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

1) Identifying patients with ILD

Specifically assess/look for:

History - Breathlessness on exertion,
Cough – dry or productive with clear sputum
Examination - Are there bibasal inspiratory crackles?

Patients presenting to Rheumatology or other speciality with possible SSc

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: O₂ 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 & O₂ sats

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

No

Continue to monitor

Yes

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