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VIEWPOINT

Multidisciplinary team discussion: the emerging gold standard for management of cardiopulmonary complications of connective tissue disease

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Key words

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Abstract

Cardiopulmonary complications of connective tissue diseases (CTDs), particularly pulmonary arterial hypertension (PAH) and interstitial lung disease (ILD), are major determinants of morbidity and mortality. Multidisciplinary meetings may improve diagnostic accuracy and optimise treatment. We review the literature regarding multidisciplinary meetings in CTD-ILD and PAH and describe our tertiary centre experience of the role of the multidisciplinary meeting in managing CTD-PAH.

Introduction

Connective tissue diseases (CTDs) are complex autoimmune conditions with multiple potential cardiopulmonary complications, particularly pulmonary arterial hypertension (PAH) and interstitial lung disease (ILD). Cardiopulmonary complications often determine prognosis and treatment decisions.^{1–3} In systemic sclerosis (SSc), PAH is implicated in up to 30% of deaths,⁴ and optimal management improves outcomes.⁵ PAH diagnosis can be challenging, with long delays between symptom onset and diagnosis.⁶ Some CTDs exhibit a high prevalence of cardiopulmonary complications, warranting screening of asymptomatic cohorts for PAH and ILD⁷ to facilitate early detection and,

thus, early treatment to prevent or delay progression.^{8,9} PAH screening in SSc is recommended annually with transthoracic echocardiography and pulmonary function testing (PFT),¹⁰ with right heart catheterisation (RHC) to confirm a diagnosis if these are abnormal. Multiple algorithms exist to guide the clinician regarding the need for RHC, generally based on a combination of cardiac biomarkers, echocardiography, PFT and other clinical features.¹¹ PAH screening in SSc has been shown to be cost-effective¹² and improve overall survival after accounting for lead-time bias.^{7,13}

Multidisciplinary meetings are the accepted gold standard for the diagnosis and management of many complex conditions. In oncology, multidisciplinary discussions improve survival.^{14,15} In ILD, multidisciplinary meetings are critical in establishing a consensus diagnosis and are a requirement prior to prescribing antifibrotic therapy in Australia.¹⁶ The important role of multidisciplinary meetings is also increasingly recognised in pulmonary hypertension.^{17,18} Pulmonary hypertension may occur as primary, pre-capillary PAH or secondary to other cardiopulmonary disease. Optimal treatment depends on the dominant aetiology of pulmonary hypertension, which

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may be complex if other cardiopulmonary CTD complications coexist (particularly left heart disease and ILD). Key considerations ideally suited to multidisciplinary discussion include interpreting investigation results to identify pre-capillary PAH-dominant disease from other causes of pulmonary hypertension,^{19,20} determining the optimal timing of invasive investigations^{19,20} and refining treatment/prognostic decisions²¹ including the risk benefit of pulmonary vasodilators. This review describes the complexity and challenges of diagnosing and managing CTD-associated pulmonary hypertension, current knowledge of the impact of multidisciplinary meetings in pulmonary hypertension and a tertiary-centre experience of CTD-pulmonary hypertension multidisciplinary meetings.

Diagnosis and classification

Establishing the dominant driver of both pulmonary hypertension and a person's symptomatology is essential to treat optimally CTD-associated pulmonary hypertension. However, this can be challenging because multiple potential contributors to pulmonary hypertension may coexist in CTDs. In SSc, up to 75% of individuals will demonstrate ILD on chest high-resolution computed tomography (HRCT).^{22,23} Data from the Australian Scleroderma Cohort study showed that 22% of 479 people with SSc-ILD also fulfilled RHC criteria for pulmonary hypertension.²⁴ Furthermore, concurrent left heart disease is common in SSc (e.g. left ventricular diastolic dysfunction in 20–30%^{25–27}), which can contribute to both the development of pulmonary hypertension and breathlessness.^{25–27} Chronic thromboembolic pulmonary hypertension (CTEPH) may also occur because of concurrent antiphospholipid syndrome.²⁸ Accordingly, approximately 60–70% of pulmonary hypertension in SSc is considered World Society of Pulmonary Hypertension Group I PAH, around 20% Group III pulmonary hypertension because of hypoxaemic lung disease and a further 10–20% Group II pulmonary hypertension because of cardiac disease^{29,30} (Fig. 1).

Additionally, deconditioning and inactivity, ILD, left heart disease and other CTD manifestations (e.g. myositis or anaemia) may all contribute to worsening dyspnoea and exercise intolerance. Thus, individuals with SSc and pulmonary hypertension may have multiple other concurrent cardiopulmonary diseases, especially in older persons, given that PAH is often a late complication of SSc. It is therefore challenging to ascertain the degree to which both symptoms and changing haemodynamic parameters are caused by pulmonary vascular disease or another CTD feature.

Accordingly, a comprehensive cardiopulmonary assessment is required to identify potential contributors to both pulmonary hypertension and breathlessness in CTDs.

Multidisciplinary meetings are ideally suited to these complex diagnostic and therapeutic dilemmas. In ILD, multidisciplinary meetings can improve diagnostic confidence^{31,32} and accuracy.²¹ ILD diagnosis changes in 41–53% of patients after multidisciplinary discussion,^{33,34} with increases in CTD-associated ILD and hypersensitivity pneumonitis³³ and reductions in unclassifiable ILD.³³ In one study, over 60% of those labelled as idiopathic pulmonary fibrosis (IPF) were reclassified, thereby altering management and prognosis.³⁴ Furthermore, multidisciplinary review of clinical information and imaging may reduce the requirement for surgical lung biopsy for ILD diagnosis and, thus, potential surgical complications.³³

While there is a paucity of data exploring the role of multidisciplinary discussion in pulmonary hypertension, there are multiple potential benefits. In SSc-associated pulmonary hypertension, multidisciplinary discussions have been shown to be less sensitive but more specific than diagnostic algorithms.¹¹ Thus, through the more targeted use of RHC,¹¹ multidisciplinary discussions may reduce healthcare costs. Furthermore, multidisciplinary discussions allow multi-speciality perspectives to establish the severity and trajectory of each disease manifestation. For example, is there radiographic progression of ILD to explain worsening dyspnoea? Is the trend of the diffusing capacity for carbon monoxide out of keeping with spirometry results suggesting pulmonary vascular dysfunction? Is the degree of left heart disease adequate to explain a rising right ventricular systolic pressure? These are complex assessments ideally suited to multidisciplinary discussions, to facilitate both accurate classification of pulmonary hypertension and symptom attribution.¹⁷

Treatment optimisation

Pulmonary vasodilator treatment may be expensive, invasive and complicated by adverse effects.^{17,35} Limited evidence exists to guide decision-making in 'overlap' situations of concurrent ILD and pulmonary hypertension, particularly in CTD cohorts where there are few randomised trials, and concerns about the generalisability of evidence from non-CTD populations. In ILD without pulmonary hypertension, endothelin receptor antagonists show no benefit in treating ILD.^{36,37} Ambrisentan was associated with disease progression in ILD without pulmonary hypertension.³⁸ In idiopathic interstitial pneumonia and pulmonary hypertension, riociguat was associated with clinical worsening.³⁹ In concurrent ILD and pulmonary hypertension, pulmonary vasodilator treatment may improve haemodynamics,⁴⁰ but little evidence supports benefit in symptoms or quality of life.⁴¹

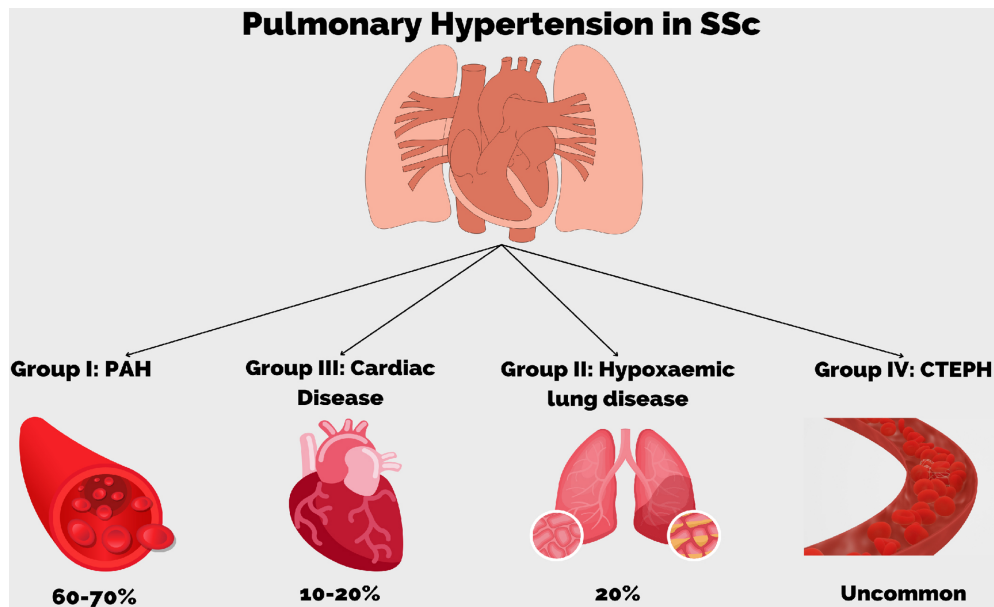


Figure 1 Possible contributors to pulmonary hypertension in SSc. CTEPH, chronic thromboembolic pulmonary hypertension; PAH, pulmonary arterial hypertension; SSc, systemic sclerosis.

In concurrent emphysema and pulmonary hypertension, pulmonary vasodilators may worsen ventilation-perfusion mismatch and exacerbate hypoxia,⁴² although this has not been demonstrated in ILD-associated pulmonary hypertension.⁴³ Accordingly, treatment decisions in those with pulmonary hypertension and ILD are complex and depend on accurate classification of pulmonary hypertension.¹⁷ The 2022 European Society of Cardiology/European Respiratory Society guidelines recommend that patients are referred to a pulmonary hypertension centre for individualised decision-making.¹⁷

ILD-multidisciplinary team (MDT) meetings have been shown to help refine treatment decisions²¹ and improve adherence to guideline-based management, including increased use of non-corticosteroid immunosuppression, antifibrotic agents and supplemental oxygen, and reduce corticosteroid prescription.³³ Pulmonary vasodilator use in ILD was increased after the MDT discussion, underscoring their utility in complex decision-making.³³ ILD-MDT meetings also increased referral to clinical trials,³³ thereby helping to bridge differences in access between centres. In CTEPH, rates of RHC and pulmonary endarterectomy were significantly increased following multidisciplinary implementation,⁴⁴ indicating improved adherence with best-practice guidelines. It is also likely that an understanding of disease complexity and the limitations of available therapies may increase timely referral for lung transplantation, a treatment shown to increase survival in ILD.⁴⁵

MDT composition in CTDs

Optimal MDT composition varies between conditions. ILD-multidisciplinary meetings commonly involve respiratory physicians, radiologists and pathologists.²¹ Palliative care involvement has also increased advanced care planning discussions and improved access to community services.⁴⁶ Ideal PAH-MDT composition is not well described. When managing cardiopulmonary complications of CTDs with multiple possible contributors, it follows that multidisciplinary meetings should include at least a respiratory physician, cardiologist and radiologist, as well as a CTD specialist.

CTD specialists are a less common fixture in multidisciplinary meetings; only 24% of ILD-multidisciplinary meetings may involve a rheumatologist.²¹ However, evidence suggests that rheumatologists may help to tailor immunosuppression in CTD-ILD.³³ In one study, rheumatology involvement facilitated treatment modification in 80% of cases with CTD-ILD and 27% with IPF.^{21,47} Rheumatologists may also assist in re-evaluating cases and establishing a rheumatological diagnosis in 20–30% of ILD cases,⁴⁸ thereby preventing invasive investigation including bronchoscopy or lung biopsy.^{21,48} Rheumatologists are often involved in diagnosing CTD-PAH or ILD through monitoring symptom trajectory and disease progression and coordinating cardiopulmonary screening investigations. Familiarity with a person's immunomodulation, particularly hepatotoxicity and myelotoxicity, may assist in therapeutic decision-making, since some pulmonary

vasodilators share these potential toxicities. Physical function may also affect the feasibility of some treatments, for example, intravenous epoprostenol may require dexterity of hand function limited by SSc-related contractures. In cases of advanced disease, a multisystem and longitudinal perspective may assist when considering transplantation referral or symptom management, including palliative care referral. Finally, patients often have a high degree of trust in their treating rheumatologist, and so their involvement in these difficult treatment decisions may offer reassurance.⁴⁹

Impact on outcomes

The prognostic impact of multidisciplinary discussion in both PAH and ILD is poorly described. In PAH, two studies report the impact of multidisciplinary care on pregnancy outcomes in non-CTD cohorts.^{18,50} In one study, 22 women with pulmonary hypertension predominantly because of congenital heart disease were managed by a MDT composed of physician experts, obstetricians, intensivists, anaesthetists and neonatologists.¹⁸ Of these, 91.7% of women and 85.7% of newborns survived, compared to previous mortality rates of 12–56%.¹⁸ Other data demonstrate reduced maternal mortality, neonatal mortality and emergency caesarean section following MDT implementation in 103 pregnancies in women with pulmonary hypertension, despite more women having severe pulmonary hypertension in the post-implementation group.⁵⁰ In ILD, multidisciplinary meetings have improved diagnostic accuracy,³⁴ allowing both more accurate prognostication and the opportunity for earlier introduction of targeted disease-modifying therapy. In multiple cancers, multidisciplinary discussion improves survival.^{14,15} While further data are required to define the survival benefits of multidisciplinary discussions in CTD-PAH and ILD, it may be surmised that their diagnostic and therapeutic impacts would confer a better prognosis.

Our experience

Our institution is a tertiary SSc referral centre where SSc-associated PAH has been predominantly managed by rheumatologists supported by a multidisciplinary meeting for over 10 years. The MDT comprises rheumatologists specialising in SSc, cardiologists specialising in PAH, senior respiratory physicians, a palliative care physician and radiologist, as well as relevant registrars/fellows. Attending specialists devote 1–1.5 h every 6 weeks to multidisciplinary discussion of complex CTD cases. Reflecting the importance of these discussions, there is now a Medicare item number for this service. Since the COVID-19 pandemic, meetings have been conducted virtually, facilitating the

attendance of a broader range of specialists, including those from centres with less SSc experience. Patients do not attend these meetings, but outcomes are discussed with the patient by the treating rheumatologist after the meeting and recorded in the patient record.

Cases with suspected or confirmed CTD-associated pulmonary hypertension are discussed. Cases are nominated for discussion based on the discretion of the treating clinician; it is not mandated that all PAH cases be discussed in this meeting. For example, cases requiring urgent investigation may be discussed among subspecialty colleagues between meetings. A mixture of simple and complex cases is generally presented. Investigation results are reviewed, including echocardiography, PFT, chest HRCT, NT-pro-BNP and functional testing (6-min walk or cardiopulmonary exercise test). The rheumatologist describes the person's functional status, symptomatology, disease features and therapies. The cardiologist reviews the echocardiography assessing direct and indirect markers of pulmonary hypertension and cardiac function. Respiratory investigations, including chest HRCT, CT pulmonary angiography, ventilation-perfusion scanning and PFT, are then reviewed with an expert pulmonologist and thoracic radiologist. A consensus recommendation is made about proceeding to RHC, more frequent echocardiographic monitoring, or further investigation of other contributors, for example, ILD progression. Following PAH diagnosis and treatment, cases are discussed as required to assess treatment response and the need for further investigation or escalation of therapy. These meetings offer an opportunity for inter-disciplinary discussions regarding prognosis and suitability for/timing of other therapies, including clinical trials, lung transplantation or palliative care.

Such meetings are invaluable in optimising CTD-associated pulmonary hypertension management in our institution. Given the longstanding therapeutic relationship between patient and rheumatologist, the treating rheumatologist coordinating PAH care supported by a MDT has been a successful model, with assistance from cardiology and respiratory colleagues as needed, depending on the clinical scenario. For many patients, multidisciplinary discussion reduces the number of specialist appointments required. We acknowledge that a model wherein cardiologists or respiratory physicians primarily manage the patient's PAH with co-management of PAH, other CTD-associated complications and overall coordination of care by the rheumatologist could also work effectively. These meetings are also a valuable opportunity for teaching and knowledge transfer and a pathway for accelerating care for deteriorating patients. For those patients from rural and remote areas, a discussion of the practicality of coordinating

certain aspects of care (e.g. PFT or medication administration) locally is also helpful.

Conclusion

Cardiopulmonary complications of CTDs are major contributors to both morbidity and mortality. ILD-MDT meetings have been shown to improve diagnostic accuracy and alter treatment decisions. In pulmonary hypertension, potential benefits include more accurate PAH classification and optimising treatment despite limited evidence. Further research is required to quantify the benefits of multidisciplinary discussion to both patients and the health system, including enhancing therapeutic

decisions, streamlining care, optimising healthcare utilisation and cost-saving.

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