The Australian Scleroderma Interest Group and database: 10 years of screening to save lives

To the Editor: As 2017 marks the 10-year anniversary of the Australian Scleroderma Interest Group (ASIG), we want to raise awareness of the significant morbidity and mortality associated with systemic sclerosis (SSc), a chronic multisystem autoimmune disorder characterised by vasculopathy and fibrosis, with a particular focus on SSc-related pulmonary arterial hypertension (PAH).[1]

SSc-PAH is the leading cause of SSc-related death with a survival, once symptomatic, of only 2–3 years without treatment.[2] Survival may be improved by annual screening with algorithms incorporating transthoracic echocardiogram and pulmonary function tests — even after adjustment for lead-time bias — and with early implementation of specific PAH therapies available through the Pharmaceutical Benefits Scheme (PBS).[3] Thus, annual screening is recommended[4] to identify patients who should undergo right heart catheterisation to confirm the diagnosis.

Despite the documented benefits of PAH screening, physician adherence in Australia is suboptimal, with over 40% not adhering to annual screening.[5] Therefore, the Australian Scleroderma Cohort Study (ASCS) was established in 2007 as a web-based screening platform for the cardiorespiratory manifestations of SSc, particularly PAH.

The ASCS is a nationwide project where patients with SSc are recruited from 13 participating centres across Australia. Physicians who are not part of ASIG and care for patients with SSc are invited to refer patients for the screening service. Over the past 9 years, 1636 patients with SSc have been recruited and, as a result of annual screening, 194 patients (11.9%) have been diagnosed with PAH. All patients have received a specific PAH therapy.

In Australia, the PBS only subsidises monotherapy (treatment with one PAH therapy at any one time) in patients with PAH who are in the World Health Organization (WHO) functional classes III or IV. International data have shown that patients treated earlier, such as those in the WHO functional class II, before damage to the right ventricle has occurred, have an improved survival rate over those treated later and those in a worse WHO functional class. Furthermore, patients treated with combination therapy (treatment with two specific PAH agents at the same time) have a survival benefit compared with those treated with monotherapy only.[6] In the coming years, ASIG will continue to campaign to optimise screening and the treatment of SSc-PAH through efforts directed at closing the evidence–practice and evidence–policy gap in Australia, with the goal of improving outcomes for patients who have this incapacitating disease.

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References are available online at www.mja.com.au.

1 Marie I, Jouen F, Hellot MF, Levesque H. Anticardiolipin and anti-beta2 glycoprotein I antibodies and lupus-like anticoagulant: prevalence and significance in systemic sclerosis. *Br J Dermatol* 2008; 158: 141-144.

2 Tyndall AJ, Bannert B, Vonk M, et al. Causes and risk factors for death in systemic sclerosis: a study from the EULAR Scleroderma Trials and Research (EUSTAR) database. *Ann Rheum Dis* 2010; 69: 1809-1815.

3 Lau EM, Manes A, Celermajer DS, Galiè N. Early detection of pulmonary vascular disease in pulmonary arterial hypertension: time to move forward. *Eur Heart J* 2011; 32: 2489-2498.

4 McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc.; and the Pulmonary Hypertension Association. *J Am Coll Cardiol* 2009; 53: 1573-1619.

5 Mangat P, Conron M, Gabbay E, Proudman SM. Scleroderma lung disease, variation in screening, diagnosis and treatment practices between rheumatologists and respiratory physicians. *Intern Med J* 2010; 40: 494-502.

6 Galiè N, Barberà JA, Frost AE, et al. Initial use of ambrisentan plus tadalafil in pulmonary arterial hypertension. *N Engl J Med* 2015; 373: 834-844. ■