# Attitudes and barriers to pulmonary arterial hypertension screening in patients with systemic sclerosis: a survey of UK-based rheumatology clinicians

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**Abstract** (n=249/250)

**Objectives.** To explore rheumatologists' current clinical screening practices of pulmonary arterial hypertension (PAH) in patients with systemic sclerosis (SSc) in the UK. Also, to identify barriers to screening and consider potential solutions.

**Methods.** A survey of 31 questions was developed and included six sections: clinician demographics, the importance of screening, screening practices, barriers to screening, treatment, and patient education. The survey was disseminated among rheumatologists working in the UK.

**Results.** Fourty-four rheumatologists working in the UK participated to the study, and the majority completed all the questions. Around one-third (37%) worked in specialised SSc unit (university or general hospital (54.5% and 45.4%, respectively). The majority recognised that SSc-PAH is a major cause of morbidity and mortality. Over half (60%) reported using the DETECT algorithm to screen for SSc-PAH, although other algorithms were also sometimes used. All of the respondents utilised transthoracic echocardiogram, and almost all (95%) performed pulmonary function tests for screening purposes. Various challenges and barriers were identified relating to SSc-PAH screening, with the difficulty in interpreting results from other hospitals and extended wait times for diagnostic tests being the most reported (76% and 74%, respectively). Most respondents agreed that access to key investigations (87%), ongoing clinician education (82%), multidisciplinary meetings (79.5%), and a better understanding of proposed screening algorithms (79.5%), could be potential solutions.

**Conclusion.** Screening SSc patients for PAH is crucial to improve survival, but variable practices exist between UK rheumatologists. Solutions include educating healthcare providers on guidelines, sharing information between centres, and integrating care services.

**Key words:** Systemic Sclerosis; Scleroderma; Pulmonary Arterial Hypertension; Rheumatologists; Screening; United Kingdom.

**Key messages:**

- Rheumatologists recognise the importance of PAH in patients with systemic sclerosis and screening improves outcomes.
- There are important differences between UK rheumatologists in systemic sclerosis-PAH screening practices.
- Educational programs, easier sharing of information between centers, and integrated care services, are potential solutions.

## Introduction

Systemic sclerosis (SSc) is a rare autoimmune rheumatological disease that affects an estimated 235.5 adults per million in the UK (1). Vascular disease is a cardinal feature in the complex aetiopathogenesis of SSc (2–5) and is associated with significant morbidity and morbidity, in particular, from pulmonary hypertension (PH) (2,3,5).

PH in patients with SSc can manifest as any one of or a combination of the five World Health Organization PH groups. Pulmonary arterial hypertension (PAH) (group 1) most commonly occurs in patients with SSc and the consequence of isolated PH affecting small pulmonary arteries or pulmonary veno-occlusive disease. Group 2 or 3 may also occur, which reflects chronic left-heart disease, including PH secondary to myocardial fibrosis/left ventricular dysfunction, and PH related to interstitial lung disease (ILD), respectively (6–8).

The estimated prevalence of PAH in patients with SSc is around 10% (7,9,10) and is a leading cause of death in SSc (2,5). However, SSc-PAH diagnosis may be delayed as patients are often asymptomatic despite significant disease; moreover, symptoms may be attributed to other conditions either directly- (e.g., ILD) or unrelated-related (e.g., physical conditioning or left heart disease) to SSc (11).

Targeted and proactive screening for PAH significantly improves early survival from SSc-PAH (10,12–17); however, longer-term survival currently remains poor. Fortunately, an early diagnosis of SSc-PAH can be actively achieved, enabling specialist referral, facilitating the introduction of approved PAH-targeted (including combination) therapies (8,11,18–24).

The current European Society of Cardiology/European Respiratory Society (ESC/ERS) guidelines for the diagnosis and treatment of PH, recommend annual *multimodal* screening for

PAH in patients with SSc, with or without symptoms, including transthoracic echocardiogram (TTE) followed by the evaluation of biomarkers and pulmonary function testing (PFT). Furthermore, the guidelines also recommend stratifying patients with SSc into those at the highest and lowest risk of developing PAH (6). Furthermore, there are combined models that have been proposed to increase the predictive effectiveness of the diagnosis and early identification of SSc-PAH (25–27) , such as the DETECT (16,28) and Australian Scleroderma Interest Group (ASIG) algorithms (29). Furthermore, given the significant international variation observed in the approaches adopted to screening, and barriers to facilitating timely screening in routine clinical practice (17,30,31), the absence of dedicated UK-based guidance is of practical concern. In addition, the current British Society of Rheumatology (BSR) guidelines for the treatment of SSc (currently under revision) (32,33), makes no specific recommendations concerning either the frequency or specific investigations for SSc-PAH screening.

Against this background, the aims of our study were to explore clinical screening practices of PAH in patients with SSc of UK-based rheumatologists, to identify barriers to screening for SSc-PAH, and to consider potential solutions.

## Materials and methods

### Study design

The questions were developed with expert input from a Steering Committee of 10 SSc- PAH specialists: clinicians with an interest in SSc (YA, MHB, FDG, MH, DK, JDP), respiratory experts with a specialist interest in pulmonary vascular disease (GC, DK, SR), and a highly specialised cardiac physiologist with expertise in adult echocardiography (IA). Relevant national and international guidelines were reviewed, including (but not limited to) the BSR guideline/European League Against Rheumatism recommendations for SSc, ESC/ERS guidelines for the diagnosis and treatment of PH, and the DETECT and ASIG algorithms (6,16,29,32,33).

### Survey questions

The survey consisted of 31 questions (see **supplementary data**, available at *Rheumatology* Online), which encompassed 6 sections: clinician demographics, importance of screening, screening practices, barriers to screening, treatment, and patient education.

### Survey distribution and responses

The survey was hosted in SurveyMonkey® and disseminated in June 2023 using a link or QR code. The survey was shared with rheumatologists (consultants and trainees) working in the UK via the local networks of the Steering Committee, as well as a UK rheumatologist mailing list developed through GlobalData Plc. The survey was also disseminated widely via social media (i.e., Twitter ®). The survey remained open between 1st June to 3rd July 2023. The survey was voluntary. The responses were completely anonymous as no personal identifying information was collected at any point, and consent was implied by participating in the survey. The participants could stop the survey at any point.

### Statistical analysis

Data are provided as descriptive statistics. The denominator is provided where this was less than the total number of rheumatologists who participated to the survey. Data were analysed using Microsoft Excel®.

## Results

### Clinician demographics

The survey was completed by 44 rheumatologists based in the UK. The clinician demographics are presented in Table 1. Over half of respondents were female (56.8%). Three quarters of respondents were aged between 31 and 50 years (41 to 50 years: 41%, 31 to 40 years: 34.1%). Two-thirds of respondents (65.9%) had ≥5 years of experience in Rheumatology. Most respondents clinical practice was broad in the North West (31.8%), South West (25%), or Yorkshire (13.6%) of England; however, there was broad geographical representation. Around half either worked in a university (54.6%) or general hospital (45.5%), and few in private practice (6.8%). Just over a third (38.6%) worked in a specialised SSc unit. There was wide

variation in the number of patients with SSc under respondents care (e.g., 50% had <25 patients, whereas 25% had >100 patients).

**The burden of PAH and importance of screening in SSc**

Over two-thirds (68.2%) of respondents believed that PAH occurs in ≤10% of *their* patients with SSc, and a third (31.8%) between 11-25%. However, they indicated that they considered that PAH was *overall* more common in in SSc, with around two-thirds (63.6%) believing that PAH affects 11–25% of *all* patients with SSc. Most respondents (84%) reported being familiar with the different types of PH (7% unfamiliar, 9% unsure). The majority (88.6%) of patients under the respondents, with SSc-PAH were under the care of a specialist PH centre.

Respondents were asked their opinion on the importance of PAH as a major cause of morbidity and mortality in patients with SSc on a categorical scale (strongly disagree, disagree, neutral, agree, strongly agree) (Figure 1). The majority (91%) believed that SSc-PAH was a major cause of morbidity (43.2% agree, 47.8% strongly agree) and mortality (41% agree, 50% strongly agree). Similarly, the vast majority indicated that they considered that PAH screening in SSc patient would help to achieve earlier diagnosis (97.7% [47.7% agree, 50% strongly agree]), earlier treatment intervention (97.8% [36.4% agree, 61.4% strongly agree]), reduce morbidity rate (95.5% [54.5% agree, 41% strongly agree]), and most (88.7% [52.3% agree, 36.4% strongly agree]) considered would reduce mortality rate (Figure 1).

**Clinical presentation and risk factors for PAH**

All the respondents associated shortness of breath, the vast majority fatigue (95%), leg/ankle swelling (87.5%), and syncope (67.5%), and around half, chest pain (57.5%) and palpitations (52.5%) with PAH. Furthermore, half (52.5%) considered that PAH can be asymptomatic.

Respondents (n=40) were questioned about factors which would lead them to consider a patient with SSc to be at high risk of PAH (Table 2). The most strongly considered factors were the forced vital capacity (FVC)/diffusing capacity for carbon monoxide (DLCO) (87.5%), B-type natriuretic peptide (NT-pro-BNP) (80%), and limited cutaneous SSc (77.5%). Furthermore,

most placed emphasis on longer disease duration (72.5%), anticentromere antibody (ACA) positivity (72.5 %), and presence of telangiectasias (70%). In addition, one-third (30%) considered that diffuse cutaneous SSc was a significant risk factor for SSc-PAH.

**PAH screening**

The screening practices of rheumatologists (n=40) are presented in Figure 2. Most (85%) respondents screen patients with SSc for PAH every 12 months, and half if the clinical situation changes (i.e., the development of new signs/symptoms). Fewer screen more often (every 6 months: 7.5 %) or less frequently (every 2 years: 12.5 %). Over half (60%) reported using the DETECT algorithm for SSc-PAH screening, and with variable use of other available algorithms: ECS/ERS guidelines (10%) or ASIG algorithm (2.5%). Furthermore, some (15%) respondents indicated using other screening algorithms (e.g., Sheffield Pulmonary Vascular Disease Unit screening protocol) in their practice. One-fifth (20%) reported using none of the aforementioned approaches to screening.

**Screening investigations**

Figure 3A depicts which investigations the respondents ordered for the screening of PAH in patients with SSc. TTE and PFT were the most commonly ordered tests, with 100% and 95% of respondents selecting them, respectively. Less commonly requested tests were NT-pro-BNP (62.5%), serum urate/uric acid (42.5%), and electrocardiogram (42.5%). Less frequently investigations are displayed in Figure 3, and cardiopulmonary MRI was not utilised for screening.

Survey recipients were also asked to score their confidence interpreting tests for SSc-PAH on a scale of 1 to 10 (1 = “not confident at all” to 10= “completely confident”), and where the cut-off point for being ‘confident’ was defined as ≥7 out of 10. Respondents felt most confident interpreting PFTs (82.5 %), TTE (79.5%), chest radiography (75%), NT-pro-BNP (72.5 %) and troponin 70%, and least confident interpreting right heart catheterisation (RHC) (33.3%), cardiopulmonary MRI (26.3%), and exercise echocardiography (18.4 %). Detailed results are presented in Figure 3B.

**Transthoracic echocardiography**

Most (80%) respondents (n=40) believed that TTE could predict the likelihood of PAH in SSc patients with an intermediate probability, and fewer with either a low or high probability (7.5% and 12.5%, respectively). Almost all (97.4%) respondents (n=38) considered the tricuspid regurgitation jet a predictor of PH. Most also believed that the tricuspid annular plane systolic excursion/systolic pulmonary artery pressure (TAPSE/SPAP) (73.7%) and right atrium area (67.6 %) predicted SSc-PH. Whereas, only fewer considered that either the left ventricular systolic (11.8 %) or diastolic function (23.5%) was a predictor of PH in SSc patients. Over half (57.6%) were uncertain of the relevance of inferior vena cava dimensions.

Less than three-quarters (70%) of the rheumatologists (n=40) surveyed had access to dedicated PH/cardiac echocardiography technicians in their centres. Around three-quarters of respondents (n=40) either believed 'agreed' (50%) or 'strongly agreed' (22.5%) that the diagnosis can only be established once right heart catheterisation has been performed. However, there was difference in opinion with one-fifth who either disagreed (15%) or 'strongly disagreed' (5%), and some were of a neutral opinion (15%). Respondents (n=40) mostly (82.5%) either agreed (27.5%) or strongly agreed (55%) that patients with SSc-PAH should receive treatment from a specialised PH centre, whereas, some were neutral (10%) to this proposal, or strongly disagreed (7.5%).

**Barriers to screening**

We asked respondents (n=38) what they considered were the barriers to screening for PAH in patients with SSc (Figure 4), on a scale of 1 to 10 (1 = "not at all important" to 10= "extremely important"), and where the cut-off point for being 'important' was defined as ≥7 out of 10). These include difficulties interpreting results from other hospitals (76%), extended wait times for diagnostic tests (74%), limited time in clinical practice (47%), and unfamiliarity with screening algorithms (50%). In addition, around one-third (34.2 %) considered the requirement for invasive testing was a barrier to SSc-PAH screening.

To overcome obstacles to screening, respondents (n=39) agreed with the following strategies (Figure 4): access to key investigations (e.g., TTE and PFTs) (87.1%), ongoing clinical education e.g., educational programmes (82 %), multidisciplinary meetings with SSc-PAH specialists (79.5 %), a better understanding of proposed screening algorithms (79.5 %), better identification of patients at high risk (74.4 %), and better communication between specialised centres (71.8 %) (Figure 4).

Any difference between University versus general hospital?

Or between Specialised versus non specialised SSc unit?

**Diagnosis of PAH**

Respondents were asked their level of agreement whether the diagnosis of SSc-PAH can only be established after RHC. Around three-quarters either agreed (22.5%) or strongly agreed (75.0%) with this proposal, 10% were neutral, or 'disagreed' (15%) or 'strongly disagreed' (5%).

**Treatment**

We asked respondents (n=39) how confident they were about the pharmacological (treatment/drug) landscape of PAH in SSc patients, on a scale of 1 to 10 (1 = "not confident at all" to 10= "extremely confident" and where the cut-off point for being 'confident' was defined as ≥7 out of 10). All of the respondents (n=38, 100%) felt most confident with phosphodiesterase type 5 inhibitors and the majority with endothelin receptor antagonists (97.4%) and prostaglandins (84.2%) for the treatment of PAH in patients with SSc, with less importance attributed to calcium channel blockers (36.8%) and anticoagulants (34.2%). (See supplementary Figure 1, available at *Rheumatology* Online).

**Patient Education**

Around half (48.7%) of respondents (n=39) provide PAH education at the time of diagnosis. Where provided, there was variation in practice (n=19), most of which was delivered verbally

(73.7%), and around half facilitated referral to patient-led organisations (52.6%) or provided written educational materials (47.4%).

## Discussion

To our knowledge, this is the first study to comprehensively examine rheumatologists approaches and barriers to PAH? Or PH? screening in patients with SSc in the UK. A key finding of our study is that rheumatologists recognise the broad burden and impact of SSc-PAH including mortality, and the need for systematic screening, including to facilitate earlier referral to specialised PH referral centres, and the introduction of treatment intervention.

Of concern, although most respondents considered that the *diagnosis* of PAH can only be established by performing RHC, around one-quarter (27.5%) were either 'neutral' or disagreed/strongly agreed with this proposal. Similarly, while most respondents believed that patients with SSc-PAH should receive care from a specialised PH referral centre, around one-fifth were either 'neutral' (10%) or 'strongly disagreed' (7.5%) concerning this. There also, appears to be a number of misconceptions and knowledge gaps related to PH in SSc. For example, around-one third of respondents considered that a barrier to screening for SSc-PAH is a requirement for invasive testing, which is incorrect. s Another example is that the respondents were 'confident' that calcium channel blockers (36.8%) and anticoagulants (34.2%) were of similar treatment importance , which does not reflect clinical practice.

The majority (85%) of respondents reported that they performed annual screening for SSc-PAH, and half sooner, if the clinical situation were change in the interim. The annual screening rate reported in our study is comparable to that in the extant literature. For example, the rate reported by Morrisroe *et al*. (17), which found 84.4% adherence to annual screening guidelines via the algorithm proposed by ASIG; however, to highlight, that the adherence rate to annual screening in non-ASIG rheumatologists was much lower (38.5–58.9%). Quinn *et al*. (31) found a somewhat lower screening rate (71.9%) for Canadian rheumatologists performing annual TTE and PFT. However, in contrast, Wigger *et al*. (34) reported a much lower screening rate (58%) with both the same annual modalities (TTE and PFTs) in a specialised PH and SSc centre in Ohio, US. Furthermore, relevant to our study, in a previous UK-based study, Pauling

*et al*. (35) conducted a retrospective evaluation of PAH screening practices in patients with SSc in a secondary and tertiary referral centre in the South West England identified that PFTs (53.1%) and, to a lesser extent, TTE (34.7%) were mainly requested on an annual basis.

The majority of respondents reported using some form of SSc-PAH screening algorithm in their clinical practice, with over half (60%) using the DETECT algorithm. Few (≤10%) used the ECS/ERS guidelines or ASIG algorithm, and some (15%) respondents indicated that they used other screening algorithms. A possible hypothesis as to why DETECT is applied more widely in the UK than other algorithms could be that specific barriers to using TTE (with or without) PFTs as a first step including (but not limited to) access to dedicated PH/cardiac echocardiography technicians in their local centres, financial constraints, and long waiting lists.

There was strong recognition by the surveyed rheumatologists concerning the presence of symptomatic PAH; however, half also considered that PAH can be asymptomatic. Indeed, this highlights the need for systematic screening in *all* patients with SSc, as up to 22% of patients are asymptomatic at diagnosis of PAH (14,36). Furthermore, respondents recognised the importance of a number of risk factors (clinical factors and investigations) identify patients with SSc at high risk of PAH. In particular, these were FVC/DLCO, NT-pro-BNP, and limited cutaneous SSc, and to a lesser extent, longer disease duration ACA, and presence of telangiectasias.

As might be expected, TTE and PFTs were the most commonly ordered screening tests. All respondents reported ordering TTE, compared with 40% for ECG or urate analysis; this is despite the inclusion of ECG and urate analysis in the first step of the DETECT algorithm, which 60% of respondents reported using. These conflicting results may reflect that step 1 of the DETECT algorithm is likely calculated in still currently calculated in routine clinical practice performed *after* the TTE (step 2) has already been performed, rather than being applied systematically at regular time points. Specifically concerning TTE, most respondents believed this can predict the probability of SSc-PAH with either ‘intermediate’ (80%) or ‘high’ (12.5%) confidence.

Another explanation for prioritising TTE might be that heart may be affected in SSc outside PH and that the physician refer for TTE for PH screening but also myocardium, pericardium and valvular investigations.

There was variable confidence indicated by the respondents in interpretation of other tests used for SSc-PAH screening. Respondents felt most confident interpreting PFTs, NT-pro-BNP, troponin, TTE and chest radiography, and least confident interpreting cardiopulmonary MRI, RHC and exercise echocardiography. In contrast, Morrisroe *et al*. (17) found that 80% of the rheumatologists surveyed were hesitant to interpret screening studies, which was one of the reasons why they did not follow screening guidelines.

The main barriers to screening included organisational and external factors such as difficulty in visualising results from other centres, current (long) waiting lists, and limited time during daily clinical practice. Conversely, (30) only 13.5% of Canadian rheumatologists surveyed by Quinn *et al*. (31), reported problems with access to screening investigations as a barrier. However, cost was reported less frequently as a barrier in our study (34%) than in the Australian study (17), which was one of the main barriers identified in their research (60%). As for the rheumatologists' knowledge concerning screening, half of the respondents identified unfamiliarity with the proposed algorithms as a barrier, which is higher than previously reported in rheumatologists surveyed in Ohio (33%) (34) or Canada (21.6%) (31). Interestingly, we found that one-third of respondents considered that screening requires invasive testing, which may reflect respondents incorrectly conflating or confusing diagnostic testing with screening.

The majority of respondents agreed with a number of possible solutions to screening barriers, including easier access to requested tests, ongoing clinical education, multidisciplinary meetings with SSc-PAH specialists, better understanding of the proposed algorithms, better identification of high-risk patients, and better communication between specialist centres. A study by Wigger *et al* (34)m identified the main barriers to screening encountered by US physicians to be: a lack of knowledge, variation in practices and difficulty in ordering tests. This led to an education programme for physicians, (instructional sessions and sharing educational materials) and the development of an alarm system in the electronic medical record to remind physicians of upcoming screening appointments, and with a subsequent 14% increase

in screening rate. However, other barriers, such as those identified in this study, may require other strategies to be overcome.

Just under half of the respondents indicated that they provide PAH education for patients with SSc-PAH at diagnosis, and was delivered by a variety of methods, namely by provision of verbal (73.7%) or written (46.4%) information, or through referral to patient-led organisations (53%). This is somewhat difficult to interpret, as patients (in the UK) will receive formal PAH education after comprehensive evaluation and confirmation of the diagnosis via a National PH Service commissioned centre. Furthermore, it is important to highlight that patients with SSc are increasingly using internet/online for sources of information and support; however, the readability and quality is often inadequate or poor (37), and therefore patients should be signposted to appropriate resources, including trusted sources of information (e.g., patient-led organisations).

Our study has several notable strengths. Firstly, our sample comes from different (SSc specialist and non-specialist) centres and geographical regions throughout the UK, which provides a national cross-section of different practices. Secondly, we assembled a large multidisciplinary Steering Committee of clinicians with an interest in, respiratory experts with a specialist interest in pulmonary vascular disease, and a highly specialised cardiac physiologist with high expertise in adult echocardiography, to both design and interpret the survey findings. However, our study we need to acknowledge the potential limitations of this study. Firstly, our sample size was relatively limited, and there may have been a potential recruitment bias of clinicians with an interest in SSc and/or pulmonary vascular disease. However, only over a third of respondents worked in a specialist SSc centre, and half (50%) had only <25 patients with SSc under their care. Secondly, since we conducted a survey, the responses we received were based on theoretical knowledge and self-reported clinical practice. Future research including larger numbers of respondents is warranted to confirm the results of this survey, including examining the opinion of other key stakeholders (e.g., respiratory/pulmonary vascular experts and PH/cardiac echocardiography technicians).

In conclusion, rheumatologists recognise the importance and value of screening for PAH in patients with SSc, however, clinicians vary in their practice and there are a number of important barriers, and have identified a number of possible solutions. Taken together, these data confirm that there is an unmet need for the development of UK-based guidelines for SSc-PAH screening.

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**Data availability statement**

The data underlying this article will be shared on reasonable request to the corresponding author.

## Supplementary data

Supplementary data are available at *Rheumatology* online.

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| | n (%) |
|---|---|
| Gender | |
| Female | 25 (56.82) |
| Male | 18 (40.91) |
| Other | 1 (2.27) |
| Age | |
| 20–30 | 0 (0) |
| 31–40 | 15 (34.09) |
| 41–50 | 18 (40.91) |
| 51–60 | 8 (18.18) |
| 61–70 | 3 (6.82) |
| >70 | 0 (0%) |
| UK Region of Practice | |
| London | 2 (4.55) |
| North East | 1 (2.27) |
| North West | 14 (31.82) |
| Yorkshire | 6 (13.64) |
| East Midlands | 2 (4.55) |
| West Midlands | 3 (6.82) |
| South East | 2 (4.55) |
| East of England | 1 (2.27) |
| South West | 11 (25) |
| Wales | 0 (0) |
| Scotland | 2 (4,55) |
| Northern Ireland | 0 (0) |
| Work in a specialised SSc unit (yes) | 17 (38.64) |
| Type of hospital of work | |
| University hospital | 24 (54.55) |
| General hospital | 20 (45.45) |
| Private practice | 3 (6.82) |
| Years of experience in rheumatology | |
| 0–5 | 15 (34.09) |
| 5–10 | 5 (11.36) |
| 11–20 | 19 (43.18) |
| 21–30 | 4 (9.09) |
| >30 | 1 (2.27) |
| Number of SSc patients under their care | |
| <25 | 22 (50) |
| 26–50 | 8 (18.18) |
| 51–100 | 3 (6.82) |
| 101–200 | 4 (9.09) |
| >201 | 7 (15.91) |
| Percentage of PAH in their SSc patients | |
| <1% | 3 (6.82) |
| 1–5% | 9 (20.45) |
| 6–10% | 18 (40.91) |
| 11–25% | 14 (31.82) |
| 26–50% | 0 (0) |

Table 1. Respondent demographics. PAH: pulmonary arterial hypertension, SSc: systemic sclerosis

| | n (%) |
|---|---|
| Symptoms associated with SSc-PAH | |
| Shortness of breath | 40 (100) |
| Fatigue | 38 (95) |
| Syncope | 37 (67.50) |
| Leg/ankle swelling | 35 (87.50) |
| Chest pain | 23 (57.50) |
| Palpitations | 21 (52.50) |
| None – asymptomatic | 21 (52.50) |
| Other[a] | 2 (5) |
| Risk factors they associate with SSc- PAH | |
| PFT: FVC/DLCO | 35 (87.50) |
| Elevated levels of NT-pro-BNP | 32 (80) |
| Limited cutaneous SSc | 31 (77.50) |
| Longer disease duration | 29 (72.50) |
| Presence of anticentromere antibody (ACA) | 29 (72.50) |
| Presence of telangiectasias | 28 (70) |
| Diffuse cutaneous SSc | 12 (30) |
| Other[b] | 2 (5) |

Table 2. Symptoms and risk factors rheumatologists associate with PAH in patients with SSc. DLCO: diffusing capacity for carbon monoxide, FVC: forced vital capacity, PAH: Pulmonary arterial hypertension, PFT: pulmonary function test, NT-pro-BNPP: N-terminal pro-B-type natriuretic peptide SSc: systemic sclerosis. [a]Cough, recurrent digital ulcers and telangiectasias. [b]Shortness of breath in the absence of interstitial lung disease, digital ulcer disease and high uric acid.

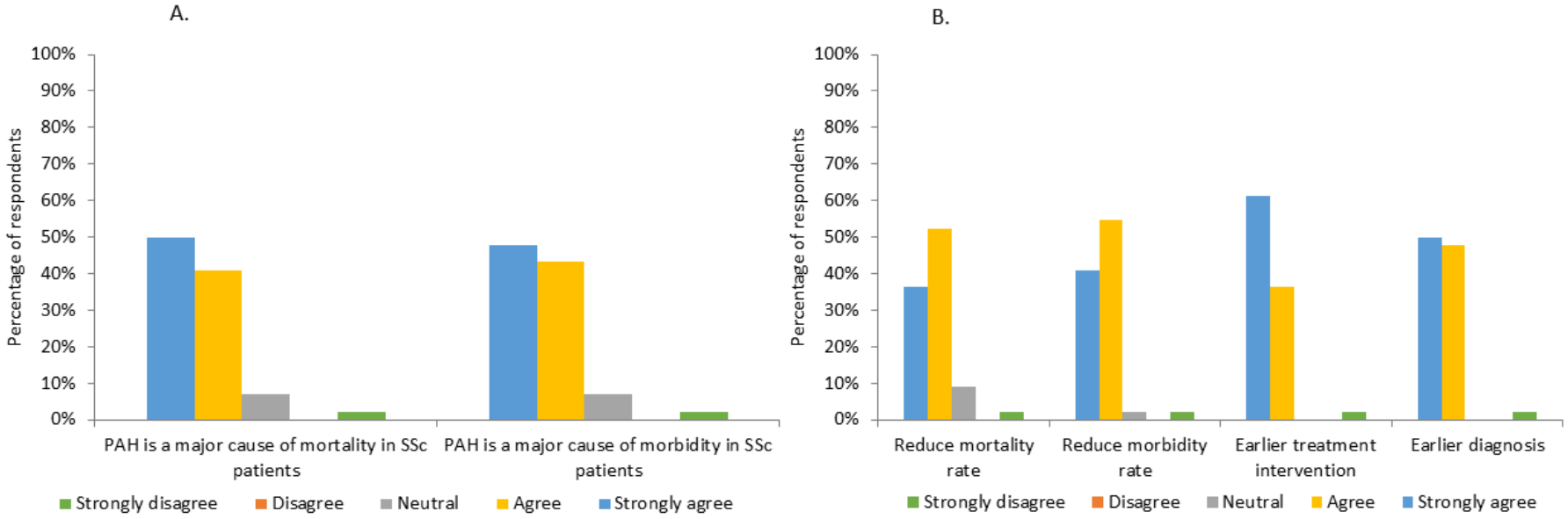



Figure 1. A: The impact of PAH as a major cause of morbidity and mortality in patients with SSc. B: The impact that PAH screening in SSc patients would help to achieve. PAH: pulmonary arterial hypertension, SSc: systemic sclerosis.

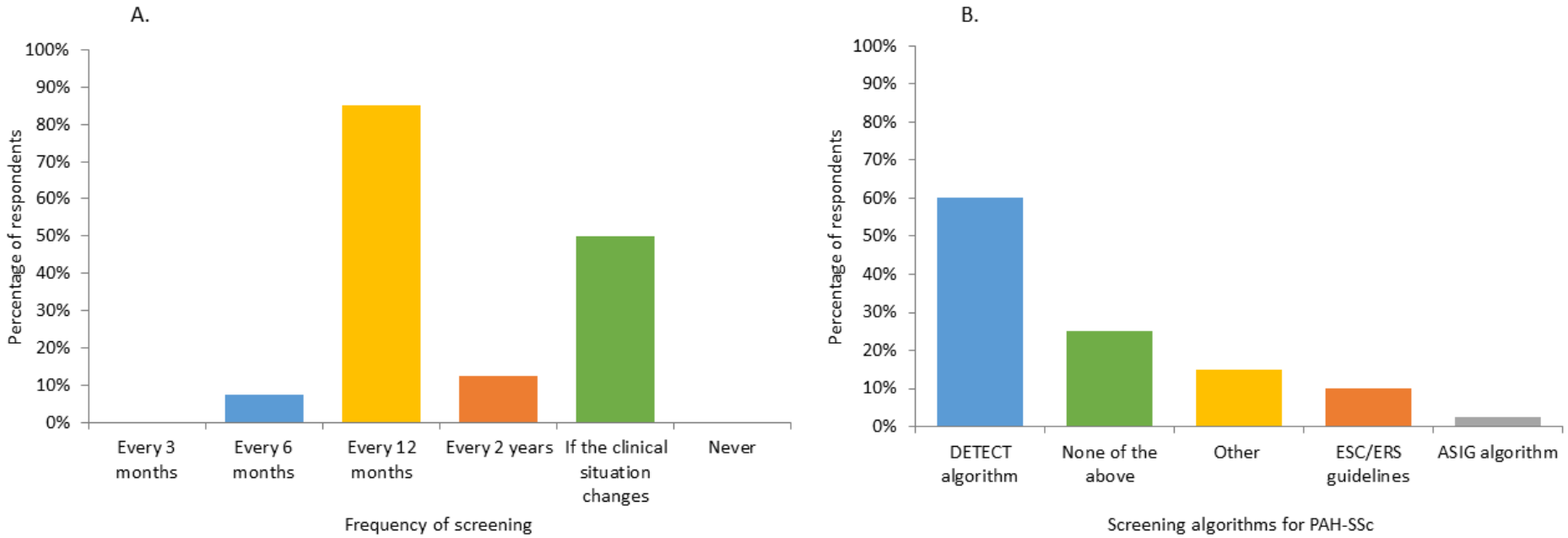



Figure 2. Screening practices for PAH in patients with SSc. A: Frequency of screening for PAH in patients with SSc. B: Screening algorithms used among the UK rheumatologists surveyed. DLCO: diffusing capacity for carbon monoxide; ECG: electrocardiogram; PFT: pulmonary function tests; TTE: transthoracic echocardiogram.

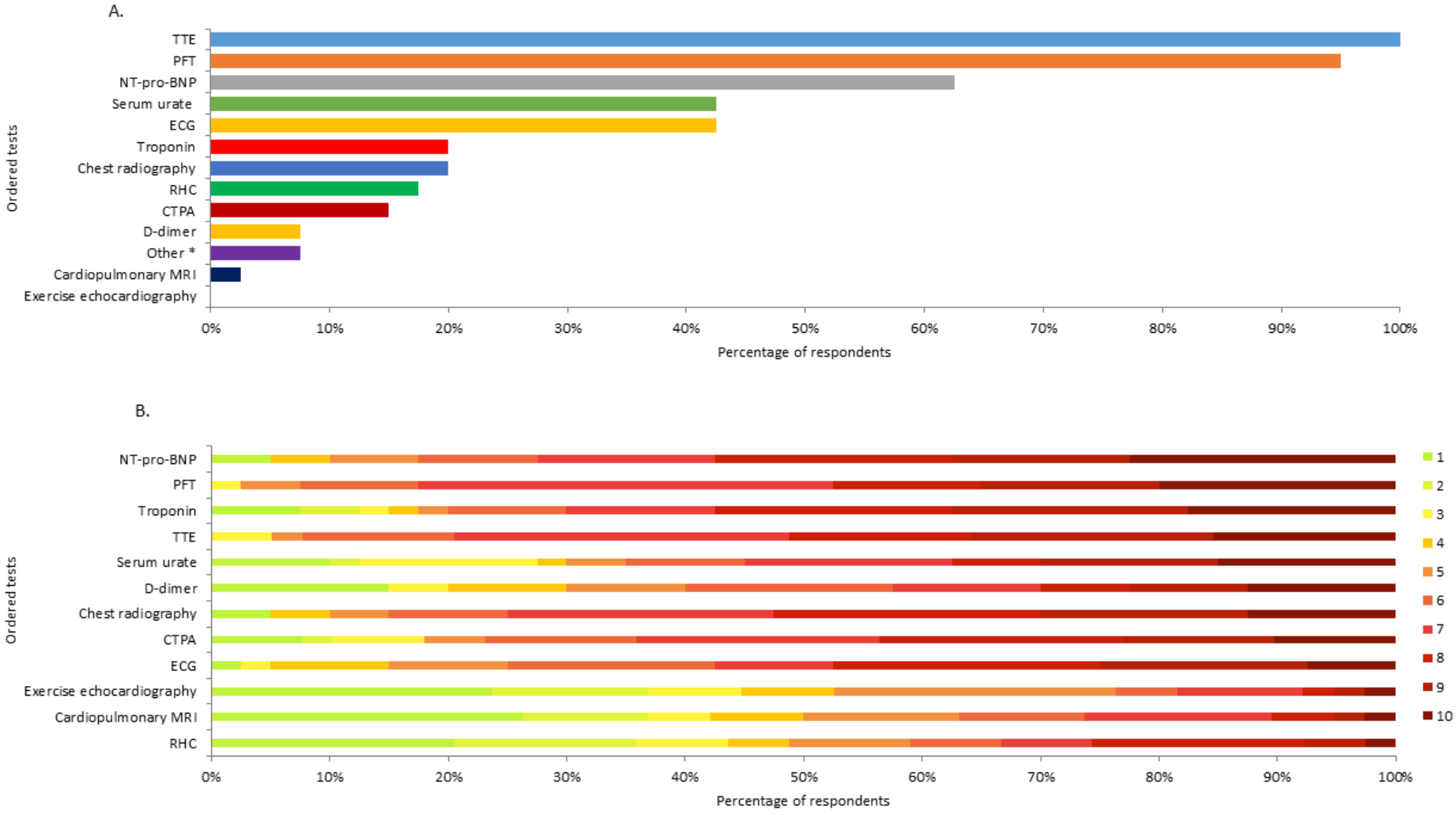
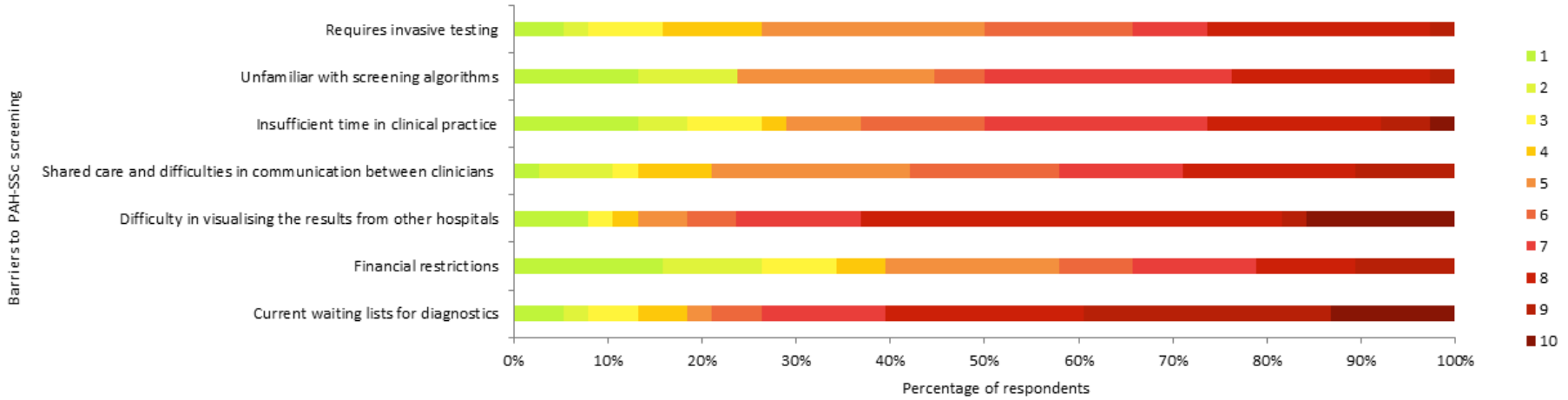

A.
TTE
PFT
NT-pro-BNP
Serum urate
ECG
Troponin
Chest radiography
RHC
CTPA
D-dimer
Other *
Cardiopulmonary MRI
Exercise echocardiography
Ordered tests
0%
10%
20%
30%
40%
50%
60%
70%
80%
90%
100%
Percentage of respondents
B.
NT-pro-BNP
PFT
Troponin
TTE
Serum urate
D-dimer
Chest radiography
CTPA
ECG
Exercise echocardiography
Cardiopulmonary MRI
RHC
Ordered tests
0%
10%
20%
30%
40%
50%
60%
70%
80%
90%
100%
Percentage of respondents
1
2
3
4
5
6
7
8
9
10

Figure 3. Screening investigations performed by rheumatologists (n=40) for PAH in patients with SSc. Tests ordered (A) and confidence (B) of the respondents in interpreting the test results, where 1 = “not confident at all” and 10 = “extremely confident”. CTPA: computed tomography pulmonary angiogram, ECG: electrocardiogram, MRI: magnetic resonance imaging, NT-pro-BNP: N-type natriuretic peptide; PFT: pulmonary function tests, RHC: right heart catheterization, TTE: transthoracic echocardiogram.


Barriers to PAH-SSc screening
Requires invasive testing
Unfamiliar with screening algorithms
Insufficient time in clinical practice
Shared care and difficulties in communication between clinicians
Difficulty in visualising the results from other hospitals
Financial restrictions
Current waiting lists for diagnostics
1
2
3
4
5
6
7
8
9
10
0%
10%
20%
30%
40%
50%
60%
70%
80%
90%
100%
Percentage of respondents

B.

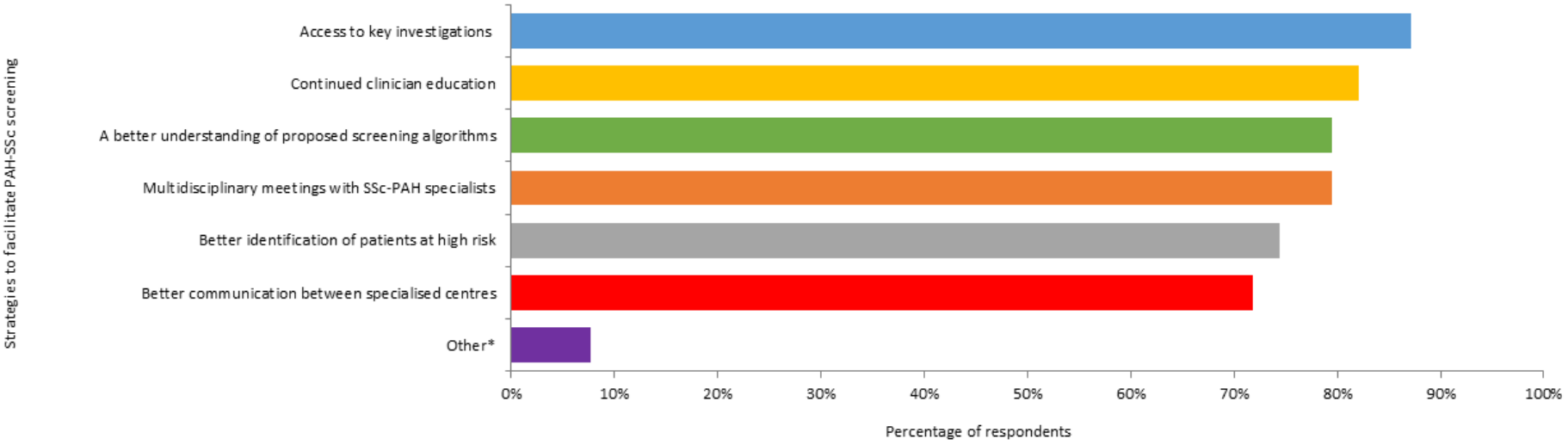

Strategies to facilitate PAH-SSc screening
Access to key investigations
Continued clinician education
A better understanding of proposed screening algorithms
Multidisciplinary meetings with SSc-PAH specialists
Better identification of patients at high risk
Better communication between specialised centres
Other*
0%
10%
20%
30%
40%
50%
60%
70%
80%
90%
100%
Percentage of respondents

Figure 4. Barriers and strategies to facilitate PAH screening in patients with SSc. Barriers (A) to screening and strategies (B) to facilitate PAH screening in patients with SSc, where 1 = “not confident at all” and 10 = “extremely confident”.