Recognition of pulmonary hypertension in the rheumatology community: lessons from a Quality Enhancement Research Initiative

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ABSTRACT

Objective. *The aim of this study was to utilise the Quality Enhancement Research Initiative in Systemic Sclerosis (QuERI-SSc) to measure and reduce a perceived gap in the diagnosis of pulmonary arterial hypertension (PAH) associated with systemic sclerosis (SSc).*

Methods. *Rheumatologists enrolled patients with SSc (aged ≥18 years) and provided data on a panel of diagnostic tests over 3 years. Pulmonary function testing, echocardiography, 6-minute walk distance, N-terminal pro-brain natriuretic peptide assays, high-resolution computed tomography of the lungs, and ventilation/perfusion scan plus right heart catheterisation (RHC; when appropriate) were emphasised. Exclusion criteria included previously documented PAH, interstitial lung disease, and SSc overlapping with other connective tissue disease.*

Results. *Participating rheumatologists enrolled 207 patients with SSc (90% female; 80% white), with a median age of 57 years and median disease duration of 5 years. A total of 82% of patients were classified as New York Heart Association functional class I and II; of these patients, 177 had an echocardiogram at enrolment and 191 at any time during the study. Of those who met study-specified criteria for RHC at enrolment, only 3 of 7 patients underwent RHC.*

Conclusion. *The screening algorithm was successful in identifying patients with mild impairment. Although specific tools were recommended for screening PAH in patients with SSc, results indicate that significant diagnostic care gaps still exist in the general rheumatology community. Better understanding and adherence to guidelines could improve the care and, ideally, outcomes of these high-risk patients.*

Introduction

Pulmonary involvement in systemic sclerosis (SSc) is the leading cause of morbidity and mortality in affected patients (1, 2). This condition is characterised by pulmonary hypertension (PH) or interstitial lung disease (ILD), or a combination of both (3). PH in patients with SSc is further categorised as either pulmonary arterial hypertension (PAH), PH due to left-sided heart disease, or PH due to associated ILD (3, 4).

PAH risk factors in patients with SSc include older age, limited cutaneous SSc, and a reduced or declining diffusing capacity for carbon monoxide (DL_{CO}) (5, 6). Published studies have shown that patients with SSc-associated PAH (SSc-PAH) have less robust and less durable outcomes than patients with idiopathic PAH (7-10). This includes measures of survival, function, and quality of life. Constitutive features of SSc are undoubtedly contributory, including the multisystem nature of the underlying disease (presence of underlying heart disease and ILD), the high proportion of elderly patients, reduced capacity for right ventricular (RV) adaptation, and the presence of aberrant pulmonary vasculopathy (including pulmonary veno-occlusive disease) (7-10).

These findings support the need for aggressive strategies to identify PAH in patients with SSc at an early stage. To this end, Hachulla *et al.* (11) developed a screening algorithm for application to a nationwide multicentre SSc population in France. This algorithm, based on dyspnea, Doppler echocardiographic evaluation of the velocity of tricuspid regurgitation (TR), and right heart catheterisation (RHC), enabled early detection of PAH at a mild stage in patients with SSc enrolled in the French ItinerAIR-Sclerodermie study. Early detection of PAH in this cohort

resulted in a survival estimate of 91.1% compared with 56.3% in patients with baseline PAH (11).

Consensus guidelines published by the American College of Chest Physicians (ACCP) and the American College of Cardiology Foundation/American Heart Association recognise the risk of PAH in SSc and provide recommendations for diagnostic screening of suspected PAH and management of patients with a confirmed PAH diagnosis with the intent to bring a more unified approach and improve the management of these high-risk patients (12-14). The 2004 ACCP guidelines recommend Doppler echocardiography to detect elevated pulmonary artery pressure (PAP) in high-risk individuals, including those with SSc (14). More recent guidelines specify annual echocardiography in SSc followed by RHC if the echocardiogram reveals evidence of PAH (elevated estimated RV systolic pressure [RVSP] or right heart enlargement) (12). Guidelines also recommend the use of pulmonary function testing (PFT) with DL_{CO} every 6 to 12 months to detect pulmonary involvement in patients with SSc (13, 14). Furthermore, a study by Humbert *et al*. (15), albeit with confounding limitations of lead time and length bias, demonstrated that active detection protocols identify patients with PAH secondary to SSc with less advanced pulmonary vascular disease and suggested dramatic gains in survival attending earlier intervention.

The Quality Enhancement Research Initiative in Systemic Sclerosis (QuERI-SSc) aimed to facilitate a guidelines-based approach to PAH management in the context of individual patient care in a real-world health care setting. To optimise patient care, QuERI-SSc provides physicians with a patient care map with practical recommendations for early PAH diagnosis and management in patients with SSc and suspected PAH or PH: laboratory tests including N-terminal pro-brain natriuretic peptide (NT-proBNP), PFTs, echocardiogram, pulmonary artery systolic pressure (PASP), RVSP, 6-minute walk distance (6MWD), ventilation/perfusion (V/Q) scan, high-resolution computed tomography (HRCT) of the lungs, and RHC when appropriate. There are 2 essential components to QuERI-SSc: (1) a Research Initiative component, based on analysis of the data, which monitors management and outcomes in real-world practice; and (2) a Quality Enhancement component, which aims to define existing practice patterns and current variations from guidelines (care gap) and implement interventions set to ultimately close this gap. To this end, participating physicians receive feedback and insight generated through patient follow-ups and outcomes. This allows physicians to compare their management style to that of scleroderma experts, as well as national and regional averages obtained through data collection and analysis. Earlier data analyses from the PAH Quality Enhancement Research Initiative (PAH-QuERI), another initiative in patients with World Health Organization (WHO) group I PAH, indicated that a diagnostic care gap is apparent, such that certain essential diagnostic tests may be underutilised and guidelines not universally followed (16). Here, we report the results of a data analysis regarding current management of PAH and 3-year outcomes of patients with SSc enrolled in QuERI-SSc at community and academic rheumatology practice settings.

Materials and methods

Study design

Rheumatologists practicing in the community and in academic centres (27 rheumatologists from 22 community practice settings and 5 academic centres) in the United States were invited to participate in the QuERI-SSc initiative. Participating physicians were provided with structured guidelines on the diagnosis and management of PAH based on a consensus of clinical evidence and expert opinion (14, 17). Guidance on patient evaluation was provided in the form of an algorithm developed by the steering committee that incorporated available published data regarding the screening of patients with SSc-PAH (Fig. 1) and which preceded consensus guidelines of both the US and EU expert communities (12, 18). Between July 2006 and December 2007, rheumatologists consecutively enrolled patients with previously or newly diagnosed SSc and recorded data on a recommended panel of diagnostic tests. They were asked to complete an electronic case report form (eCRF) to document the screening and management of patients suspected of having PAH. Patients were enrolled and followed for 3 years. Rheumatologists were given electronic feedback recommending RHC based on their case report forms.

Patients

Male and female patients aged ≥18 years were included if they met criteria for SSc based on American College of Rheumatology guidelines (19). Exclusion criteria included the following: unavailable for follow-up; previously documented PH/PAH (RHC-determined mean PAP >25 mm Hg, pulmonary vascular resistance >3 Wood units, and pulmonary capillary wedge pressure >15 mm Hg); severe ILD (forced vital capacity [FVC] <45% predicted); and overlap with other connective tissue disease.

Data collection

The following information was collected for all patients at enrolment: demographics, organ involvement, clinical and laboratory variables, current medications including PAH-specific therapies, and diagnostic tests including PFT, echocardiogram, and 6MWD when available. Additional tests including V/Q scanning, HRCT, and RHC were performed when appropriate (12). RHC was recommended, but not mandated, when a patient exhibited dyspnea associated with a New York Heart Association (NYHA) functional class (FC) ≥II and 2 of the following 4 "triggers":

A. Doppler echocardiogram estimated RVSP >40 mm Hg or TR jet velocity >3.0 m/sec;
B. DL_{CO} <55% of the predicted value and an FVC/DL_{CO} ratio of >1.4;
C. NT-proBNP plasma concentration >140 pg/mL;
D. Physical examination evidence of PAH including increased pulmonary component of second heart sound (P2), RV gallop, TR murmur, and evidence of right-sided heart failure.

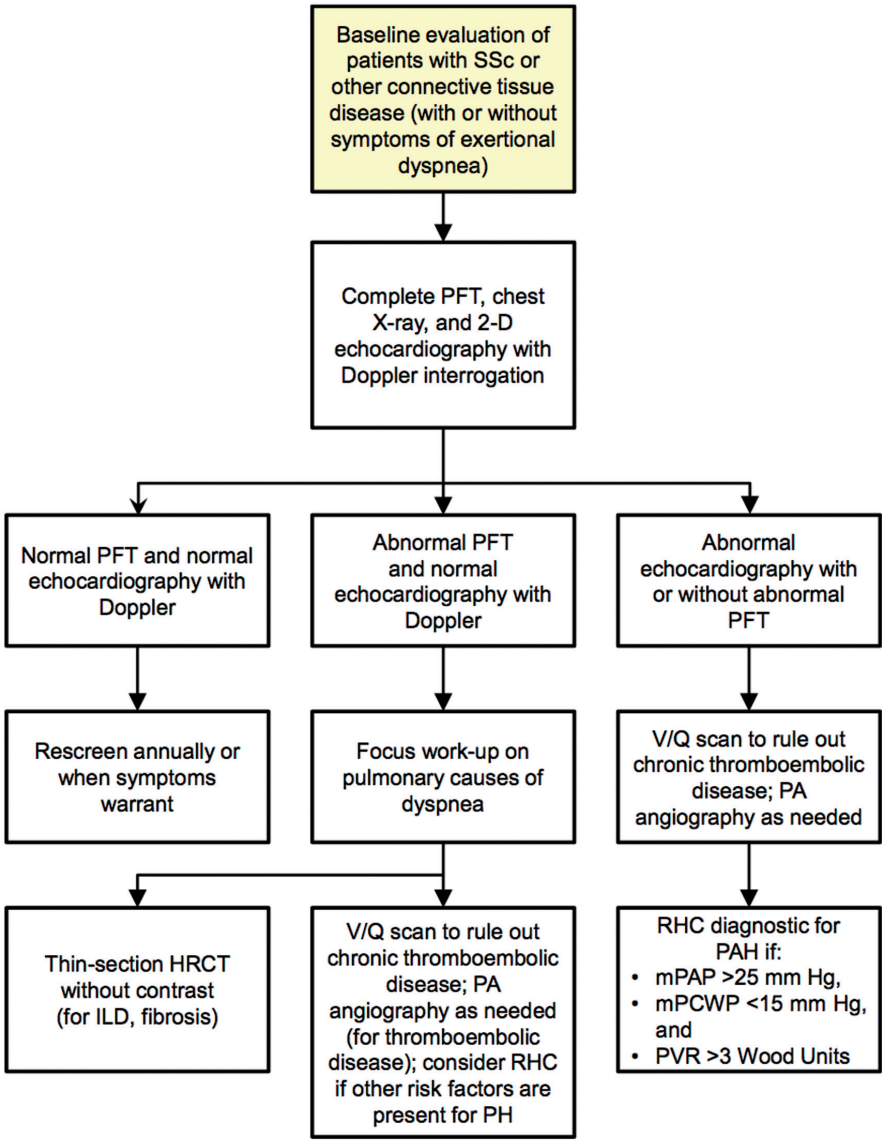

Fig. 1. Algorithm for evaluating patients with suspected pulmonary arterial hypertension (PAH). HRCT: high-resolution computed tomography; ILD: interstitial lung disease; mPAP: mean pulmonary artery pressure; mPCWP: mean pulmonary capillary wedge pressure; PA, pulmonary artery; PFT: pulmonary function test; PVR: pulmonary vascular resistance; RHC: right heart catheterisation; SSc: systemic sclerosis; V/Q: ventilation/perfusion.

These criteria were chosen based on previous studies showing an association with PAH (12-14, 20). NT-proBNP was measured by local laboratories. HRCT was recommended in any patient with physical evidence of ILD, including inspiratory crackles, an FVC of <90% of the predicted value, or a DL_{CO} of <55% of the predicted value. These criteria were recommended as a floor for consideration of RHC but did not preclude performance of RHC based on less complete data, eg, a grossly abnormal echocardiographic study or conspicuous clinical findings.

Physicians completed the eCRF at enrolment, at 6 months, and yearly for up to 3 years. The results of all assessments were recorded together with data regarding the therapy given, diagnosis, and mortality over the 3 years of follow-up. Physicians were provided with computerised "real-time" feedback to demonstrate how their patient management compared with national and regional averages and showed variance with scleroderma experts and evidence-based guideline recommendations. Investigators received modest payment for the completion of eCRFs. Investigators were further compensated for attendance at annual meetings held in conjunction with the National Scientific Meetings of the American College of Rheumatology where study goals and interim results were discussed.

The study protocol was approved centrally (Western Institutional Review Board, Olympia, WA, USA) and by the institutional review boards of each participating institution prior to commencement of the study. Participation in the study was voluntary, and all patients were given full and adequate verbal and written information regarding the study objectives and procedures. All patients provided written informed consent prior to study entry.

Statistical analyses

Analysis of the QuERI-SSc data was exploratory in nature. Numerical values were summarised as medians (25^{th} percentile – 75^{th} percentile), and categorical variables were presented as absolute frequencies and proportions (n [%]). A two-sided *p*-value <0.05 was considered significant. All statistical analyses were performed using SAS version 9.2 (SAS Institute Inc., Cary, NC, USA).

Results

Patient demographics, functional characteristics, and symptoms at enrolment

A total of 214 patients were recruited, 7 of whom did not meet the inclusion criteria. Demographics and characteristics at enrolment of the 207 patients screened and entered in the study are summarised in Table I. The median time from diagnosis of SSc was 5 years (interquartile range, 1–10 years).

Compliance with protocol recommendations for diagnostic work-up

Table II shows the percentage of patients having recommended diagnostic tests at enrolment. Echocardiography at enrolment was conducted in 177 patients (86%) and RHC in only 11 patients (5%), 2 of whom were in NYHA FC I, 6 in FC II, and 2 in FC III; FC was not known for 1 patient.

Echocardiographic findings at enrolment for the 177 patients are summarised in Table III. Echocardiogram

Table I. Demographic characteristics at time of enrolment.

Characteristic, n (%)	n=207
Age, years (range)*	57 (49–66)
Female sex	186 (89.9)
White	166 (80.2)
African American	17 (8.2)
Hispanic	9 (4.4)
Asian	3 (1.5)
Pacific Islander	2 (1.0)
Other	4 (1.9)
Unknown	6 (2.9)
Type of SSc	
Limited SSc	126 (60.9)
Diffuse SSc	66 (31.9)
Unknown	15 (7.2)
Signs/symptoms at enrolment	
Raynaud's phenomenon	181 (87.4)
Digital ulcers	64 (30.9)
Dysphagia	76 (36.7)
Heartburn	125 (60.4)
Proximal muscle weakness	36 (17.4)
NYHA functional class	
I	97 (46.9)
II	72 (34.8)
III	14 (6.8)
IV	3 (1.4)
Unknown	21 (10.1)
Physical examination	
BMI, kg/m^2*	25.1 (22.4–29.4)
Systolic BP, mm Hg*	120 (110–132)
Diastolic BP, mm Hg*	70 (66–80)
Heart rate, bpm*	76 (68–83)
Increased P2	12 (5.8)
RV gallop	1 (0.5)
Tricuspid regurgitation murmur	5 (2.4)
Peripheral oedema	35/192 (18.2)
Rales	31 (15.0)

*Reported as median (interquartile range).
BMI: body mass index; BP: blood pressure; bpm: beats per minute; NYHA: New York Heart Association; P2: pulmonary component of second heart sound; RV: right ventricular; SSc: systemic sclerosis.

readings performed by the patients' community cardiologists were interpreted by participating physicians to determine whether they were suggestive of PH, based on guidance provided in the study protocol: an echocardiogram with a TR jet velocity >3.0 m/sec or an estimated RVSP >40 mm Hg. Based on the response to the question in the echocardiography eCRF "Echocardiogram consistent with diagnosis of PH, yes/no?," 31 of the patients with an enrolment echocardiogram were identified by a rheumatologist as having results suggestive of PH. Among the 31 defined with echocardiogram results

Table II. Diagnostic work-up at enrolment.

Parameter, n (%)	n=207
PFT	172 (83.1)
Echocardiogram	177 (85.5)
HRCT or chest x-ray	128 (61.8)
Chest x-ray	92 (44.4)
Normal chest x-ray (n=92)	59 (64.1)
HRCT	66 (31.9)
6MWD	22 (10.6)
Serum BNP, yes	51 (24.6)
Serum NT-proBNP, pg/mL*	48 (21–74)
RHC, total	11 (5.3)
V/Q lung scan	2 (1.0)

*Reported as median (interquartile range).
6MWD: 6-minute walk distance; BNP: brain natriuretic peptide; HRCT: high-resolution computed tomography; NT-proBNP, N-terminal pro-BNP; PFT: pulmonary function test; RHC: right heart catheterisation; V/Q: ventilation/perfusion.

consistent with PH, 9 (29%) had RV enlargement and 7 developed PAH according to the physicians' classification of PAH etiology collected at the end of the study (not shown). Of particular note, TR jet velocity and estimated RVSP/PASP were reported in only 89 (50%) and 123 (70%) of these 177 patients, respectively; 102 patients had RVSP/PASP ≤40 mm Hg and 21 had RVSP/PASP >40 mm Hg (Table III).

Analysis of compliance with protocol recommendations for performing an RHC showed that, among 89 patients presenting with dyspnea and NYHA FC ≥II at enrolment, 7 (8%) satisfied any 2 or more of the 4 "trigger" parameters and thus met the criteria for a recommended RHC. However, only 3 of these 7 patients in whom the RHC was recommended were referred for this procedure (Table IV). Five other patients with NYHA FC ≥II but who did not meet the protocol-based criteria for referral for RHC nevertheless underwent RHC at enrolment based on investigator clinical judgment or other laboratory information.

During the 3-year follow-up period, a total of 27 patients (13%) underwent RHC. Of these, 19 were in NYHA FC ≥II, but only 6 met the recommended RHC "trigger" criteria, as 1 patient underwent RHC performed at both enrolment and the 6-month visit (Table IV).

Final PAH etiological classification

Based on clinical impressions of the

Table III. Echocardiogram results at enrolment.

Parameter, n (%)	n=177
RV enlargement	15 (8.5)
RV diameter, mm (n=15)*	27.0 (22.0–39.0)
RV hypertrophy (n=148)	4 (2.7)
Septal flattening	1 (0.6)
Tricuspid regurgitation	89 (50.3)
Jet velocity, m/sec (n=45)*	2.3 (2.0–2.6)
Maximum jet velocity >3 m/sec (n=45)	5 (11.1)
RVSP/PASP, mm Hg (n=123)*	30.0 (24.0–38.0)
No RVSP/PASP recorded	54 (30.5)
Analysis of patients with reported RVSP/PASP	
RVSP/PASP ≤40 mm Hg	102 (57.6)
RVSP/PASP >40 mm Hg with RVSP/PASP value >40 mm Hg (n=21)	21 (11.9)
RV enlargement	5/21 (23.8)
RV hypertrophy	2/15 (13.3)
Septal flattening	0 (0.0)
LV systolic dysfunction (n=163)	2 (1.2)
LV diastolic dysfunction (n=162)	17 (10.5)
Pericardial effusion (n=162)	12 (7.4)

*Reported as median (interquartile range).
LV: left ventricular; PAH: pulmonary arterial hypertension; PASP: pulmonary artery systolic pressure; RHC: right heart catheterisation; RV: right ventricular; RVSP: RV systolic pressure.

attending physician, 30 patients were stated to have developed PAH during the 3-year follow-up. However, only 10 had confirmatory RHC, and only 5 of these were considered the result of protocol-based "trigger" criteria for RHC. We did not record the RHC haemodynamics or the details of investigator basis for assigning a diagnosis of PH. In addition, we did not inquire about the WHO classification of PH.

Discussion

QuERI-SSc was an initiative to help improve the gap in early diagnosis and treatment of PAH in patients with SSc. This study showed that, despite all efforts to bring a more unified approach to the diagnosis and management of these high-risk patients, diagnostic gaps still exist in the rheumatology community, and certain essential and recommended diagnostic tests seem under- or inappropriately utilised. Overall, only 5% (n=11/207 [8 with FC II and other triggers for RHC]) of patients underwent RHC at baseline, and 13% (n=27/207 [25 RHC in 19 unique patients with FC

Table IV. Compliance with protocol recommendations for performing RHC in patients with NYHA functional class ≥II in each visit.

Trigger parameters for RHC (at least 2 of 4 required for RHC) NYHA functional class ≥II[†]	Enrolment (n=89)	6 months (n=71)	1 year (n=50)	2 years (n=40)	3 years (n=42)
A. Echocardiography-estimated RVSP >40 mm Hg or TR jet velocity >3.0 m/sec	11 (12.4)	2 (2.8)	4 (8.0)	3 (7.5)	4 (9.5)
B. DL_{CO} <55% and FVC/DL_{CO} ratio >1.4	14 (15.7)	8 (11.3)	3 (6.0)	1 (2.5)	6 (14.3)
C. NT-proBNP >140 pg/mL	5 (5.6)	1 (1.4)	2 (4.0)	2 (5.0)	0 (0.0)
D. Physical examination*	11 (12.4)	9 (12.7)	5 (10.0)	4 (10.0)	4 (9.5)
Patients with ≥2 trigger parameters[‡]	7 (7.9)	3 (4.2)	1 (2.0)	1 (2.5)	3 (7.1)
RHC performed in patients with ≥2 trigger parameters[§]	3 (3.4)	1 (1.4)	1 (2.0)	1 (2.5)	1 (2.4)
Total RHC performed[¶]	8 (9.0)	6 (8.5)	4 (8.0)	3 (7.5)	4 (9.5)

*Physical examination (increased pulmonary component of second heart sound, right ventricular gallop, tricuspid regurgitation [TR] murmur, and evidence of right-sided heart failure) was only performed at time of enrolment; thus, such patients who attended follow-up visits were included. Not all patients attended every visit.
[†]All percentages were calculated based on the N denoted in the column.
[‡]Sum of all visits = 15, but only 10 unique patients (*i.e.* no patient was counted twice).
[§]Sum across visits = 6 unique patients (patient at 6-month visit was the same patient who underwent right heart catheterisation [RHC] at enrolment).
[¶]Sum across visits = 19 unique patients.
DL_{CO}: diffusing capacity for carbon monoxide; FVC: forced vital capacity; NT-proBNP: N-terminal pro-brain natriuretic peptide; NYHA: New York Heart Association; RVSP: right ventricular systolic pressure.

II and other triggers for RHC]) during the 3-year follow-up. Among the 89 patients with NYHA FC ≥II at enrolment, 8% (n=7/89) met the recommended, but not mandatory, protocol-based criteria for RHC, yet only 3 of these 7 patients underwent RHC. It seems that diagnostic recommendations were not appropriately followed by the rheumatology community, and thus this particular initiative towards a more unified approach to clinical assessment did not succeed in closing the diagnostic gap.

The guidelines clearly state that all patients considered to have "probable PAH" after non-invasive screening should undergo RHC, and RHC is the gold standard for assessment of haemodynamics in PAH (12). The study by Humbert *et al*. (15) offers a good demonstration of the effectiveness and benefits of RHC testing, which allowed the identification of patients with milder forms of PAH and better outcomes.

In the Humbert study, consecutive patients with SSc who entered a systematic PAH surveillance programme using echocardiography as screening were compared with a group of patients with SSc-PAH diagnosed via routine clinical practice. Those in the detection programme had better NYHA FC and milder pulmonary haemodynamic abnormalities at enrolment than those in the routine practice cohort, consistent with the idea that they were detected earlier. In addition, patients in the detection programme had significantly better survival rates at 1, 3, 5, and 8 years (100%, 81%, 73%, and 64%, respectively) than the routine practice group (75%, 31%, 25%, and 17%; p=0.0037) (15). These data suggest improved outcomes in patients who are identified earlier via systematic screening programmes.

The adherence of rheumatologists to the proposed algorithm was unreliable. A total of 14% of patients did not have an echocardiogram, 75% did not have NT-proBNP testing, and 17% did not have the PFT with DL_{CO} at baseline. However, it should be noted that NT-proBNP testing is not broadly covered by insurance in the United States for patients without heart failure, and may not be easily obtained through community laboratories.

Even more concerning is that only 10 of the 30 patients who were said to have developed PAH during the course of this study actually underwent an RHC to establish the diagnosis. The exact reason for this gap between recommended and actual testing for the diagnosis of PAH in SSc is not clear. Lack of knowledge regarding the predictors of PAH in the rheumatology community or cardiologists'/pulmonologists' decision not to perform RHC in these patients may be relevant issues. We did not explicitly evaluate the reason for the disparity between established guidelines and real-life practice, but our findings do indicate the need for an interdisciplinary model promoting collaboration between different subspecialists caring for patients with PAH. This should be a continued goal and could be the focus of future research. One might also argue that patients with SSc would benefit from referral to a specialised scleroderma or PH centre where guidelines might more likely be followed.

The previous analysis of management and outcomes of patients with PAH enrolled in the PAH-QuERI study showed that, despite the availability of detailed and clear guidelines and at least one automatic reminder to physicians, the use of recommended diagnostic tests for PAH also deviated from the recommendations (16). Similarly, in a comparative study of patients with idiopathic PAH and those with SSc enrolled in PAH-QuERI, Clements *et al*. found significant differences in the evaluation and treatment of patients with SSc. Patients with SSc were less likely to be treated with parenteral prostanoids and had a significantly lower survival rate compared with patients with idiopathic PAH (21).

NYHA FC is an independent predictor of survival in PAH. Hachulla *et al*. found that prognosis was more severe among patients in NYHA FC IV *versus* patients in NYHA FC II or III during a 3-year follow-up in patients with SSc (22). The association of NYHA FC II and survival has also been demonstrated in other PAH cohorts (23-26). QuERI-SSc was successful in recruiting patients with milder NYHA functional limitations (82% in NYHA FC I and II). This is consistent with findings from

Hachulla *et al*. (27) where a screening algorithm was able to recruit 55% of patients with NYHA FC I and II compared with usual referral to PAH clinics (1% and 24% with FC I and II, respectively) (28) in France.

Transthoracic echocardiograms were performed by community cardiologists in the majority of cases. An estimate of RVSP and TR jet velocity was reported in only 70% and 50% of patients, respectively. In a large point prevalence survey of echocardiography in SSc, 19% of the patients could not be evaluated for RVSP due to lack of a TR jet. Although the current recommendation mentions elevated RVSP or right-sided heart enlargement (12) as signs of PAH on echocardiography that should initiate a PAH work-up, our algorithm failed to include the latter. Presence of RV enlargement (n=15/177) or hypertrophy (n=4/177) may explain RHCs in QuERI-SSc that did not meet the "trigger" criteria. The proposed RHC algorithm also points to the possible importance of PFT with DL_{CO} as a screening modality for PAH. Steen *et al*. (6) demonstrated that a DL_{CO} of 52% five years before the diagnosis of PAH or a FVC/DL_{CO} ratio of 1.6 was a predictor of future PAH. In Steen's study, the majority of patients met this criterion for RHC. In another study by Allanore *et al*. (28), baseline elevated NT-proBNP and low DL_{CO} were associated with developing PAH during follow-up (hazard ratio: 47).

Our study has important limitations. Although we were able to recruit patients with mild functional impairment, we did not capture the method used for assignment of a PAH diagnosis without RHC. In addition, because ascertaining information regarding treatment, hospitalisations, and causes of death were not purposes of this study, these data were not recorded. Furthermore, the study did not assess the reasons for nonadherence to the criteria for RHC; however, this information would have been useful so that future studies could address issues of non-adherence to the criteria for RHC.

Conclusions

The ultimate goal of the QuERI-SSc initiative was to close the gap between recommended and actual care in the diagnosis and management of PAH in patients with SSc. In this study, the effort had mixed results. On the one hand, the diagnostic algorithm was associated with the recruitment of milder functional impairments. On the other hand, significant gaps in the diagnosis of PAH in patients with SSc still exist in the rheumatology community. These gaps include underuse of RHC and other diagnostic criteria such as physical examination, NT-proBNP assays, and appropriate use and reporting of echocardiography. Thus, a call to action is necessary to implement interdisciplinary teamwork and education between rheumatologists, pulmonologists, and cardiologists. A better understanding and stricter adherence to the published guidelines could improve the quality of care and outcomes in high-risk patients with SSc (30).

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