Scleroderma Classification Criteria: Developing Methods for Multi-criteria decision analysis

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On behalf of the
ACR-EULAR SSc Classification Criteria Committee

ACR Meeting 2012

Disclosures

AMERICAN COLLEGE OF RHEUMATOLOGY
EDUCATION • TREATMENT • RESEARCH

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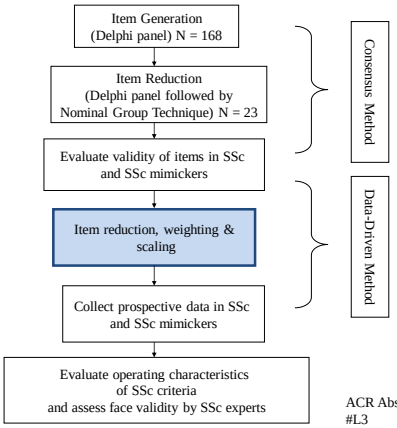


ACR Abstract, 2012
#L3

Aim

- Define a system of criteria, which produces a measure of the relative probability that a particular case (combination of clinical features) has SSc
- Reduce and weight the candidate criteria

Objectives

- SSc specific instrument
  - Develop
  - Evaluate: Sensibility
- Multi-criteria decision analysis
  - Reduce
  - Weight
- Explore agreement among SSc experts

Instrument

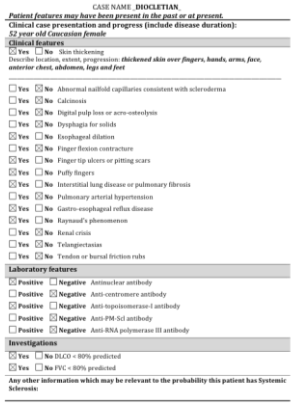
Design

- Format
- Visual presentation
- Response options

Sensibility

- Comprehensibility
- Clarity
- Face validity
- Content validity
- Feasibility

Dillman. Tailored Design Method. 2009
Feinstein. Clinimetrics. 1987



CASE NAME: **DIOCLETIAN**

Patient features may have been present in the past or at present.

Clinical case presentation and progress (include disease duration):

52 year old Caucasian female

Clinical features

☑ Yes ☐ No Skin thickening

Describe location, extent, progression: ***thickened skin over fingers, hands, arms, face, anterior chest, abdomen, legs and feet***

☐ Yes ☑ No Abnormal nailfold capillaries consistent with scleroderma
☐ Yes ☑ No Calcinosis
☐ Yes ☑ No Digital pulp loss or acro-osteolysis
☐ Yes ☑ No Dysphagia for solids
☑ Yes ☐ No Esophageal dilation
☐ Yes ☑ No Finger flexion contracture
☑ Yes ☐ No Finger tip ulcers or pitting scars
☑ Yes ☐ No Puffy fingers
☑ Yes ☐ No Interstitial lung disease or pulmonary fibrosis
☐ Yes ☑ No Pulmonary arterial hypertension
☑ Yes ☐ No Gastro-esophageal reflux disease
☑ Yes ☐ No Raynaud's phenomenon
☐ Yes ☑ No Renal crisis
☐ Yes ☑ No Telangiectasias
☐ Yes ☑ No Tendon or bursal friction rubs

Laboratory features

☑ Positive ☐ Negative Antinuclear antibody
☐ Positive ☑ Negative Anti-centromere antibody
☑ Positive ☐ Negative Anti-topoisomerase-I antibody
☐ Positive ☑ Negative Anti-PM-Scl antibody
☐ Positive ☑ Negative Anti-RNA polymerase III antibody

Investigations

☑ Yes ☐ No DLCO < 80% predicted
☑ Yes ☐ No FVC < 80% predicted

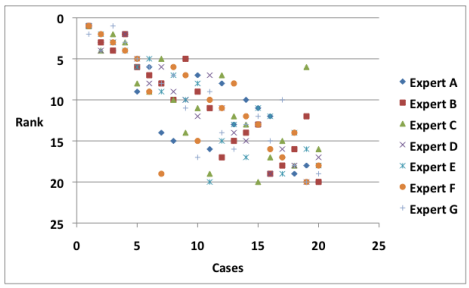
Any other information which may be relevant to the probability this patient has Systemic Sclerosis:

Sensibility

Attribute	Endorsement n = 6
Clarity and navigation of the form	83%
Clarity of the instructions	100%
Clarity of the response option	100%
Median time to completion	10 minutes (10 -20 minutes)

SSc Experts
Ranking and Multi-criteria decision analysis

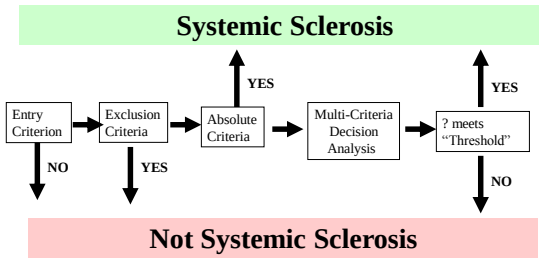
Attribute	n = 8
Male sex	63%
Median years in practice	30 (range 13 – 40 years)
Practice location	50% Europe 50% North America
Involvement in previous phases of criteria development	38%



1st Ranking. Experts' rankings of the relative probability that the case has systemic sclerosis.
The cases ranked from highest (rank = 1) to lowest probability (rank = 20) on the Y-axis.

ICC_{All} = 0.73 (95% CI 0.58, 0.86)

ICC_A = 0.68 (95% CI 0.48, 0.84) ICC_B = 0.76 (95% CI 0.60, 0.88)



PAPRIKA method
Potentially All Pairwise RanKings of All hypothetically-possible patients

Which patient ('Left' or 'Right') has the higher probability of being classified as systemic sclerosis?
(given they are identical in all other aspects)

Left	Right
Raynaud's phenomenon	**Raynaud's phenomenon**
GERD	SSc specific antibodies

The overall ranking of ***all*** hypothetically-possible patients is arrived at by asking experts to make ***tradeoffs between 2 criteria at a time***

Item Reduction

- **Exclusion criterion**
  - *Skin thickening sparing the fingers*
  - If present, the use of the SSc classification criteria should not proceed further
- **Absolute criterion**
  - *Skin thickening proximal to the MCP joints*
  - If present, the patient could be classified as SSc

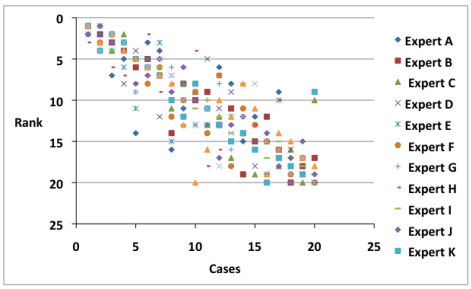
Item Reduction: Low weights

- FVC
- DLCO
- Dysphagia for solid foods
- GERD
- Anti-PM-ScL antibody
- ANA

Item reduction: Criterion revision

- ***Skin thickening of the fingers***
  - a) distal to MCP, or b) distal to PIP joint.
- ***Finger tip lesions***
  - a) pitting scars, b) digital tip ulcers, or c) clinical evidence of acro-osteolysis.
- ***Scleroderma specific antibodies***
  - anti-topoisomerase-1, anticentromere or anti-RNA polymerase III antibody

Criteria	Sub-criteria	Weight
Skin thickening of the fingers *(count only one of these 2)*	Distal to PIP only	14
	Whole Finger, distal to MCP	22
Finger tip lesions *(count only one of these 3)*	Digital Tip Ulcers	9
	Pitting Scars	16
	Clinical evidence of acro-osteolysis	21
Finger flexion contractures		16
Telangiectasia		10
Abnormal nailfold capillaries		10
Puffy fingers		5
Calcinosis		12
Raynaud's phenomenon		13
Tendon or bursal friction rubs		21
Interstitial lung disease (ILD) or pulmonary fibrosis (PF)		14
Pulmonary Hypertension (without ILD/PF)		11
Renal crisis		11
Esophageal dilatation		7
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase1 [anti-Scl 70], anti-RNA polymerase III)		15
TOTAL SCORE:		



Experts' rankings of the relative probability that the case has systemic sclerosis in second ranking exercise.
The cases ranked from highest (rank = 1) to lowest probability (rank = 20) on the Y-axis.

ICC_{All} = 0.80 (95% CI 0.68, 0.90)

Summary

- Reduced the number of candidate criteria
- Indicated relative weights.

- Experts had substantial overall agreement in rank order of the relative probability that each case can be classified as having SSc

- **Defined a system of criteria, which produces a measure of the relative probability that a particular case (combination of clinical features) has SSc**

Strengths

- **Methodologic rigor**
  - Bias reduction strategies

- **Diverse methodology**
  - Consensus methods
  - Measurement science
  - Decision analysis

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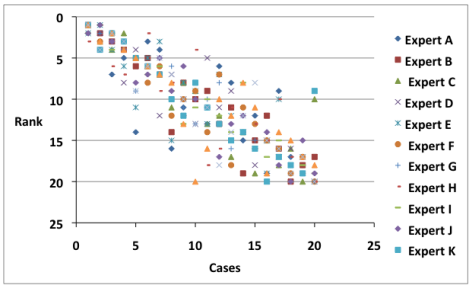

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Next Phases

- Need for further item reduction
- Possible re-weighting and scaling
- Threshold to classify a patient as having SSc
- Validation of criteria

- Face validity
- External validation

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