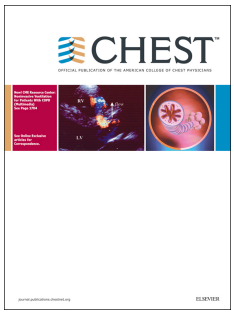
Accepted Manuscript



Pulmonary hypertension in parenchymal lung diseases: any future for new therapies?

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PII: S0012-3692(17)31077-2

DOI: 10.1016/j.chest.2017.06.008

Reference: CHEST 1158

To appear in: *CHEST*

Received Date: 4 October 2016

Revised Date: 16 May 2017

Accepted Date: 7 June 2017

Please cite this article as: Harari S, Elia D, Humbert M, Pulmonary hypertension in parenchymal lung diseases: any future for new therapies?, *CHEST* (2017), doi: 10.1016/j.chest.2017.06.008.

Pulmonary hypertension in parenchymal lung diseases: any future for new therapies?

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Conflicts of Interest: SH has relationships with Actelion, Roche, Boehringer Ingelheim and Intermune. In addition to being investigator in trials involving these companies, he is involved in lectures and is a member of scientific advisory boards. DE has nothing to disclose. MH reports personal fees from Actellion, Bayer, GSK, Pfizer and Roche.

ABSTRACT

Pulmonary hypertension (PH) due to chronic lung diseases is associated with a poor prognosis, regardless of the underlying respiratory condition. Updated PH guidelines recommend optimal treatment of the underlying lung disease, including long-term oxygen therapy, in patients with chronic hypoxaemia despite the lack of randomized controlled clinical trials supporting this statement. So far, randomized controlled trials on drugs approved for pulmonary arterial hypertension (PAH) have yielded discouraging results in both interstitial lung diseases (ILD) and chronic obstructive pulmonary diseases (COPD) with PH. In some cases, the trials were terminated because of an increase in death and other major adverse events in the active treatment arm versus placebo. In cases of PH due to idiopathic pulmonary fibrosis (IPF), new investigative therapies use a combination of novel antifibrotic treatments and other treatments approved for PAH. The choice of robust end points as well as a target group of patients with specific haemodynamic criteria may help in the selection of innovative therapeutic strategies. The aim of this review is to discuss recent studies and clinical trials for the treatment of PH due to the main chronic respiratory diseases and discuss possible future scenarios for the evaluation of new therapeutic strategies.

Introduction

Precapillary pulmonary hypertension (PH) is defined by a mean pulmonary artery pressure (mPAP) ≥ 25 mmHg at rest with a normal pulmonary capillary wedge pressure (i.e. ≤ 15 mmHg). A normal mPAP (± standard deviation) is equal to 14 ± 3 mmHg. Thus, an mPAP of 21–24 mmHg at rest is above the upper limit of normal but does not qualify for the diagnosis of PH (1). PH due to