

ACR/EULAR Classification Criteria for Systemic Sclerosis (SSc, Scleroderma)



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Disclosure

- This project was jointly funded by
 - American College of Rheumatology (ACR)
 - European League Against Rheumatism (EULAR)

Objectives

1. To understand the strengths and weaknesses of the current classification criteria in SSc
2. To become familiar with how to use the new criteria in SSc



Arthritis & Rheumatism

An Official Journal of the American College of Rheumatology
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SPECIAL ARTICLE

AQ: 1

2013 Classification Criteria for Systemic Sclerosis

An American College of Rheumatology/European League
Against Rheumatism Collaborative Initiative

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Jacob M. van Laar,¹⁸ James R. Seibold,¹⁹ Laszlo Czirjak,²⁰ Virginia D. Steen,²¹ Murat Inanc,²²
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Barri J. Fessler,³⁰ Serena Guiducci,⁷ Ariane Herrick,³¹ Vivien M. Hsu,³² Sergio Jimenez,³³
Bashar Kahaleh,³⁴ Peter A. Merkel,³⁵ Stanislaw Sierakowski,²³ Richard M. Silver,³⁶
Robert W. Simms,³⁵ John Varga,³⁷ and Janet E. Pope³⁸

This criteria set has been approved by the American College of Rheumatology (ACR) Board of Directors and the European League Against Rheumatism (EULAR) Executive Committee. This signifies that the criteria set has been quantitatively validated using patient data, and it has undergone validation based on an external data set. All ACR/EULAR-approved criteria sets are expected to undergo intermittent updates.



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An American
League Against Rheumatism

Criteria

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2013 classification criteria for systemic sclerosis: an American college of rheumatology/European league against rheumatism collaborative initiative

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ACR/EULAR SSc Classification Criteria Committee

Co-convenors

- Frank van den Hoogen (EULAR)
- Janet E Pope (ACR)

Members

- Jaap Fransen
- Sindhu R Johnson
- Alan Tyndall
- Murray Baron
- Marco Matucci-Cerinic
- Dinesh Khanna

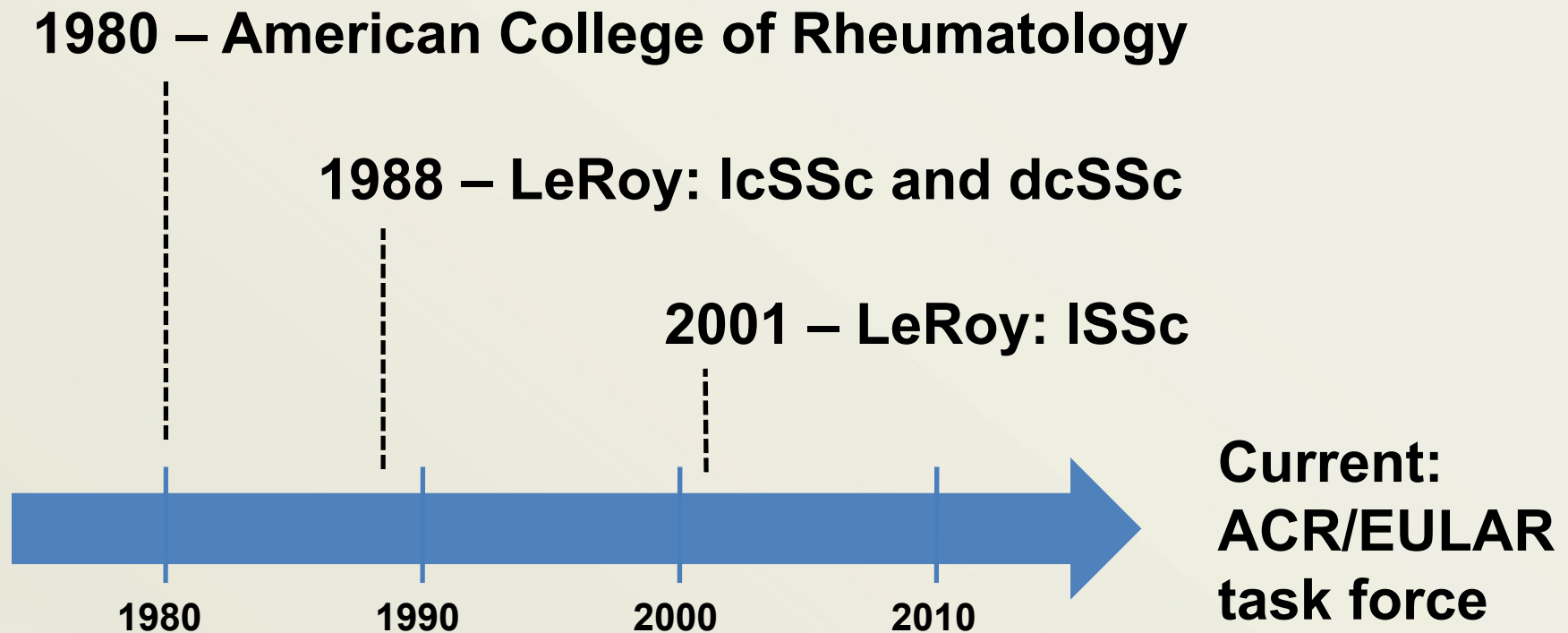
SSc Criteria were developed collaboratively

Frank van den Hoogen, Dinesh Khanna, Jaap Fransen, Sindhu R. Johnson, Murray Baron, Alan Tyndall, Marco Matucci-Cerinic, Raymond Naden, Thomas A Medsger Jr., Patricia Carreira, Gabriela Riemekasten, Phillip Clements, Christopher P Denton, Oliver Distler, Yannick Allanore, Daniel E Furst, Armando Gabrielli, Maureen Mayes, Jacob M van Laar, James R Seibold, Laszlo Czirjak, Virginia Steen, Murat Inanc, Otylia Kowal-Bielecka, Ulf Müller-Ladner Gabriele Valentini, Douglas Veale, Madelon Vonk, Ulrich A Walker, Lorinda Chung, Dave Collier, Mary Ellen Csuka, Barry J Fessler, Serena Guiducci, Ariane Herrick, Vivian Hsu, Sergio Jimenez, Bashar Kahaleh, Peter Merkel, Stanislav Sierakowski, Richard Silver, Rob Simms, John Varga, Janet E Pope

Why do we need SSc classification criteria?

1. To enroll SSc patients in studies (i.e., meeting classification criteria)
2. To differentiate SSc from other mimickers
3. To help to understand pathophysiology, subsets of patients with various features

Classification schemes – Overview



Which patient(s) meet the 1980 ACR SSc classification criteria?

1. Sclerodactyly, Raynaud's, positive anti-centromere, pulmonary arterial hypertension
2. Sclerodactyly, GERD, dilated esophagus, dysphagia RNA polymerase III, Scleroderma renal crisis
3. Sclerodactyly, Raynaud's, anti-centromere, dysphagia, dilated nailfold capillaries, calcinosis
4. Modified Rodnan skin score of 20 including finger involvement
5. Modified Rodnan skin score of 20 and no finger involvement ever, no Raynaud's Phenomenon and negative ANA

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90

Which patient(s) meet the 1980 ACR SSc classification criteria?

1. Sclerodactyly, Raynaud's, positive anti-centromere, pulmonary arterial hypertension **NO**
2. Sclerodactyly, GERD, dilated esophagus, dysphagia RNA polymerase III, Scleroderma renal crisis **NO**
3. Sclerodactyly, Raynaud's, anti-centromere, dysphagia, dilated nailfold capillaries, calcinosis **NO**
4. **Modified Rodnan skin score of 20 including finger involvement**
5. Modified Rodnan skin score of 20 and no finger involvement ever, no Raynaud's Phenomenon and negative ANA **NO**

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90

1980 ACR Scleroderma Classification Criteria

- **Major criterion:** proximal cutaneous sclerosis/skin thickening (non-pitting) proximal to the MCPs (metacarpophalangeal joints), affecting other parts of the body
 - usually bilateral, symmetrical, and almost always including sclerodactyly
 - **The major criterion is sufficient.**
- **Minor criteria: 2 or 3 are needed if no major criterion:**
 - sclerodactyly
 - digital pitting scars of fingertips or loss of substance of the distal finger pad
 - bibasilar pulmonary fibrosis

1 major criterion or ≥ 2 minor criteria were found in 97% of definite SSc cases (sensitivity 97%) but in only 2% of the comparison patients (specificity 98%)

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90.

What are the limitations of the 1980 ACR classification criteria for SSc?

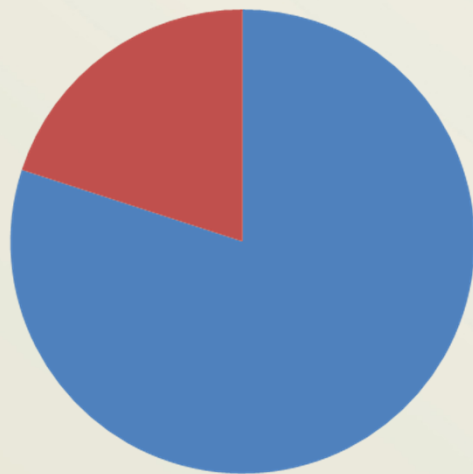
1. They don't classify some of those with limited cutaneous SSc
2. They don't always classify early SSc
3. They don't include antibodies that are common in SSc such as anti-centromere
4. They don't include nailfold changes that could help differentiate SSc from primary Raynaud's Phenomenon (RP)

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90

Limitations of the 1980 ACR Scleroderma classification

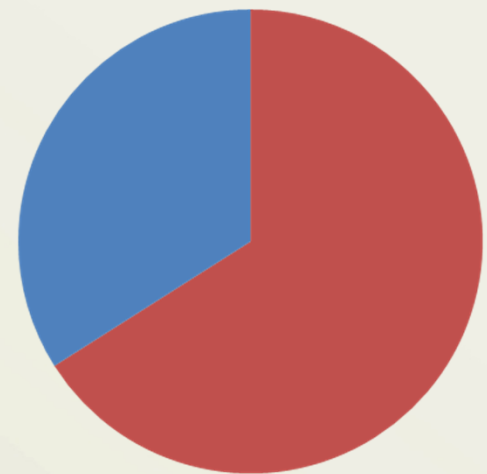
- ♦ Pittsburgh cohort of 639 patients with SSc¹

- ♦ 20% of limited SSc patients did not meet ACR criteria



- ♦ French Canadian cohort of 259 patients with SSc²

- ♦ 66% of limited SSc patients did not meet ACR criteria



■ ACR criteria met
■ ACR criteria not met

- ♦ **10 to 20% of limited SSc patients do not meet criteria for SSc³**
- ♦ Many SSc patients diagnosed by expert clinicians would be excluded based on ACR criteria

1. Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90 2. Lonzetti LS, et al. *Arthritis Rheum* 2001; 44:735-6. 3. J Walker, J

Pope Clinical Rheumatol 2007

Development of Classification and Response Criteria for Rheumatic Diseases

CLASSIFICATION AND RESPONSE CRITERIA SUBCOMMITTEE OF THE AMERICAN COLLEGE OF RHEUMATOLOGY COMMITTEE ON QUALITY MEASURES

Collaboration between experts and clinical epidemiologists

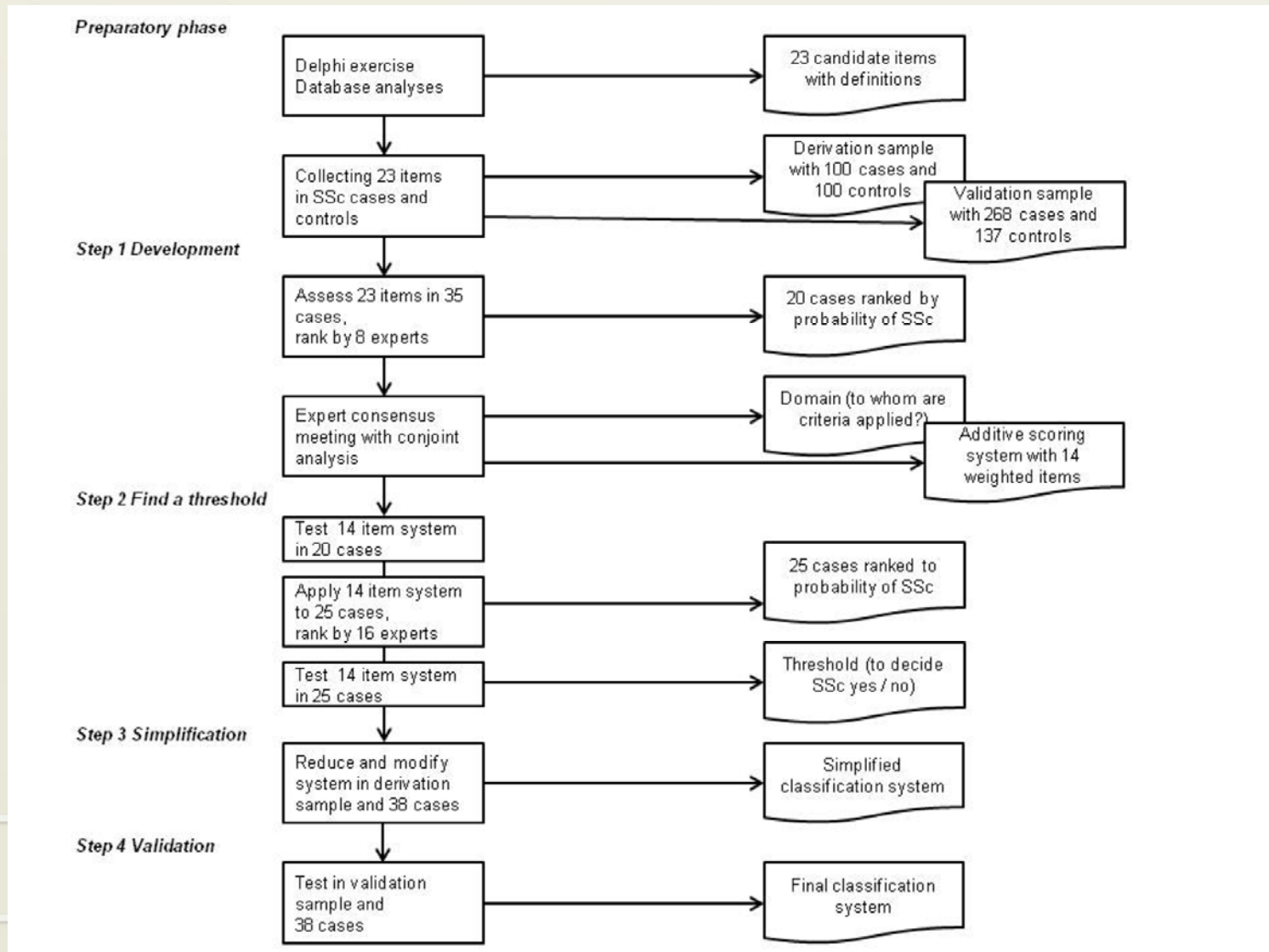
Balance of expert based and data driven methods

Avoid circularity of reasoning

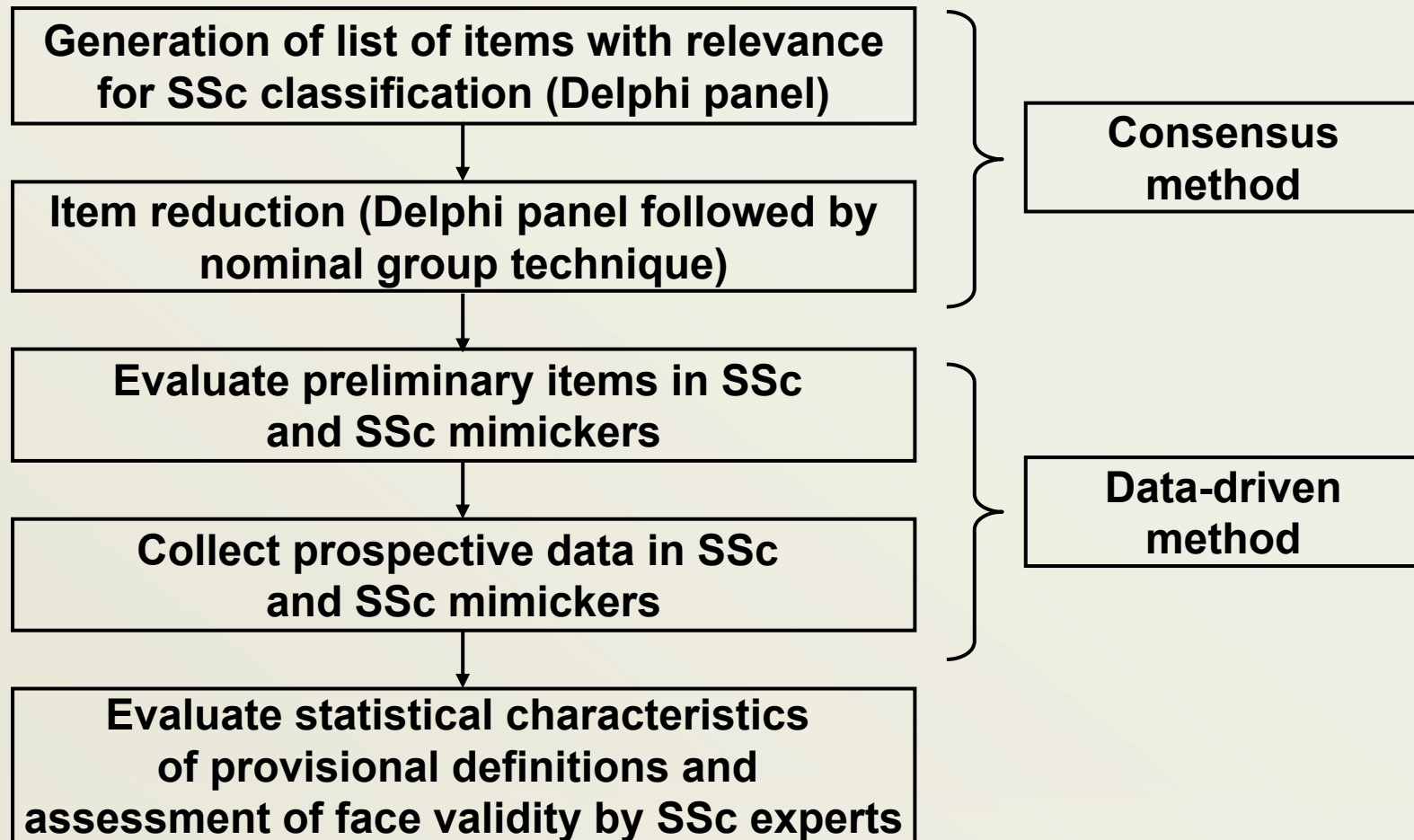
Evaluate psychometric properties of candidate criteria

Singh JA, et al. Arthritis Rheum. 2006 Jun 15;55(3):348-52.

Overview of the ACR / EULAR SSc Classification Criteria Development



Reclassification of SSc by the ACR / EULAR Task Force



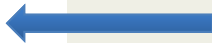
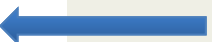
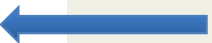
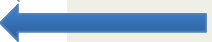
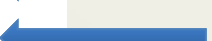
SSc mimickers = eosinophilic fasciitis, generalised morphea, other connective tissue disease, scleromyxedema, etc.

Delphi results: Items to be tested for the new ACR / EULAR SSc criteria

Item	Appropriateness score* (median)	TOP score	Rank
Positive antitopoisomerase I	9	73%	1
Presence of scleroderma	9	70%	2
Abnormal nailfold capillary pattern	9	67%	3
Positive anticentromere	9	62%	4
Positive anti-RNA polymerase III	8	43%	5
Finger tip ulcers or pitting scars	8	34%	6
Raynaud's phenomenon	7	33%	7
Interstitial lung disease/pulmonary fibrosis	7	28%	8
Renal crisis	8	27%	9

*Scored from 1 to 9, with a higher score denoting greater appropriateness for the classification of SSc

**DATA
driven
validation
of top SSc
items for
criteria
from
DELPHI
exercise**

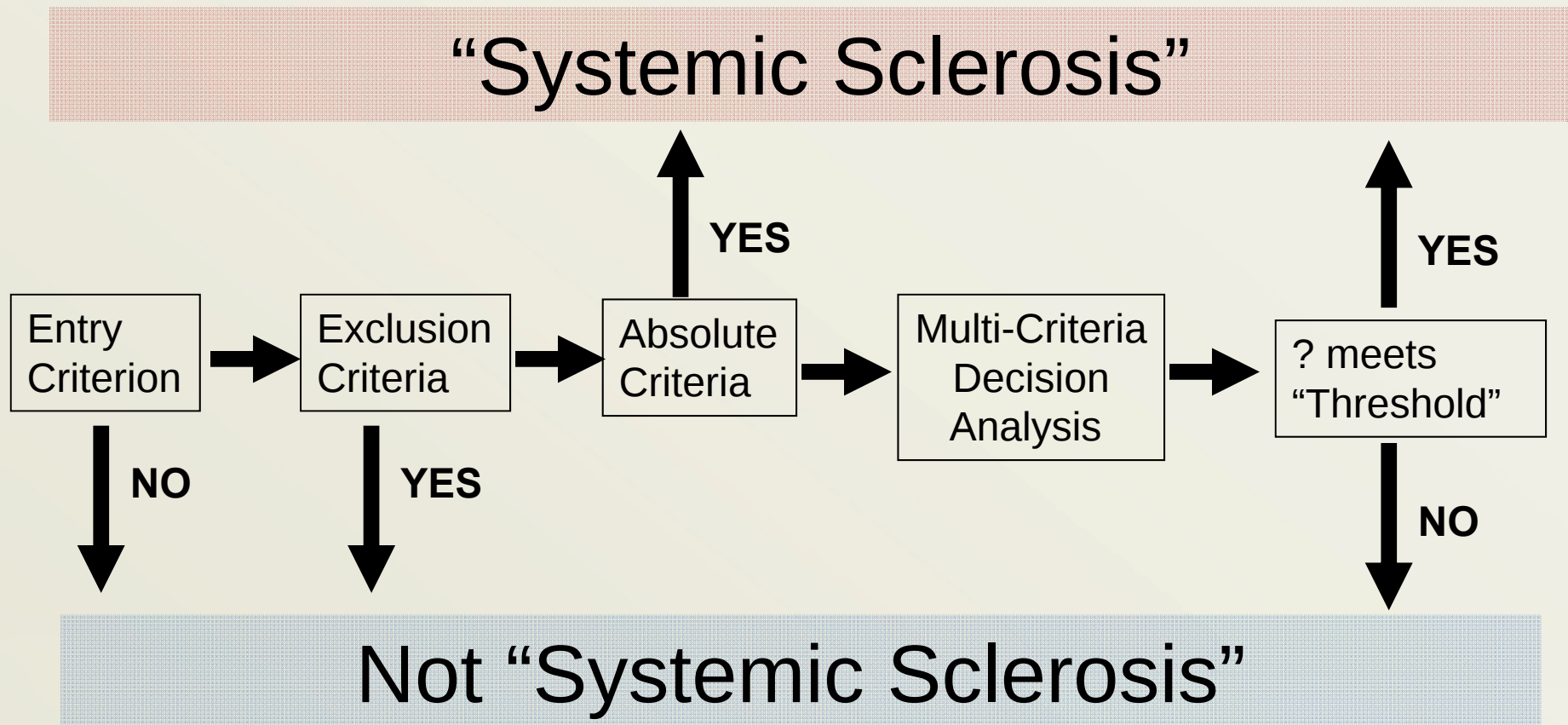
Criterion	Pooled mean OR (95% CrI)	Empiric ranking	Expert based ranking
Renal crisis	NC	1 ^{ab}	9
Digital pulp loss or acro-osteolysis	NC	1 ^{ab}	13 ^b
Scleroderma	426.7 (256.5, 691.2)	2	1 
Telangiectasia	91.4 (57.6, 154.5)	3	11
Anti-RNA polymerase III antibody	75.4 (13.2, 312.6)	4	6
Puffy fingers	34.9 (24.0, 49.2)	5	12
Finger or wrist flexion contracture	29.0 (17.8, 46.2)	6	19
Tendon or bursal friction rubs	26.81 (2.4, 91.9)	7	10
Anti-topoisomerase-1 antibody	24.9 (12.7, 48.0)	8	2 
Raynaud's phenomenon	24.1 (15.3, 37.5)	9	7
Finger tip ulcers or pitting scars	19.3 (12.7, 28.8)	10	5 
Anti-centromere antibody	13.8 (9.0, 21.0)	11	3 
Abnormal nailfold capillary pattern	10.4 (6.9, 15.1)	12	4 
Gastro-esophageal reflux disease	7.8 (6.1, 10.0)	13	17
Antinuclear antibody	6.06 (4.1, 8.8)	14	13 ^b
Calcinosis	6.05 (3.4, 10.5)	15	18
Dysphagia	5.7 (4.2, 7.7)	16	19
Esophageal dilatation	5.6 (2.9, 10.2)	17	14
Interstitial lung disease	4.5 (3.4, 5.8)	18	8
Anti-PM-SCL antibody	2.4 (1.9, 7.1)	19	20 ^b
Pulmonary arterial hypertension	1.2 (0.9, 1.5)	20	15
Carbon monoxide diffusion capacity (DLCO)	1.5 (1.1, 2.0)	21	16
Forced vital capacity (FVC)	0.9 (0.6, 1.3)	22	20 ^b

NC= Not Calculable

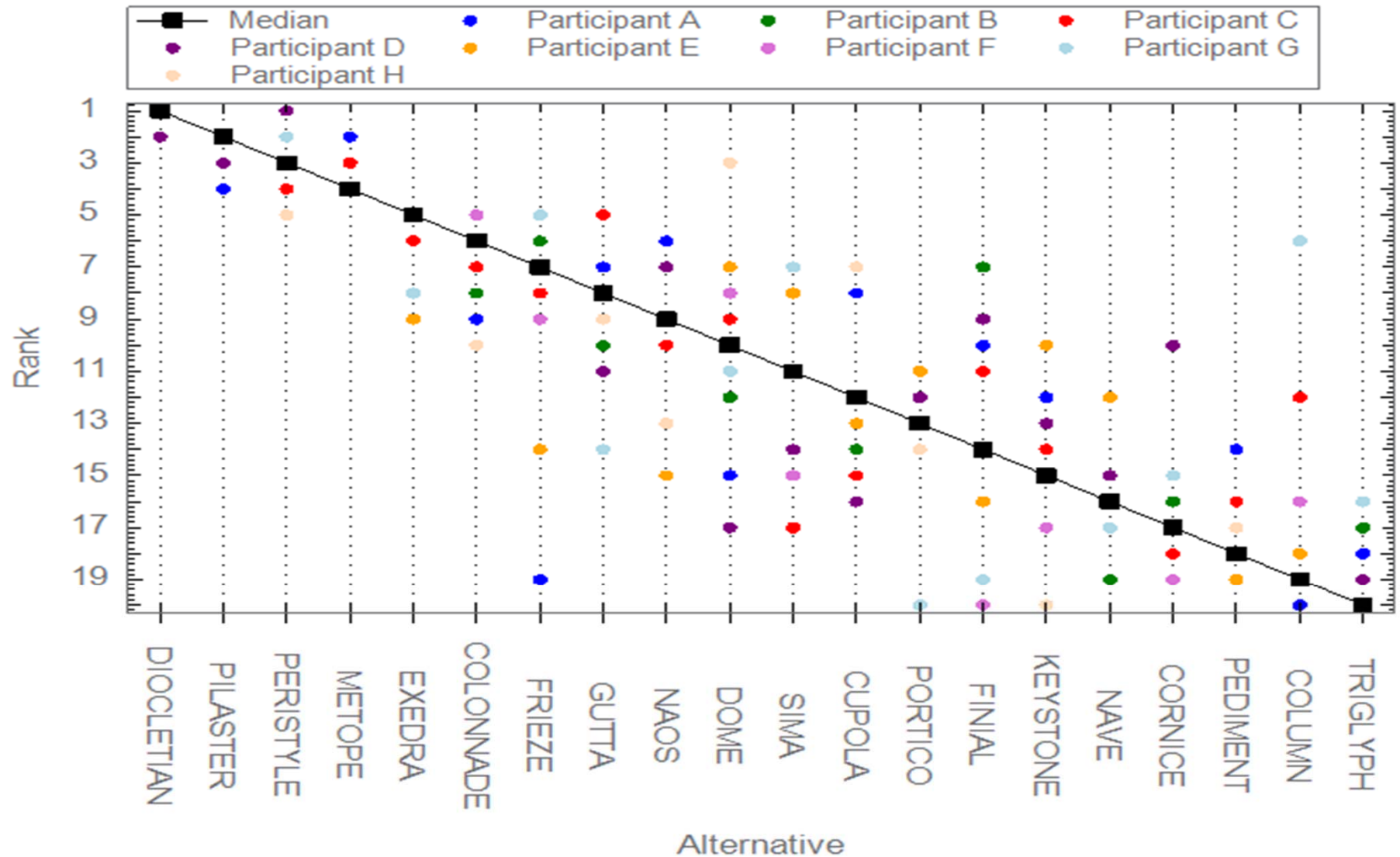
Johnson SR, et al. Arthritis Care Res (Hoboken). 2012 Mar;64(3):358-67.

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Classification of “Systemic Sclerosis” ACR / EULAR



Ranking of cases with a spectrum of probabilities as having SSc



Item	Derivation sample			Validation sample		
	SSc	Scleroderma-like disorder	p-value	SSc	Scleroderma-like disorder	p-value
N=	100	100	--	268	137	--
Age	55 (13)	51 (15)	0.05	54 (13)	52 (15)	0.17
Female	86 (86%)	79 (79%)	0.25	221 (83%)	101 (75%)	0.08
Region						
North America	50	50	--	191 (68%)	91 (32%)	0.32
Europe	50	50		77 (63%)	46 (37%)	
Time since onset of Raynaud's (years)	13 (7-18)	12 (4-18)	0.42	9 (5-18)	10 (4-22)	.40
Time since first non-Raynaud's symptom (years)	10 (4-13)	9 (2-14)	0.58	7 (3-12)	7 (3-15)	.89
Time since diagnosis (years)	8 (3-12)	6 (1-9)	0.10	5 (2-11)	4 (1-7)	.016
Scleroderma-like disorders		28 (28%)			32 (23%)	
systemic lupus erythematosus						
polymyositis/dermatomyositis		23 (23%)			21 (15%)	
primary Raynaud's syndrome		19 (19%)			7 (5%)	
mixed connective tissue disease		9 (9%)			14 (10%)	
undifferentiated connective tissue disease		8 (8%)			17 (12%)	
eosinophilic fasciitis		6 (6%)			16 (12%)	
nephrogenic sclerosing fibrosis		3 (3%)			3 (2%)	
generalized morphea		5 (5%)			8 (6%)	
scleromyxedema		1 (1%)			3 (2%)	
graft versus host disease		3 (3%)			3 (2%)	
other diagnoses		8 (8%)			13 (9%)	
Manifestations						
Raynaud's phenomenon	91 (91%)	49 (49%)	<0.0001	257 (96%)	63 (46%)	<0.0001
Autoantibodies	68	7 (7%)	<0.0001	137	15 (11%)	<0.0001

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Sensitivity and Specificity on ACR-EULAR Classification Criteria for SSc

	Derivation sample (N=200)		Validation sample (N=405)	
	Sensitivity (95% CI)	Specificity (95% CI)	Sensitivity (95% CI)	Specificity (95% CI)
1980 ARA SSc Criteria	.80 (.72-.87)	.77 (.68-.84)	.75 (.70-.80)	.72 (.64-.79)
2001 LeRoy and Medsger criteria	.76 (.68-.84)	.69 (.68-.84)	.75 (.70-.80)	.78 (.70-. .85)
2013 SSc Criteria	.95 (.90-.98)	.93 (.86-.97)	.91 (.87-.94)	.92 (.86-.96)

Item	Definition
Skin thickening	Skin thickening or hardening not due to scarring after injury, trauma, etc.
Puffy fingers	Swollen digits - a diffuse, usually nonpitting increase in soft tissue mass of the digits extending beyond the normal confines of the joint capsule. Normal digits are tapered distally with the tissues following the contours of the digital bone and joint structures. Swelling of the digits obliterates these contours. Not due to other reasons such as inflammatory dactylitis.
Finger tip ulcers or pitting scars	Ulcers or scars distal to or at the PIP joint not thought to be due to trauma. Digital pitting scars are depressed areas at digital tips as a result of ischemia, rather than trauma or exogenous causes.
Telangiectasia	Telangiectasia(e) in a scleroderma like pattern are round and well demarcated and found on hands, lips, inside of the mouth, and/or large matt-like telangiectasia(e). Telangiectasiae are visible macular dilated superficial blood vessels; which collapse upon pressure and fill slowly when pressure is released; distinguishable from rapidly filling spider angiomas with central arteriole and from dilated superficial vessels.
Abnormal nailfold capillary pattern consistent with SSc	Enlarged capillaries and/or capillary loss with or without peri-capillary hemorrhages at the nailfold and may be seen on the cuticle.
Pulmonary arterial hypertension	Pulmonary arterial hypertension diagnosed by right heart catheterization according to standard definitions.
Interstitial lung disease	Pulmonary fibrosis on HRCT or chest radiograph, most pronounced in the basilar portions of the lungs, or presence of 'Velcro' crackles on auscultation not due to another cause such as congestive heart failure.
Raynaud's phenomenon	Self report or reported by a physician with at least a two-phase color change in finger(s) and often toe(s) consisting of pallor, cyanosis and/or reactive hyperemia in response to cold exposure or emotion; usually one phase is pallor.
Scleroderma specific antibodies	Anti-centromere antibody or centromere pattern on antinuclear antibody (ANA) testing; anti-topoisomerase I antibody (also known as anti-Scl70 antibody); or anti-RNA polymerase III antibody. Positive according to local laboratory standards.

Defini- tions of terms

Classification criteria

- They are not diagnostic criteria but hopefully are close
- Thus early patients may not be classified
- Classification criteria do not equal diagnostic criteria, so it is possible that some patients may be diagnosed with SSc but don't meet the ACR / EULAR SSc classification criteria

Use of the ACR / EULAR SSc classification criteria

- **Exclusion criteria**

- You know there is a different disease
- Absence of sclerodactyly (ever) if skin involvement is elsewhere

- **Absolute inclusion criterion**

- Scleroderma skin involvement with sclerodactyly and contiguous skin involvement proximal to the MCPs



Caveats for ACR / EULAR SSc classification criteria

- Mark highest score in each category
- Not all centers have access to RNAPolymerase III Ab
- Don't apply criteria if you don't think patient has SSc or have a better explanation
 - Such as generalized morphea
- You can have more than one connective tissue disease (CTD) so a patient can have an overlap with SSc as one of the diseases and some patients with mixed connective tissue disease may be classified with SSc

Conclusions

- There is a need to effectively classify SSc patients
- Classification systems have evolved over the years with advancing technology
 - Capillaroscopy, autoantibodies, new methodologies
- Patients can be classified with more than one disease with the ACR / EULAR SSc classification criteria (i.e., overlaps)
- You have to do some math!
- Current classification includes more patients who would not have been classified by previous criteria

Conclusions about 2013 ACR/EULAR SSc classification criteria

- Items have good face validity
- Items have good discriminant validity
- Items have good construct validity
- The sensitivity and specificity of the ACR/EULAR SSc classification criteria are better than previous criteria

Using the 2013 ACR / EULAR SSC Classification Criteria

- Apply the criteria for SSc, if appropriate



1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of 9 or more are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly,
Raynaud's,
+ anti-centromere,
pulmonary arterial
hypertension**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
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**Sclerodactyly 4,
Raynaud's 2,
+ anti-centromere 3,
pulmonary arterial
hypertension, ILD 2**

Score 11

**Yes, criteria are
met**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

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Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly,
GERD, dilated
esophagus,
dysphagia
RNA polymerase III,
Scleroderma
renal crisis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Sclerodactyly, 4
GERD, 0
dilated esophagus, 0
dysphagia 0
RNA polymerase III
3,
Scleroderma renal
crisis 0

Total score 7,
SSc criteria not
met

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly just to PIP,
GERD, dilated esophagus,
dysphagia
RNA polymerase III,
RNP+
Scleroderma renal crisis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Sclerodactyly
just to PIP, 0
GERD 0,
dilated esophagus
0,
dysphagia 0
RNA polymerase
III 3,
RNP+ 0
Scleroderma renal
crisis 0

Total 3

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly,
Raynaud's,
+ anti-
centromere,
dysphagia,
dilated nailfold
capillaries,
calcinosis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly 4,
Raynaud's 3,
+ anti-
centromere 3,
Dysphagia 0,
dilated nailfold
capillaries 2,
calcinosis 0**

Total score 12

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '***Skin thickening sparing the fingers***' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of 9 or more are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Modified
Rodnan skin
score of 20
including
finger
involvement
Raynaud's
ANA
nucleolar
GERD
ILD**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Modified Rodnan skin score of 20 including finger involvement 9 Raynaud's 3 ANA nucleolar 0 GERD 0 ILD 2

Total score 14

NB criterion is met for skin thickening proximal to MCP so criteria don't need to be applied

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Modified Rodnan skin score of 20 and no finger involvement ever

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Modified Rodnan skin score of 20 and no finger involvement ever

Don't apply the criteria N/A Skin thickening sparing the fingers are not cl

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

No skin involvement ever, Telangiectasia, PAH, Raynaud's, +ANA

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (<i>only count the highest score</i>)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (<i>only count the highest score</i>)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

No skin involvement ever 0, Telangiectasia 2, PAH 2, Raynaud's 3, +ANA 0

Total score 7

SSc Criteria are not currently met

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

No skin involvement ever, Telangiectasia, PAH, Raynaud's, +ANA, +anti-centromere Ab

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

No skin involvement ever 0, Telangiectasia 2, PAH 2, Raynaud's 3, +ANA 0, +anti-centromere Ab 3

Total score 10

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (<i>only count the highest score</i>)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (<i>only count the highest score</i>)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**No skin
involvement
ever,
Telangiectasia,
ILD, PAH,
Raynaud's,
+ANA
+ScI70 (topo1)**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

No skin involvement ever 0, Telangiectasia 2, ILD and PAH 2, Raynaud's 3, +ANA 0, +ScI70 (topo1) 3

Total score 10

Not PAH and/or ILD count total of 2

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1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

No skin involvement ever, Telangiectasia, PAH, Raynaud's, +ANA, dilated capillaries at nailbeds in past (well documented)

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**No skin involvement ever, 0
Telangiectasia, 2
PAH, 2
Raynaud's, 3
+ANA, 0
dilated capillaries at nailbeds in past (well documented) 2**

**Total score 9
Criteria are met**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers,
Raynaud's,
Abnormal
nailfold cap,
+ANA
New dysphagia
retrosternally,
Scleroderma
renal crisis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Puffy fingers, 2
Raynaud's, 3
Abnormal
nailfold cap, 2
+ANA 0
New dysphagia
retrosteronally, 0
Scleroderma
renal crisis 0

Total score 7
Criteria are not
met

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers,
Raynaud's,
Abnormal
nailfold cap,
+ANA
New dysphagia
retrosterally,
Scleroderma
renal crisis,
tendon friction
rubs**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of 9 or more are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Puffy fingers, 2
Raynaud's, 3
Abnormal nailfold cap, 2
+ANA 0
New dysphagia retrosternally, 0
Scleroderma renal crisis, 0
tendon friction rubs 0

Total score 7
Criteria are not met

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers,
Raynaud's,
Abnormal
nailfold cap,
+ANA 0
New dysphagia
retrosternally, 0
Scleroderma
renal crisis, 0
+RNApol III**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Puffy fingers, 2
Raynaud's, 3
Abnormal nailfold cap, 2
+ANA 0
New dysphagia retrosternally, 0
Scleroderma renal crisis, 0
+RNApol III 3

Total score 10

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (<i>only count the highest score</i>)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (<i>only count the highest score</i>)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers,
Raynaud's,
Abnormal
nailfold cap,
+ANA
New dysphagia
retrosternally,
Scleroderma
renal crisis,
+RNApol III,
tendon friction
rubs,
Digital ulcers**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Puffy fingers, 2
Raynaud's, 3
Abnormal
nailfold cap, 2
+ANA 0
New dysphagia
retrosternally, 0
Scleroderma
renal crisis, 0
+RNApol III, 3
tendon friction
rubs, 0
Digital ulcers 2

Total score 12

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of 9 or more are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
dital to PIPs,
Raynaud's,
Abnormal
nailfold cap in
past, +ANA with
anti-centromere
pattern,
Digital pitted
scars**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
distal to PIPs, 0
Raynaud's, 3
Abnormal
nailfold cap in
past, 2
+ANA with anti-
centromere
pattern, 3
Digital pitted
scars 3**

Total score 11

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
distal to PIPs,
Raynaud's,
+ANA with anti-
centromere
Telangiectasia,
calcinosis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
dital to PIPs, 0
Raynaud's, 3
+ANA with anti-
centromere 3
Telangiectasia, 2
calcinosis 0**

**Total score 8
SSc criteria not
met**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
whole fingers
and tight mouth
and face,
Raynaud's,
+ANA with anti-
centromere
Telangiectasia,
calcinosis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
whole fingers and
tight mouth and
face, 4
Raynaud's, 3
+ANA with anti-
centromere 3
Telangiectasia, 2
calcinosis 0**

Total score 12

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sclerodactyly
whole fingers
and continuous
proximal to
MCPs to hand,
Raynaud's,
+ANA
Telangiectasia,
calcinosis
Needing repeat
esophageal
dilations**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Sclerodactyly whole fingers and continuous proximal to MCPs to hand, 9
Raynaud's, 3
+ANA 0
Telangiectasia, 2
calcinosis 0
Needing repeat esophageal dilations 0

Total score 14

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Tight skin but sparing fingers and hands,
Monoclonal gammopathy,
neg ANA,
Neg ENA
No Raynaud's
Weight loss
No dysphagia
No dilated capillaries**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Tight skin but sparing fingers and hands, Monoclonal gammopathy, neg ANA, Neg ENA No Raynaud's Weight loss No dysphagia No dilated capillaries

Do not apply criteria, no hand involvement ever with skin involvement elsewhere – suspect another diagnosis

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**SLE with +ANA,
anti-DNA,
+centromere Ab
Also finger tip
ulcers,
Sclerodactyly
whole fingers
Raynaud's,
Telangiectasia,
calcinosis
Needing repeat
esophageal
dilations**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**SLE with +ANA, 0
anti-DNA, 0
+centromere Ab 3
Also finger tip
ulcers, 2
Sclerodactyly
whole fingers 4
Raynaud's, 3
Telangiectasia, 2
calcinosis 0
Needing repeat
esophageal
dilations 0**

**Total score 14
Patient has SLE
and SSc overlap**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sjögren's with
+ANA,
centromere Ab
+Ro, La, RF
Puffy whole
fingers
Raynaud's,
Telangiectasia,
ILD**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerasel [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Sjögren's with +ANA, 0 centromere Ab 3 +Ro, La, RF 0 Puffy whole fingers 2 Raynaud's, 3 Telangiectasia, 2 ILD 2

Total score 12 Patient has Sjogren's and SSc overlap

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sjögren's with
+ANA,
centromere Ab
+Ro, La, RF
Puffy whole
fingers
Raynaud's,
Telangiectasia,
dilated
capillaires**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sjögren's with
+ANA, 0
centromere Ab 3
+Ro, La, RF 0
Puffy whole fingers
2
Raynaud's, 3
Telangiectasia, 2
dilated capillaires 2**

Total score 12

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (<i>only count the highest score</i>)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (<i>only count the highest score</i>)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sjögren's with
+ANA,
centromere Ab
+Ro, La, RF
Puffy whole
fingers
Raynaud's**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Sjögren's with
 +ANA, 0
 centromere Ab
 3
 +Ro, La, RF 0
 Puffy whole
 fingers 2
 Raynaud's 3**

**Total score 8,
 criteria not met
 for SSc**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Dermatomyositis+
ANA, +Jo1
Puffy whole
fingers
Raynaud's
Digital ulcer
Dilated capillaries
which resolved
with treatment of
myositis
Cape like Rash,
photosensitivity.
ILD**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Dermatomyositis+
ANA, +Jo1 0
Puffy whole
fingers 2
Raynaud's 3
Digital ulcer 2
Dilated capillaries
which resolved
with treatment of
myositis 2
Cape like Rash, 0
photosensitivity. 0
ILD 2**

**Total score 11,
BUT** you may
choose not to apply
criteria if SSc
overlap is not
suspected and you
think the features
are due to
dermatomyositis

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-Scl 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Dermatomyositis+
ANA, +Jo1
Sclerodactyly
distally to PIPs
Raynaud's
Dilated capillaries
which resolved
with treatment of
myositis
Cape like Rash,
photosensitivity.
ILD**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

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	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Dermatomyositis+
ANA, +jo1
Raynaud's
Digital ulcer
Dilated capillaries
which resolved
with treatment of
myositis
Cape like Rash,
photosensitivity.
ILD
Tight face and
tight fingers
proximal to MCPs**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

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Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Dermatomyositis+A
NA, +Jo1 0
Raynaud's 3
Digital ulcer 2
Dilated capillaries
which resolved with
treatment of
myositis 2
Cape like Rash, 0
photosensitivity. 0
ILD 2
Tight face and tight
fingers proximal to
MCPs 9

Total score 18
Overlap of
dermatomyositis
and SSc

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
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	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers+ANA,
Raynaud's
Dysphagia
Scleroderma
Renal crisis**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

Puffy fingers 2
+ANA, 0
Raynaud's 3
Dysphagia 0
Scleroderma
Renal crisis 0

Total score 5
SSc criteria not met

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerasel [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers 2
+ANA, 0
Raynaud's 3
Dysphagia 0
Scleroderma
Renal crisis 0**

**Patient returns
in two months
with skin
tightening to
above the
elbows
and tendon
friction rubs**

1. These criteria are applicable to any patient considered for inclusion in a SSc study.
2. These criteria are **not applicable** to patients having a systemic sclerosis-like disorder better explaining their manifestations, such as: nephrogenic sclerosing fibrosis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft versus host disease, and diabetic chierarthropathy. Patients with '**Skin thickening sparing the fingers**' also are not classified as having SSc.

Items	Sub-items	Weight / Score
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Skin thickening of the fingers (only count the highest score)	Puffy fingers	2
	Whole Finger, distal to MCP	4
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2
	Pitting Scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerase I [anti-ScL 70], anti-RNA polymerase III)		3
TOTAL SCORE^:		
Patients having a total score of <u>9 or more</u> are being classified as having definite systemic sclerosis. ^ Add the maximum weight (score) in each category to calculate the total score.		

**Puffy fingers 2
+ANA, 0
Raynaud's 3
Dysphagia 0
Scleroderma
Renal crisis 0**

**Patient returns
in two months
with skin
tightening to
above the
elbows 9
and tendon
friction rubs 0**

**Total score 14
Criteria now met**

Backup Slides

Early symptoms of SSc

- Raynaud's phenomenon
 - May precede skin changes and visceral disease by several years especially in limited SSc
 - In dcSSc, RP may begin shortly after or coincident with the onset of skin disease
 - SSc patients may have less hyperaemia than in primary Raynaud's due to structural vasculopathy in SSc
- Puffy fingers may be the presenting sign in dcSSc
- Malaise, fatigue, arthralgia, myalgia
- Gastroesophageal symptoms or dyspnoea may be the presenting symptoms

Which patient does **not** meet 1980 ACR SSc classification criteria?

1. Sclerodactyly and digital pits and Raynaud's, positive anti-centromere
2. Sclerodactyly and dysphagia and pulmonary fibrosis
3. Sclerodactyly, Raynaud's, anti-centromere, dysphagia, dilated nailfold capillaries, PAH and calcinosis
4. Modified Rodnan skin score of 20 including finger involvement

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90

Which patient does **not** meet 1980 ACR SSc classification criteria?

1. Sclerodactyly and digital pits and Raynaud's, positive anti-centromere
2. Sclerodactyly and dysphagia and pulmonary fibrosis
3. **Sclerodactyly, Raynaud's, anti-centromere, dysphagia, dilated nailfold capillaries, PAH and calcinosis**
4. Modified Rodnan skin score of 20 including finger involvement

Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. *Arthritis Rheum* 1980; 23:581-90

LeRoy criteria (2001): Classification criteria early SSc

ISSc	Raynaud's phenomenon (objective) Plus any one: SSc-type nailfold capillary pattern or SSc selective autoantibodies
	OR
	Raynaud's phenomenon (subjective only) Plus both SSc-type nailfold capillary pattern and SSc selective autoantibodies
IcSSc	Criteria for ISSc plus distal cutaneous changes
dcSSc	Criteria for ISSc plus proximal cutaneous changes

LeRoy EC and Medsger TA, Jr. *J Rheumatol* 2001; 28:1573-6.

Subsets of SSc

Puffy fingers, RP,
dilated capillaries
This patient has
limited SSc

Limited SSc

Limited
cutaneous SSc

Diffuse cutaneous SSc

SSc sine scleroderma

? Indeterminate subset,
? Overlaps

Adapted from Hachulla E, et al. *Clinical Rev Allergy Immunol* 2010; Epub ahead of print.



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