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BRIEF COMMUNICATION

Predictive accuracy of the ASIG algorithm in a prospective systemic sclerosis cohort undergoing annual screening for pulmonary arterial hypertension

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Abstract

The Australian Scleroderma Interest Group (ASIG) algorithm for screening pulmonary arterial hypertension (PAH) in systemic sclerosis (SSc) requires only respiratory function tests and serum N-terminal pro-brain natriuretic peptide as first-tier tests, and is recommended in international guidelines. In this communication, we present the findings of the application of the ASIG screening algorithm to a Singaporean cohort undergoing prospective annual screening for PAH, which shows a high negative predictive value. The ASIG algorithm may offer an alternative to more complex and costly SSc-PAH screening algorithms.

Systemic sclerosis (SSc) is a multisystem autoimmune disease with a high burden of mortality resulting from cardiopulmonary complications.[1] Multinational registry data have shown that the predominant causes of SSc-related deaths remain pulmonary arterial hypertension (PAH) and interstitial lung disease (ILD), and that PAH occurs in SSc with a prevalence of 8.6–15.3%.[2]

An early diagnosis of PAH, made through systematic screening, is associated with improved long-term outcomes and enables early implementation of effective therapy.[3] Annual screening for all patients with SSc and SSc-spectrum disorders is the recommended standard of care following the sixth World Symposium on Pulmonary Hypertension in 2018.[4]

Recommended modalities for screening include annual transthoracic echocardiography (TTE), or the use of composite screening algorithms, such as the DETECT algorithm and the Australian Scleroderma Interest Group (ASIG) algorithm.[4,5] The DETECT algorithm relies on nomograms to score clinical, biochemical, electrocardiography and spirometry parameters in step one, which, in turn, determines those who proceed to TTE in step 2. TTE parameters are then used to determine referral for right heart catheterisation (RHC), the gold standard diagnostic test for PAH (Fig. 1).[6]

The ASIG algorithm evaluates only two of the variables from step one of the DETECT algorithm: serum N-terminal

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pro-brain type natriuretic peptide (NT-proBNP) and spirometry parameters. If either parameter is 'positive' according to the specified cutoffs, it is recommended that these patients proceed to further investigation of possible pulmonary hypertension (PH) with TTE, pulmonary radiography and/or RHC if indicated (Fig. 1).[7,8]

Whilst the DETECT algorithm has been widely validated, the ASIG algorithm, derived in an Australian SSc cohort, has only one external retrospective evaluation of 117 consecutive patients with SSc in a cohort attending Rennes University Hospital, France, of whom only 16 underwent RHC.[9,10] The ASIG and DETECT algorithms were positive in all patients with diagnosed PAH, meaning there were no missed cases of PAH. Two patients had false-positive ASIG and DETECT algorithm results and a normal RHC result.[10]

Given the paucity of external validations of the ASIG algorithm, we evaluated the sensitivity of the ASIG algorithm for the detection of PAH in a Singaporean SSc cohort undergoing prospective annual screening. We were unable to evaluate the specificity of the ASIG algorithm as not all patients in this cohort underwent the gold-standard diagnostic test, RHC, which was requested only for those in whom there was clinical suspicion of possible PAH.

We analysed data from patients enrolled in the Systemic Sclerosis Cohort Singapore (SCORE) who had undergone RHC resulting in a diagnosis of PAH and had sufficient data to apply the ASIG algorithm retrospectively. From 2008, patients with SSc meeting the American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) SSc classification criteria or Very Early Diagnosis of Systemic Sclerosis (VEDOSS) criteria in this cohort have undergone annual screening with TTE, respiratory function test and serum NT-proBNP.[11,12] Data are recorded annually according to a protocol that has been approved by local institutional ethics review committees.[13]

Referral for RHC in the SCORE cohort occurs when there is unexplained dyspnoea together with findings from TTE showing a systolic pulmonary artery pressure ≥40 mmHg and/or a ratio of forced vital capacity (FVC) % predicted to diffusion capacity for carbon monoxide (DLCO) % predicted of >1.4 (FVC/DLCO ratio). In 2018, the protocol was revised such that referral for RHC occurs when the European Society of Cardiology 2015 guidelines for the diagnosis and management of PH probability for PH is intermediate or high by TTE parameters; or dyspnoea is present with DLCO <60% and/or FVC/DLCO ratio >1.6; or in asymptomatic individuals with DLCO <60% and/or FVC/DLCO ratio >1.6 and NT-proBNP >2 times the upper limit of normal.[14]

At the time of study census (4 August 2022), there were 472 patients enrolled in the SCORE cohort and 106 (22.5%) had undergone the first RHC. At this time in the SCORE cohort, 52 (11.0%) patients had PAH confirmed on RHC, defined as mean pulmonary artery pressure >20 mmHg, pulmonary artery wedge pressure ≤15 and pulmonary vascular resistance ≥3 WU.[15]

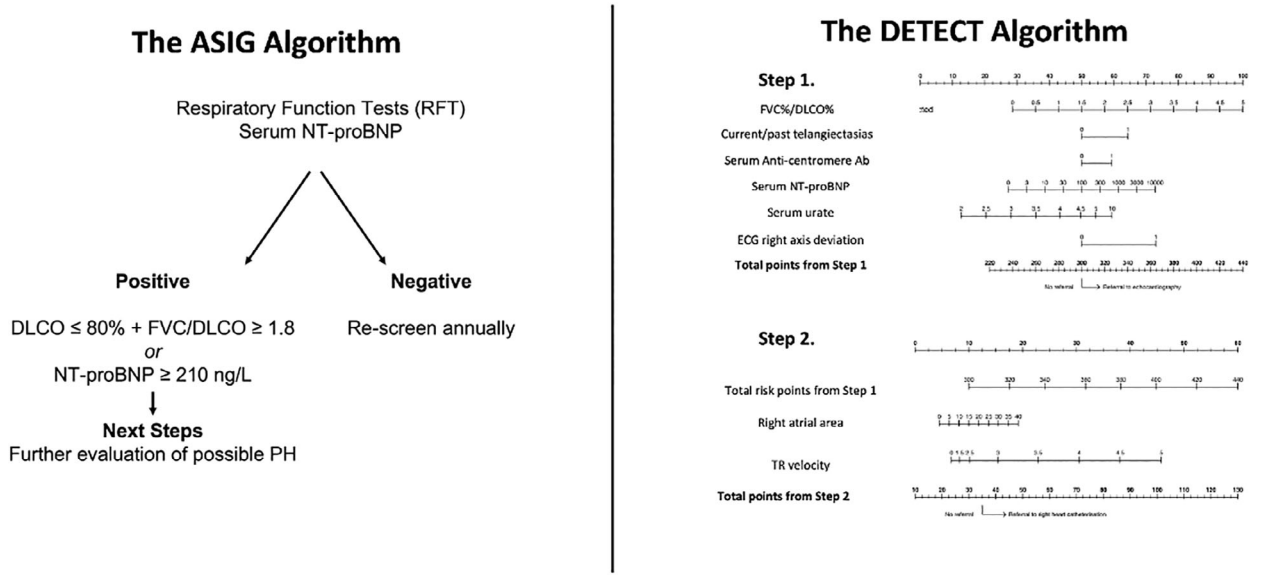

Figure 1 The Australian Scleroderma Interest Group (ASIG) and DETECT algorithms. DLCO, diffusion capacity for carbon monoxide; ECG, electrocardiography; FVC, forced vital capacity; NT-proBNP, N-terminal pro-brain type natriuretic peptide; PH, pulmonary hypertension; TR, tricuspid regurgitant. The ASIG algorithm adapted from: reference.[8] The DETECT algorithm adapted from: reference.[6]

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We present here 39 patients with sufficient data to apply the ASIG algorithm within 12 months of the first RHC. Of these, 27 (69.2%) were diagnosed with PAH (Table 1).

The cohort was predominantly female (36 (92.3%)), with a median age at the time of screening of 62.5 years (interquartile range = 51.3–72.5 years). Eleven patients (37.9%) had the diffuse cutaneous disease subtype and 18 (62.1%) had the limited cutaneous disease subtype (Table 1). ILD by high-resolution computed tomography (HRCT) chest was present in 32 (82.1%) patients. Autoantibody profile and SSc disease duration are reported in Table 1.

There were 35 (89.7%) patients with a positive ASIG screen result who underwent RHC, and 25 (92.6%) were diagnosed with PAH. There were two patients with a negative ASIG screen who were diagnosed with PAH. The PAH characteristics at the time of diagnosis in this cohort are shown in Table 1. A comparison of those with and without PAH is shown in Supplementary Table S1.

One case with a false-negative ASIG screen result had a serum NT-proBNP level of 192 ng/L, DLCO of 51.0% predicted and FVC/DLCO ratio of 1.6. By TTE parameters, the tricuspid regurgitant (TR) jet velocity was not detectable. When TTE was performed 2 months prior to RHC, the right ventricular systolic pressure (RVSP) was 36 mmHg.

The second case with a false-negative ASIG screen result underwent screening in November 2018 and had a serum NT-proBNP level of 80.0 ng/L, DLCO of 53.0% predicted and FVC/DLCO ratio of 1.7. At this time, the TTE demonstrated an RVSP of 34 mmHg, TR jet velocity of 2.7 m/s and right atrial area of 11 cm^2. This patient was diagnosed with PAH in October 2019 (less than 12 months after a negative screen result), 1 month prior to their scheduled annual screening review. A visit performed 1 month after PAH diagnosis demonstrated a serum NT-proBNP level of 76.8 ng/L, DLCO of 49.0% and FVC/DLCO ratio of 1.9.

We cannot determine the true performance characteristics of the algorithm, such as specificity in this work, as not all patients in the cohort undergoing screening had an RHC. However, only two missed cases of PAH indicate that the ASIG algorithm is likely to have a high negative predictive value. This is consistent with the recent French evaluation and with the performance of the ASIG algorithm in the derivation cohort (ASIG algorithm sensitivity 100%, specificity 54.5%).[10,16]

Among those who did have an RHC within 12 months of a screening visit in this cohort ($n = 39$), the sensitivity of the ASIG algorithm to identify PAH was 92.59% (95% confidence interval (CI) = 75.71–99.09), specificity 16.67% (95% CI = 2.09–48.41%), positive predictive value 71.4% (95% CI = 53.70–85.36) and negative predictive value 50.0% (95% CI = 6.76–93.24).

The strength of this analysis is that it has provided insights into the predictive accuracy of the ASIG algorithm in a diverse population, phenotypically different from the Australian cohort in which it was derived, with more patients with diffuse disease (37.9%), anti-topoisomerase antibody (anti-Scl-70) (39.5%) and anti-U1 ribonucleoprotein antibody (anti-U1RNP) (29.7%).[2] The SCORE cohort also included four patients with positive anti-double-stranded deoxyribonucleic acid (anti-dsDNA) antibodies, which could represent an overlap with systemic lupus erythematosus (SLE), although clinical data concerning possible SLE were not available for the purposes of this analysis.

The prevalence of PAH in this cohort (11.0%) is consistent with international SSc cohorts, and highlights the critical role for screening this at-risk population in order to identify and treat PAH before symptoms develop.[17,18] Consistent with previous reports, there was a high frequency of ILD (82.1%) with a moderately reduced median FVC% (63.50% (51.0–76.0)) in the SCORE cohort.[2] The reduced median FVC% automatically makes it more difficult to reach a high FVC/DLCO ratio, one of the components in both the DETECT and ASIG algorithms. In this cohort, the majority of positive cases were therefore on the basis of a positive NT-proBNP component.

As we have defined PAH based on haemodynamic criteria, some of these cases may represent group 3 PH because of ILD or a combination of group 1 PAH and group 3 PH because of ILD. The purpose of the ASIG algorithm is to detect those likely to have PH. Deciphering the aetiology of PH and determining the most suitable approach to treatment is informed by ancillary investigations (in addition to right heart catheterisation) performed at the physician's discretion. This highlights the importance of multidisciplinary case review of patients with SSc presenting with PH to elucidate the predominant contributing pathology to elevated pulmonary pressures and to determine appropriate therapeutic strategy.[5] Overall, our analyses indicate that the ASIG algorithm may have limitations when applied to screening for PAH in those with SSc-ILD.

To the authors' knowledge, this is the largest external evaluation of the ASIG algorithm in which patients had undergone confirmatory RHC. We were not able to compare this with the performance of the DETECT algorithm as serum uric acid and electrocardiograms, which are components of the DETECT algorithm, are not routinely performed in this cohort. However, as demonstrated in this study, variables that are required to apply the ASIG algorithm tend to be readily available.

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Table 1 Characteristics of patients with SSc in the SCORE cohort who underwent RHC and those in whom PAH was diagnosed

Variable	SCORE cohort who underwent RHC ($n = 39$) *n* (%) Median (IQR)	SCORE patients with PAH on RHC ($n = 27$) *n* (%) Median (IQR)
Age at most recent review (years)	62.5 (51.7–72.5)	64.5 (55.3–73.4)
SSc disease duration from first non-RP SSc manifestation (years to census)	5.93 (2.5–9.3)	5.1 (2.1–9.3)
Sex		
Male	3 (7.7%)	2 (7.4%)
Female	36 (92.3%)	25 (92.6%)
Disease subclassification		
Diffuse SSc	11 (37.9%)	8 (40.0%)
Limited SSc	18 (62.1%)	12 (60.0%)
Disease subclassification		
DcSSc	11 (28.2%)	8 (29.6%)
LcSSc (proximal to MCPJ/MTPJ and distal to elbow/knee)	17 (43.6%)	12 (44.4%)
MCTD	3 (7.7%)	2 (7.4%)
SSc overlap (DcSSc/RA)	1 (2.6%)	-
SSc overlap (LcSSc/Sjogren syndrome)	2 (5.1%)	1 (3.7%)
SSc overlap (SSc/RA)	1 (2.6%)	1 (3.7%)
SSc overlap/MCTD	2 (5.1%)	1 (3.7%)
Sine scleroderma	2 (5.1%)	2 (7.4%)
ASIG screen		
Positive	35 (89.7%)	25 (92.6%)
Negative	4 (10.3%)	2 (7.4%)
NT-proBNP ≥210 ng/L	35 (89.7%)	25 (92.6%)
FVC/DLCO ≥1.8 and DLCO <70%	1 (2.6%)	1 (3.7%)
PAH	27 (69.2%)	27 (100%)
Autoantibodies		
ANA nucleolar	9 (25.7%)	6 (25.0%)
ACA	5 (15.2%)	3 (14.3%)
ENA Scl-70	15 (39.5%)	9 (34.6%)
ENA Sm	5 (13.5%)	3 (12.0%)
ENA Ro	15 (39.5%)	12 (46.2%)
ENA U1RNP	11 (29.7%)	8 (32.0%)
Anti-dsDNA	4 (12.1%)	4 (18.2%)
Rheumatoid factor	10 (50.0%)	9 (60.0%)
Ever had ILD on HRCT	32 (82.1%)	24 (88.9%)
Respiratory function tests		
FVC%	63.5 (51.0–76.0)	62.0 (44.0–76.0)
DLCO%	54.5 (50.0–64.0)	51.0 (51.0–58.0)
NYHA functional class		
Class I	13 (33.3%)	8 (29.6%)
Class II	12 (30.8%)	6 (22.2%)
Class III	12 (30.8%)	11 (40.7%)
Class IV	2 (5.1%)	2 (7.4%)
6-min walk distance (m)	296.0 (285.2–411.0)	292.0 (190.0–383.00
NT-proBNP (ng/L)	420.0 (245.0–941.0)	405.0 (252.0–923.0)
Echocardiographic (TTE) parameters		
Right atrial area (cm^2)	13.1 (11.4–16.1)	13.1 (11.0–15.3)
TR velocity (m/s)	2.97 (2.5–3.1)	3.1 (2.5–3.1)
Systolic pulmonary pressure (mmHg)	46.0 (38.0–60.0)	52.0 (42.0–63.0)
Pericardial effusion	16 (41.0%)	12 (44.4%)
RHC parameters		
RAP (mmHg)	7.0 (3.0–10.0)	7.0 (3.0–13.0)
mPAP (mmHg)	27.0 (24.0–34.0)	29.0 (25.0–35.0)

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Table 1 *Continued*

Variable	SCORE cohort who underwent RHC ($n = 39$) *n* (%) Median (IQR)	SCORE patients with PAH on RHC ($n = 27$) *n* (%) Median (IQR)
PAWP (mmHg)	10.0 (7.0–13.0)	9.0 (7.0–12.0)
Cardiac output (L/min)	3.9 (3.5–4.8)	3.9 (3.5–4.4)
Cardiac index (L/min m^2)	-	-
PVR (WU)	4.2 (2.6–6.4)	5.9 (4.1–7.0)

The table contains key patient characteristics. For categorical variables, the number (percentage) is presented, and for nonnormally distributed variables, the median (interquartile range (IQR)) is presented.

ACA, anticentromere antibody; ANA, anti-nuclear antibody; anti-dsDNA, anti-double-stranded deoxyribonucleic acid; ASIG, Australian Scleroderma Interest Group; DcSSc, diffuse systemic sclerosis; DLCO, diffusion capacity for carbon monoxide; ENA, extractable nuclear antigen; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; LcSSc, limited systemic sclerosis; MCPJ, metacarpophalangeal joint; MCTD, mixed connective tissue disease; mPAP, mean pulmonary artery pressure; MTPJ, metatarsophalangeal joint; non-RP; non-Raynaud phenomenon; NT-proBNP, N-terminal pro-brain type natriuretic peptide; NYHA, New York Heart Association; PAH, pulmonary arterial hypertension; PAWP, pulmonary arterial wedge pressure; PVR, pulmonary vascular resistance; RA, rheumatoid arthritis; RAP, right atrial pressure; RHC, right heart catheterisation; RNP, anti-U1 ribonucleoprotein; Ro, anti-Ro52 antibody; Scl-70, topisomerase; SCORE, Systemic Sclerosis Cohort Singapore; Sm, anti-Smith antibody; SSc, systemic sclerosis (scleroderma); TTE, transthoracic echocardiography; TR, tricuspid regurgitant; U1RNP, U1 ribonucleoprotein antibody.

The performance metrics in this analysis may not truly be reflective of the performance of the algorithm as all patients in this cohort had been referred for RHC and therefore had a high pre-test probability of having PAH.

Whilst there is a need for further external validation of the ASIG algorithm, this analysis provides reassurance that the ASIG algorithm performs well to identify SSc-PAH outside of an Australian SSc cohort.

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Supporting Information

Additional supporting information may be found in the online version of this article at the publisher's web-site:

Supplementary Table S1: Characteristics of patients who underwent RHC and did not have PAH compared to those with PAH.

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