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EDITORIAL

Pulmonary hypertension: basic concepts and practical management

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Not even 3 yrs after the *European Respiratory Journal* concluded its last series of review articles on pulmonary hypertension [1], the Journal now launches a new review series dealing with pulmonary vascular disease. Why is this?

The simple reason is that the field moves at a rapid pace, which still seems to be accelerating instead of slowing down. Therefore, both researchers and clinicians are constantly faced with new pathophysiological concepts, new drugs and new therapeutic frontiers. The field of pulmonary hypertension is large and encompasses a broad range of diseases and conditions affecting hundreds of thousands of patients worldwide [2]. For a single but rather rare form of pulmonary hypertension, called pulmonary arterial hypertension (PAH), several classes of drugs are now available. These drugs have a substantial impact on the survival of patients with PAH. For instance, 3-yr survival rates have increased from ~40% in the 1980s [3] to ~80% in current case series [4–6]. But what is the current state of knowledge in treating other forms of pulmonary hypertension that pulmonologists frequently encounter in chronic thromboembolism, obstructive lung disease or pulmonary fibrosis, among other conditions? As we are aware that many *European Respiratory Journal* (*ERJ*) readers have a special interest in pulmonary vascular disease, we felt that the time was right to summarise the latest information. The idea was to combine an overview of the current understanding of basic concepts with an update on the latest therapeutic advances. We have invited experts in the field, both from basic science and clinical practice backgrounds, to work together in summarising and commenting on the latest developments in research and medical therapy.

The series will start with an article by DUPUIS and HOEPER [7] in the current issue of the *ERJ* dealing with endothelin receptor antagonists. Two substances, bosentan and sitaxsentan, have already been approved in Europe and a third compound, ambrisentan, is expected to be available soon. As always when several players struggle for their place in a rather small market, there is proper and improper information about the advantages and disadvantages of each drug, and we hope that this article will help our readers to make their own judgements. A similar situation is currently evolving in the field of prostanoids where, next to inhaled iloprost and intravenous epoprostenol, more and more players enter the field. H. Olschewski and M. Gomberg-Maitland will review the current data in this area. Phosphodiesterase-5 inhibitors, another important class of substances that play an increasingly important role in the management of pulmonary hypertension will be covered by A. Ghofrani and M. Wilkins in the third article in this series.

These first three articles will deal mostly with the field of PAH, since the vast majority of clinical trials with these compounds have been performed in this patient population. The series will then move towards some orphan conditions, *i.e.* those forms of pulmonary hypertension for which no treatment has yet been studied in randomised controlled clinical trials, including pulmonary hypertension in patients with chronic obstructive lung disease (A. Chaouat and R. Naeije) and interstitial lung disease (J. Behr and J. Ryu). There is a strong medical need to develop treatments for these conditions but the review articles will show that it may not be prudent to translate current therapies used to treat PAH to other forms of pulmonary hypertension without appropriate clinical studies in the specific conditions.

The next two articles will address the very rare yet still important conditions, pulmonary veno-occlusive disease (D. Montani, P. Dorfmueller, F. Laenger, M.M. Hoeper and M. Humbert) and pulmonary complications of hereditary haemorrhagic telangiectasia (M. Faughnan, L. Young and J. Granton). Pulmonary veno-occlusive disease remains one of the most malignant and most difficult to treat forms of pulmonary hypertension but the understanding of this disease has substantially increased in recent years. There are now some tools that help to establish a diagnosis of pulmonary veno-occlusive disease during life-time [8], which have certainly improved the overall management of these patients. Hereditary haemorrhagic telangiectasia, another rare disease in which recent years have witnessed a tremendous improvement in the understanding of its pathogenesis, presents with various cardiopulmonary complications, some of which can be successfully managed these days.

Two other articles will cover biomarkers in pulmonary hypertension (D. Yates) and end-points in pulmonary hypertension trials (A. Peacock, R. Naeije, N. Galiè and L. Rubin).

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The series will then be concluded by a review of pulmonary vascular signalling and its implications for future treatments (A.T. Dinh-Xuan).

Our experts have been asked to provide the best currently available overview on their topics and we are certainly optimistic that they reach this ambitious goal, not least with the help of the editorial team and rigorous peer reviewers. We welcome any feedback from our readership as we stay tuned to share with you the latest developments in pulmonary vascular disease.

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