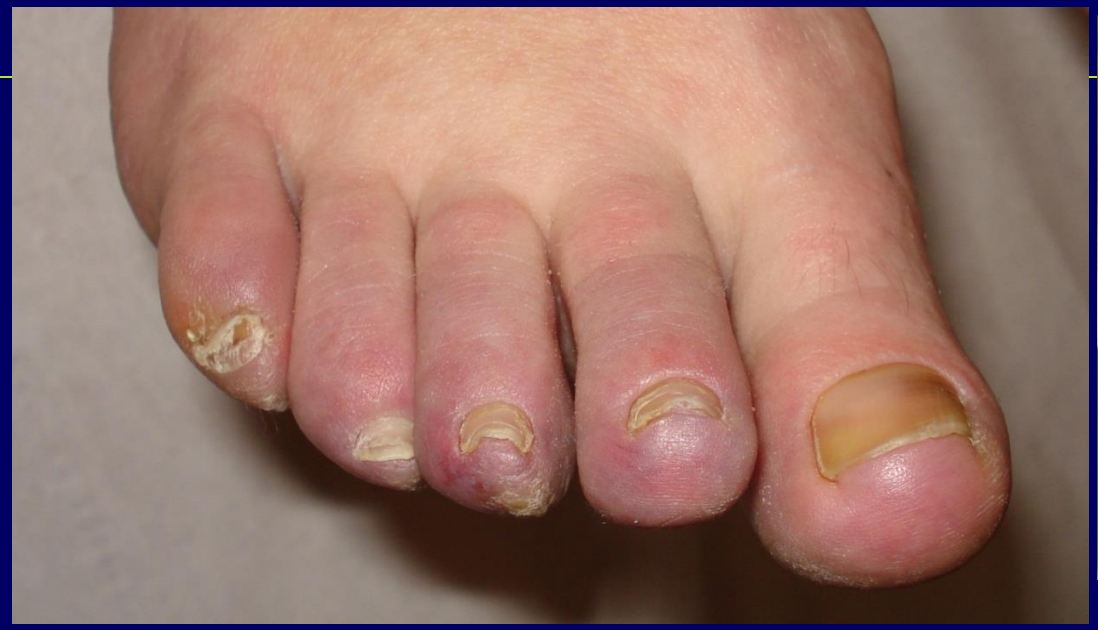


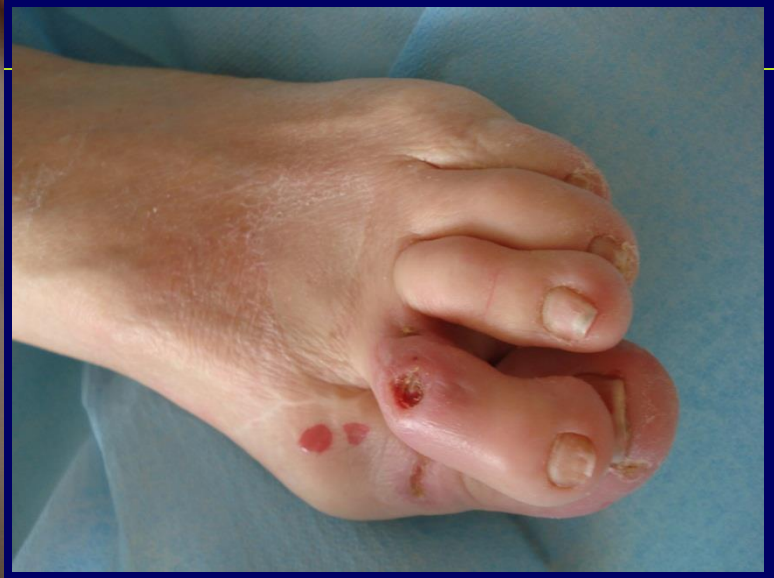


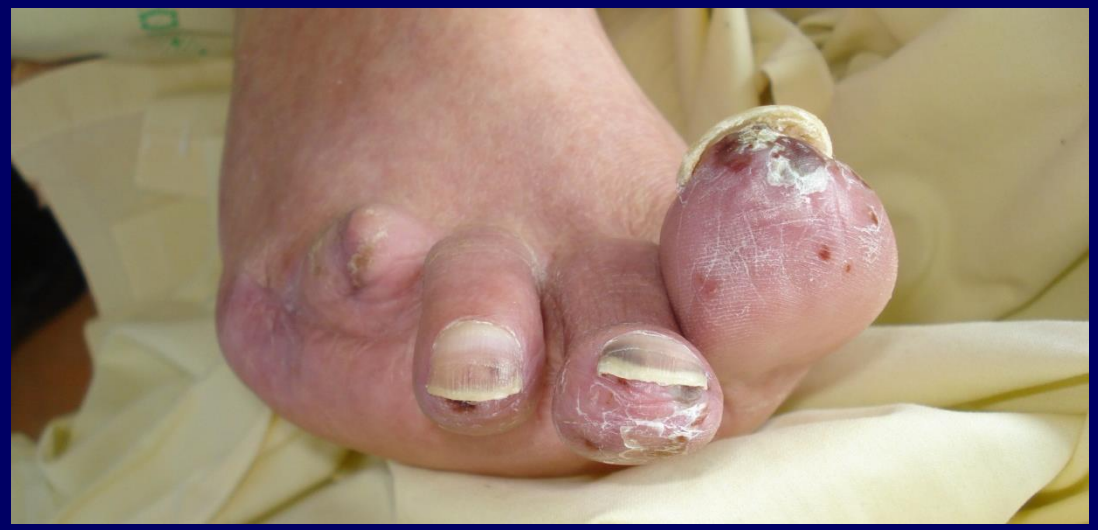
Les pieds

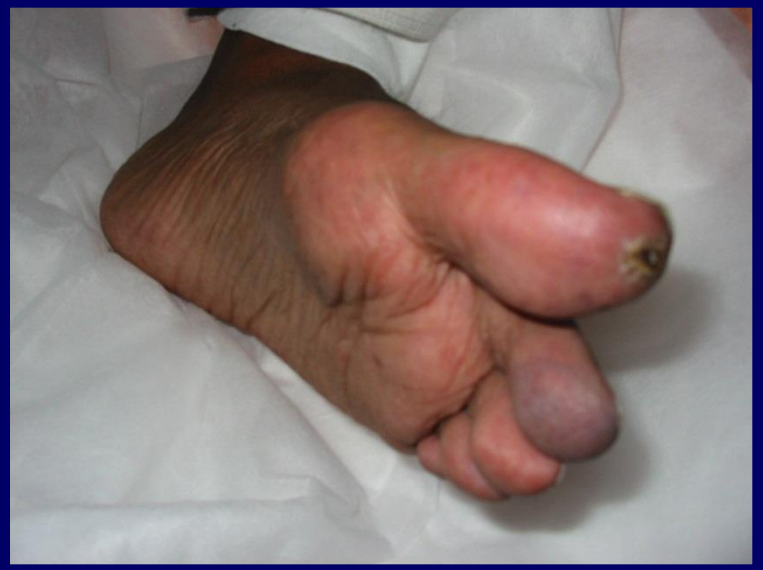
- Déformations
- Durillons plantaires
- Ulcérations

Digital ulcers: Also on the feet!









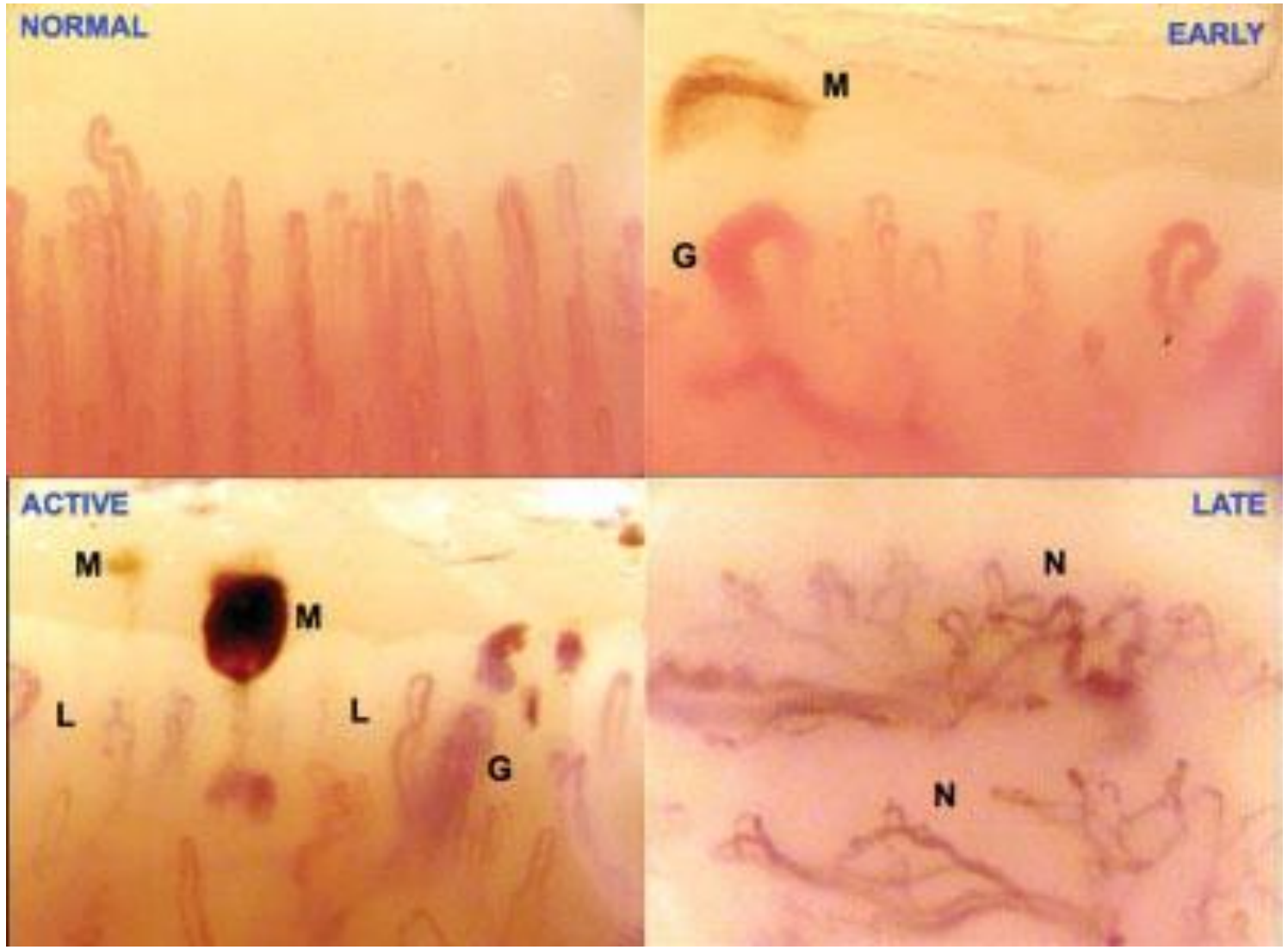
Sclérodermie systémique: bilan initial (examens biologiques)

- **NFS plaquettes**
- **Ionogram, creatinin**
- **CRP**
- **Liver enzymes**
- **uricemia**
- **CK**
- **Serum protein electrophoresis**
- **NT pro-BNP**
- **ANA**
- **Anti-ECT**
- **Anti-RNA polymerase III**
- **Anti-fibrillarine**

Sclérodermie systémique: bilan initial (autres examens complémentaires)

- Capillaroscopie
- ECG
- Radiographie des mains de face
- Echographie cardiaque avec mesure PAPs (insuffisance tricuspidienne)
- EFR avec mesure DLCO
- Scanner thoracique coupes fines haute résolution
- Fibroscopie oeso-gastro-duodenale: dans les 5 premières années
- Echo-doppler artériel membres supérieurs

Capillaroscopie



Herrick A & Cutolo M A&R 2010

Anomalies électrocardiographiques au cours de la sclérodermie systémique

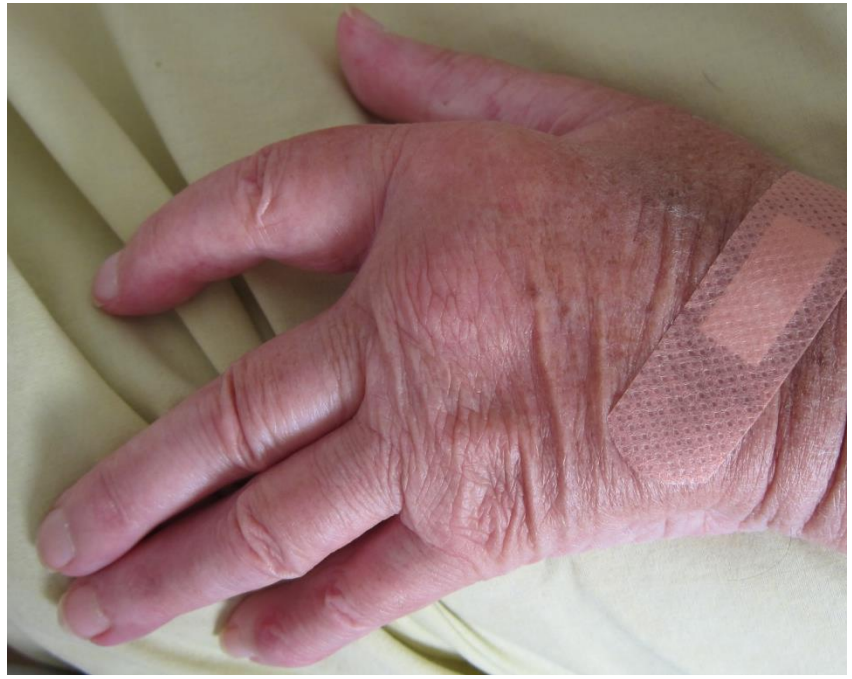
Anomalie	Incidence
Anomalies non-spécifiques du segment ST	12-15%
Micro-voltage	10-28%
Hypertrophie ventriculaire gauche	2-35%
Hypertrophie ventriculaire droite	4-9%
Onde P bifide ou biphasique	45-60%
Hypertrophie auriculaire	3%
Bloc auriculo-ventriculaire (premier degré)	6-10%
Bloc auriculo-ventriculaire (2ème et 3ème degrés)	0-2%
Bloc de branche droit	3-6%
Bloc de branche gauche incomplet	7-16%
Bloc de branche gauche complet	3-6%
Anomalies de conduction ventriculaire non-spécifiques	2-3%
Infarctus septal	10%
Arythmie auriculaire	4%
Arythmie ventriculaire	4-6%

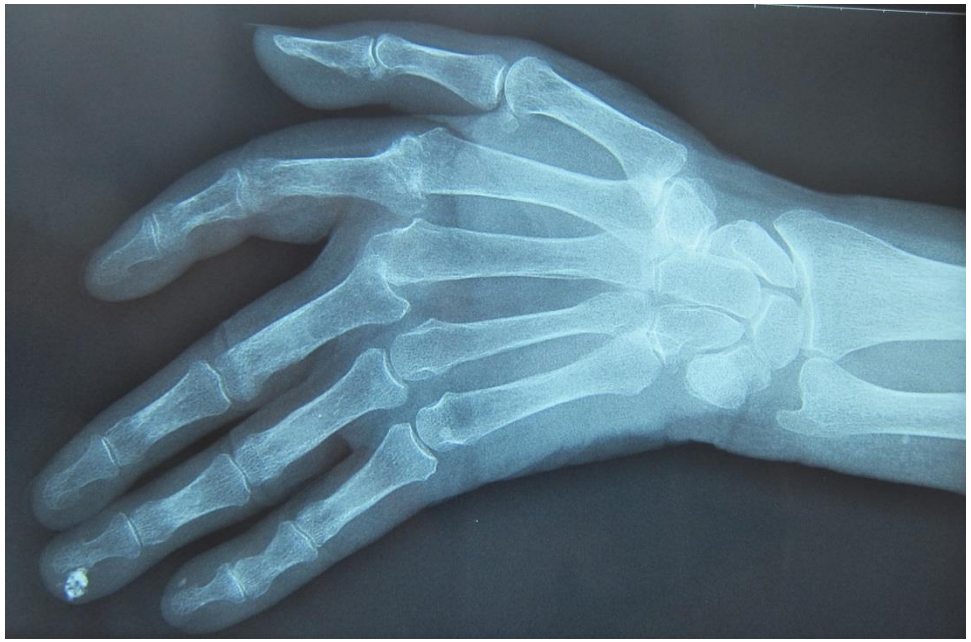
Radiological hand involvement in systemic sclerosis

- 120 consecutive SSc patients
- Radiological abnormalities in SSc:
  - Erosion (21%)
  - Joint space narrowing (28%)
  - Arthritis (erosion and joint space narrowing) (18%)
  - Demineralisation (23%)
  - Acro-osteolysis (22%)
  - Flexion contracture (27%)
  - Calcinosis (23%)

Avouac J, *et al. Ann Rheum Dis* 2006;

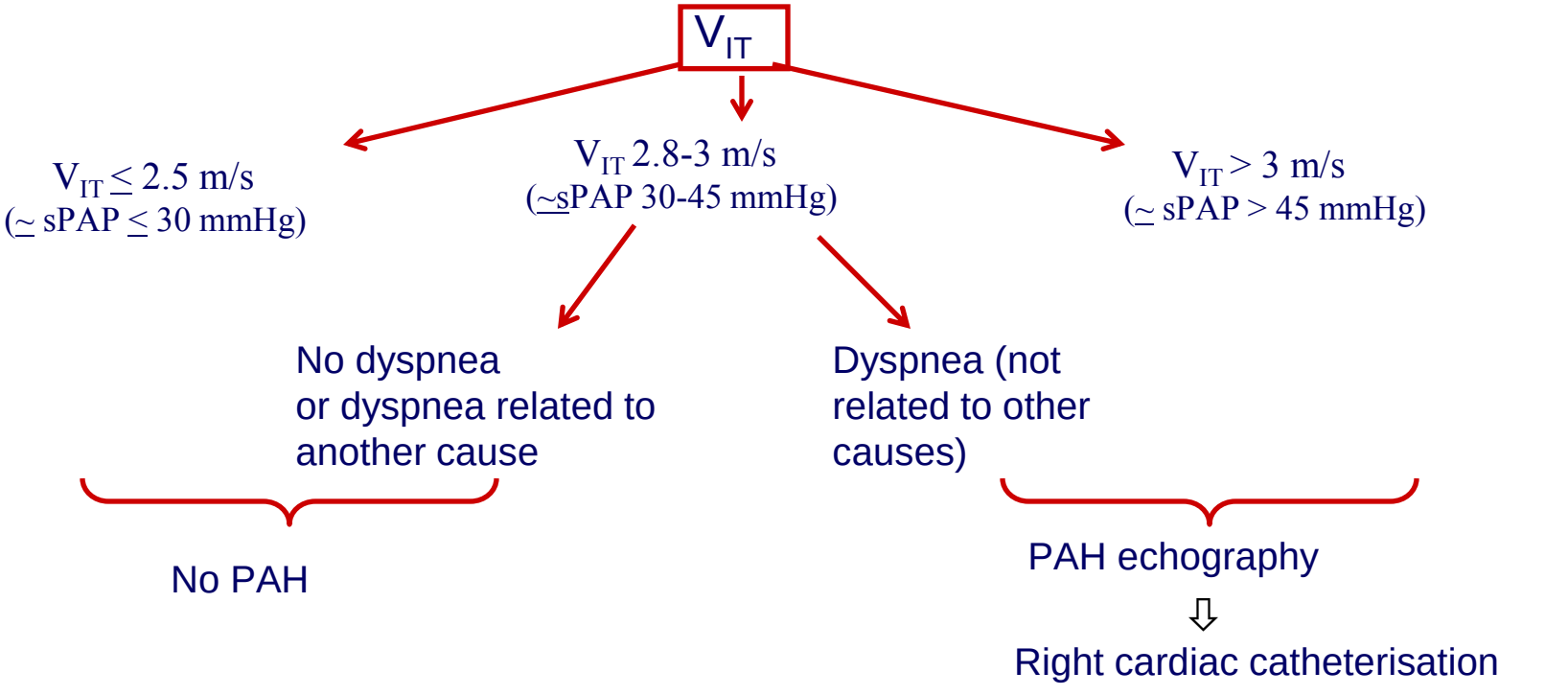
Radiographie des mains





Sclérodermie systémique, arthropathie, calcinose

Cardiac EchoDoppler
PAH definition

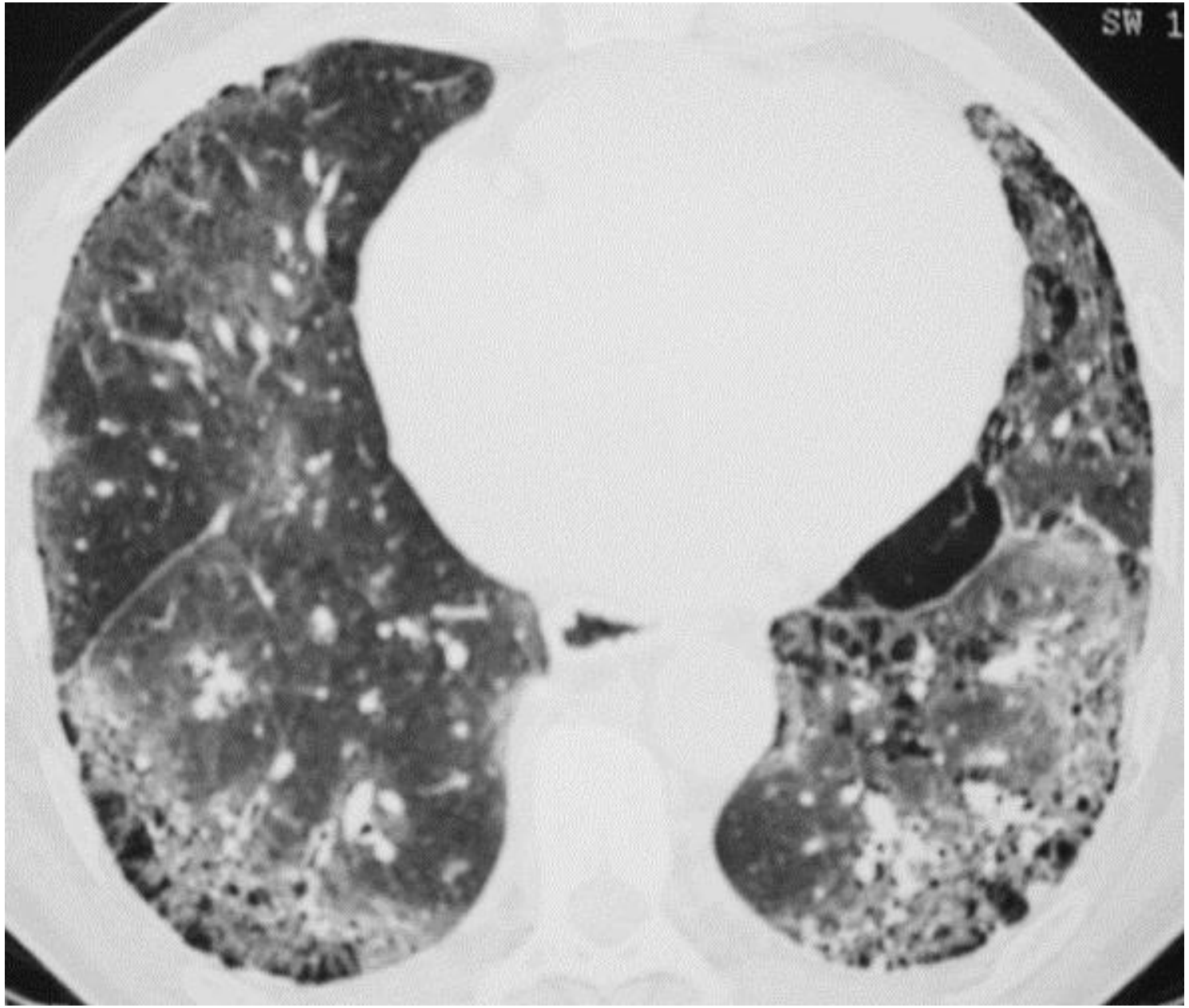


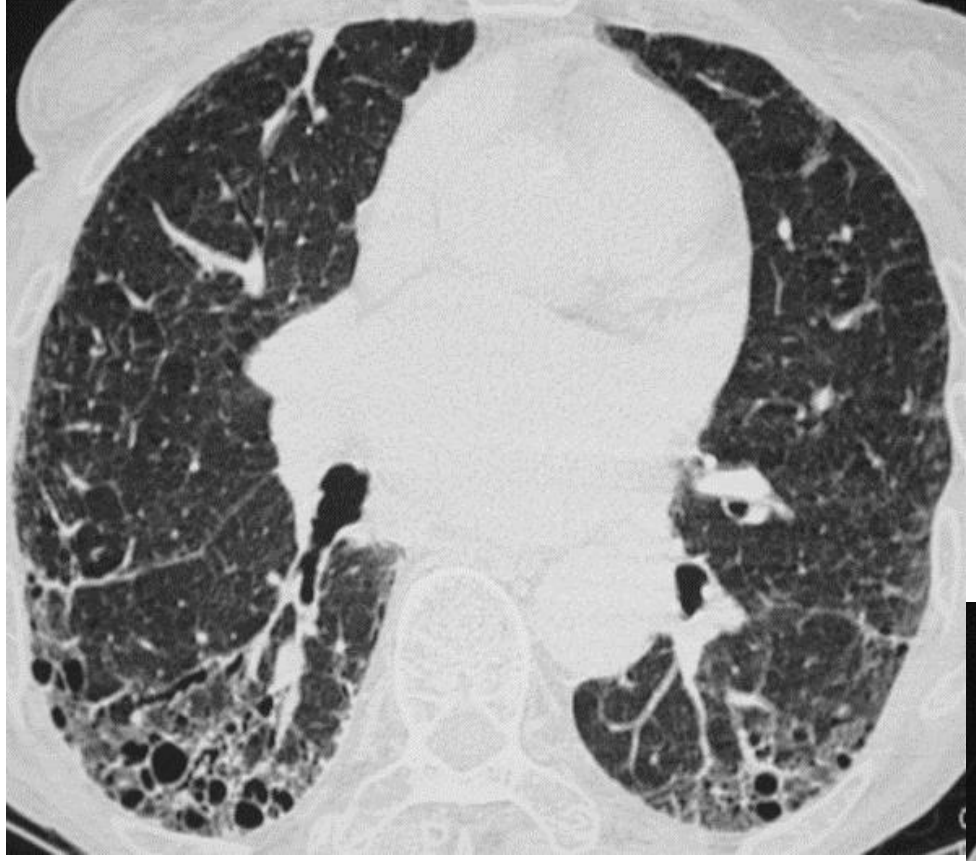
Hachulla et al. Arthritis Rheum 2005
Hachulla et al. Arthritis Rheum 2009

PID: Examens complémentaires

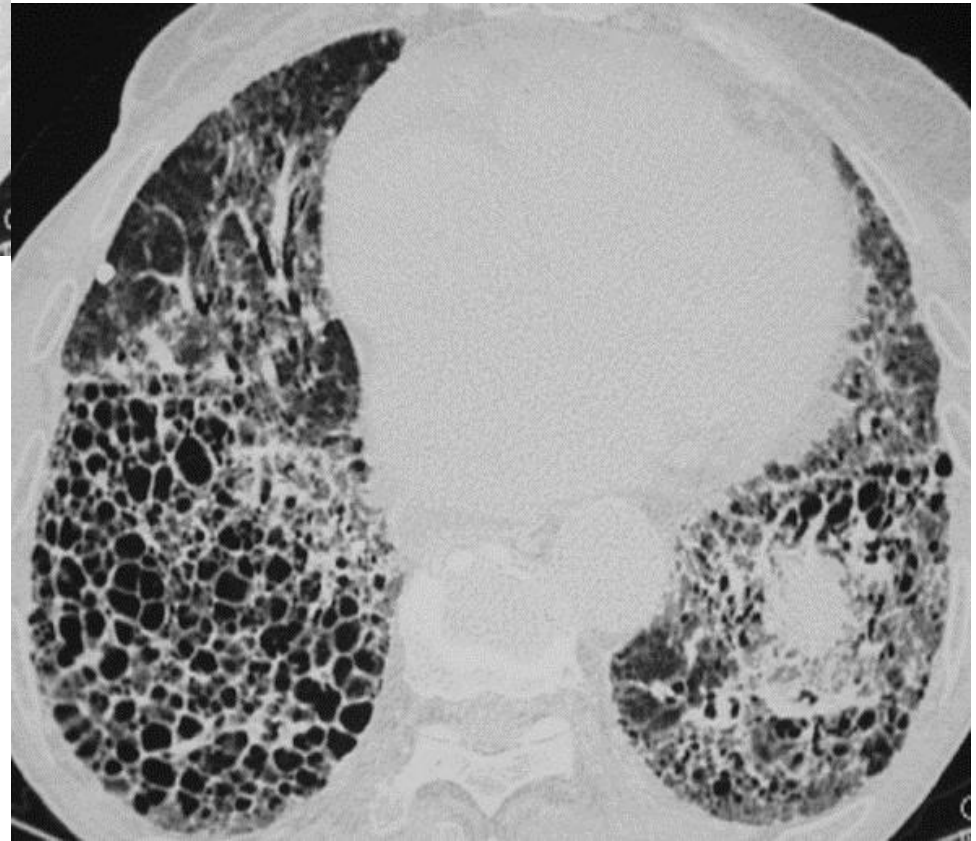
- Le mauvais pronostic de la PID au cours de la ScS impose son **dépistage systématique**.
- Pas de recommandations consensuelles sur les examens de dépistage et la fréquence à laquelle les renouveler.
- Le bilan doit comporter:
  - tomodensitométrie thoracique haute résolution (TDMHR)
  - épreuves fonctionnelles respiratoires (EFR) avec mesure du coefficient de transfert du monoxyde de carbone (DLCO)
  - test de marche de 6 min avec mesure de la saturation en oxygène et l'estimation de la dyspnée à l'aide de l'indice de Borg.

Non specific interstitial pneumonia

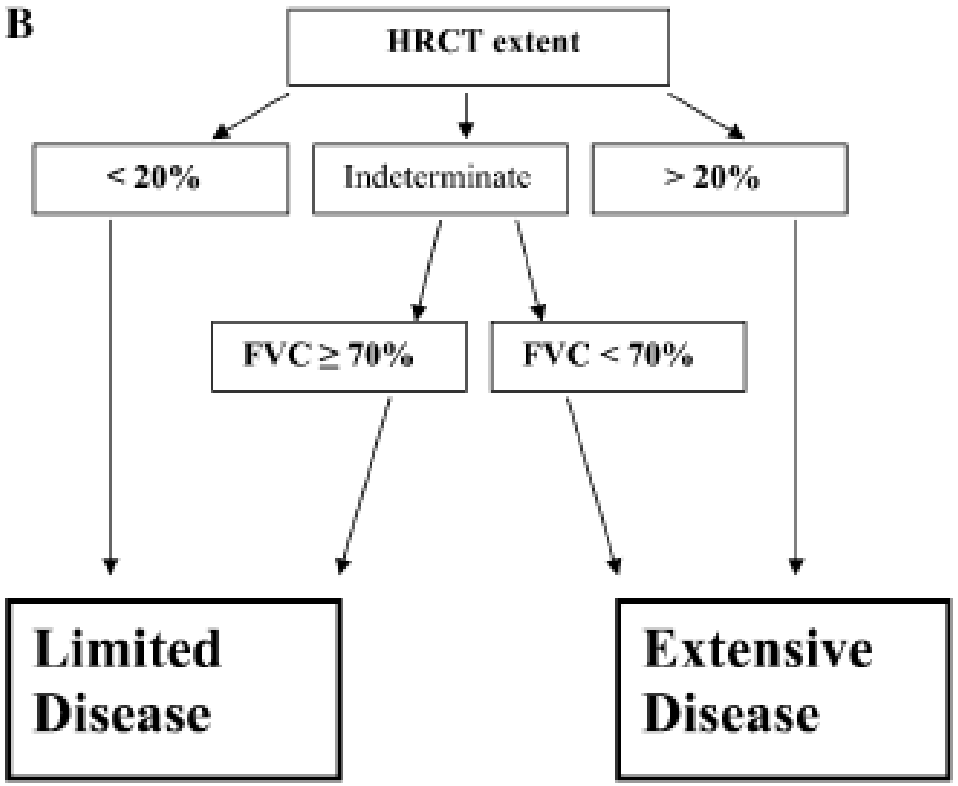


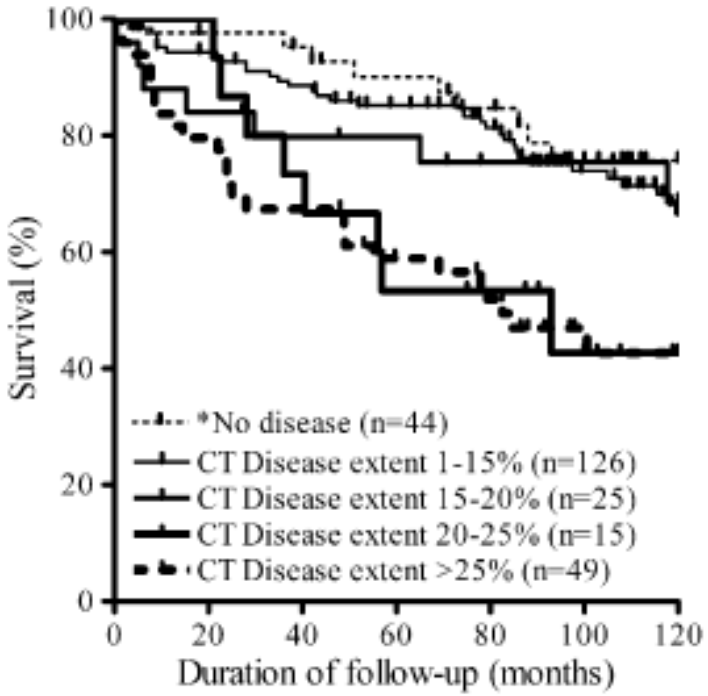


Usual interstitial pneumonia



Interstitial Lung Disease in Systemic Sclerosis
A Simple Staging System
Goh NSL, AJRCCM 2008





Combined Pulmonary Fibrosis and Emphysema Syndrome in Connective Tissue Disease

Table 1. Classification of connective tissue diseases in the 34 study patients*

Rheumatoid arthritis	18 (53)
Systemic sclerosis	10 (29)
Diffuse cutaneous	3 (9)
Limited cutaneous	7 (20)
Mixed connective tissue disease	2 (6)
Overlapping connective tissue disease	2 (6)
Sjögren's syndrome	1 (3)
Polymyositis	1 (3)

* Values are the number (%) of patients.

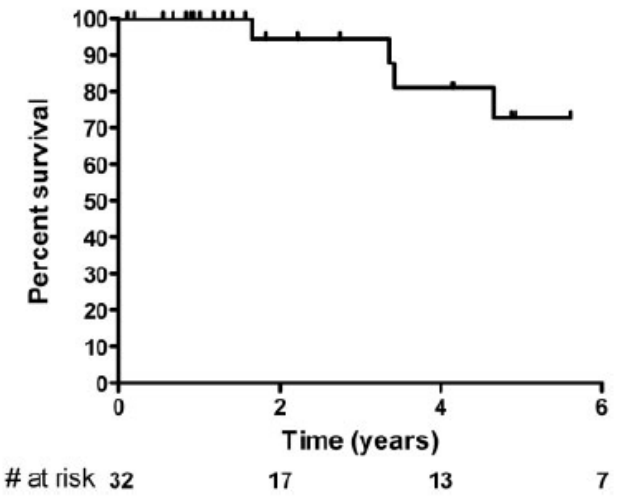
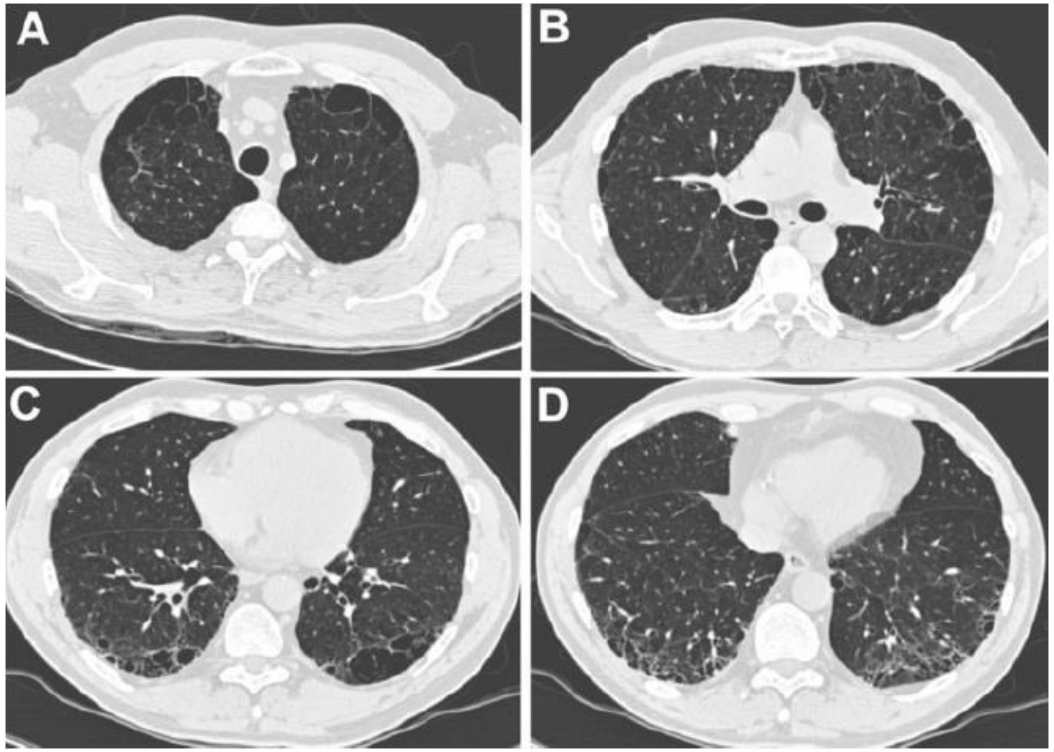
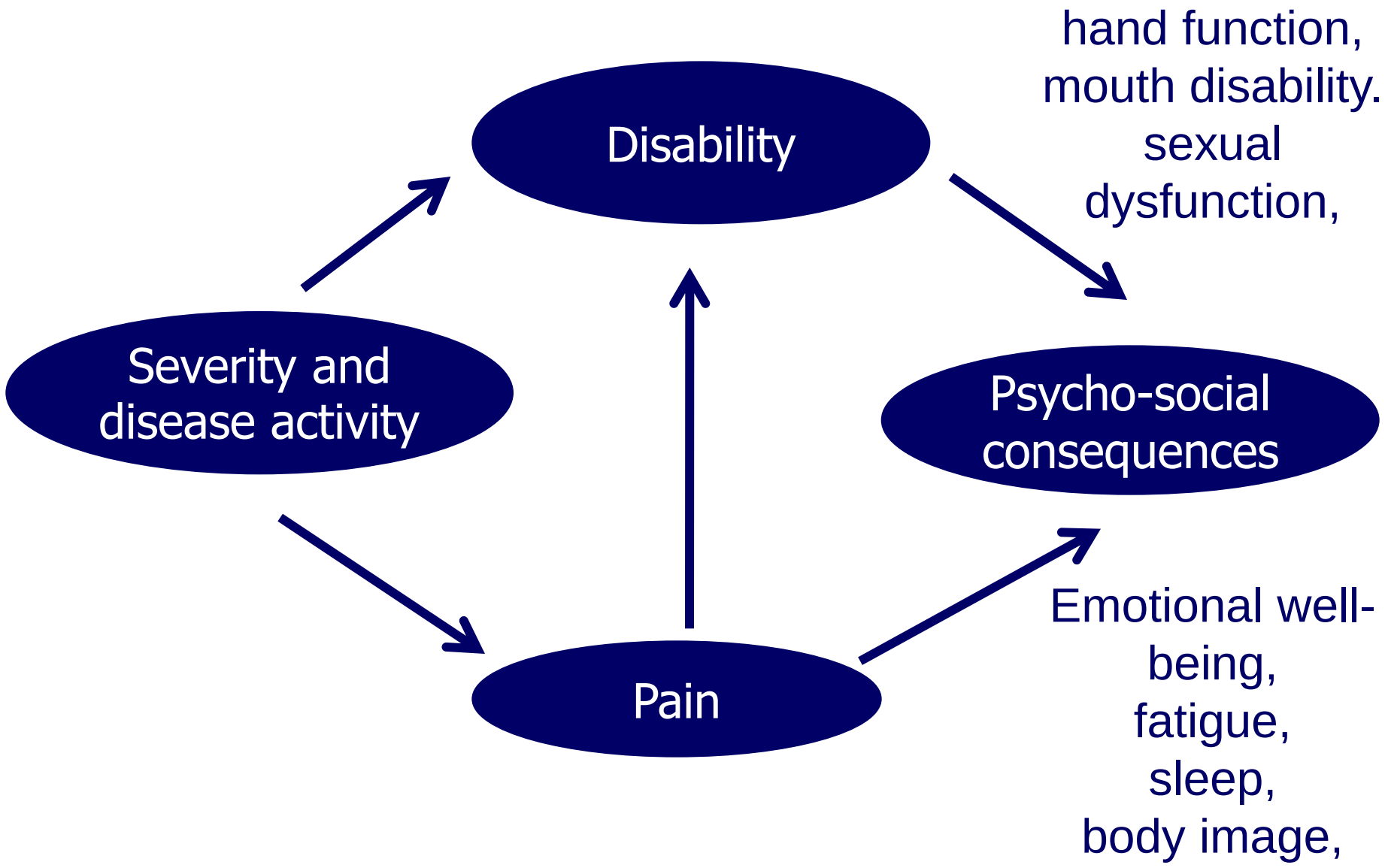


Figure 2. Kaplan-Meier estimates of survival in patients with combined pulmonary fibrosis and emphysema syndrome and connective tissue disease.



Quality of life (QOL) in SSc



Conclusion

Differentiate diffuse / limited
Initial evaluation very important to detect worsening
Perform systematic clinical examination
Perform biological exams
Perform complementary exams to detect early visceral involvement
Detect early disability

GFRS

CMR
CENTRE MALADIES RARES
VASCULARITES I SCLÉRODERMIES
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