Handicap et qualité de vie dans la sclérodermie systémique

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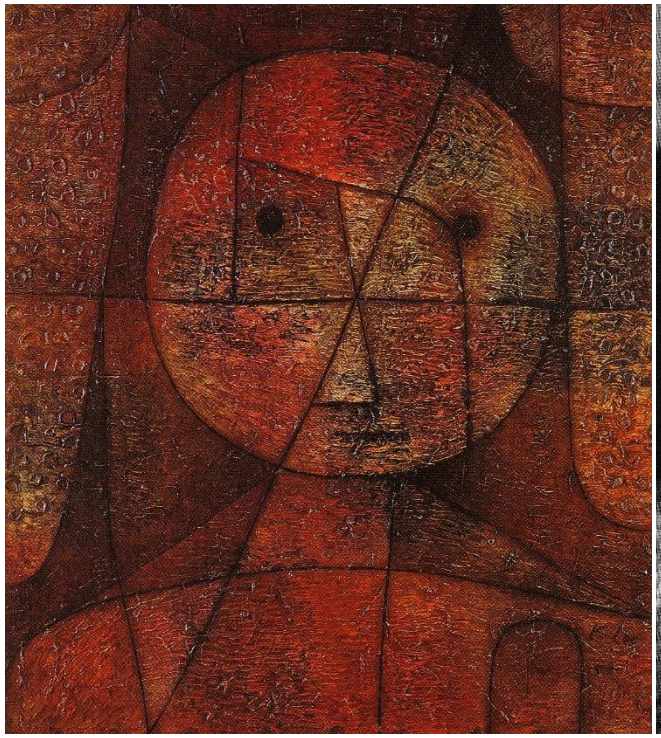
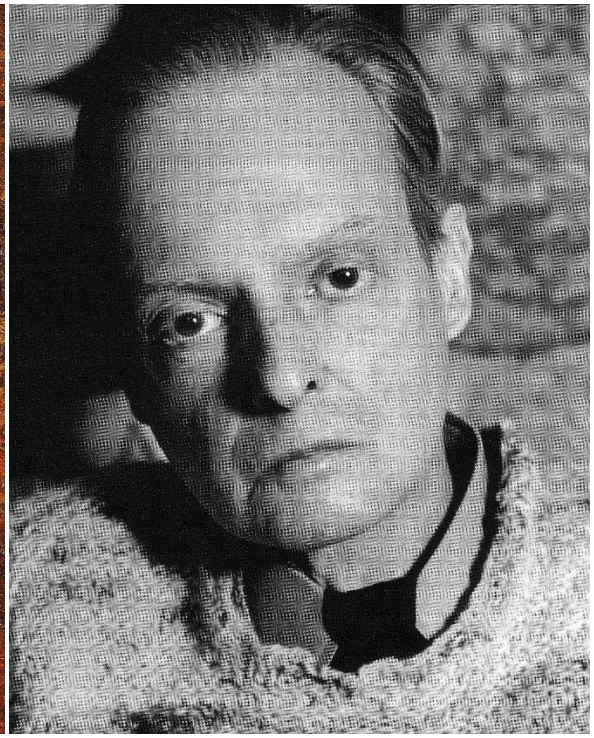
Conflicts of interest

- **Consultant**: Actelion, CSL Behring, LFB Biotechnologies, Lilly, Pfizer, Octapharma
  - Financial support to ARMIIC
- **Investigator**: Actelion, CSL Behring, Pfizer
- **Financial support (grants to ARMIIC)**: Actelion, CSL Behring, GSK, LFB Biotechnologies, Pfizer
- **Invited conference**: SOBI, Roche, Actelion, CSL Behring, Octapharma, GSK, LFB Biotechnologies, Pfizer, Lilly, UCB pharma

Paul Klee : 1879-1940 (I)





Gezeichneter 1935
„portant la marque de la mort“

1933 Raynaud‘s phenomenon

1934 Fatigue, dyspnea, thickened skin

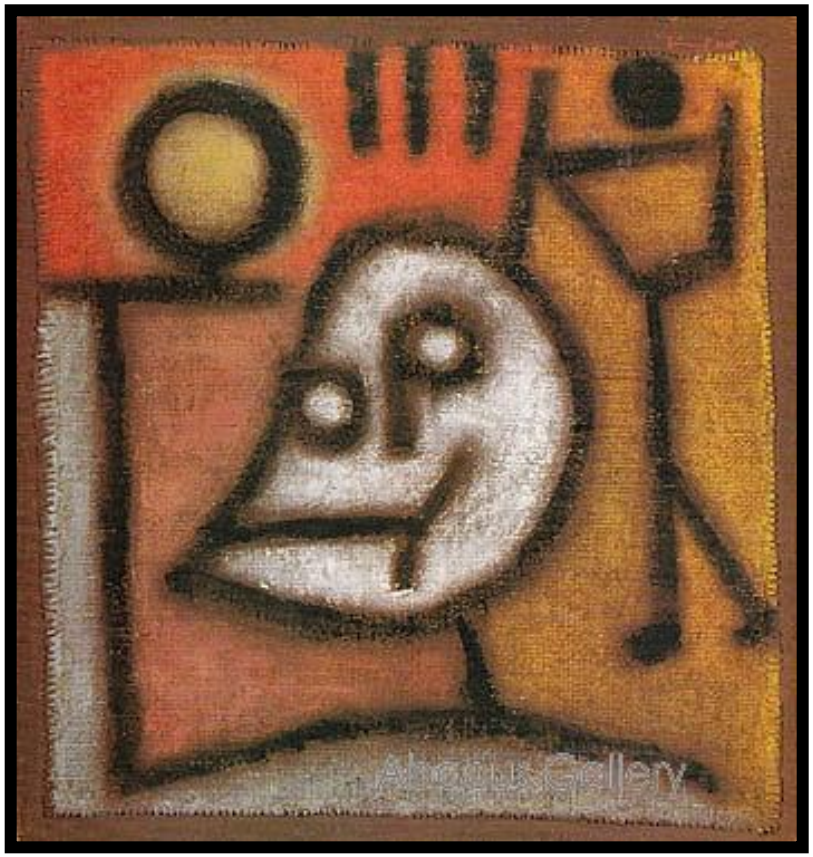
1936 Extension of skin fibrosis

1940 Hospitalisation at Sant‘ Agnes, Locarno (worsening of dyspnea)

Died in June 1940

Paul Klee: 1879-1940 (2)

Mask – 1921



Death and Fire – 1940

***Paul Klee* Polyphonies, Cité de la *musique*, Paris**
18 October 2011 – 15 January 2012

Patients expectations

Patient and practitioner views about systemic sclerosis and its management: a qualitative interview study

- Relationship with care providers, especially physicians, is a priority.
- Patients expected physicians to be human and attentive but also involved in research in the field
- They also expected more individualized management, improvements in diagnosis and follow-up organization, more efforts in education and information
- Care providers were more focused on treatment improvement, optimization of the health care system, education of general practitioners and expected increased funding for research.

Mouthon L et al. Submitted

Quality of life

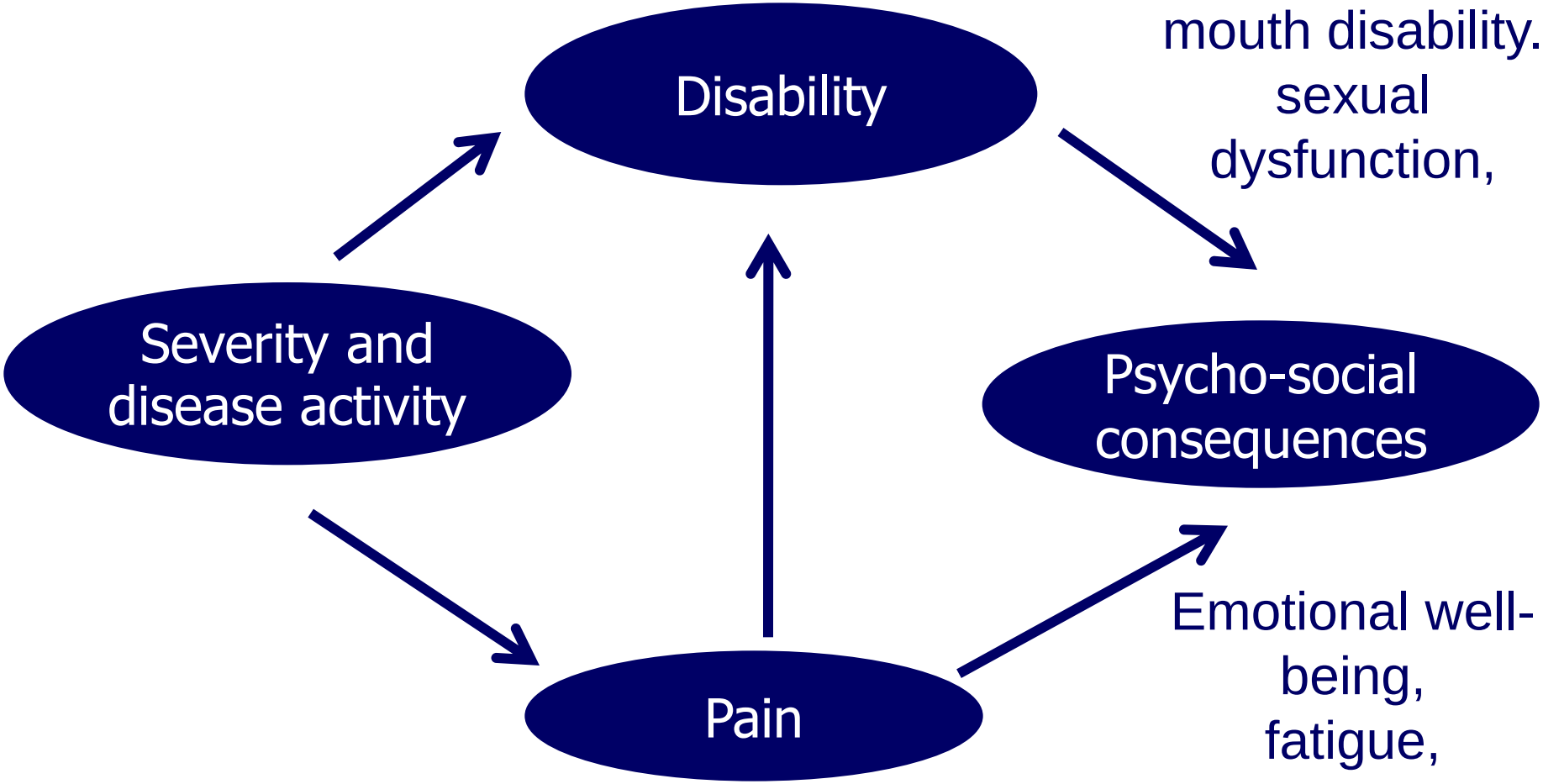
HR-QOL and SF-36 in SSc

- **HRQoL is altered in patients with SSc.**
- **Use of the PCS has been shown to discriminate between lcSSc and dSSc.**
- **In dSSc patients, SF-36 discriminate between less and more severe breathlessness in patients with lung disease.**
- **However, the relevance of using the 2 aggregate scores, PCS and MCS, in SSc has not been demonstrated.**
- **Experts have recommended additional research in this area.**

Khanna D et al. J Rheumatol. 2005
Khanna D et al. Arthritis Rheum. 2005.
Merkel PA et al. J Rheumatol. 2003.

Del Rosso A et al. Arthritis Rheum. 2004
Danieli E et al. Clin Rheumatol. 2005.
Georges C et al. Rev Med Interne. 2004

Quality of life (QOL) in SSc



Patient-Reported Outcomes Related to Psychological Health and Well-Being in Systemic Sclerosis: A Consensus Research Agenda

- Fatigue
- Pain
- Pruritus (Itching)
- Body Image
- Sexual Function
- Depression/Distress
- Other Areas

Pain in systemic sclerosis

- Visual analogic scale (P-VAS)
- Predictive factors of pain
  - High Rodnan skin score
  - Edema
  - Tendon retractions
- Pain correlate with
  - Arthralgias and synovitis
  - Digital ulcers
  - Physical component score of the SF36:
    - Bodily pain
    - Physical function
    - General health
    - Physical role

Steen et al, Arthritis Rheum, 1997
Merkel et al, Arthritis Rheum, 2002
Georges et al, Rheumatology 2006
Malcarne et al, J Rheum, 2007
Scheir et al, Arthritis Care Res 2010

Anxiety and depression

Frequency of depression in SSc

Author	Scale	Number of patients	Frequency of depression (CI 95%) and threshold used
Roca et al. Arthritis Rheum 1996	BDI	54	65% (56-74), BDI≥10
Angelopoulos et al. Psychother Psychosom 2001	DSSI/sAD	31	42% (32-51), DSSI/sAD≥4
Benrud-Larson et al. Pain 2002	BDI	142	51% (42-60), BDI≥10
Matsuura et al. J Rheumatol 2003	BDI	50	46% (37-56), BDI≥11
Legendre et al. Joint Bone Spine 2005	MADRS	42	43% (33-52), MADRS≥16
Hyphantis et al. J Psychosom Res 2007	DSSI/sAD	56	Depression score higher in SSc than controls (10,6±4,3 vs 8,5±1,8, p=0,029)
Beretta et al. Reumatismo 2006	BDI	111	56% (47-65), BDI≥11
Nietert et al. J Rheumatol 2005	CES-D	72	36% (27-45), CES-D≥16 26% (17-38), CES-D≥19
Richards et al. Arthritis Rheum 2003	HADS-D	49	38% (28-46), HADS-D≥8 17% (7-28), HADS-D≥11
Baubet et al. Presse Med 2011	MINI, BDI	100	20% (ND)
Thombs et al. Arthritis Rheum 2008	CES-D	376	35% (ND), CES-D≥16 18% (ND), CES-D≥23

Launay D et al. Presse Med 2010

Current and lifetime prevalence of DSM-IV anxiety disorders, as assessed with MINI in a cohort of 100 SSc patients

	Current	Lifetime
Panic Disorder	**6%**	**10%**
Agoraphobia	**9%**	**11%**
Social Phobia	**13%**	**15%**
Obsessive Compulsive Disorder	**2%**	**2%**
Generalized Anxiety	**13%**	**19%**
Post-Traumatic Stress Disorder	**2%**	**9%**
Any anxiety disorder	**49%**	**64%**

Baubet et al. Presse Med 2011

Association of gender with depression and anxiety in SSc

	All patients n = 381	Males n = 62	Females n = 319	p-value†
HADa (0–21) (mean [SD])	9.2 (4.5)	8.3 (5.1)	9.4 (4.4)	0.088
• HADa≥8	224/378 (59.3)	27/62 (43.5)	197/316 (62.3)	0.006
HADd (0–21) (mean [SD])	6.6 (4.2)	6.5 (4.6)	6.6 (4.1)	0.781
• HADd≥8	154/378 (40.7)	25/62 (40.3)	129/316 (40.8)	0.957
• HADa and HADd<8	129/378 (34.1)	29/62 (46.8)	100/316 (31.6)	0.021

*Values are number/number of patients for whom the data is available (%), otherwise indicated in parenthesis.
SSc: systemic sclerosis; n: number; HADa: Hospital Anxiety and Depression scale for Anxiety; HADd: Hospital Anxiety and Depression scale for Depression.

- In SSc patients, male gender tends to be associated with diffuse disease and female gender with calcinosis and self-reported symptoms of anxiety.
- Disease-associated disability and HRQoL were similar in both groups

Nguyen C et al. Plos One 2011

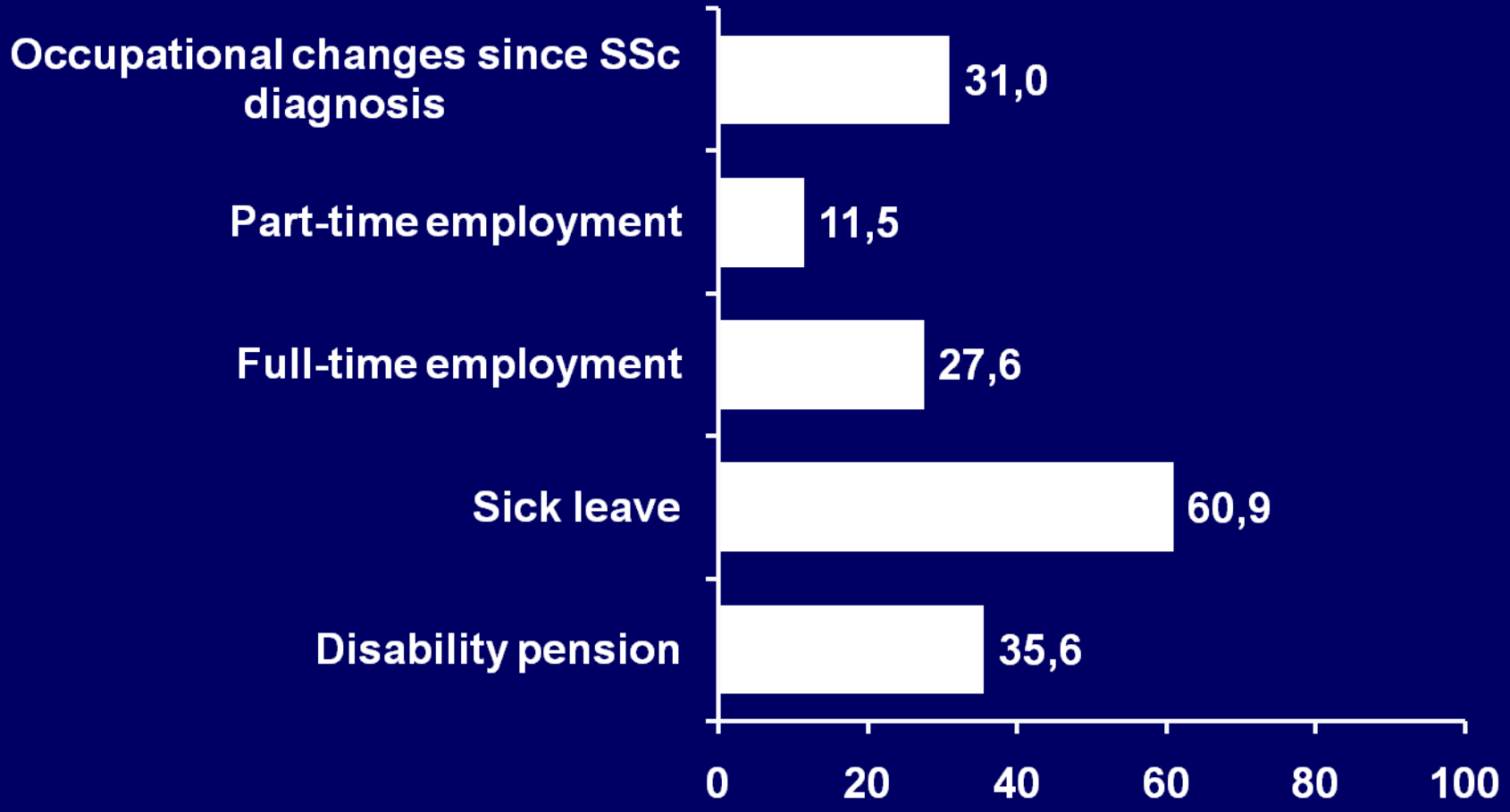
Final model of multivariable analysis of current clinically significant symptoms of depression and anxiety according to clinical, functional and health-related quality of life features of patients with SSc, with adjustment for age and sex.

Variables	Adjusted Odds Ratio	95% CI	P value
Depression symptoms correlates			
• Age	1.01	0.99–1.03	0.354
• Sex	1.17	0.55–1.86	0.768
• HAQ	2.90	0.74–1.38	$<10^{9}$
• SF36 -PCS			0.005
Anxiety symptoms correlates			
• Age	1.00	0.98–1.01	0.810
• Sex	0.49	0.57–1.75	0.003
• HAQ	1.40	0.76–1.31	0.012
• SF36 -PCS			0.002

A p-value less than 0.05 was considered statistically significant.
CI: confidence interval; HAQ: Health Assessment Questionnaire; PCS: Physical Component Score;; of SF-36: Medical Outcomes Study 36-Item Short Form Health Survey.

Nguyen et al. Plos one 2014

Employment status in SSc patients



Nguyen C, *et al. Rheumatology* 2010; 49:982-9.

Factors associated with employment status in SSc

SSc patients on sick-leave have greater global, hand and mouth handicaps and depression

Outcome measures (range)	Sick leave (*n* = 53)	No sick leave (*n* = 34)	*p* value
HAQ (0-3)	0.9 (0.7)	0.6 (0.5)	0.021
CHFS (0-90)	21.7 (18.9)	10.7 (12.1)	0.003
MHISS (0-48)	20.2 (10.8)	14.6 (10.0)	0.014
HADd (0-21)	7.1 (3.9)	4.8 (3.4)	0.003

Values represent mean (SD)
CHFS = Cochin hand function scale (higher score = increased hand disability); HADd = Depression dimension of the hospital anxiety and depression scale (higher score = increased depression); HAQ = Global disability, ranging from 0 (no disability) to 3 (maximal disability); MHISS = Mouth handicap in SSc scale (higher score = increased mouth disability).

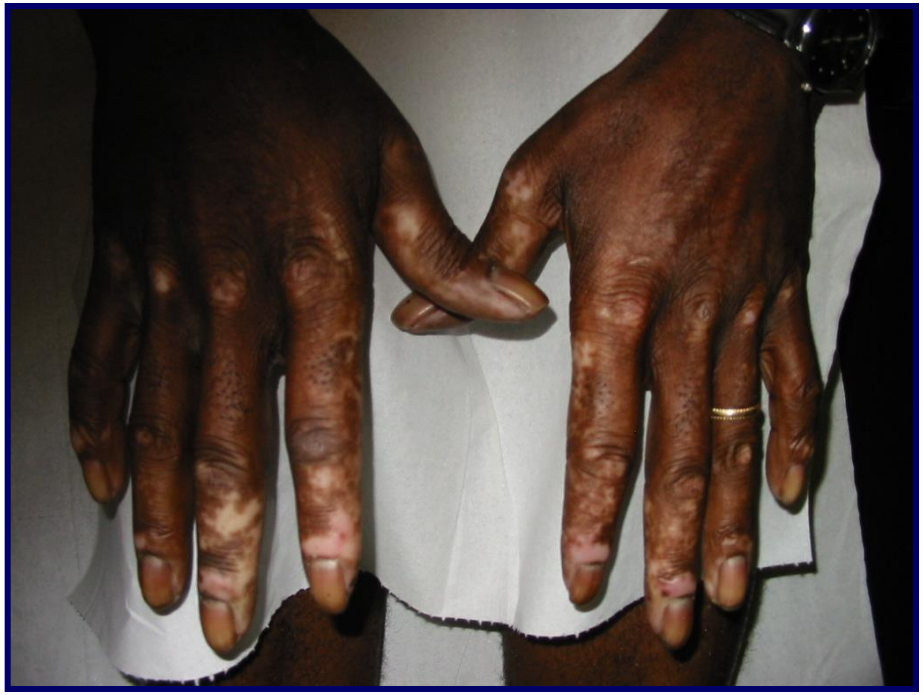
Nguyen C, *et al. Rheumatology* 2010; 49:982-9.

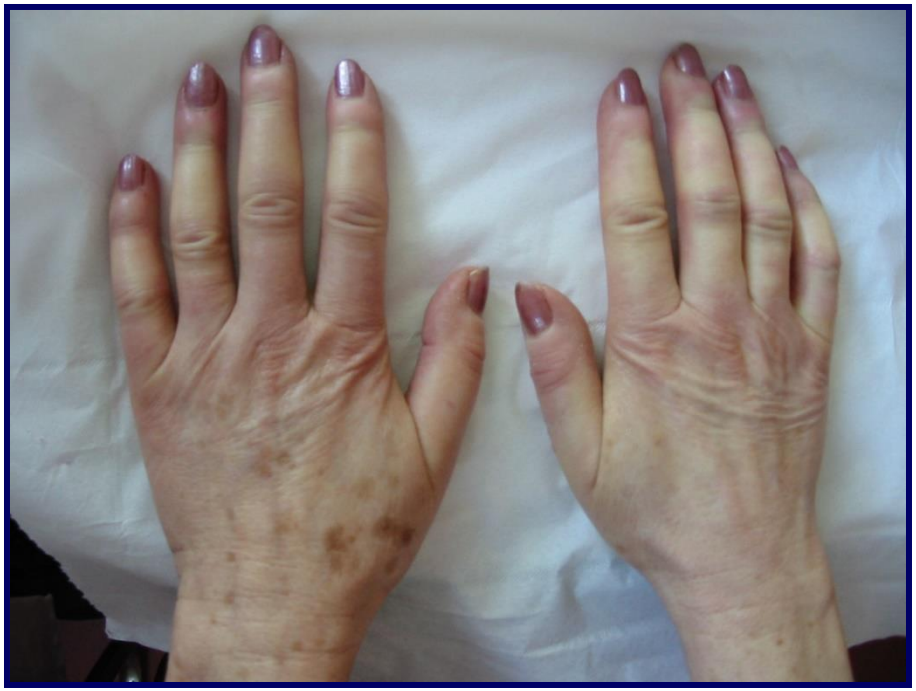
Socio-economic burden of 87 SSc patients

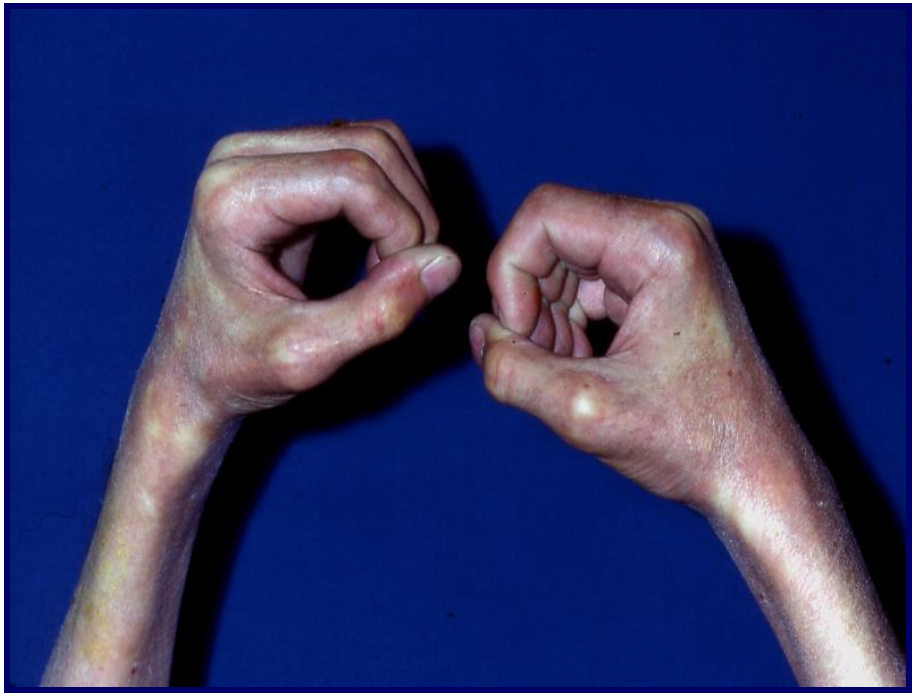
	All patients	Disability pension		*p*-value
		Disability pension	No disability pension	
Occupational changes, mean (SD)	0.6 (1.1)	0.9 (1.3)	0.4 (0.9)	0.014
Decreased income, *n* (%)	35 (40.2)	22 (71.0)	13 (23.2)	0.000
Lack of advancement, *n* (%)	38 (43.7)	22 (71.0)	16 (28.6)	0.000
Feelings of discrimination, *n* (%)	10 (11.5)	7 (22.6)	3 (5.4)	0.030

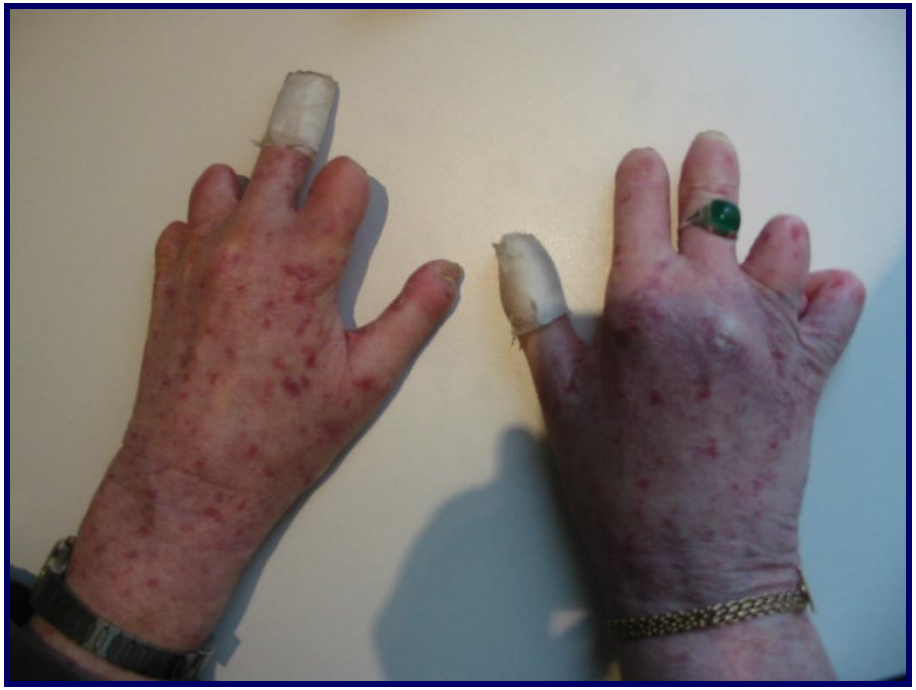
Nguyen C, *et al. Rheumatology* 2010; 49:982-9.

Disability









Hand involvement in systemic sclerosis

- Skin involvement
- Sub-cutaneous tissues
- Joint and periarticular involvement
- Bone involvement
- Tendon involvement
- Muscle involvement
- Vascular involvement
- Peripheral nervous system

Cochin hand function scale (CHFS)

Without the help of adapted instruments, in the past two weeks, did you:

- **Categories for assessment**

- **The scale is based on the following answer scores**

0 = Yes , without difficulty

1 = Yes, with a little difficulty

2 = Yes, with some difficulty

3 = Yes, with much difficulty

4 = Nearly impossible to do

5 = Impossible

Hand disability contribute to 75 % of the variance of the HAQ in SSc

Duruöz MT, *et al. J Rheumatol* 1996

Rannou *et al* Arthritis Rheum. 2007.

Table 3. Convergent and divergent validities of the Cochin Hand Function Scale, HAQ and sHAQ, and PCS and MCS scores of the SF-36 for patients with SSc (correlation with other variables)*

Scales	Spearman's correlation coefficient
Cochin Hand Function Scale	
Convergent validity	
sHAQ	0.81
HAQ	0.75
Kapandji index	0.63
HFI	0.58
SF-36 physical functioning	0.53
Perceived individualized handicap (MACTAR)	0.48
SF-36 PCS	0.45
Divergent validity	
Anxiety (HADa)	0.16
SF-36 MCS	0.14
Depression (HADd)	0.05
Disease duration	0.15
Age	0.01

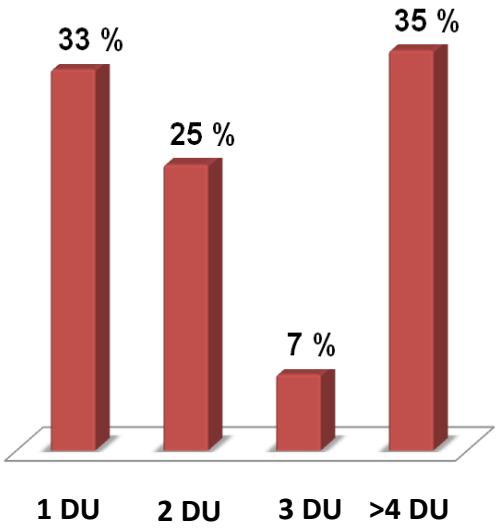
Hand disability contribute to 75 % of the variance of the HAQ

Rannou *et al* A & R. 2007

Impact of Systemic Sclerosis on Occupational and Professional Activity With Attention to Patients With Digital Ulcers

A. BÉREZNÉ,[1] R. SEROR,[1] S. MORELL-DUBOIS,[2] M. DE MENTHON,[1] E. FOIS,[1] A. DZEING-ELLA,[1] C. NGUYEN,[3] E. HACHULLA,[2] L. GUILLEVIN,[1] S. POIRAUDEAU,[3] AND L. MOUTHON[1]

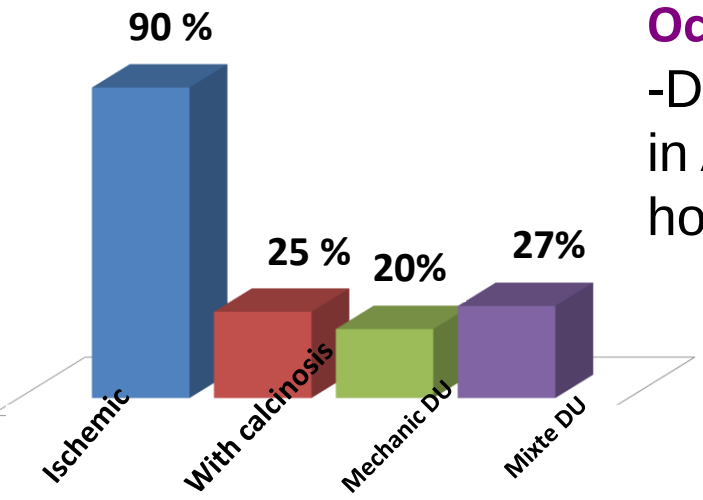
Arthritis Care & Research
Vol. 63, No. 2, February 2011, pp 277–285



At baseline 60/189 patients had ≥1 DU for a total of 221 DU, a mean of 3,7 DU/patient

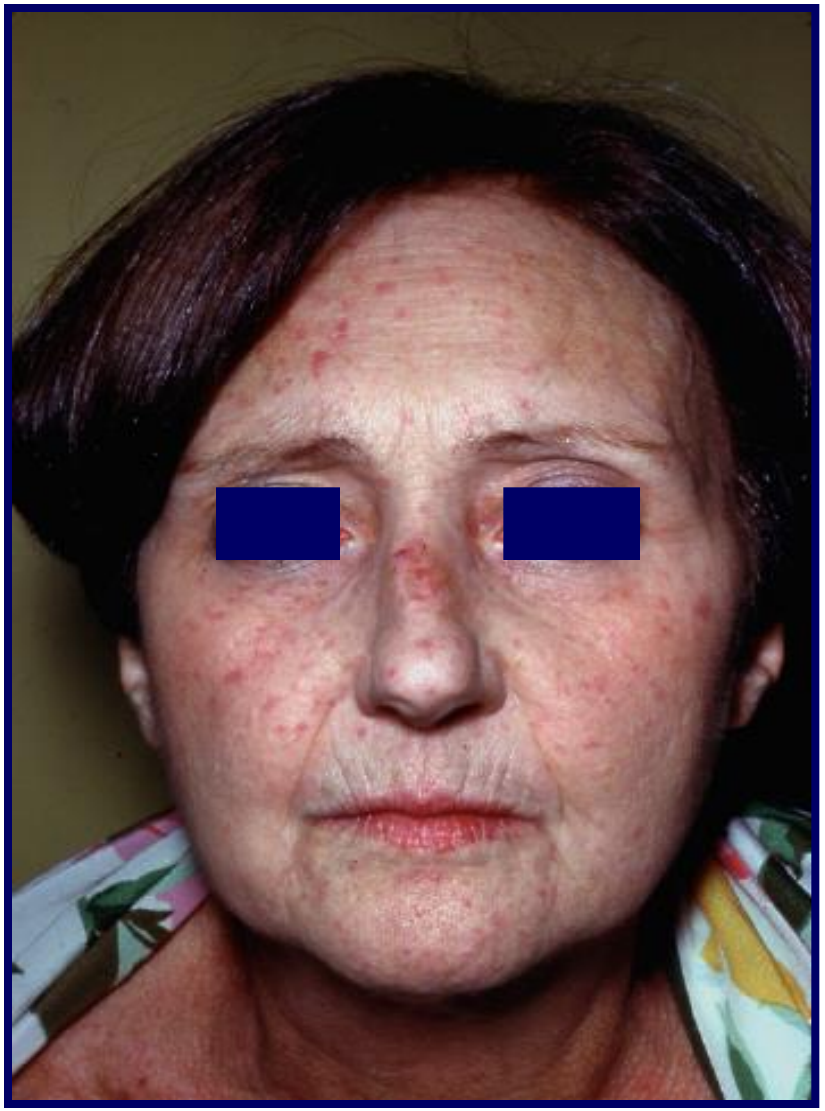
Functional impact of systemic sclerosis and DU

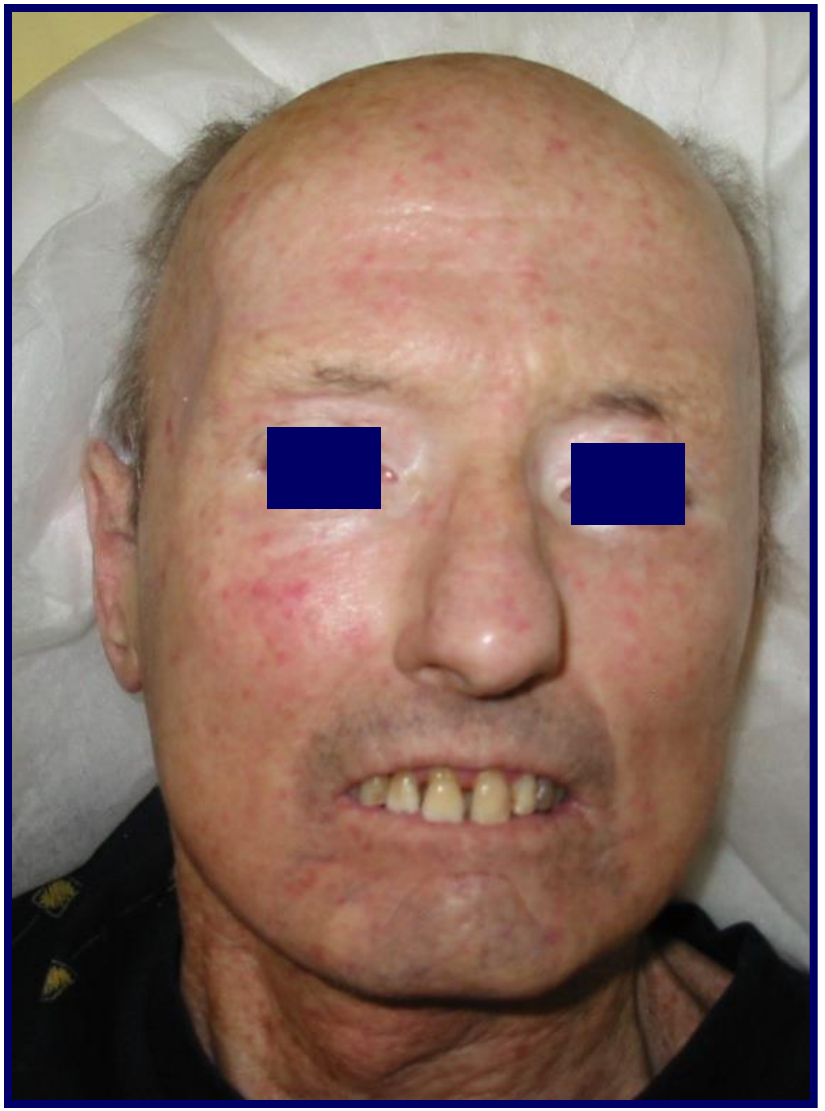
-had significantly greater HAQ (*P*=0.001), and
-Cochin Hand Function Scale (CHFS) (p<0.0001)



Occupational impact of systemic sclerosis and DU

-DUs were associated with decreased performance in ADL (p=0.006), and a need for paid and nonpaid household help (p=0.001)





Development and validation of a scale for mouth handicap in systemic sclerosis: the Mouth Handicap in Systemic Sclerosis scale

L Mouthon*, F Rannou*, A Bérezné, C Pagnoux, J-P Arène, E Foïs, J Cabane, L Guillevin, M Revel, J Fermanian, S Poiraudeau

Ann Rheum Dis 2007;**66**:1651–1655. doi: 10.1136/ard.2007.070532

Conclusion: We propose a new scale, the Mouth Handicap in Systemic Sclerosis (MHISS) scale, which has excellent reliability and good construct validity, and assesses specifically disability involving the mouth in patients with SSc.

Table A1 MHISS scale

		Never	Rarely	Occasionally	Often	Always
1	I have difficulties opening my mouth	0	1	2	3	4
2	I have to avoid certain drinks (sparkling, alcohol, acidic)	0	1	2	3	4
3	I have difficulties chewing	0	1	2	3	4
4	My dentist has difficulties taking care of my teeth	0	1	2	3	4
5	My dentition has become altered	0	1	2	3	4
6	My lips are retracted and/or my cheeks are sunken	0	1	2	3	4
7	My mouth is dry	0	1	2	3	4
8	I must drink often	0	1	2	3	4
9	My meals consist of what I can eat and not what I would like to eat	0	1	2	3	4
10	I have difficulties speaking clearly	0	1	2	3	4
11	The appearance of my face is modified	0	1	2	3	4
12	I have trouble with the way my face looks	0	1	2	3	4

As you are probably aware, your systemic sclerosis might involve your face and your mouth. This questionnaire is aimed at assessing how much your face and mouth involvement affects your daily life.

The Mouth Handicap in Systemic Sclerosis (MHISS) scale (I)

Factor 1: mouth opening		Never	Rarely	occasionally	Often	Always
1	I have difficulties opening my mouth	0	1	2	3	4
2	I have to avoid certain drinks (sparkling, alcoholised, acidic)	0	1	2	3	4
3	I have difficulties chewing	0	1	2	3	4
4	My dentist has difficulties taking care of my teeth	0	1	2	3	4
5	My dentition has become altered	0	1	2	3	4
6	My lips are retracted and/or my cheeks are sunken	0	1	2	3	4

The Mouth Handicap in Systemic Sclerosis (MHISS) scale (II)

		Never	Rarely	occasionally	Often	Always
Factor 2: sicca syndrome						
7	My mouth is dry	0	1	2	3	4
8	I must drink often	0	1	2	3	4
9	My meals consist of what I can eat and not what I would like to eat	0	1	2	3	4
10	I have difficulties with my diction	0	1	2	3	4
Factor 3: aesthetic concerns						
11	The appearance of my face is modified	0	1	2	3	4
12	I have trouble with the way my face looks	0	1	2	3	4

What needs to happen

BMJ | 29 AUGUST 2009 | VOLUME 339

We need minimally disruptive medicine

The burden of treatment for many people with complex, chronic, comorbidities reduces their capacity to collaborate in their care. **Carl May, Victor Montori**, and **Frances Mair** argue that to be effective, care must be less disruptive

ordinated and increasing workload. These problems have important connections. We propose that some non-adherence is structurally induced by the healthcare system.

Clinicians lack the tools to detect patients overwhelmed by the burdens of treatment, and they lack strategies to lift these burdens

Patient-centered Care

- Patient empowerment
- Shared decision-making
- Care plans that take into consideration patient preferences and values
- Care that meets the needs of individual patients
- In chronic disease…..
  - Comprehensive care to reduce disability and improve health-related quality of life

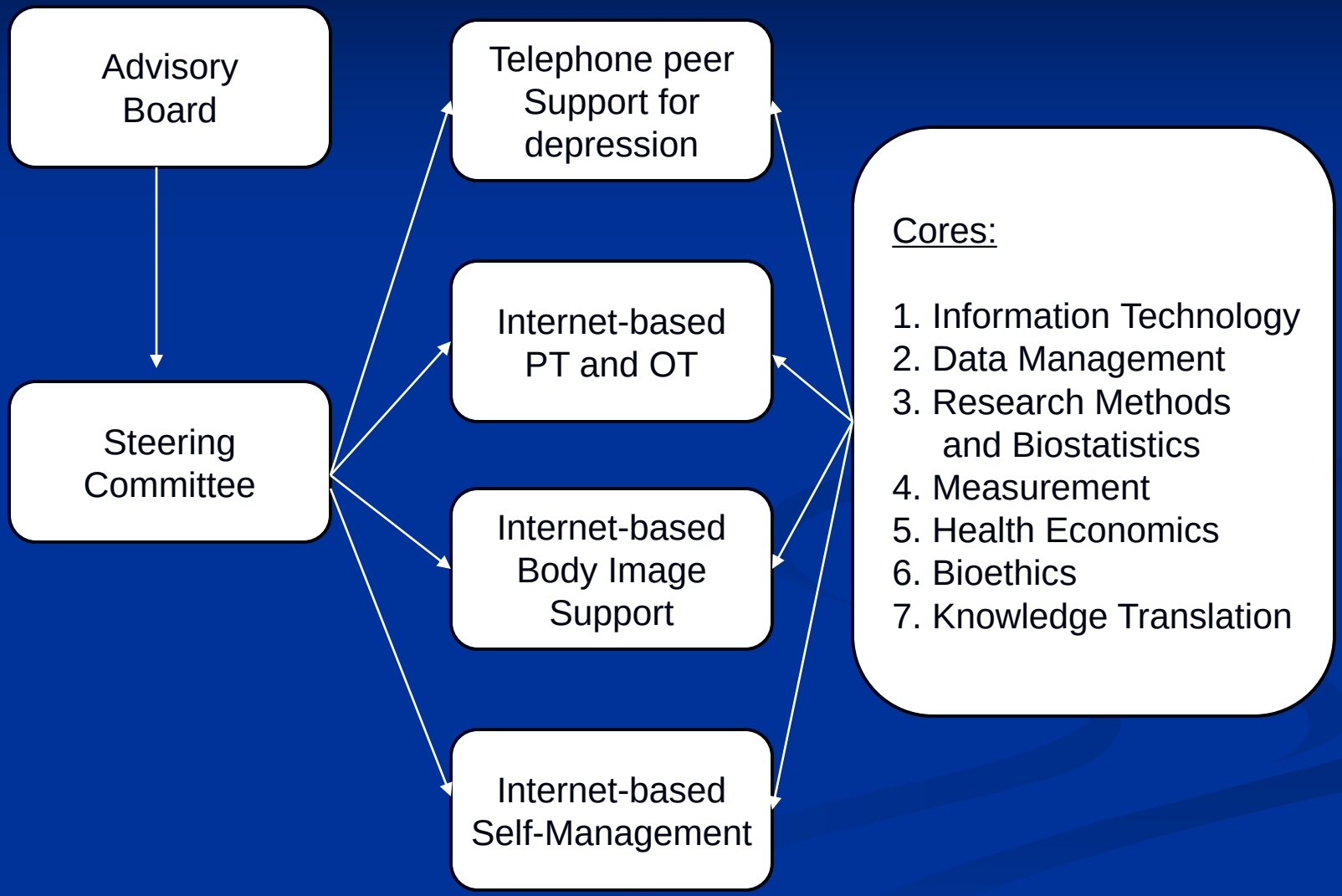
Consortium for Clinical Trials of Psychosocial, Educational, and Rehabilitation Interventions in Scleroderma

- Scleroderma Society of Canada
  - Scleroderma Society of Ontario
  - Sclérodermie Québec
- Federation of European Scleroderma Associations
- Research/Clinical Centers
  - Canada
  - USA
  - France
  - The Netherlands

SPIN

- A collaboration of people living with scleroderma, clinicians, and researchers
- Objective: to develop an infrastructure that can be used on an ongoing basis to test accessible, low-cost interventions that are effective to reduce disability and improve quality of life for people with SSc.

Scleroderma Patient-centered Intervention Network (SPIN)



Thombs et al. Clin Exp Rheumatol. 2012

SPIN Innovation: Research Methods

- Formation of a network of major scleroderma clinical research centers across Canada, the US, and Europe
- The SPIN Cohort - “cohort multiple RCT design”

SPIN Innovation: Psychosocial and Rehabilitative Care Delivery

- Leveraging technology to deliver care
- Partnering with patient organizations to deliver psychosocial and rehabilitation interventions not feasibly provided by any single centre

Conclusion

- Altered quality of life in patients with SSc
- High prevalence of clinically significant anxious and depressive symptoms among patients with SSc
- Special attention should be paid to detect clinically significant symptoms of anxiety and/or depression in patients with SSc in order to propose suitable interventions.
- Unemployment and socio-economic burden are major issues in SSc patients.
- Increased disability in SSc patients
- Supervised rehabilitation program can improve disability inpatients with SSc.
- Difficulties to perform rehabilitation autoprogram.
- SPIN: low-cost interventions that are effective to reduce disability and improve quality of life for people with SSc.

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