

Systemic Sclerosis in Australia: using the Australian Scleroderma Screening Program (ASSP) database to assist in assessment of cardiopulmonary complications.



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BACKGROUND

Systemic sclerosis (SSc), both diffuse (dSSc) and limited (lSSc), is a major risk factor for the development of pulmonary arterial hypertension (PAH). Often PAH is far advanced by the time it is detected by right heart catheterisation (RHC), severely impacting on quality of life and survival. Therapeutic options available for PAH have increased significantly over the last few years. Attempts to identify PAH early in these patients improve haemodynamics, quality of life, function and possibly survival. ILD remains a major cause of morbidity in SSc. Early detection and treatment may improve long term survival.

OBJECTIVE

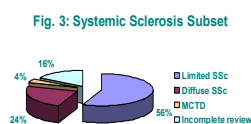
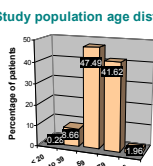
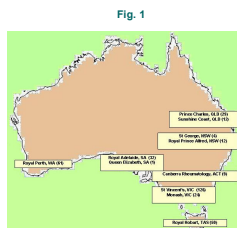
The ASSP was established to provide high quality evidenced based screening for cardiopulmonary complications of (SSc) and mixed connective tissue disease (MCTD) and to ensure patients thus identified receive appropriate and timely treatment. The secondary aim was to create a detailed electronic database to assist in the application of the screening algorithm and to promote Australian research in SSc.

METHOD

Recruitment commenced in January 2007. The screening program includes patient questionnaire, history, clinical examination, laboratory studies, pulmonary function tests (PFT), ECG, echocardiogram (ECHO) and 6 minute walk test. Data are entered into the database online. The program identifies and stratifies risk for either PAH or interstitial lung disease ILD, alerts the responsible clinician and recommends further assessment according to the algorithm.

RESULTS

As of April 2008, a total of 360 patients had been recruited from 11 participating centres in Australia (Fig.1). There were some incomplete data from patients still being reviewed. Among all scleroderma patients, 86.1% were female; with the female to male ratio 6.2:1. The age of the patients when surveyed ranged from 19.8 to 84.3 years, with a mean age 57.4 years (Fig.2). A total of 23.9% patients had dSSc, 56.1% patients had lSSc and 4.2% had MCTD (Fig.3).



Disease manifestations in various subsets of patients are shown in Table 1. The age of onset of SSc was similar in all subsets. The mean Rodnan skin score was higher (21.3) in dSSc than in lSSc (8.9) and MCTD (7.1). Musculoskeletal manifestations (joint contractures, myositis) were almost twice as common in dSSc as in lSSc. The frequency of reflux oesophagitis was similar in all SSc subsets, whereas oesophageal dysmotility was more common in dSSc (17%) compared to lSSc (10%) and MCTD (8%).

Table 1: Disease Manifestation among SSc Subset (%)

	Limited SSc	Diffuse SSc	MCTD
Gender Ratio F:M	7.4:1	2.9:1	14:01
Age of onset (years)	46	44	47
Mean Rodnan Score	8.9	21.3	7.1
Digital Ulcers	17	37	13
Joint contractures	22	74	27
Myositis	4	11	20
Reflux Oesophagitis	55	54	53
Oesophageal Dysmotility	10	17	8
Pulmonary Hypertension	15	7	20
Pulmonary Fibrosis	20	35	13
Renal crisis	1	5	0
Malignancy	13	8	20

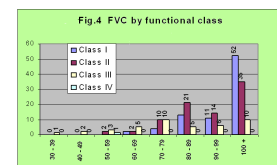
sPAP(mm Hg)	No of patients	No of patients with DLCO≤50%, FVC>85%	No. of patients for RHC	No. of patients diagnosed PAH on RHC
Unrecordable	24	2	3	0
sPAP < 40	157	4	13 (2 awaiting)	7
40 ≤ sPAP ≤ 50	29	3	8 (4 awaiting)	5
sPAP > 50	16	2	10 (6 awaiting)	9

Using the algorithm 16 patients were identified to be at high risk of PAH (sPAP ≥ 50 mmHg) (Table 2). All patients proceeded to RHC. Nine of the patients were diagnosed with PAH on RHC, 6 patients are awaiting RHC.

Twenty nine patients were at moderate of PAH (40mmHg ≤ sPAP ≤ 50mmHg), of which 3 patients also had disproportionately low DLCO (DLCO ≤ 50, FVC > 85%). 12 patients have been referred for a RHC. Of the 8 patients in whom RHC has been completed, 5 were found to have PAH. Six patients with ILD and 9 patients with Class I and II NYHA were not referred for a RHC in this group. A total of 157 patients were at low risk of PAH (sPAP ≤ 40mmHg). Indication for catheterization in this group included unexplained dyspnoea in 15 out of 157 (NYHA Class III). Seven of the 13 patients who have completed RHC were diagnosed with PAH. Twenty four patients had unrecordable sPAP on echo. Three patients with NYHA Class III proceeded to RHC. None of these patients had PAH. In total, 21 patients have been identified with PAH.

Table 3: Severity of ILD in patients, n=54

Mid	22
Moderate	24
Severe	8



Of 252 patients with recorded FVC, 69 (27.4%) were identified to have significant risk of ILD as defined by FVC < 85%. Table 3 shows the degree of severity of ILD recorded in 54 HRCT.

CONCLUSION

As a result of investigations recommended by the algorithm, 9.5% patients were identified with PAH and significant lung disease in 27.4% patients. These preliminary data demonstrate the value of the ASSP program, aided by the decision support facility of an on-line electronic database, in identifying SSc patients at high risk of cardiopulmonary complications. Furthermore the program is establishing a comprehensive and detailed data set which will enable further clinical research into this rare but severely disabling condition in Australian patients.

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