Pulmonary vascular disease for the general respiratory clinician

Brian D. Kent[1,2]

[1]Dept of Respiratory Medicine, St James' Hospital, Dublin, Ireland. [2]School of Medicine, Trinity College Dublin, Dublin, Ireland.

Corresponding author: Brian D. Kent (briankent@physicians.ie)

Shareable abstract (@ERSpublications)
Pulmonary vascular disease is relevant to many practising respiratory clinicians: ~1% of the world's population is estimated to have PH, and it is an important consequence of many other respiratory diseases (*e.g.* COPD, ILD, sleep breathing disorders) https://bit.ly/3hLqUx1

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Coinciding with the recent publication of the updated European Society of Cardiology (ESC)/European Respiratory Society (ERS) guidelines for the diagnosis and management of pulmonary hypertension (PH) [1], this issue of *Breathe* focuses on pulmonary vascular disease, from pulmonary arterial hypertension (PAH) to pulmonary vasculitides. Although sometimes considered a rather niche area of respiratory medicine, pulmonary vascular disease is highly relevant to many practising respiratory clinicians: approximately 1% of the world's population is estimated to have PH, and it is an important consequence of many other respiratory diseases, including COPD, interstitial lung disease (ILD) and sleep breathing disorders.

Characterised by the development of pulmonary vasculopathy, with intimal fibrosis, medial hypertrophy, vasoconstriction and thrombosis, PAH (World Health Organization group 1 PH) had been considered a disease that predominantly affected younger women and conferred a particularly grim prognosis [2]. As discussed by Cullivan *et al.* [3], it is now apparent that PAH is frequently diagnosed in older cohorts and in men, and is an important complication of connective tissue disease, congenital heart disease, and infectious diseases like HIV and schistosomiasis. The treatment of PAH has been transformed in the past two decades by a growing understanding of the physiological and molecular mechanisms underpinning the disease, with five classes of medication targeting different pathways now in use, and helping to dispel any sense of therapeutic nihilism that may have existed around it.

In contrast with PAH, specific therapies for most patients with PH as a consequence of lung disease or hypoxia (group 3 PH) are lacking. For example, phosphodiesterase inhibitors such as sildenafil are important agents in the management of PAH, but in group 3 PH any improvement they deliver in pulmonary haemodynamics is often offset by a worsening in gas exchange and functional status [4]. Krompa and Marino [5] discuss the identification and management of PH in chronic respiratory disease, including how current evidence suggests that care of these patients should be focussed on optimising management of the underlying respiratory disease, including use of oxygen therapy, noninvasive ventilation and pulmonary rehabilitation where appropriate. Meanwhile, recent data regarding the use of treprostinil in ILD patients with PH shows that ongoing trials of therapies specifically targeting the pulmonary vasculature remain worthwhile in patients with group 3 PH.

The bedrocks of management of chronic thromboembolic pulmonary hypertension (CTEPH; group 4 PH) are pharmacotherapy and, in selected patients, surgical pulmonary endarterectomy. However, medical therapy is not always effective in CTEPH patients, and not all patients or pulmonary vascular lesions are suitable for surgical intervention. This issue's "Landmark papers in respiratory medicine" article discusses key studies in the use of balloon pulmonary angioplasty in CTEPH, and how they have changed the management of non-operable disease [6].

PH is an important potential complication of chronic lung disease of prematurity. In a forthcoming article, S. Chan and co-workers outline what paediatric respiratory clinicians should know about PH in

children with bronchopulmonary dysplasia, emphasising the importance of the multidisciplinary team in its management.

Not all pulmonary vascular disease involves PH. Pulmonary vasculitides are uncommon, but can be devastating, and may crop up in unexpected places, like the asthma clinic or the ILD clinic. Alam and Nanzer [7] present a review of eosinophilic granulomatosis with polyangiitis (EGPA), including a discussion of how the development of eosinophil-targeting biologic agents has improved outcomes and reduced the burden of systemic corticosteroid therapy in patients with this challenging disease. In a companion article, Boyle *et al.* [8] discuss the clinical presentation, diagnosis and management of pulmonary renal syndromes, including anti-glomerular basement membrane (GBM) disease and antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitis.

In keeping with the overall theme of the issue, our forthcoming Lung function corner discusses the measurement of pulmonary haemodynamics, and we have our usual slate of content for and by our early career members, including an overview of the new ERS Clinical Practice Guidelines Methodology Network [9]. We hope you enjoy this issue of *Breathe*, and as ever, we would be delighted to hear any feedback or suggestions readers may have for future issues and content.

Conflict of interest: B.D. Kent has participated in advisory boards and/or received speaker fees from Astra Zeneca, Chiesi, GSK, Novartis, and Teva; has received educational travel bursaries from Boehringer Ingelheim, Chiesi, and Napp; and has received research funding from Itamar Medical.

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