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Systemic Sclerosis Classification Criteria: Developing methods for multi-criteria decision analysis with 1000Minds

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Abstract

Objective—Classification criteria for systemic sclerosis (SSc) are being developed. The objectives were to: develop an instrument for collating case-data and evaluate its sensibility; use forced-choice methods to reduce and weight criteria; and explore agreement between experts on the probability that cases were classified as SSc.

Study Design and Setting—A standardized instrument was tested for sensibility. The instrument was applied to 20 cases covering a range of probabilities that each had SSc. Experts rank-ordered cases from highest to lowest probability; reduced and weighted the criteria using forced-choice methods; and re-ranked the cases. Consistency in rankings was evaluated using intraclass correlation coefficients (ICC).

Results—Experts endorsed clarity (83%), comprehensibility (100%), face and content validity (100%). Criteria were weighted (points): finger skin thickening (14–22), finger-tip lesions (9–21), friction rubs (21), finger flexion contractures (16), pulmonary fibrosis (14), SSc-related antibodies (15), Raynaud's phenomenon (13), calcinosis (12), pulmonary hypertension (11), renal crisis (11), telangiectasia (10), abnormal nailfold capillaries (10), esophageal dilation (7) and puffy fingers (5). The ICC across experts was 0.73 (95%CI 0.58,0.86) and improved to 0.80 (95%CI 0.68,0.90).

Conclusions—Using a sensible instrument and forced-choice methods, the number of criteria were reduced by 39% (23 to 14) and weighted. Our methods reflect the rigors of measurement science, and serves as a template for developing classification criteria.

Keywords

Scleroderma; Systemic Sclerosis; Decision Analysis; Forced-Choice; Classification Criteria; Conjoint Analysis; Sensibility

Classification criteria for rheumatic diseases are important for research and practice. Previous iterations of classification criteria have been criticized for their lack of methodological rigor, inability to reflect the changing construct of disease or inefficiency when applied in the real world.(1–4) Application of the preliminary criteria for classification of systemic sclerosis (SSc)(5–7) for recruitment into trials results in the exclusion of approximately 20% of patients with either early SSc or the limited subtype.(8–10) As a result, new classification criteria for SSc are being developed.(11)

In phase 1, 168 candidate criteria were generated through Delphi exercises.(11) The items were reduced to 23 criteria using another Delphi exercise and nominal group technique. The

23 criteria were validated using SSc and SSc-mimicking condition cohorts. The criteria were found to have good face, discriminant and construct validity.(12) The next phase of criteria development require further item reduction, weighting and scaling. The 2010 rheumatoid arthritis classification criteria were successfully developed with a balanced use of expert-based and data-driven methods. Forced-choice methods (facilitated by 1000Minds software) allowed for item reduction and item weighting.(13, 14) In order for these methods to be utilized in the development of SSc classification criteria, a SSc-specific instrument using a standardized format based on the 23 candidate items needed to be developed.(15)

The aim of this study was to develop a SSc-specific instrument for use in the forced-choice phase of SSc criteria development, and conduct a forced-choice study to reduce and weight the criteria. The sensibility of an instrument is critical to whether it is useful or not.(15) Attributes of sensibility include comprehensibility, clarity, face validity, content validity and feasibility. If an instrument is not sensible, it does not warrant use in clinical research.(15) Therefore, the first objective was to develop and evaluate the sensibility of a SSc-specific instrument for use in a forced-choice study. The second objective was to use forced-choice methods to reduce the criteria and to valueate relative weights for each criterion. The third objective was to explore the agreement among experts on which patients are considered to have SSc. Given the heterogeneity of SSc, we hypothesized that in the absence of distal AND proximal skin thickening present at the same time, there would be variability in agreement among experts on which patients have SSc. If this hypothesis were true, it would provide justification for the need to apply standardized classification criteria for inclusion of patients into research studies.

METHODS

Candidate criteria

Items were generated through consensus exercises resulting in 168 candidate criteria. A Delphi exercise and nominal group technique reduced the candidate criteria to 23:

- antinuclear antibody;
- anti-topoisomerase-I antibody;
- anti-centromere antibody or centromere pattern on antinuclear antibody test;
- anti-RNA polymerase III antibody;
- anti-PM-Scl antibody;
- scleroderma;
- puffy fingers;
- finger flexion contractures;
- tendon or bursal friction rubs
- calcinosis;
- telangiectasia;

- abnormal nailfold capillary pattern;
- Raynaud's phenomenon;
- finger tip ulcers and/or pitting scars;
- digital pulp loss or acro-osteolysis;
- renal crisis;
- pulmonary arterial hypertension;
- interstitial lung disease or pulmonary fibrosis;
- reduced diffusion capacity for carbon monoxide (DLCO);
- reduced forced vital capacity (FVC);
- dysphagia for solid foods;
- esophageal dilatation;
- persistent, recurrent gastro-esophageal reflux disease (GERD) by history. The response option was dichotomized as present or absent.(11) SSc subjects (n=783) were compared to subjects with diseases similar to SSc (n=1071). The candidate items were found to have good face, discriminant and construct validity.(12)

SSc-specific instrument development

The purpose of the disease specific instrument was to present SSc patient-based cases that differ in clinical characteristics in a standardized format.(15) Using Dillman methods(16), a standardized instrument was developed based on the 23 candidate criteria.(11)

Evaluation of sensibility

Sensibility is critical to the usefulness of an instrument.(17) Attributes of sensibility include comprehensibility, clarity of instructions and response options, face validity, content validity and feasibility.(15) The SSc-specific instrument was independently evaluated by 4 investigators for clarity, format, visual design, organization and navigation through the form. (15) Members of the SSc classification criteria steering committee (n=6) pilot tested 6 cases and the ranking procedure. This committee was composed of North American and European experts, each with experience in SSc care and research. The SSc-specific instrument and ranking procedure were evaluated for clarity of the instructions, comprehensibility of the response option and time to completion. The cases and instructions were revised based on pilot testing results.

The SSc-specific instrument was applied to 34 cases from 20 Scleroderma Centers in North America and Europe. The cases were purposively sampled to reflect the spectrum of probability that each case could be classified as having SSc (high, intermediate or low probability) by experts. The case mix was sampled to have a balance of patients with early disease (<2 years since the onset of the first non-Raynaud's symptom) versus late disease, and those with the limited and diffuses subtypes. Cases had a spectrum of combinations of present and absent items from among the 23 criteria. Each case was assigned a unique

architecturally based identifier. This method has been demonstrated to reduce potential bias from alphanumeric labeling, and has been used in other criteria development studies.(14) Twenty of 34 cases were purposively sampled for the ranking procedure, and the remainder held in reserve. (Table 1.)

Forced-choice study

Four European and four North American SSc experts were invited to participate in the forced-choice phase.(18) Experts were selected based on geographic representation, clinical and research expertise, and availability. At least half of them had not participated significantly in previous phases of this criteria development.(11, 12)

First ranking exercise—The 20 cases, answer form, standardized definitions, and standardized instructions were sent electronically to the expert panel (Appendices 1–3). In order to minimize the presence of ordering bias, the cases were sorted randomly. A randomly selected half of the experts (group A) were given one set of cases. The cases were again randomly sorted, and given to the remaining experts (group B). Using a standardized response form, experts were asked to rank order the cases from 1 (highest probability of having SSc) to 20 (lowest probability of having SSc). Data from earlier phases of criteria development were not given to the experts to reduce the effect of anchoring bias.(19) A logic check was incorporated into the response form to reduce data errors. Experts were blinded to other experts' rankings.

Item reduction and weighting—To enumerate experts' preferences for criteria contributing to the probability of a case being classified as SSc, multi-criteria decision analysis (forced-choice methods, conjoint analysis) was used. These methods are suited to the valuation of criteria that differ, but each may contribute to the overall construct of the classification of disease.(20) The experts met for a consensus meeting and were presented their rankings. The experts discussed the need for 1) entry criterion, 2) exclusion criterion, and 3) sufficient (or absolute) criterion for inclusion. (Figure 1). The criteria were discussed individually, to determine whether any could be grouped (if they were not independent, e.g., the SSc-related antibodies) or were essentially aspects of the same essential criterion (e.g., those related to lung involvement). The definitions of the criteria were examined to ensure that they could be interpreted and applied consistently. (Appendix 4).

The experts participated in a multi-criteria decision analysis to determine the relative weighting of each of the criteria using 1000Minds software(12). The panel was presented paired scenarios differing in 2 criteria. (Figure 2). For each pair, they anonymously (using touch key pads) selected the scenario they believed had a higher probability of being classified as having SSc. The distribution of opinions on each dilemma was presented to the group. Where there was not agreement, the reasons for disagreement were discussed. Sometimes the group re-voted. Consensus was considered achieved when all experts indicated agreement or could accept the majority decision. Through iterative discrete pairwise choices, the decision analytic software was able to assign relative weights to the criteria (12). During this consensus and weighting process, criteria were eliminated and revised. The weighting exercise was repeated when all of the criteria had been agreed upon. The resulting

weights were presented to the expert panel. Our methods conform with good research practices for conjoint analysis.(20)

Second ranking exercise—The SSc-specific instrument and the initial 20 cases were revised, eliminating criteria that had been discarded and refining terms and definitions. The revised cases and a standardized answer form were sent to the experts and steering committee (n=13). Participants were asked to order the cases from 1 (highest probability of having SSc) to 20 (lowest probability of having SSc).

Analysis

Summary statistics were used to describe the data. Randomization was achieved using a computerized random number generator. The distribution of rankings for each case and each expert panel member was plotted. The consistency in rankings was evaluated using an intraclass correlation coefficient (ICC). Landis and Koch have characterized values of reliability coefficients as follows: slight (0–0.20), fair (0.21–0.40), moderate (0.41–0.60), substantial (0.61–0.80), and almost perfect (0.81–1.00).(21) The ICC (2,1) of Shrout and Fleiss was used to assess the reliability of k=8 ratings assuming that all 20 cases were rated by the same raters who are a subset of all possible raters. With 8 observations for each case and a hypothesized value of 0.4 for reliability, we had 80% power to reject a minimally acceptable level of reliability of 0.2 with alpha=0.05.(23) Analyses were performed using R (version 2.2.1, The R Foundation for Statistical Computing).

Institutional research ethics board approval was obtained for collection of patient data.

RESULTS

SSc specific instrument testing

Participants endorsed clarity and ease of navigation of the form (83% (5/6)), clarity of the instructions (100% (6/6)), and clarity of the response option (100% (6/6)). None of the participants endorsed insufficient face or content validity. The median time to completion was 10 (range 10–20) minutes.

SSc Experts

All experts invited to participate consented. Five were male. The median years treating SSc patients were 30 (range 13–40) years. Fifty percent of experts practice in Europe and 50% in North America. Thirty-eight percent (3/8) had some involvement in earlier iterations of criteria development. All the steering committee members (n=8) participated in the ranking so they had a full understanding of the methods, but their rankings were not included in the first ranking analysis in order to reduce bias.

First ranking exercise

Table 1 summarizes the case characteristics. Experts' initial ranking of cases is presented in Figure 3. For the first ranking exercise, the ICC for agreement across experts was 0.73 (95%CI 0.58, 0.86). The ICC for expert group A was 0.68 (95%CI 0.48, 0.84) and the ICC for expert group B was 0.76 (95%CI 0.60, 0.88). The overlapping confidence intervals that

include both point estimates suggest there was no significant difference in rankings between expert groups.

Item reduction and weighting

Through consensus discussion, the experts agreed on further precision in the use of the criteria. They decided that ‘skin thickening sparing the fingers’ would be an exclusion criterion. If this exclusion was present, the use of the SSc classification criteria should not proceed. The expert panel agreed that ‘skin thickening of the fingers extending proximal to the metacarpophalangeal joints’ was a sufficient criterion for classification as SSc. If present, the patient could be classified as SSc, and use of the classification criteria is not required. After the weighting exercise, FVC, DLCO, antinuclear antibody, dysphagia, persistent GERD, and digital pulp loss were discarded due to their relative low weights, difficulties in assessing reliably in a clinical context, or having low value in discriminating patients with high from low probability of SSc. Skin thickening was redefined as skin thickening of the fingers, with response options a) distal to metacarpophalangeal joints, or b) distal to proximal interphalangeal joints. Finger lesions were consolidated under ‘finger tip lesions,’ with the response options categorized as a) pitting scars, b) digital tip ulcers, or c) clinical evidence of acro-osteolysis. Serology was consolidated under ‘scleroderma related antibodies’ and categorized as positive or negative. This process reduced candidate criteria from 23 to 14. The relative weightings are presented in Table 2. The ranking of the reduced, weighted criteria was compared to the empiric ranking based on frequency of criteria in cases and controls using SSc cohorts.(12) The ICC for agreement at 0.44 ($p=0.045$)

Second ranking exercise

Experts’ second ranking of cases is presented in Figure 4. For the second ranking exercise done after the face-to-face meeting, the ICC for agreement across experts was 0.80 (95%CI 0.68, 0.90).

DISCUSSION

Methodologic rigor is necessary for quality data and valid inferences(24), and this is particularly true as we enter an era of disease classification criteria redevelopment.(1) In this paper, we outline the methods employed to develop a SSc-specific instrument and strategies used to reduce the effect of potential biases. Often, methodologic detail is excluded, thereby limiting critical evaluation of the methods employed. Our explicit methodologic detail in criteria development may inform other groups, particularly if they plan to utilize forced-choice methods endorsed by the American College of Rheumatology (ACR) for the development of rheumatoid arthritis classification criteria.(14) The methodologies outlined conform to modern standards of measurement science and the recommendations of ACR and EULAR.(2, 25)

Our methods have employed bias reduction strategies. We have used strategies to reduce potential biases resulting from lack of clarity in the instructions, misunderstanding the response option, lack of clarity of the instrument, alphanumeric labeling of cases, data entry errors, and ordering bias.(15, 16, 26) All of these potential sources of bias have been shown

to reduce the validity and/or reliability of the data.(19) Our forms have demonstrable clarity, and feasibility. In addition, blinding was used so no respondent was 'singled out' as an outlier. Although the SSc classification criteria steering committee participated in the process, their rankings were not included in this analysis. This was done to reduce potential bias as the committee members have been involved in most of the process to date. Circularity of reasoning is a bias that can occur when the same experts who contribute patients are the ones who contribute to criteria development.(1) This is a critical bias to previous iterations of classification criteria development.(1, 3) We have ensured that some experts have not been involved in previous phases of the current criteria development to avoid the bias introduced by circular reasoning. Together, our evaluation of instrument design, implementation methods and pilot testing are quality indicators of good research practice.(20)

We have detailed the methodologic steps in the item reduction phase, leading up to and including the forced-choice experiment. The ranking of cases representing the spectrum of probability of having SSc, the group discussion among experts regarding possible reasons for lack of agreement in case rankings, and the discarding of criteria that lack reliability or discriminant validity were necessary prerequisites to the forced-choice experiment. Our use of psychometric properties of the candidate criteria as a means of item reduction led to a more manageable number of criteria and informed discussion during the forced-choice experiment.

A strength of this study is the use of forced-choice methods. Forced-choice methodology allowed us to identify relatively weak criteria and to determine a hierarchy among a reduced number of criteria that are influential in SSc classification. This is a more robust, scientific approach than retaining numerous criteria that are all equally weighted (and not necessarily reflective of the construct of the disease). The use of forced-choice methods to reduce items and ascertain relative weights, instead of additional Delphi exercises with/without nominal group technique, conferred a number of advantages. 1000Minds software facilitated a systematic process for a points-based prioritization system based on clinical cases. Using experts' knowledge and preferences, this method seeks to rank *all* hypothetically-possible patients representable by a given points system. From the overall ranking, it will derive the point values for the point stem thereby matching the expert panels' knowledge and preferences. The overall ranking of all hypothetically-possible patients is arrived at by asking the expert panel to make trade-offs between 2 criteria at a time. The pairwise ranking of just 2 alternatives is *cognitively less burdensome* to derive the weights.(14) Using the property of transitivity (if $a > b$, and $b > c$, then $a > c$), tens of thousands of cases can be automatically and incontrovertibly ranked. This method is *more efficient* than others because any pairwise decisions, in which one option clearly has a high probability or where consensus has been achieved, are not presented. This method can be *administered over the internet*, and individual *criterion can be modified*, such as when new information becomes available, and the weightings recalculated without disturbing the validity of the method or the previous consensus decisions made.(14)

A potential limitation of this study is the relatively small number of cases used in the ranking procedure, and the small number of experts involved. The inclusion of additional

cases would result in a greater representation of the diversity of clinical characteristics. In contrast, a strength of our approach is that all the criteria were included in each case as present or absent and all cases had different positive and negative items that varied. Furthermore, previous work has shown that participants have difficulty performing relative rankings of more than about 20 cases. The inclusion of additional cases reduces the validity and reliability of the rankings, thereby affecting the internal validity of the study findings.

In conclusion, our SSc-specific forms have demonstrable sensibility. Forced-choice methods were successfully utilized to reduce criteria and to indicate relative weights. The work described in this paper defines a system of criteria development, which produces a measure of the relative probability that a particular case (combination of clinical features) has SSc. The next phase of SSc classification criteria development will determine the need for further item reduction, possible re-weighting and scaling.(30, 31) The appropriate threshold to classify a patient as having SSc will need to be identified. The impact of criteria elimination, criteria clustering and threshold on sensitivity and specificity of the criteria will need to be tested using data-driven methods. Our methods reflect the rigors of modern measurement science, and may serve as a template for other groups developing classification criteria.

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Appendix 1. Standardized instructions

INTRODUCTION

The aim of the workshop in Chicago is to develop a system for classifying individual patients as to whether they have sufficient clinical and laboratory criteria to justify the classification of “Systemic Sclerosis”.

The process we will follow will use clinical cases, initially to ensure that we have the appropriate criteria in a format which makes sense clinically, then to develop and refine the method of combining the criteria for weighting, and finally to validate the system.

The first step is to take a set of clinical cases and ask each of the expert panel to assess the degree of confidence they have that each case has (or does not have) Systemic Sclerosis. A systematic way to facilitate an efficient discussion of these opinions is to have each expert panelist rank the cases and to discuss the results with the panel at the beginning of the workshop. In this way we can avoid spending time on issues on which we all largely agree and focus our efforts on resolving areas of controversy.

INSTRUCTIONS

Please rank the attached cases in order of the relative probability that the patient/case has Systemic Sclerosis. Please rank the case with the highest probability as “1”, the second most probable as “2”, etc down to the case with the lowest probability as “20”.

This is a difficult exercise but it is important to make your judgments. It is necessary to rank every case above or below the other cases - you cannot give cases equal ranks.

You will probably find it easiest to print off the 20 cases as separate pages, one for each case. It is wise to shuffle the cases out of the order in which they print, to avoid a bias from this order.

Then sort these pages (cases) into an order from highest to lowest probability. It is often easiest to lay out the pages on a large table or bench or even the floor, then move them around until you are happy with the order.

Once you have the cases sorted, collect them into a pile in order of your assessed probability and then number them from “1” (the highest probability) to “20” (the lowest).

Then open up the EXCEL table and enter the number (the rank) of each case against the case name in the column under your name. (I have entered random data under my name as an example).

When you have finished, check the “CHECK SUM” total at the base of your column - if you have entered the data correctly (no duplicates, typos, etc) the sum of your ranks will be 210.

Appendix 2. Response Form

	CLINICIAN	
CASE	Your ranking	Example
COLONNADE		5
COLUMN		10
CORNICE		15
CUPOLA		20
DIOCLETIAN		4
DOVE		9
EXEDRA		14
FINIAL		19
FRIEZE		3
GUTTA		8
KEYSTONE		13
METOPÉ		18
NAOS		2
NAVE		7
PEDIMENT		12
PERISYLE		17
PILASTER		1
PORTICO		6
SIMA		11
TRIGLYPH		16
CHECK SUM	0	210

Appendix 3. Cases

CASE NAME COLONNADE

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

Clinical case presentation and progress (include disease duration):
34 year old Caucasian patient referred for evaluation of lung fibrosis.

Clinical features

☐ Yes ☒ No Skin thickening

Describe location, extent, progression:

☒ Yes ☐ No Abnormal nailfold capillaries consistent with scleroderma

☐ Yes ☒ No Calcinosis

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

Clinical case presentation and progress (include disease duration):
34 year old Caucasian patient referred for evaluation of lung fibrosis.

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

Laboratory features

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

Investigations

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

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

CASE NAME _____ **GUTTA**_____

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

Clinical case presentation and progress (include disease duration): 16 year old Caucasian patient referred for Raynaud's phenomenon for 2 years and muscle weakness.

Clinical features

☒ Yes	☐ No	Skin thickening
Describe location, extent, progression: Mild skin thickening on the face, no sclerodactyly		
☒ Yes	☐ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis

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

Clinical case presentation and progress (include disease duration): *16 year old Caucasian patient referred for Raynaud's phenomenon for 2 years and muscle weakness.*

☐ Yes	☒ No	Dysphagia for solids
☒ Yes	☐ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☒ Yes	☐ No	Interstitial lung disease or pulmonary fibrosis
☐ Yes	☒ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

Any other information which may be relevant to the probability this patient has Systemic Sclerosis: *Elevated CPK in 3,000 and responded well to high dose prednisone and IVIG therapy*

CASE NAME SIMA

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

Clinical case presentation and progress (include disease duration):
47 year old Chinese female referred with shortness of breath on exertion

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☐ Yes	☒ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids

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

Clinical case presentation and progress (include disease duration):
47 year old Chinese female referred with shortness of breath on exertion

☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☒ Yes	☐ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☒ Yes	☐ No	Pulmonary arterial hypertension
☐ Yes	☒ No	Gastro-esophageal reflux disease
☐ Yes	☒ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME KEYSTONE

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

Clinical case presentation and progress (include disease duration):
65 year old Caucasian patient referred for evaluation of positive lupus test

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☐ Yes	☒ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☒ Yes	☐ No	Esophageal dilation

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

Clinical case presentation and progress (include disease duration):
65 year old Caucasian patient referred for evaluation of positive lupus test

☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☐ Yes	☒ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☐ Yes	☒ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME __PEDIMENT__

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

Clinical case presentation and progress (include disease duration):
56 year old Caucasian male with rapidly progressive skin thickening and induration over 12 weeks

Clinical features

☒ Yes	☐ No	Skin thickening
Describe location, extent, progression: <i>Skin involvement of wrists, forearms, upper chest, proximal legs, distal legs and ankles. No skin involvement of hands, feet nor face.</i>		
☐ Yes	☒ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation

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

Clinical case presentation and progress (include disease duration):

56 year old Caucasian male with rapidly progressive skin thickening and induration over 12 weeks

☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☒ Yes	☐ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☐ Yes	☒ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME ____ **CORNICE** ____

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

Clinical case presentation and progress (include disease duration):

47 year old Caucasian female referred with shortness of breath

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☐ Yes	☒ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture

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

Clinical case presentation and progress (include disease duration):
47 year old Caucasian female referred with shortness of breath

☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☒ Yes	☐ No	Pulmonary arterial hypertension
☐ Yes	☒ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

Any other information which may be relevant to the probability this patient has Systemic Sclerosis:
anticardiolipin antibody positive, systemic hypertension

CASE NAME EXEDRA

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

Clinical case presentation and progress (include disease duration):
65 year old female with disease duration for 30 years

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☒ Yes	☐ No	Abnormal nailfold capillaries consistent with scleroderma
☒ Yes	☐ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☒ Yes	☐ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars

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

Clinical case presentation and progress (include disease duration):
65 year old female with disease duration for 30 years

☐ Yes	☒ No	Puffy fingers
☒ Yes	☐ No	Interstitial lung disease or pulmonary fibrosis
☒ Yes	☐ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☒ Yes	☐ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME CUPOLA

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

Clinical case presentation and progress (include disease duration):
60 year old Caucasian female referred with anti-centromere antibody

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☐ Yes	☒ No	Abnormal nailfold capillaries consistent with scleroderma
☒ Yes	☐ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☒ Yes	☐ No	Puffy fingers

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

Clinical case presentation and progress (include disease duration):
60 year old Caucasian female referred with anti-centromere antibody

☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☐ Yes	☒ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☐ Yes	☒ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME _____ FINIAL_____

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

Clinical case presentation and progress (include disease duration):
34 year old Caucasian patient referred for chronic intestinal pseudo-obstruction

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☒ Yes	☐ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☒ Yes	☐ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis

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

Clinical case presentation and progress (include disease duration):
34 year old Caucasian patient referred for chronic intestinal pseudo-obstruction

☐ Yes	☒ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

Any other information which may be relevant to the probability this patient has Systemic Sclerosis:
Low WBC of 3.0, +SSA, + RNP, admission of cardiac tamponade requiring window

CASE NAME PORTICO

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

Clinical case presentation and progress (include disease duration):
40 year old female with recurrent pericarditis and anti-ScL antibody

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☒ Yes	☐ No	Abnormal nailfold capillaries
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☐ Yes	☒ No	Pulmonary arterial hypertension

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

Clinical case presentation and progress (include disease duration):
40 year old female with recurrent pericarditis and anti-ScL antibody

☒ Yes	☐ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

Any other information which may be relevant to the probability this patient has Systemic Sclerosis: *recurrent pericarditis, fever, anti-Jo-1 antibody*

CASE NAME FRIEZE

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

Clinical case presentation and progress (include disease duration):
50 year old Caucasian female with cough of 6 months duration

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☐ Yes	☒ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☒ Yes	☐ No	Finger tip ulcers or pitting scars
☒ Yes	☐ No	Puffy fingers
☒ Yes	☐ No	Interstitial lung disease or pulmonary fibrosis
☒ Yes	☐ No	Pulmonary arterial hypertension
☐ Yes	☒ No	Gastro-esophageal reflux disease

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

Clinical case presentation and progress (include disease duration):
50 year old Caucasian female with cough of 6 months duration

☐ Yes	☒ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME **__ DOME**

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

Clinical case presentation and progress (include disease duration):
75 year old Caucasian referred for hypertensive emergency and sudden renal failure

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☒ Yes	☐ No	Abnormal nailfold capillaries consistent with scleroderma
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☐ Yes	☒ No	Dysphagia for solids
☐ Yes	☒ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☐ Yes	☒ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon

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

Clinical case presentation and progress (include disease duration):
75 year old Caucasian referred for hypertensive emergency and sudden renal failure

☒ Yes	☐ No	Renal crisis
☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

Any other information which may be relevant to the probability this patient has Systemic Sclerosis: Renal biopsy consistent with microangiopathy with no cellular deposition and no crescents

CASE NAME NAOS

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

Clinical case presentation and progress (include disease duration):
55 year old West Indian female

Clinical features

☐ Yes	☒ No	Skin thickening
Describe location, extent, progression:		
☒ Yes	☐ No	Abnormal nailfold capillaries
☐ Yes	☒ No	Calcinosis
☐ Yes	☒ No	Digital pulp loss or acro-osteolysis
☒ Yes	☐ No	Dysphagia for solids
☒ Yes	☐ No	Esophageal dilation
☐ Yes	☒ No	Finger flexion contracture
☐ Yes	☒ No	Finger tip ulcers or pitting scars
☐ Yes	☒ No	Puffy fingers
☐ Yes	☒ No	Interstitial lung disease or pulmonary fibrosis
☐ Yes	☒ No	Pulmonary arterial hypertension
☒ Yes	☐ No	Gastro-esophageal reflux disease
☒ Yes	☐ No	Raynaud's phenomenon
☐ Yes	☒ No	Renal crisis

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

Clinical case presentation and progress (include disease duration):
55 year old West Indian female

☐ Yes	☒ No	Telangiectasias
☐ Yes	☒ No	Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

Any other information which may be relevant to the probability this patient has Systemic Sclerosis: recurrent small bowel bacterial overgrowth syndrome; difficulty maintaining BMI > 20 due to involuntary vomiting and diarrhea; requires g-tube for nutrition; history of chronic obstructive pulmonary disease and smoking

CASE NAME NAVE

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

Clinical case presentation and progress (include disease duration):
36 year old African-American female with arthralgia

Clinical features

☐ Yes	☒ No	Skin thickening
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Describe location, extent, progression:

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

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

Clinical case presentation and progress (include disease duration):
36 year old African-American female with arthralgia

☐ Yes ☒ No Tendon or bursal friction rubs

Laboratory features

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

Investigations

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

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

CASE NAME _PILASTER

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

Clinical case presentation and progress (include disease duration):
72 year old female

Clinical features

☒ Yes ☐ No Skin thickening
Describe location, extent, progression: *grade 3 thickening over fingers, grade 1 thickening over hands, face and legs*

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

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

Clinical case presentation and progress (include disease duration):
 72 year old female

Laboratory features

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

Investigations

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

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

CASE NAME _COLUMN_

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

Clinical case presentation and progress (include disease duration):
 62 year old female

Clinical features

☐ Yes	☒ No	Skin thickening
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Describe location, extent, progression:

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

Laboratory features

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

Clinical case presentation and progress (include disease duration):
62 year old female

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

Investigations

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

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

CASE NAME _TRIGLYPH_

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

Clinical case presentation and progress (include disease duration):
24 year old female

Clinical features

☐ Yes	☒ No	Skin thickening
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Describe location, extent, progression:

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

Laboratory features

☒ Positive	☐ Negative	Antinuclear antibody
☐ Positive	☒ Negative	Anti-centromere antibody

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

Clinical case presentation and progress (include disease duration):
24 year old female

☐ Positive	☒ Negative	Anti-topoisomerase-I antibody
☐ Positive	☒ Negative	Anti-PM-Scl antibody
☐ Positive	☒ Negative	Anti-RNA polymerase III antibody

Investigations

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

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

CASE NAME DIOCLETIAN

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

Clinical case presentation and progress (include disease duration):
52 year old Caucasian female

Clinical features

☒ Yes	☐ No	Skin thickening
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Describe location, extent, progression: *thickened skin over fingers, hands, arms, face, anterior chest, abdomen, legs and feet*

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

Laboratory features

☒ Positive	☐ Negative	Antinuclear antibody
☐ Positive	☒ Negative	Anti-centromere antibody
☒ Positive	☐ Negative	Anti-topoisomerase-I antibody

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

Clinical case presentation and progress (include disease duration):
52 year old Caucasian female

☐ Positive	☒ Negative	Anti-PM-Scl antibody
☐ Positive	☒ Negative	Anti-RNA polymerase III antibody

Investigations

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

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

CASE NAME _METOPE_

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

Clinical case presentation and progress (include disease duration):

Clinical features

☒ Yes	☐ No	Skin thickening
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Describe location, extent, progression:

Face and neck, both arms from fingers up to elbows.

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

Laboratory features

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

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

Clinical case presentation and progress (include disease duration):

Investigations

☒ **Yes** ☐ **No** DLCO < 80% predicted
☐ **Yes** ☒ **No** FVC < 80% predicted

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

CASE NAME ___**PERISTYLE**___

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

Clinical case presentation and progress (include disease duration):

Clinical features

☒ **Yes** ☐ **No** Skin thickening

Describe location, extent, progression:

Both fingers and arms above elbows, both feet and lower legs

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

Laboratory features

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

Investigations

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

Clinical case presentation and progress (include disease duration):

☐ Yes ☒ No DLCO < 80% predicted

☐ Yes ☒ No FVC < 80% predicted

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

Appendix 4. Candidate criteria definitions

Item	Definition
Abnormal nailfold capillary pattern consistent with SSc	Enlarged capillaries and/or capillary loss with or without peri-capillary hemorrhages.
Anti-centromere antibody or centromere pattern on antinuclear antibody test	Positive according to local laboratory standards
Anti-topoisomerase I antibody	Positive according to local laboratory standards
Anti-RNA polymerase III antibody	Positive according to local laboratory standards. May not be available in all labs as part of extractable nuclear antigen (ENA)) testing
Calcinosis	Detected either clinically or by radiographically. Calcinosis is defined as palpable, dermal and/or subcutaneous or intramuscular deposits. It is usually located in digits or over large proximal joints or extensor surfaces of distal extremities.
Clinical evidence of acro-osteolysis	Shortening of the distal phalanx clinically or on radiograph consistent with acro-osteolysis (distal tuft resorption)
Dysphagia for solid foods	By history, as substernal discomfort on swallowing or sensation of food being held up or "stuck" in a retrosternal location.
Esophageal dilatation	Esophagus dilated by imaging (barium swallow, chest radiograph, or high resolution computed tomography (HRCT) of the chest).
Bilateral finger flexion contractures	Inability to extend fingers to neutral position not likely due to another condition such as arthritis deformities, Dupuytren's, diabetic cheiropathy, or fascial changes.
Finger tip ulcers or pitting scars	Ulcers or scars not thought to be due to trauma. Digital pitting scars are depressed areas at the tips as a result of ischemia, rather than trauma or exogenous causes.
Interstitial lung disease or pulmonary fibrosis	Pulmonary fibrosis on HRCT or chest radiograph, most pronounced in the basilar portions of the lungs, or presence of 'velcro' crackles on auscultation.
Persistent or recurrent gastro-esophageal reflux disease	By history, endoscopy or imaging.
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.
Pulmonary arterial hypertension	Pulmonary arterial hypertension diagnosed by right heart catheterization.
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.
Renal crisis	New onset of a systolic blood pressure (SBP) ≥ 140 mmHg and diastolic blood pressure (DBP) ≥ 90 mmHg; OR a rise in SBP ≥ 30 mmHg compared to usual and rise in DBP ≥ 20 mmHg compared to usual, AND at least 1 of these features:

Item	Definition
	1. Serum creatinine: increase of 50% above usual level; 2. Proteinuria: 2+ by dipstick confirmed by protein: creatine ratio > ULN; 3. Hematuria: 2+ by dipstick or > 10 RBCs/HPF (without menstruation); 4. Thrombocytopenia: < 100,000 /mm ³ ; 5. Hemolysis (fragmented RBC: by blood smear or increased reticulocyte count.
Scleroderma (skin thickening)	Skin thickening or hardening anywhere but not due to scarring after injury, trauma, etc.
Telangiectasias	Telangiectasias in a scleroderma like pattern are round and well demarcated and found on hands, lips, inside of the mouth, and/or large matt-like telangiectasias. Telangiectasias 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.
Tendon or bursal friction rubs	One or more friction rubs detectable at places such as shoulders, olecranon bursae, wrists (flexor or extensors), fingers (flexor or extensor), knees, ankles (Achilles, peroneal, posterior tibial, or anterior tibial tendons).

Figure 1.
Conceptual framework for SSc classification criteria development

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

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

Figure 2.

Example of how questions were posed to participants in the discrete choice experiment.

Figure 3.

Experts' rankings of the relative probability that each of the 20 cases has systemic sclerosis in first ranking exercise.

Note:

Cases are presented in order of the median rank (X-axis). The ranks (Y-axis) for each case are those assigned by each expert (A – G). Some points shown represent multiple coinciding data points.

Figure 4.

Experts' rankings of the relative probability that each of the 20 cases has systemic sclerosis in second ranking exercise.

Note:

Cases are presented in order of the median rank (X-axis). The ranks (Y-axis) for each case are those assigned by each expert (A – K). Some points shown represent multiple coinciding data points.

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Table 2

Relative weighting of reduce classification criteria.

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

'Skin thickening sparing the fingers' is an exclusion criterion. If present, the use of the SSc classification criteria should not proceed further.

'Skin thickening of the fingers and proximal to the metacarpophalangeal joints' is an absolute criterion. If present, the patient could be classified as SSc, and further use of the classification criteria is not required.