Defining quality standards of care in CTD-PAH and management best practices: a Delphi panel consensus

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Abstract

Objective: This study aimed to develop a consensus of expert opinion on the quality standards of care and outline management best practices for pulmonary arterial hypertension (PAH) in systemic sclerosis (SSc) and other connective tissue diseases (CTDs).

Methods: Twenty physicians and four nurse practitioners (NPs) managing patients with CTD-PAH (including SSc-PAH) from six European countries participated in a modified Delphi panel. Consensus was defined as ≥80% agreement among the physicians.

Results: The importance of a multi-modal approach to screening and early detection of PAH, through combining echocardiography, biomarker tests and symptom evaluation, was emphasised. Consensus agreement was also reached on the usefulness and increased access of echocardiography for optimising screening and early detection processes, noting the importance of cardiology expertise for accurate assessment of right-heart variables. Panellists reached consensus agreement on the importance of a multi-disciplinary approach to managing SSc- and other CTD-PAH, through collaboration between rheumatologists and pulmonary hypertension (PH) specialists, NPs, and patients. Rheumatologists aligned on the usefulness of the DETECT screening algorithm, whereas cardiologists and pulmonologists remained divided, with consensus not being reached, thus highlighting the lack of agreement regarding the relevance of a stepwise approach. Similarly, no consensus was reached on the impact of the new haemodynamic definition of PH in CTD-PAH management, where greater evidence is required.

Conclusions: Consensus was reached on key recommendations for optimising CTD-PAH management, including the standardisation of multi-modal screening and promotion of closer collaboration between healthcare specialties and patients.

Lay summary

What does this mean for patients?

Pulmonary arterial hypertension (PAH) is a serious, long-term disease resulting in the arteries of the lungs narrowing, which can lead to right heart failure and death. PAH is often linked to connective tissue diseases (CTDs), such as systemic sclerosis (SSc). CTD-PAH is a complex disease that can be difficult to detect at an early stage due to the gradual onset of non-specific symptoms and limitations of routine screening tools. To explore how care could be improved, 24 experts (doctors and nurse practitioners) from six European countries reviewed current practices and provided recommendations for optimising CTD-PAH management. The experts agreed that screening for PAH should use a combination of tests, including echocardiography, blood biomarkers, and symptom monitoring. They also emphasised the involvement of heart specialists to interpret heart scans accurately. A team-based approach to care was strongly supported, where rheumatologists, lung and heart specialists, nurse practitioners, and patients work closely together. These recommendations could help ensure that patients with CTDs receive faster, more accurate PAH diagnoses and more coordinated, personalised care. This study also highlighted where further research and guidance is needed for improved patient management.

Keywords: Pulmonary arterial hypertension, systemic sclerosis, connective tissue disease, management, consensus

Key messages:

- The importance of a multi-modal approach to screening and early detection of PAH was emphasised.
- Improved patient management and outcomes are supported by healthcare specialties working synergistically.
- Greater evidence on the impact of the new haemodynamic definition of PH on management is required.

Introduction

Pulmonary arterial hypertension (PAH) is a chronic, progressive, and life-threatening disease characterised by increased pulmonary arterial pressure, leading to right ventricular failure.(1,2) Connective tissue diseases (CTDs) are the most common comorbidity associated with PAH, accounting for approximately 25% of the PAH patient population.(1,3)

Systemic Sclerosis (SSc) accounts for the majority of cases of CTD-PAH in Europe and North America, followed by Systemic Lupus Erythematosus (SLE) and Mixed CTD (MCTD).(1,4) A meta-analysis in SSc reported a PAH prevalence of 6.4% and incidence of 18.2 cases per 1,000 person-years.(5) Patients with CTD-PAH have both a poorer prognosis and overall survival.(6,7) To mitigate late diagnosis and prompt earlier intervention, there is a need to adopt more effective screening programmes, supported by evidence-based guidance.(8)

The most recent guidelines (2022 European Society of Cardiology [ESC] and European Respiratory Society [ERS]) highlight the importance of timely and accurate screening and early detection, referral, diagnosis, treatment, and follow-up of patients with PAH.(7) By understanding current standards of care (SoC) in CTD-PAH, opportunities for optimising disease management in clinical practice may emerge.(9-11) Furthermore, the implications of the new haemodynamic definition of PH on healthcare decision-making for CTD-PAH remain uncertain and should be further explored to outline areas where additional guidance is necessary.(7,12)

Given the complex nature of CTD-PAH, disease management requires a multidisciplinary approach, and shared decision-making with patients.(13) In recent years, the value of a collaborative approach when managing CTD-PAH is increasingly recognised.(7) Several studies have demonstrated that working within multi-disciplinary teams (MDTs) improves the efficiency and accuracy of patient management, resulting in improved diagnostic accuracy, treatment decisions, and patient outcomes.(14-16) A better understanding of current collaboration between rheumatology and PAH specialists could identify strategies and best practices to optimise the management of CTD-PAH, facilitating earlier detection and intervention, leading to improved patient outcomes.

The Delphi panel methodology was used to identify areas for improvement when considering existing CTD-PAH (with a focus on SSc) management and to provide expert guidance on management best practices. Delphi panels are a well-established, appraised methodology for eliciting consensus where evidence is limited and have been used within the fields of PAH(17-19) and CTD(20,21) previously. This Delphi panel also aimed to inform best practices for collaborative clinical management processes between rheumatology and PAH specialists, beyond diagnosis, to improve efficiencies for both patients and physicians.

Methods

Modified Delphi methodology

A modified Delphi panel methodology was adopted for the management of SSc- and other CTD-PAH, using an expert panel of physicians and nurse practitioners (NPs) specialising in cardiology, pulmonology, and rheumatology. The modified approach adapts the conventional Delphi technique by introducing a final consensus meeting after the initial two survey rounds (Figure 1).(22) Panellists were required to provide consent for participation at each round of the study.

Panellists

A Steering Committee provided suggestions regarding potential panellists to participate in the study, supplemented through free-found recruitment via a third-party agency. The Delphi panel consisted of healthcare professionals from Belgium, France, Germany, Italy, Spain, and the United Kingdom (UK).

Physicians and NPs were recruited according to their healthcare expertise, years of experience managing CTD-PAH, patient caseload, and familiarity with the 2022 ESC/ERS guidelines.

Data collection and analysis

Surveys were developed in Microsoft Word, programmed using the SogoSurvey™ platform, and links were distributed via email by a third-party agency.

Qualitative and quantitative responses were collected from the panel. Qualitative insights from free-text responses captured the panel's experience and opinions, while quantitative data gathered through rating scales, ranking, and numerical inputs, were leveraged to elicit consensus.

Consensus was defined as ≥80% of panellists in 'agreement' (rated between 7-9) or 'disagreement' (rated between 1-3) based upon a nine-point Likert scale. Where consensus was reached in the first-round survey, statements were excluded from the second round. Where statements neared consensus**,** these were re-presented against individual and group mean responses to encourage consensus by group norming, and rationales for answers further explored.

A final consensus meeting, comprising physicians only, was held via web-conference to discuss consolidated responses and provide final conclusions and recommendations on management best practices for CTD-PAH.

Ethical compliance

This research is based solely on the opinions of a group of experts, as part of a market research activity, which falls outside the remit of the governance arrangements for research ethics committees. No patient data or information were collected for the purpose of this study. Therefore, this study did not require ethical committee approval. The study was conducted in accordance with ethical principles that have their origin in the Declaration of Helsinki and that are consistent with Good Pharmacoepidemiology Practices and applicable laws and regulations of the EU, as appropriate. The study remained compliant with the General Data Protection Regulation (GDPR).

Limiting study bias

The study sponsor, investigators, and panellists remained double-blinded throughout the conduct of the study to mitigate bias during analysis and reporting of results. To maintain blinding, a third-party agency managed the participant screening, recruitment and distribution of study materials to panellists.

Results

A total of 20 physicians and four NPs were included in this study and completed the first-round survey (**Table 1**). Nineteen physicians and four NPs completed the second round, while 11 physicians participated in the consensus meeting. Inclusion in the consensus meeting was dictated by physicians' availability. NPs were only able to be recruited from Italy and the UK, and due to the small sample size, consensus was not sought amongst this group. Therefore, consensus was solely determined by physician responses, and insights captured during survey rounds from NPs supplemented and validated physician responses.

Consensus on management of SSc- and other CTD-PAH

In accordance with the 2022 ESC/ERS guidelines, key recommendations on the quality SoC in patients with SSc- and other CTD-PAH from this Delphi panel are highlighted in Figure 2 and described in detail below.

Screening and early detection of PAH in SSc and other CTDs

Across 69 key statements, 27 (39%) reached consensus agreement (**Supplementary Table S1-11**).

Guidelines for screening and early detection of PAH

The panel agreed that the 2022 ESC/ERS guidelines for screening and early detection of PAH in SSc are easy to implement into clinical practice; however, consensus was not reached for the same statement for CTDs other than SSc. Screening was recommended for SSc; the panel noted that additional guidance for screening and early detection of PAH in patients with MCTD, particularly those with features of SSc, would also be useful. Annual screening in MCTD was recommended, as for SSc, when considering patients presenting with ribonucleoprotein (RNP) antibodies and vascular components (e.g., abnormal capillaroscopy).

Screening and early detection tools for PAH

When rating the usefulness of seven tools used for screening and early detection of PAH, the panel only reached agreement for echocardiography. Other tools, such as biomarker tests, were considered useful and cost-effective but non-specific for early detection of PAH when used in isolation. The importance of a multi-modal approach to screening and early detection was emphasised; combining the application of echocardiography, biomarker tests, and symptom evaluation was considered by some panellists to be superior to echocardiography alone. The panel stressed that the accuracy of echocardiography interpretation is contingent on the availability and quality of cardiologist expertise. Greater standardisation in approach and reporting of echocardiography outcomes, conducted by PAH specialist cardiologists, was identified as key for

mitigating late and inaccurate PAH diagnosis. The panel additionally highlighted that strong collaboration across healthcare specialties is critical when determining next steps for disease management.

When exploring optimisation strategies for improving timely screening and early detection of PAH, consensus agreement was reached on the implementation of an automated reminder system for SSc, and a flowchart on echocardiography outlining what to do, when to do it, and how to interpret results. The latter strategy was considered particularly valuable for any managing rheumatologist, non-PH specialist, or those with limited PAH awareness. Other statements reaching consensus in agreement for optimising screening and early detection are shown in **Table 2**.

DETECT screening algorithm for PAH in SSc

Consensus was not reached on the usefulness of the DETECT algorithm for PAH screening in SSc; a dichotomy of opinion was identified regarding its applicability in clinical practice. When assessed according to speciality, DETECT was considered as 'very useful' by 100% of rheumatologists, compared to only 55% of cardiologists and pulmonologists. While some panellists highlighted that the accuracy of DETECT is superior to echocardiography alone, it was countered that the two-step process of DETECT could be time-consuming and impractical for everyday clinical settings, thus limiting its use. The panel highlighted that as annual echocardiography is standard for all patients with SSc, regardless of PH suspicion, the step-by-step procedure may be of limited relevance and may introduce delays in care.

Impact of concomitant PAH drug use on screening and early detection

The panel identified a need for further evidence and guidance on the use of concomitant PAH drugs for other conditions, prior to developing PAH, in patients with SSc and other CTDs. Throughout the study, the panel remained divided on whether concomitant PAH drug use may mask PAH detection. It was agreed that screening and early detection approaches should not be altered based on concomitant PAH drug use for other conditions.

Referral of patients with SSc and other CTDs

Across 18 key statements, 8 (44%) reached consensus agreement (**Supplementary Table S2**).

The panel did not reach agreement on whether referrals for PAH evaluation are delayed, noting that this was dependent on the level and quality of communication between managing rheumatology and PAH centres. Consensus agreement was reached on the usefulness of shared protocols across multi-disciplinary networks to improve the referral for PAH evaluation. However, agreement was not reached on whether Patient Advocacy Group (PAG) recognition of such protocols would be beneficial. This contrasted to the opinions of the NPs, who supported this approach. The proposed strategies for improving the coordination of MDTs at referral that reached consensus agreement are summarised in

Table 3.

Diagnosis of PAH in SSc and other CTDs

Across four key statements, three (75%) reached consensus agreement (**Supplementary Table S3**).

Echocardiography, pulmonary function tests (PFTs), and right-heart catheterisation (RHC) were agreed as the most useful tools for PAH diagnosis in SSc and other CTDs. Conversely, no consensus was reached on the usefulness of imaging tools, such as computed tomography (CT), magnetic resonance imaging (MRI) or ventilation perfusion (V/Q) scans, for the diagnosis of PAH.

Consensus was not reached for directly referring patients for RHC in the interest of timely and accurate diagnosis, as RHC should be dependent on the individual's disease profile and resources available.

Treatment decision-making in SSc- and other CTD-PAH

Across 19 key statements, 11 (63%) reached consensus agreement (**Supplementary Table S4**).

Multi-disciplinary management

The panel agreed that PAH-related treatment decisions should be delegated to the PH specialist and always disclosed to the managing rheumatologist. Similarly, patients should also remain informed and involved in their own healthcare decision-making, supporting a holistic approach to disease management. While the involvement of NPs in treatment decision-making did not reach consensus agreement, their role in patient education, organisation, and treatment monitoring was acknowledged.

Treating PAH in SSc and other CTDs in patients with other conditions

The panel remained uncertain on the management of SSc- and other CTD-PAH in patients with other conditions. Although the impact of other conditions, such as cardio-respiratory comorbidities, on treatment decisions and consideration of risk-status and disease phenotype in the guidelines was acknowledged, no specific recommendations could be made for the management of patient subgroups with comorbidities. Instead, the panel highlighted that management of such patients requires a nuanced, patient-centred approach and close monitoring is required to mitigate adverse patient outcomes.

Impact of the new haemodynamic definition of PH

None of the six key statements exploring the impact of the new haemodynamic definition of PH on management of PAH in SSc and other CTDs reached consensus in agreement.

The panel did not reach consensus on whether the new haemodynamic definition of PH would prompt earlier treatment in SSc- and other CTD-PAH. While earlier treatment was considered possible, it is not recommended based on current guidance. Additionally, close monitoring of patients with a mean pulmonary arterial pressure (mPAP) of 21–24 mm Hg is required for early treatment to be considered on a case-by-case basis. Among patients who do not receive early treatment, 53% of panellists suggested re-evaluation (with echocardiography) every 6 months to assess disease progression. The panel indicated they were hesitant to apply the new definition of PH to the DETECT screening algorithm in clinical practice, raising concerns regarding potential loss of sensitivity.

Follow-up of patients with SSc- and other CTD-PAH

Across 30 key statements, 12 (40%) reached consensus agreement (**Supplementary Table S5**).

The panel agreed with follow-up evaluation every 3–6 months (at a minimum) after diagnosis, as per the 2022 ESC/ERS guidelines, although this should remain flexible according to the patient's condition, disease stability, and resources available. Continued communication between MDTs and patients following diagnosis remained important for ensuring timely intervention and management upon disease worsening or progression. Of the ten proposed strategies for optimising current follow-up processes, 40% were agreed as 'very useful' in clinical practice.

Discussion

The overarching aim of this Delphi panel was to establish expert consensus on the acceptability and feasibility of best practice recommendations for CTD-PAH screening and management. Findings indicated strong agreement on the importance of timely and accurate screening, early detection, and diagnostic processes for PAH in CTD. Key areas for optimising current management practices were identified.

Most panellists were aware that a multi-modal approach to screening and early detection of PAH is superior to echocardiography alone. Leveraging echocardiography, alongside biomarker measurements, haemodynamic assessments, and symptom evaluation, supports timely and accurate diagnosis for earlier intervention and improved patient outcomes.(23) Echocardiography is universally recognised as important for PAH screening and early detection, however, it lacks specificity. Therefore, echocardiography is not sufficient as a standalone for diagnosis and, hence, RHC is required, which is considered the "gold standard" for diagnosis of PAH.(7) A lack of consensus on the usefulness of other screening tools reflects heterogeneity in panellists' expertise and preferences towards specific tools and highlights a need for greater awareness of their value. To maximise the informative value of right-heart echocardiography results, correct expertise is understood to be crucial. Physicians should comprehensively assess and report key echocardiographic parameters, including (but not limited to) the tricuspid regurgitation velocity, tricuspid annular plane systolic excursion to systolic pulmonary arterial pressure ratio, and right atrial area.

The importance of leveraging several tools for PAH diagnosis in SSc and other CTDs (specifically echocardiography, PFT, and RHC) was highlighted. While consensus on the usefulness of individual imaging tools, including CT, MRI, and V/Q scans, was not met, their role in a multi-modal approach to diagnosis, and monitoring of disease progression is clearly relevant in CTD-PAH.(24-26) The use of these imaging tools would support the mitigation of late referral and diagnosis, and should be applied to best practices as per 2022 ESC/ERS guidance.(7,27) Timely and accurate echocardiography assessment supports CTD-PAH management and provides greater holistic management of patients with other comorbidities. Echocardiography should be repeated with any new or worsening of symptoms, or at the first positive DETECT screen.

A suggested approach for standardising SoC was the development of a best practice screening and early detection flowchart for echocardiography (Figure 2). For example, in rheumatology, a fast-track algorithm for discerning non-scleroderma from scleroderma patterns through capillaroscopy was developed by the EULAR Study Group on Microcirculation in Rheumatic Diseases, supporting assessment by clinicians of any experience.(28) Similar guidance could assist shortening PAH diagnosis pathways, reducing inconsistencies in the measurement and interpretation of variables, and improving patient experiences. Another potential strategy for optimising current SoC is the implementation of an automated reminder system, to support physicians identifying 'at risk' patients for screening. This tool would be most impactful if it considered key patient details, such stage of disease, to identify suitable candidates for RHC. Similar systems have promoted timely screening in alternative indications, and may encourage further collaboration between rheumatology and PAH expert centres.(29,30)

The DETECT screening algorithm for PAH is a proven tool for early detection of PAH in SSc, comprised of a two-step process, first evaluating non-echocardiographic variables, then echocardiography based on the determined risk of PAH.(31) DETECT's accuracy was recognised by those with experience in using the tool (rheumatologists) to be superior to echocardiography alone, however the usefulness of DETECT was questioned. Since annual echocardiography is standard for all patients with SSc, some panellists felt that the two-step process was time-consuming and impractical for real-world practice. The dichotomy of opinion (where 100% of rheumatologists versus only 55% of cardiologists and pulmonologists rating DETECT as 'very useful') likely reflects heterogeneity in their real-world clinical practice, with rheumatologists primarily assessing symptomatic and asymptomatic patients for screening and cardiologists/ pulmonologists focusing on diagnosis. The utility of DETECT in clinical practice may also be influenced by the attitudes and awareness of managing physicians, as well as expertise and tools available.(32) When considering the new haemodynamic definition of PH, the panel remained hesitant to apply DETECT processes, due to concerns on the potential loss of accuracy. Recent evidence has demonstrated that the accuracy of DETECT, when applied to the new haemodynamic definition, is maintained.(33) The panel's lack of consensus on the impact of the new definition on disease management highlights the need for greater awareness from healthcare professionals.

A recurrent theme throughout the Delphi panel was the importance of a multi-disciplinary approach, alongside increased patient involvement, in the management of PAH in SSc and other CTDs. The quality of care and clinical outcome for patients with SSc- and other CTD-PAH is contingent on healthcare specialties working synergistically with one another, as well as the level and quality of communication shared. Strong collaboration between managing physicians and patients can improve quality of care, patient satisfaction, therapeutic compliance, and facilitate earlier diagnosis and intervention, leading to improved patient outcomes.(7,34-37) Furthermore, the importance of engaging NPs in MDTs is often overlooked in clinical practice, reflected by a dichotomy of views

within this panel. Despite these differences of opinion, the role of dedicated NPs can be extensive, from coordination of care, patient advocacy, education, and being the primary point of contact for patients.(38) Utilising NP's expertise in clinical practice may improve the quality of care received, as well as patient satisfaction, adherence, and enhanced patient outcomes.

As key stakeholders in disease management, patients should be made to feel empowered to have a meaningful role in setting the SoC they receive.(38) While the importance of the patient voice in disease awareness, care, and research is increasingly acknowledged, patient involvement is often overlooked in healthcare decisions.(13,39) Although consensus was not met for PAG recognition of MDT protocols, the value of the patient voice in the management of PAH is clearly demonstrated by the involvement of PAG representatives in the development of the 2022 ESC/ERS guidelines.(7) Providing a comprehensive educational package to patients, upon diagnosis of SSc and other CTDs, would support their role in prompting timely screening and early detection of PAH.

The complexity of effectively managing PAH in patients with SSc and other CTDs is reflected in the poor prognosis and overall survival of patients.(6,7) Patients presenting with comorbidities, and/or already receiving concomitant drugs that can treat PAH, introduce further complexity in management decision-making for SSc- and other CTD-PAH. The panel discussed that, regardless of comorbidities or concomitant drug use, slowing disease progression and improving patient outcomes should be the priority, compared to an overly cautious approach. Maintaining a rigorous approach to the management of PAH in patients with SSc and other CTDs may support improved patient prognosis and outcomes. Although this study focused on optimising PAH management strategies in SSc and other CTDs, it should be acknowledged that therapeutic requirements vary across CTD subtypes. Future research should explore how treatment options can be effectively integrated into comprehensive CTD-PAH management pathways to validate these recommendations in clinical practice.

There were some limitations to this study. Firstly, the influence of peer pressure may have introduced bias, however maintaining anonymity for participants mitigated this risk. Secondly, as the study was conducted with 24 participants across six countries, subjectivity and generalisation should be considered. However, the study involved an equal balance of participants across countries, ensuring sufficient representation across Europe and range of opinion. Furthermore, as this study relied solely on expert opinion, findings must be validated through clinical research to determine applicability in real-life healthcare settings. Lastly, due to challenges in recruitment for NPs, this sample was too small to conclude consensus; results cannot also be considered widely generalisable as NPs were only recruited from Italy and the UK.

Conclusion

This Delphi panel reached consensus on several aspects of CTD-PAH management, highlighting key recommendations for improving the quality SoC , including the standardisation of a multi-modal screening approach, improved synergy between managing healthcare specialties, and increased recognition on the value of the patient voice and NP involvement, as well as for PAH expert cardiologists to conduct right-heart echocardiography. These findings can support the optimisation of future management best practices, to improve clinical efficiencies and patient outcomes.

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Tables/Figures

Table 1. Summary of panellist characteristics

Criteria		Number of panellists (n)
Physicians (n=20)		
Specialty area	**Pulmonology**	7
	Cardiology	6
	Rheumatology	7
Country	**Belgium**	2
	France	4
	Germany	3
	Italy	4
	Spain	3
	UK	4
NPs (n=4)		
Specialty area	**Pulmonology**	1
	Cardiology	1
	Rheumatology	1
	Cardio-pulmonology*	1
Country†	**Italy**	1
	UK	3

NP: Nurse practitioner; UK: United Kingdom.
*One nurse practitioner selected both cardiology and pulmonology.
†NPs from Germany were not recruited due to restrictions on pharmaceutical companies contacting nurses.

Table 2. Statements for improving screening and early detection processes reaching consensus agreement (≥80%)

Category	Statement	Degree of agreement (%)	Stage consensus reached
Population	The following factors are very important to trigger case findings in patients with SSc: ✓ Clinical subset of limited cutaneous SSc (lcSSc) ✓ Disease-specific profile of antibodies (e.g. anti-centromere (ACA), RNA polymerase III (RNAP3), Fibrillarin- Th/To) ✓ High FVC%/ DLCO% ratio >1.6 ✓ Isolated reduction in DLCO	100	Consensus meeting
Screening tools	✓ Assessing the echocardiography TAPSE/sPAP ratio variable, would improve the timely and accurate detection of PAH in SSc, if the necessary expertise were available	100	Consensus meeting
	✓ Treatment escalation is required in patients with SSc and other CTDs, who are already treated with CCBs or other PAH drugs for Raynaud's phenomenon or digital ulcers and develop PAH	84	Second round
Treatment	✓ 2022 ESC/ERS guidelines do not adequately address the use of concomitant PAH drugs in patients with SSc and other CTDs, prior to a formal PAH diagnosis, during the screening and early detection process	100	Consensus meeting

ACA: anti-centromere; CCB: calcium channel blocker; CTD: connective tissue disease; DLCO: diffusing capacity for carbon monoxide; FVC: forced vital capacity; LcSSc: limited cutaneous systemic sclerosis; MDT: multi-disciplinary team; PAH: pulmonary arterial hypertension; RNA: ribonucleic acid; sPAP: systolic pulmonary artery pressure; SSc: systemic sclerosis; TAPSE: tricuspid annular plane systolic excursion.

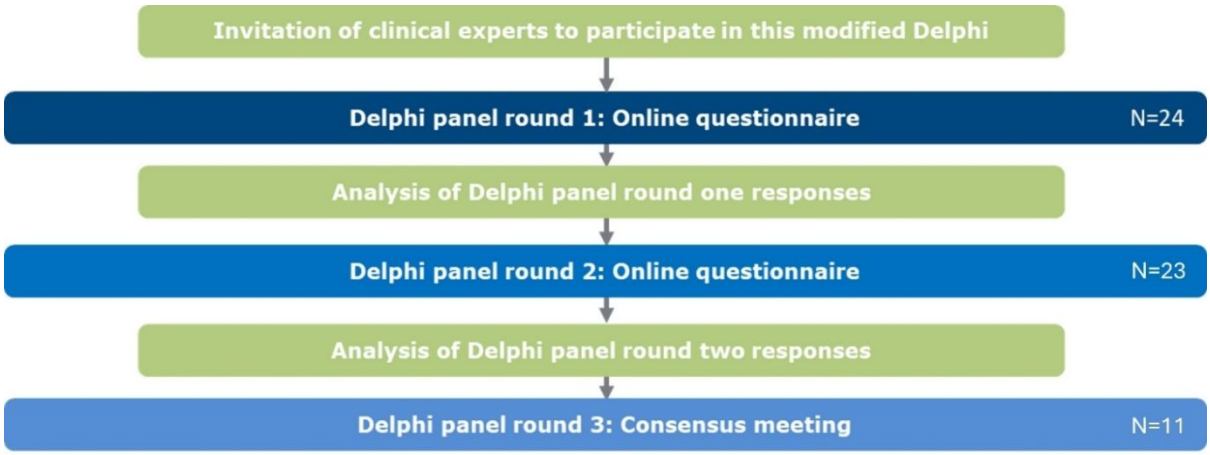
Table 3. Strategies for improving the coordination of MDTs at referral reaching consensus in agreement (≥80%)

Statement	Degree of agreement (%)	Stage consensus reached
✓ Clear definition of the roles of each stakeholder in the multi- disciplinary care pathway	84	Second round
✓ The fear of losing patients at follow-up can be overcome by proper communication and definition of tasks*	95	Second round
✓ Involvement of NPs within the MDT for management of SSc- and other CTD-PAH	100	Consensus meeting
✓ Involvement of rheumatologists in the MDT for management of SSc- and other CTD-PAH	100	Consensus meeting
✓ General management of SSc and other CTDs should remain with the rheumatologist	100	Consensus meeting

CTD: connective tissue disease; MDT: multi-disciplinary team; NP: nurse practitioner; PAH: pulmonary arterial hypertension; SSc: systemic sclerosis

*Among referring physicians worried about the potential loss at follow-up or in patients being more at-risk of being lost at follow-up.

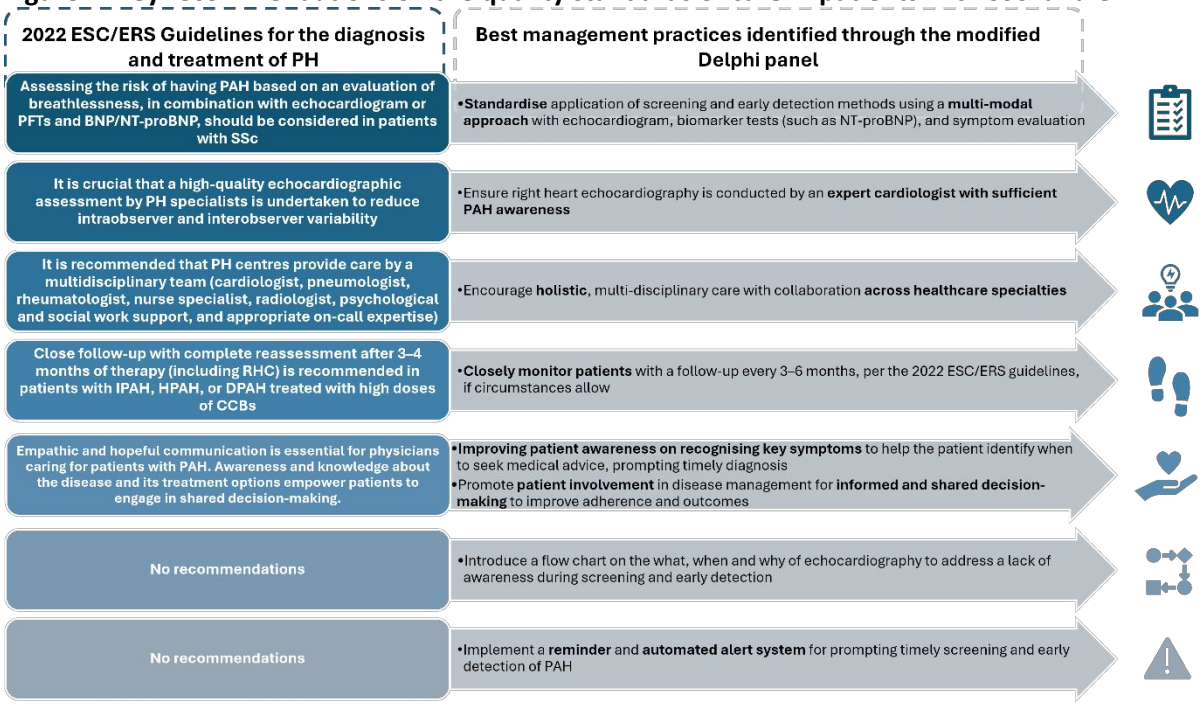
Figure 1. Overview of modified Delphi methodology



The first-round survey included 20 physicians and four nurse practitioners. The second-round survey included 19 physicians and four nurse practitioners. The consensus meeting only included physicians.

ALT TEXT: Flow diagram presenting an overview of the modified Delphi panel methodology, which comprised of two survey rounds (round 1 and 2) and a final consensus meeting (round 3), and the number of participants at each round.

Figure 2. Key recommendations on the quality standards of care in patients with SSc- and CTD-PAH



Abbreviations: BNP: B-type natriuretic peptide; CCB: calcium channel blocker; DPAH: drug-associated pulmonary arterial hypertension; ERS: European Respiratory Society; ESC: European Society of Cardiology; IPAH: idiopathic pulmonary arterial hypertension; HPAH: heritable pulmonary arterial hypertension; NT-proBNP: N-terminal pro-B-type natriuretic peptide; PAH: pulmonary arterial hypertension; PFT: pulmonary function test; PH: pulmonary hypertension; RHC: right-heart catheterisation; SSc: systemic sclerosis.

ALT TEXT: Diagram showing key recommendations for best standards of care in the management of patients with SSc- and CTD-PAH identified during the modified Delphi panel versus the 2022 European Society of Cardiology (ESC)/ European Respiratory Society (ERS) guidelines for the diagnosis and treatment for pulmonary hypertension.