La sclérodermie systémique

Divi Cornec, Rhumatologie, Brest

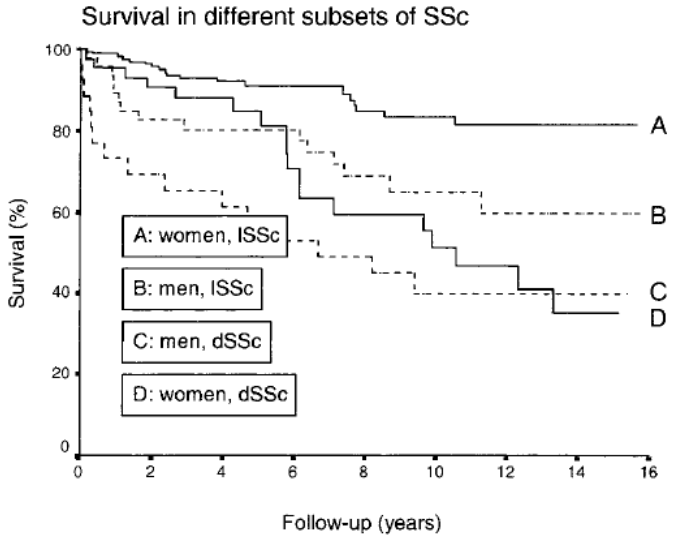
Christophe Gut-Gober, Pneumologie, Brest

Deux types de sclérodermie systémique

LeRoy et Metsger, J Rheumatol 2001;28:15736.

SSc limitée

- Distale (<coudes/genoux, face)
- Anticorps anti-centromères
- « CREST »
- HTAP tardive



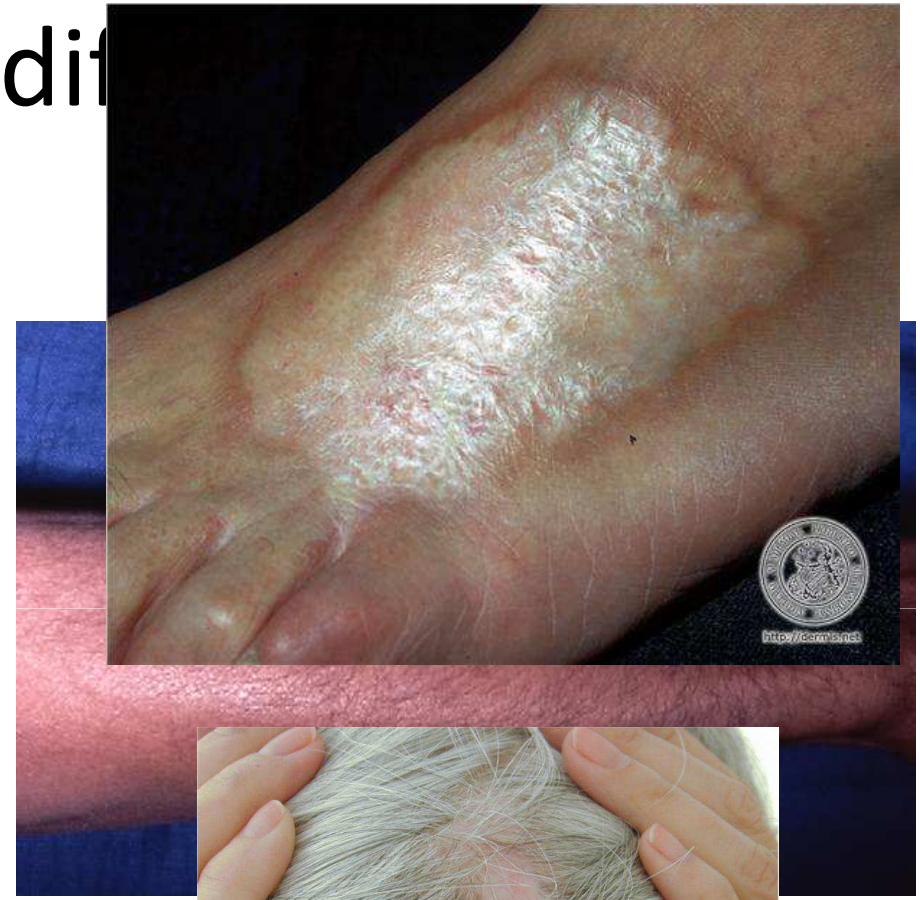
SSc diffuse

- Proximale (>coudes/genoux, tronc)
- Anti-topoisomérase (Scl70)
- Atteinte d'organes
- Traitement immunosuppresseur

Hesselstrand et al, Rheumatology (Oxford). 2003 Apr;42(4):534-40.

Diagnostics dif

- Sclérodermie localisée (morphée, en coup de sabre)
- Fasciite à éosinophiles (Shulman)
- Fibrose néphrogénique (gadolinium, IRC)
- GVH cutanée



Description de la maladie : cohorte de l'EUSTAR

- **7655 patients inclus en 2011**
- Sex ratio : F 86%, H 14%
- Raynaud 96%
- Début du Raynaud : 42 ans
- RGO 67%
- Ulcères digitaux : 36% (actifs 17%)
- FAN 96%, ACA 32%, anti-Scl70 37%

- Traitements :
  - IPP 65%
  - Inhibiteur calcique 53%
  - Prednisone 45%
  - IEC 21%
  - Endoxan 16%
  - AINS 15%
  - Methotrexate 14%
  - Diurétiques 13%
  - Bosentan 10%

Meier at al, Ann Rheum Dis. 2012 Aug;71(8):1355-60.

Complications sévères de la sclérodermie : la règle des 15%

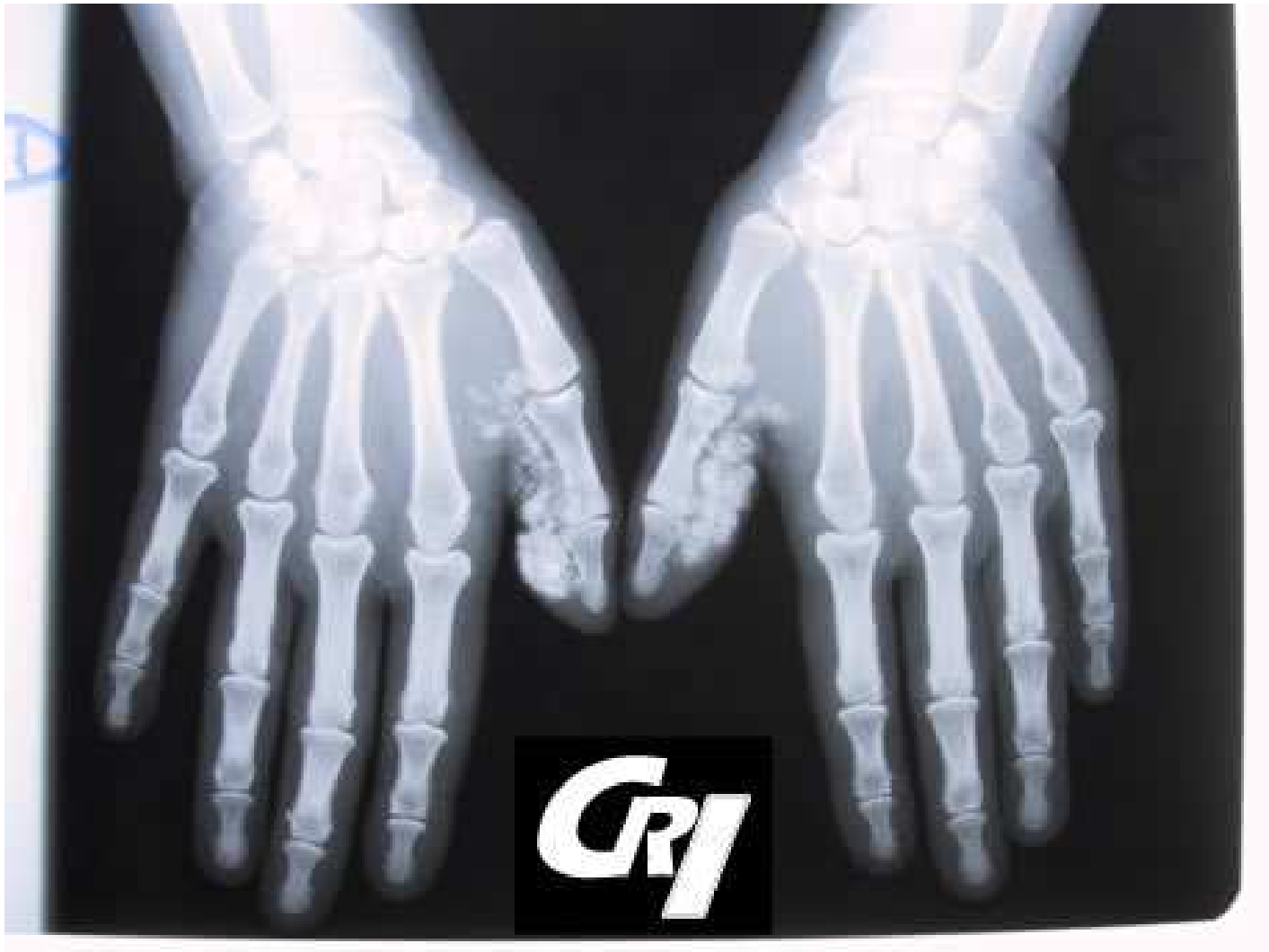
- Méta-analyse sur 69 articles
- Prévalence des complications
  - Atteinte myocardique : 15%
  - HTAP (KT droit) : 15%
  - Pneumopathie intestitielle (CVF<70%) : 15%
  - Myosite : 13%
  - Arthrite clinique : 12%
  - Ulcères digitaux : 15%
  - Sjögren secondaire : 13%
  - *Crise rénale : 3%*

Muangchan C; Canadian Scleroderma Research Group, J Rheumatol. 2013 Sep;40(9):1545-56

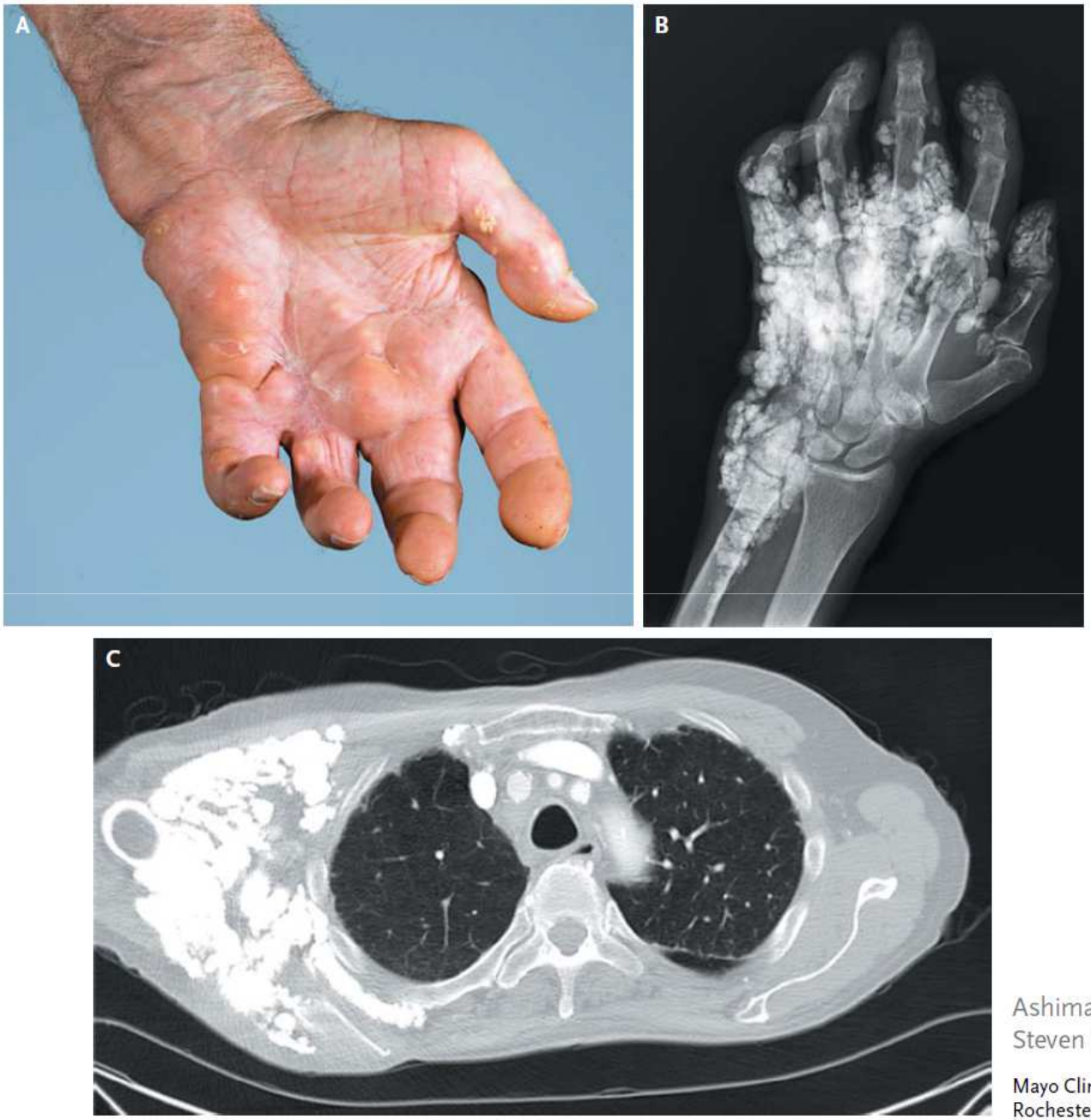
Traitement de l'arthrite inflammatoire

- AINS : tolérance digestive…
- Corticoïdes : pas plus de 10 mg/j !
- Methotrexate ?
  - Pas de preuve de son efficacité
  - Un essai randomisé négatif dans la scérodermie (Pope et al, Arthritis Rheum. 2001 Jun;44(6):1351-8.)
- Anti-TNF ?
  - Déconseillé par des experts de l'EUSTAR (Distler et al, Clin Exp Rheumatol. 2011 Mar-Apr;29)
- Rituximab ?
  - Pas d'effet sur la fibrose cutanée (Lafyatis et al, Arthritis Rheum. 2009 Feb;60(2):578-83.)
  - Un essai contre placebo débute en France (étude RECOVER, NCT01748084, Pr Allanore à Cochin)
- Tocilizumab ou abatacept ?
  - 1 série de 20 patients (Elhai et al., Ann Rheum Dis 2013;72:1217–1220.)
  - Efficacité sur la polyarthrite
  - Pas d'effet apparent sur fibrose
  - bien tolérés

La calcinose



N Engl J Med. 2011 Jun 9;364(23):2245, Bisson-Vaivre et al, Diagn Interv Imaging. 2013 Jun;94(6):645-7, Aggarwal, N Engl J Med. 2013 May 23;368(21):e28.



Ashima Makol, M.D.
Steven R. Ytterberg, M.D.

Mayo Clinic
Rochester, MN

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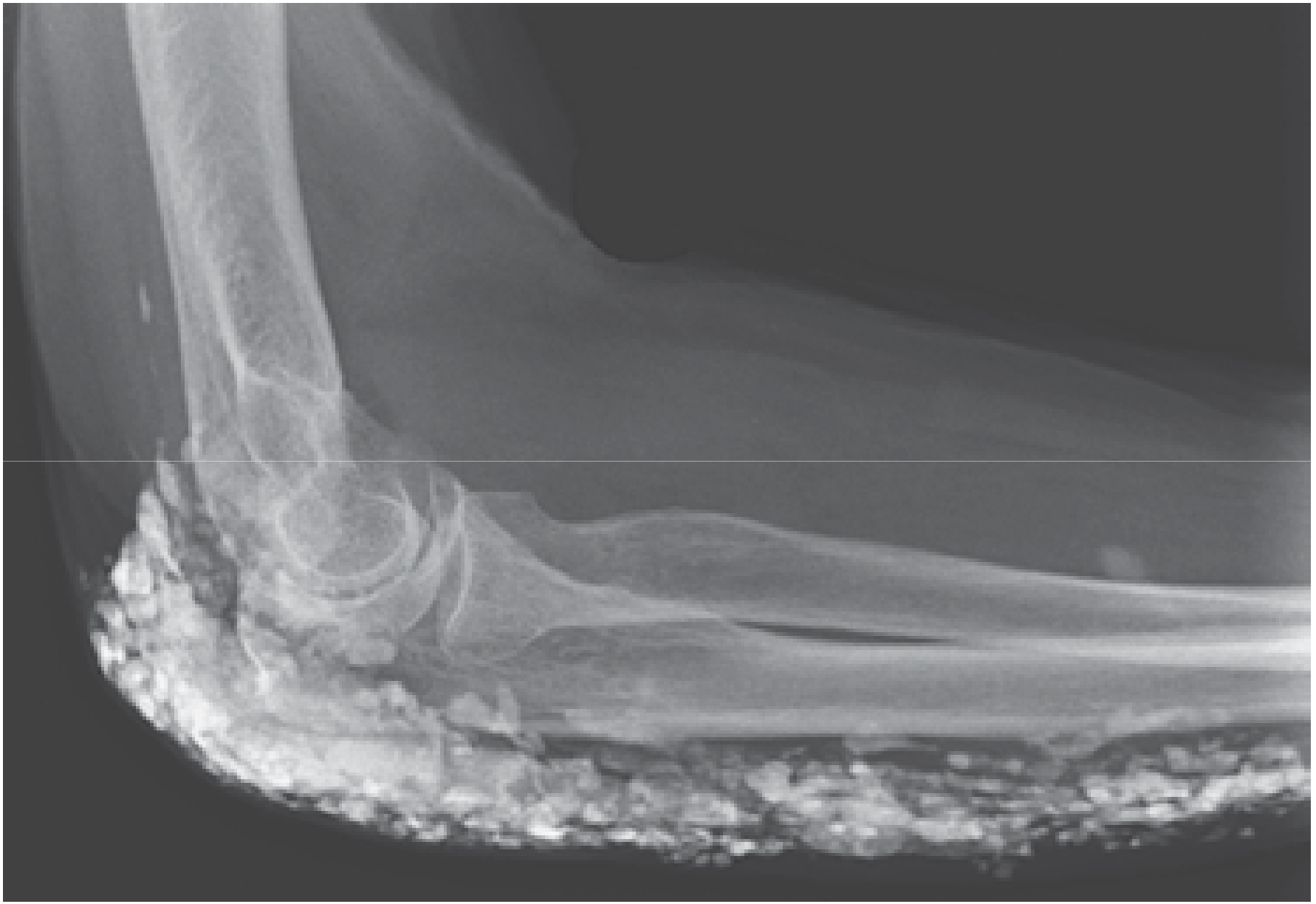

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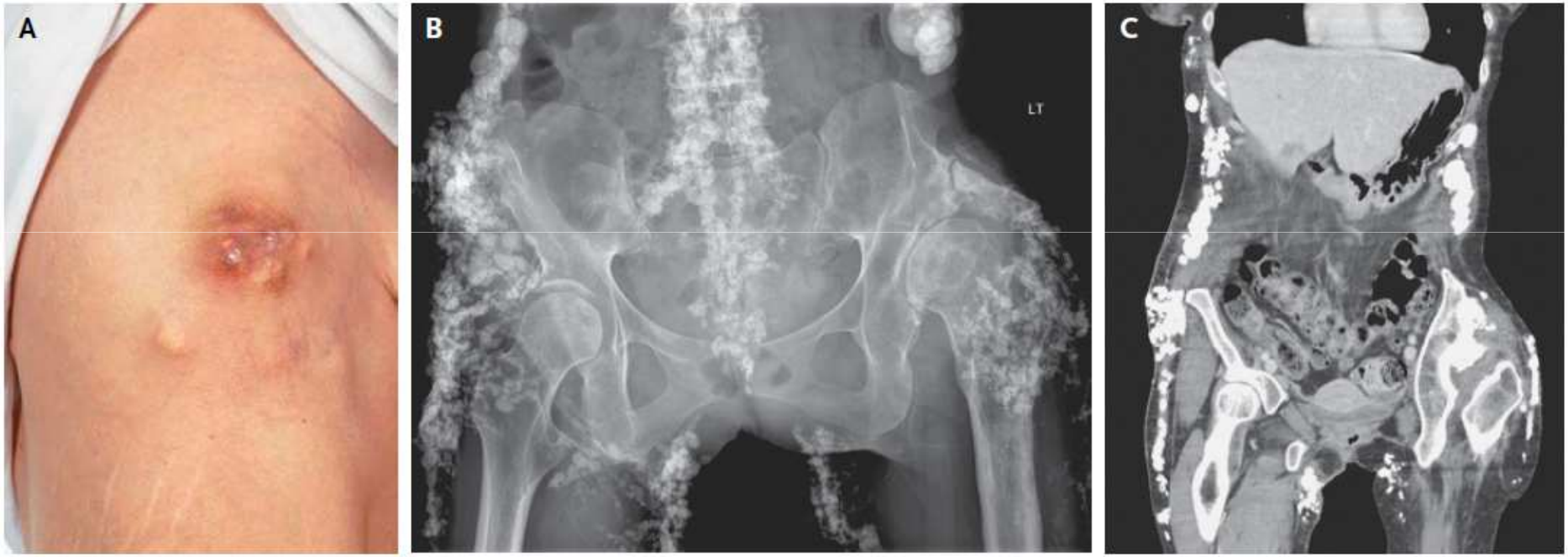

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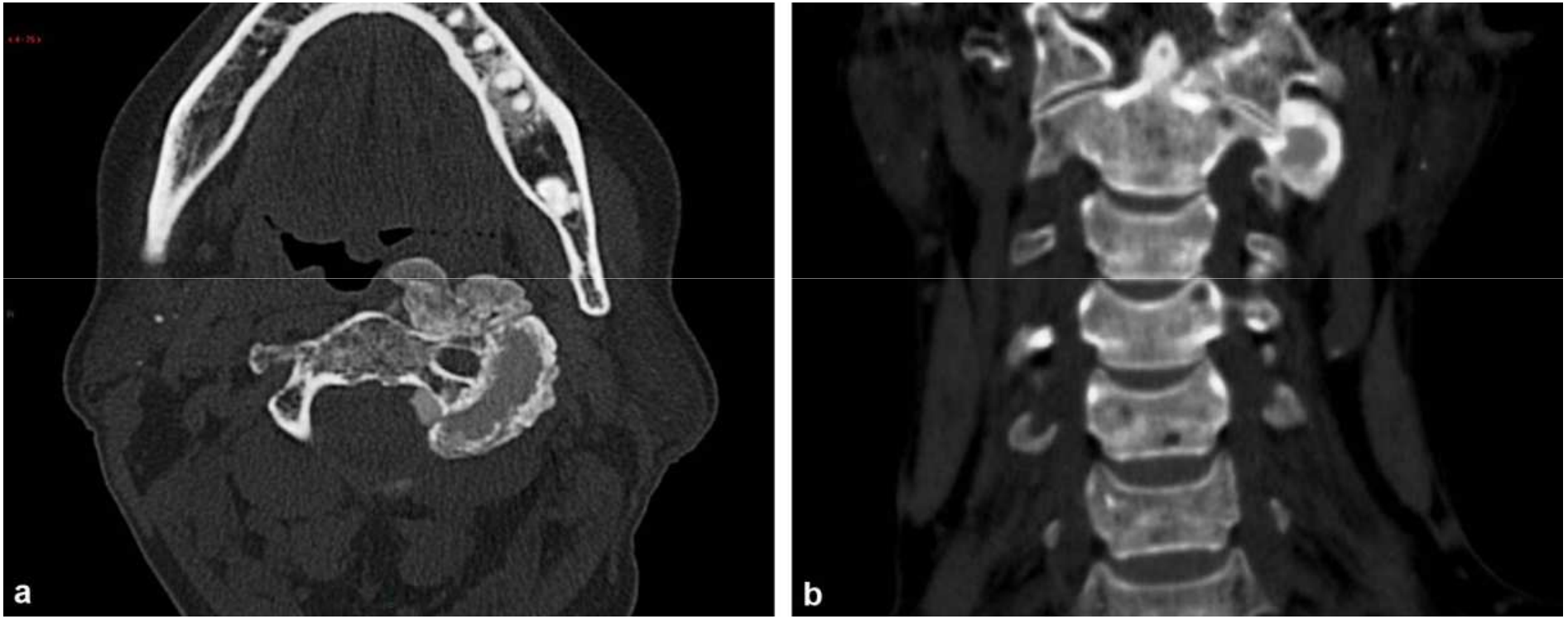

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Traitement de la calcinose

- Aucun traitement n'a prouvé son efficacité…
- A priori pas efficaces : bisphosphonates, calcitonine, IVIg, anticoagulants, immunosuppresseurs
- Peuvent être envisagés
  - Traitement du Raynaud +++, protection froid/trauma
  - AINS/Antalgiques/Corticoïdes
  - Colchicine
  - Chirurgie si gêne fonctionnelle majeure (récidive…)

Ulcérations digitales

- Prévention +++ (éviction froid, traumatismes, médicaments favorisants)
- ARRET DU TABAC !
- Inhibiteurs calciques, IEC/ARA2, (kardégic ?)
- Bosentan : en prévention secondaire, diminue le nombre de récidive
- Traitement curatif :
  - Soins locaux
  - Iloprost IV

Difficultés sexuelles

Arthritis Care & Research
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ORIGINAL ARTICLE

Rates and Correlates of Sexual Activity and Impairment Among Women With Systemic Sclerosis

BROOKE LEVIS,[1] MARIE HUDSON,[1] RUBY KNAFO,[1] MURRAY BARON,[1] WARREN R. NIELSON,[2] MARILYN HILL,[2] BRETT D. THOMBS,[1] AND THE CANADIAN SCLERODERMA RESEARCH GROUP (CSRG)

Table 1. Rates of sexual activity without impairment by age group and marital status

	N	No. (%) active	No. (%) not impaired among active	No. (%) active without impairment
Age group, years				
18–39	25	13 (52)	7 (54)	7 (28)
40–49	103	67 (65)	39 (58)	39 (38)
50–59	171	81 (47)	27 (33)	27 (16)
≥60	248	65 (26)	22 (34)	22 (9)
Marital status*				
Married	373	189 (51)	75 (40)	75 (20)
Not married	167	33 (20)	18 (55)	18 (11)
Total	547	226 (41)	95 (42)	95 (17)

* Seven patients did not report marital status.

Table 3. Unadjusted and adjusted ORs for reporting sexual relations with a partner in the last 4 weeks (n = 237)*

	Unadjusted OR	95% CI	*P*	Adjusted OR	95% CI	*P*
Age group, years						
18–39	0.39	0.06–2.41	0.310	0.25	0.03–1.97	0.190
40–49	1.00	Reference	–	1.00	Reference	–
50–59	0.42	0.17–1.05	0.062	0.24	0.08–0.73	0.012
≥60	0.31	0.12–0.76	0.011	0.12	0.04–0.37	< 0.001
Not married	2.90	0.64–13.04	0.166			
Nonwhite race/ethnicity	0.85	0.29–2.51	0.763			
Alcohol consumption						
None	1.00	Reference	–			
<1 drink per day	2.41	1.28–4.54	0.007			
≥1 drink per day	8.32	1.88–36.75	0.005			
Smoking status						
Never	1.00	Reference	–			
Former	1.02	0.56–1.88	0.945			
Current	0.71	0.26–1.93	0.498			
Body mass index, kg/m^2						
<25	1.00	Reference	–			
25–29.9	0.49	0.25–0.97	0.041			
≥30	0.29	0.14–0.62	0.001			
Skin scores	0.97	0.93–1.00	0.045	0.98	0.94–1.03	0.382
Disease duration, years	1.00	0.97–1.04	0.827			
Fingertip-to-palm distance	0.90	0.77–1.05	0.191	0.89	0.73–1.09	0.268
Hemoglobin, gm/liter	1.02	1.00–1.04	0.083	1.01	0.99–1.04	0.336
Severity of GI symptoms	0.74	0.65–0.84	< 0.001	0.80	0.69–0.94	0.005
Pulmonary hypertension	0.21	0.08–0.55	0.001			
Interstitial lung disease	0.61	0.34–1.12	0.111	0.81	0.39–1.67	0.569
Finger ulcers	0.79	0.44–1.41	0.424			
Severity of Raynaud's phenomenon symptoms	0.78	0.71–0.87	< 0.001	0.82	0.72–0.93	0.003
Severity of breathing problems	0.81	0.72–0.90	< 0.001	0.95	0.82–1.09	0.450
Menopause	1.20	0.62–2.32	0.584			

Dysfonction érectile

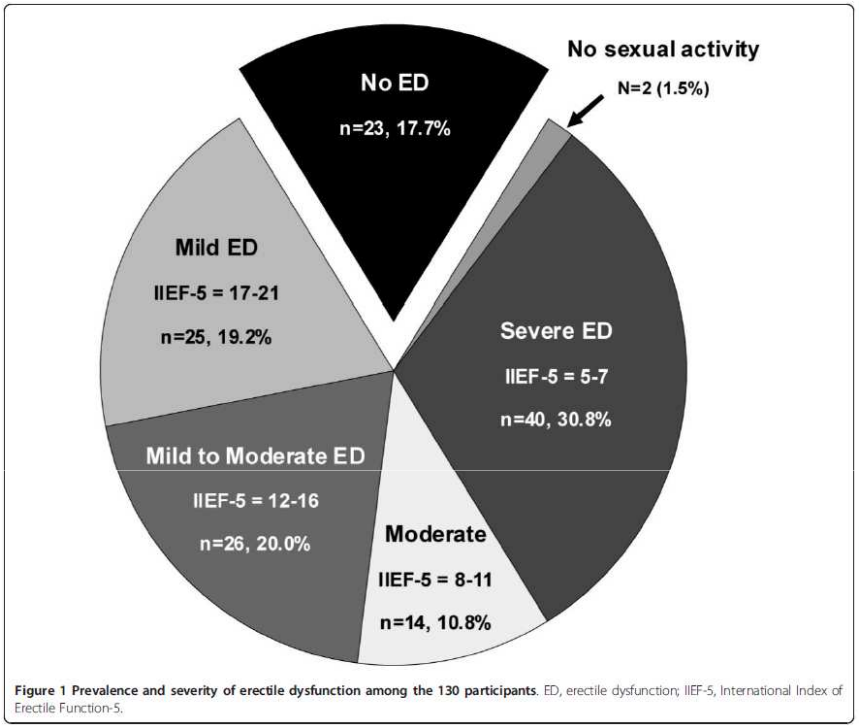

Figure 1 Prevalence and severity of erectile dysfunction among the 130 participants. ED, erectile dysfunction; IIEF-5, International Index of Erectile Function-5.

Table 2 Comorbidities of the participants

	No erectile dysfunction (*n* = 23)	Erectile dysfunction (*n* = 105)	Odds ratio (95% confidence interval)
Cardiovascular risk factors			
Systemic arterial hypertension	14.3	24.1	1.11 (0.92 to 1.34)
Diabetes mellitus	4.4	6.9	1.08 (0.82 to 1.42)
Coronary heart disease	4.4	13.3	1.17 (0.98 to 1.39)
Hypercholesterolaemia	19.1	13.3	0.92 (0.70 to 1.21)
History of smoking	31.8	42.6	1.09 (0.92 to 1.29)
Cigarette smoking (pack-years)	15 (10 to 21)	20 (9 to 30)	*P* = 0.69
Medication			
Antidepressant, sedative, neuroleptic or antiepileptic	4.4	9.1	1.12 (0.89 to 1.40)
Thiazides or spironolactone	4.4	7.0	1.08 (0.82 to 1.43)
Alcohol consumption (> 2 units/day)	0	13.7	1.27 (1.15 to 1.40)*
Other			
Depression	4.6	9.0	1.11 (0.88 to 1.39)
Central nervous system problems	0	3.9	1.23 (1.13 to 1.35)*
Prostatic disease	0	8.4	1.24 (1.13 to 1.36)*
Hormonal (hypogonadism, hyperprolactinaemia)	0	2.8	1.23 (1.11 to 1.36)
Number of comorbidities per patient			
At least one comorbidity	52.2	61.5	1.07 (0.90 to 1.28)
At least two comorbidities	13.0	36.5	1.21 (1.05 to 1.40)*
At least three comorbidities	4.4	14.4	1.17 (1.00 to 1.34)*
At least four comorbidities	0	5.8	1.23 (1.13 to 1.35)*
At least five comorbidities	0	1	1.22 (1.13 to 1.33)*

Table 5 Treatment of erectile dysfunction

	Erectile dysfunction severity				
	Mild (*n* = 25)	Mild to moderate (*n* = 26)	Moderate (*n* = 14)	Severe (*n* = 40)	All (*n* = 105)
Sildenafil	20	8	31	11	15
Tadalafil	12	8	15	11	11
Vardenafil	0	4	0	0	1
Alprostadil urethral	0	0	0	0	0
Alprostadil cavernous	0	0	15	0	2
Vacuum device	0	0	0	0	0
Penile prosthesis	0	0	0	5	2

Data represent percent of men on each treatment modality. A total of 3 men received combination therapy.

Foocharoen et al, Arthritis Res Ther. 2012 Feb 20;14(1):R37.