UK Scleroderma Study Group (UKSSG)
Consensus Best Practice

Pathways of care for systemic sclerosis
developed by consensus within UKSSG multidisciplinary groups, with external comment and involvement of patients, support groups and non-medical professionals.

Investigation and treatment in primary, secondary or tertiary care.

• NOT – new guidelines – simply agreed practice across the UK SSc specialist centres
• UK specific – replicating success of the UK pulmonary hypertension network
• Complementary to evidence based treatment recommendations
Pathways of care for systemic sclerosis – a UKSSG initiative to establish a comprehensive management framework for scleroderma

- Establish “consensus best practice”
- Goal is non-expert specialists – secondary level care
  - Roadmap for management
  - Harmonise care
  - Identify a stakeholder in each hospital
- Implications for specialist commissioning
- Complementary to existing and planned recommendations
Management of critical digital ischaemia in SSc

1. Establish diagnosis and identify any treatable contributory cause

No contributory cause

2. Treat any contributory cause
   - Large (proximal) vessel disease
   - Vasculitis
   - Coagulopathy
   - Thromboembolism
   - Smoking

3. Admit for IV prostanoid and analgesia

   Ineffective

4. ± Antiplatelet therapy

   Effective

5. ± Consider statin

6. ± Antibiotic if any possibility of infection

7. Optimise oral vasodilator therapy (consider PDE5 inhibitor)

8. Consider digital sympathectomy

9. ± Surgical debridement if necrotic tissue

10. ± Short term anticoagulation

UKSSG Consensus Best Practice recommendations
(Herrick, Denton, Ong et al, 2012)
Management of digital ulceration (DU) in SSc

1. Establish diagnosis early

2. Treat any contributory cause e.g. infection, large vessel disease

3. Optimal wound care and analgesia

4. Optimise oral vasodilators or IV prostanoids

5. Consider surgical debridement in patients with necrotic tissue or underlying calcinosis

6. Antiplatelet and/or statin therapy

7. Repeat IV prostanoids or PDE5 inhibitor or ERA

8. Consider digital sympathectomy

UKSSG Consensus Best Practice recommendations (Herrick, Denton, Ong et al, 2012)
Management of Raynaud’s Phenomenon

1. Establish diagnosis and identify any underlying cause amenable to treatment

   - No underlying cause amenable to treatment

2. General/lifestyle measures:
   - Patient education
   - Avoid cold, keep warm
   - Stop smoking
   (Complementary therapies)

   - Treat underlying cause e.g. cryoglobulinaemia

3. Drug therapy: first line
   - CCB, ARB, SSRI alpha blockers, ACE inhibitors, topical nitrates

   - Ineffective oral therapies/refractory disease

4. Antiplatelet and/or statin therapy
   - Effective

5. Drug therapy: refractory
   - IV prostanoid

   - Ineffective

6. PDE5 inhibitor

   - Progression to digital ulceration and/or critical ischaemia

UKSSG Consensus Best Practice recommendations
(Herrick, Denton, Ong et al, 2012)