UKSSG Best Practice Recommendations

Guidance on SSc PAH, supporting information.

Screening is worthwhile as 10-15% of SSc patients have PAH. The DETECT study – a cross-sectional screening study in SSc provides a solid evidence base for initial population screening[1]. After initial screening individuals with normal pulmonary pressures are unlikely to develop PAH in the next 5 years, while those with modestly abnormal pressure have a high likelihood of progression over three years[2]. We propose follow-up focusing on the dynamic elements of the evidence-based protocol relying on the natural variation and test-re-test accuracy to determine thresholds for re-investigation. Other protocols are also available based on combining echocardiography, lung function testing and symptomatic assessment. Referral for diagnosis is based on the ESC/ERS guidelines[3], with additional guidance on how to determine who is a very high risk based on experience. Referral to a nationally designated PH centre should not be delayed to complete investigations, particularly where PAH is likely..
Diagnosing PH and identifying a cause for PH is usually straightforward and safe, in experienced centres but requires cardiac catheterization. However, excluding left heart/lung disease related PH and pulmonary veno-occlusive disease may be challenging[4]. The table highlights findings on investigations that suggest PH if present, is less likely to be due to PAH.
Therapy and follow up should be as for other PAH patients. The evidence for anticoagulation is weaker than for idiopathic PAH[5]. Criteria for a satisfactory response require slight modification, because of co-morbidity. SSc patients may be breathless on exertion without PH and the mean 6MWD is lower in the 'healthy' SSc population[2]. The cardiac index associated with prognostic benefit on treatment is a little higher in SSc[6]. Cardiopulmonary exercise testing is less often useful in SSc patients, while PVOD is more common and needs to be actively sought.

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[3] Galie N, Hoeper MM, Humbert M, et al. Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Respir J 2009;34:1219–63.

[4] Hurdman J, Condliffe R, Elliott C et al. Aspire registry: Identifying the spectrum of pulmonary hypertension identified at a referral centre. Eur Respir J 2012; 39: 945 - 55

5 Olsson KM, Delcroix M, Ghofrani HA, et al. Anticoagulation and Survival in Pulmonary Arterial Hypertension: Results from the COMPERA Registry. Circulation 2013 e pub.

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sildenafil. Rheumatology 2010;49:490–500.