Pulmonary hypertension: a paradigm for rare pulmonary diseases

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Pulmonary hypertension has become a paradigm for a successful approach to rare diseases in the respiratory field http://ow.ly/7y3l30gN0hq

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This year's final issue of the *European Respiratory Review* (*ERR*) features a series of articles on pulmonary hypertension, selected from some of the contributions presented in March 2017 in Vienna (Austria) at the Pulmonary Hypertension Forum organised yearly by Actelion Pharmaceuticals Ltd (Allschwil, Switzerland). A reader might wonder why our journal, like many others in the respiratory and cardiological fields, dedicates so much attention to pulmonary hypertension (an important but rare condition) compared to more common diseases. The answer lies in the great advances that research has made in this field over the last two decades. These advances have been translated into a real revolution in the approach to the disease and the patients, with important improvements in prognosis and quality of life, as already highlighted in a recent editorial [1]. Thanks to the joint efforts of the medical community and of the industry, in a healthy and transparent collaboration, pulmonary hypertension has become both an example and a paradigm of a successful approach to rare diseases, with the development of new effective drugs, the involvement of patient associations, and attention raised among the scientific community, which is often focused on less complex and more widespread diseases.

In the respiratory field, another disease seems to be closely following the steps of pulmonary hypertension, namely idiopathic pulmonary fibrosis (IPF), which, together with other fibrosing lung diseases, represents a group of conditions that regularly attracts the interest of researchers and specialists, as well as of an increasingly strong and cohesive international scientific community. However, it is still too early to predict whether the results in IPF will be as successful as they have been in pulmonary hypertension.

In this issue of the *ERR*, outstanding experts from around the world discuss pulmonary hypertensive conditions. In their review, Hemnes and Humbert [2] investigate some lesser-known pathobiological pathways responsible for the development of pulmonary arterial hypertension (PAH), outside the three better-known systems of nitric oxide, endothelin and prostacyclin. Indeed, these newly recognised pathways could become targets for the development of new drugs in the near future. Chan and Rubin [3] describe the metabolic mechanisms that are known to be dysregulated in pulmonary hypertension, by exploring advances in diagnostic testing and imaging modalities currently under development to improve the capability to efficiently diagnose this disease. They also discuss emerging drugs targeting these

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metabolic pathways. Gaine and McLaughlin [4] raise the need for personalised and "holistic" approaches to patients with PAH, integrating and going beyond the mere observance of the international guidelines. Madani *et al.* [5] review the latest developments in the rapidly evolving field of chronic thromboembolic pulmonary hypertension: the improvements in imaging modalities and advances in surgical and interventional techniques, as well as the efficacy and safety of medical therapies targeted to the treatment of this condition. Finally, Bartolome *et al.* [6] tackle the difficult topic of the role of mechanical circulatory support and lung transplantation in the clinical management of patients with advanced PAH. All in all, the various contributions published in this issue contain the most up-to-date information available on pulmonary hypertensive diseases, while being dispensed and discussed by renowned experts in an original and stimulating way.

The Editor in Chief and all the staff at the *ERR* wish everyone good reading and a Happy New Year!

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