

# The Investigation, Management and Treatment of Interstitial Lung Disease (ILD) in Patients with Systemic Sclerosis (SSc)

## 1) Identifying patients with ILD

Patients presenting to Rheumatology or other speciality with **possible SSc**

*Specifically assess/look for:*

**History** - Breathlessness on exertion,

Cough – dry or productive with clear sputum

**Examination** - Are there bibasal inspiratory crackles?

Patients presenting to Respiratory or other speciality with **possible ILD**

*Specifically assess /look for:*

**History** - Raynaud's, skin thickening, digital ulcers, oesophageal reflux, joint pain and/or muscle weakness

**Examination** - telangiectasia, Raynaud's, calcinosis, skin thickening/puffy fingers, nailfold changes, proximal myopathy and/or arthropathy

If SSc ILD in differential diagnosis

## 2) Investigation of patients with potential ILD

### Tests

CXR – evidence ILD but may be normal

Full lung function including transfer factor – restrictive defect with reduced DLco

ECHO – to exclude PAH and/or diagnose coexisting PAH

HRCT chest scan

Other: O<sub>2</sub> sat room air, 6MWT

Suggestive of SSc: +ve ANA, anti-Scl 70 (anti-topoisomerase), anti-centromere antibody, raised CK

**Red flags – raised CRP without infection, desaturation on exercise**

## 3) Management of patients with ILD

### a. Monitoring

4-6 monthly lung function, annual ECHO & O<sub>2</sub> sats

Is there significant deterioration (e.g. VC > 10% decrease & DLco >15% decrease) from baseline?

No

Yes

Continue to monitor

Consider HRCT chest, referral to ILD MDT and treatment

### b. Who to treat

Progressive and/or extensive disease e.g. early with extensive disease on HRCT, progressive (with rapidly falling VC and DLco)

## 4) Treatment

- Immunosuppressive e.g. cyclophosphamide (IV), mycophenolate mofetil, corticosteroids (low dose oral or IV but beware risk of acute renal crisis).
- Consider co-prescribing proton pump inhibitor (especially if using steroids)
- Non-pharmacological measures