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Scleroderma Patient-centered Intervention Network (SPIN)

Additional File 2: SPIN Medical Data Form

- 1. Patient ID**
- 2. Date** (dd/mm/yyyy)
- 3. Sex** **Male** **Female**
- 4. Date of Birth** (dd/mm/yyyy)
- 5. Height** **cm** **Not available**
- 6. Weight** **kg** **Not available**
- 7. Raynauds** **Yes** **No** **Not available**

8. Date of onset of Raynaud's

(if exact date is not known, provide best approximate estimate)

(dd/mm/yyyy) **Not available**

9. Date of onset of first non-Raynaud's disease manifestation

(if exact date is not known, provide best approximate estimate)

(dd/mm/yyyy) **Not available**

10. Date of SSc diagnosis *(if exact date is not known, provide best approximate estimate)*

(dd/mm/yyyy) **Not available**

11. Disease Subset

Diffuse – skin sclerosis involving the limbs proximal to the elbows and knees and/or the chest and/or trunk, at any time

Limited – skin sclerosis confined to the limbs distal to the elbows and knees and/or face

Limited **Diffuse** **Sine** **Not available**

12. Autoantibodies

a. Antinuclear Antibody (ANA)

By Indirect immunofluorescence (IIF)

Positive **Negative** **Not done**

If ANA positive, titre > 1:160

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Yes **No**

b. Anti-centromere
By IIF or other antigen-based methods

Positive **Negative** **Not done**

c. Anti-topoisomerase I (also called Scl-70)

Positive **Negative** **Not done**

d. Anti-RNA polymerase III

Positive **Negative** **Not done**

13. Modified Rodnan Skin Score (MRSS, range 0-51)

Score **Not done**

14. Puffy fingers

Enlarged/swollen fingers, which may be pitting or non-pitting

Yes **No** **Not available**

15. Sclerodactyly

Symmetric thickening and non-pitting induration of the skin of the digits (at any time, now or in the past)

Yes **No** **Not available**

16. Sclerodermatous skin involvement proximal to the metacarpophalangeal joints (MCPs)

Yes **No** **Not available**

17. Digital pitting scars

Depressed areas at tips of digits or loss of digital pad tissue as a result of digital ischemia rather than trauma or exogenous causes (at any time, now or in the past)

Yes **No** **Not available**

18. Digital ulcers

Areas of de-epithelialization that can vary from focal infarcts to extensive gangrene of the fingers, and that are substituted by scars when healed, regardless of etiology (ischemia, calcinosis, trauma), at any time, now or in the past.

a. Digital pulp (volar), distal to distal interphalangeal joints (DIPs)

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Yes **No** **Not available**

b. Dorsal finger, distal to DIPs

Yes **No** **Not available**

c. Between the MCPs and DIPs, volar or dorsal

Yes **No** **Not available**

19. Telangiectasias

Visible macular dilatation of superficial cutaneous blood vessels that collapse upon pressure and fill slowly when pressure is released (do not include spider angiomas [point-like, dilated arterioles] or linear superficial dilated vessels that are found in sun-exposed areas or in areas of venous dependency, mostly the legs)

a. Any

Yes **No** **Not available**

b. On the face

Yes **No** **Not available**

20. Abnormal nailfold capillaries

Yes **No**

Specify method of detection:

- Naked eye**
- Dermatoscope**
- Ophthalmoscope**
- Widefield microscopy**
- Videocapillaroscopy**
- Other**

21. Abnormal skin pigmentation

Hyperpigmentation, often but not necessarily containing areas of punctuate or patchy hypopigmentation or depigmentation (“pepper and salt”), thought to be related to SSc

a. Any

Yes **No** **Not available**

b. On the face

Yes **No** **Not available**

22. Tendon friction rubs

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Palpable crepitus over flexor or extensor tendons, particularly common over the wrists, fingers, knees and ankles

- Currently, with or without past**
- In the past, but not currently**
- Never**
- Not available**

23. Joint contractures

Limitation of range of motion of a joint secondary to tightening around the joint

a. Small joints

DIP, PIPs (proximal interphalangeal joints), MCPs and/or wrists

- Yes** **No** **Not available**

a. Large joints

Elbows, knees, hips, ankles

- Yes** **No** **Not available**

24. Gastrointestinal tract

a. Esophageal

Dysphagia, heartburn and/or reflux, due to SSc (or requiring medications to alleviate these symptoms)

- Yes** **No** **Not available**

b. Stomach

Early satiety and/or vomiting, due to SSc (or requiring medications to alleviate these symptoms)

- Yes** **No** **Not available**

c. Intestinal

Diarrhea, bloating and/or constipation, due to SSc (or requiring medications to alleviate these symptoms)

- Yes** **No** **Not available**

25. Interstitial lung disease

Bilateral fine reticular, reticulonodular or honeycombing markings which are most pronounced in basilar portions of the lungs as detected by chest radiography or HRCT; should not be attributable to another primary lung disease

- Yes** **No** **Not available**

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26. Pulmonary arterial hypertension (PAH) by right heart catheterization (RHC)
MPAP (mean pulmonary artery pressure) \geq 25 mmHg and PCWP (pulmonary capillary wedge pressure)/LVEDP (left ventricular end diastolic pressure) \leq 15 mmHg

Yes **No** **Not done**

27. History of scleroderma renal crisis

Yes **No** **Not available**

28. Overlap syndrome

a. Systemic lupus erythematosus (*by ACR classification criteria*)

Yes **No** **Not available**

b. Rheumatoid arthritis (*by ACR/EULAR 2010 classification criteria*)

Yes **No** **Not available**

c. Sjogren's syndrome

(positive for at least two of three objective diagnostic tests: 1) anti-SS-A/B blood test. There are two scenarios: a) Positive serum levels of either the SSA and/or SSB antibody and/or b) positive serum levels of the rheumatoid factor antibody (RA) and elevated antinuclear antibody (ANA) titers; 2) ocular surface staining (measures the dissipation rate of a specialized dye that is applied to the tear film that bathes the surface of the eye; a score of three or more is considered to be positive); 3) salivary gland biopsy (one or more sites of inflammation per four millimeters squared area is considered positive)

Yes **No** **Not available**

d. Idiopathic inflammatory myositis

Either definite or probable polymyositis or dermatomyositis, according to the following criteria:

- 1. symmetric proximal muscle weakness muscle biopsy evidence of myositis*
- 2. elevation in serum skeletal muscle enzymes*
- 3. characteristic EMG pattern of myositis*
- 4. typical rash of dermatomyositis (heliotrope rash or Gottron's papules).*

All of criteria 1-4 or any 3 of criteria 1-4 are required for definite or probable polymyositis, respectively. Criteria 5 plus any 3 of criteria 1-4 or criteria 5 plus any 2 of criteria 1-4 are required for definite or probable dermatomyositis, respectively. The absence of other forms of myopathy, including inclusion body, metabolic, inherited or infectious forms, is also

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required to diagnose PM/DM.

Yes **No** **Not available**

e. Primary biliary cirrhosis (*diagnosed by the presence of: cholestatic indices, antimitochondrial antibodies, and liver histology diagnostic or compatible with PBC*)

Yes **No** **Not available**

f. Autoimmune thyroid disease (*with a diagnosis supported by clinical presentation and the presence of either thyroid peroxidase (TPO) or thyroglobulin (Tg) antibodies*)

Yes **No** **Not available**