beneficial in this setting. As stated at a recent international conference on PH, PH-specific drugs are not recommended in patients with PH associated with chronic respiratory diseases (group 3), and treatment should focus on managing the underlying disease [4]. Oxygen supplementation may stabilise PH in patients with COPD [5], and NIPPV may improve haemodynamics in patients with alveolar hypoventilation [1]. Although off-label treatment is sometimes considered in individuals with severe PH when functional limitation is considered to be related to pulmonary vascular disease, there is currently little if any evidence that PH therapy may be beneficial in this setting, with further anecdotal evidence that treatment may increase hypoxaemia through ventilation/perfusion mismatch.

We therefore strongly agree with HELD *et al.* [1] that correction of hypoventilation should be the main objective of management in chronic respiratory insufficiency, even in the presence of severe PH.

@ERSpublications
Correction of hypoventilation should be the main objective in chronic respiratory insufficiency, even with severe PH http://ow.ly/wxvcN

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Received: March 21 2014 | Accepted after revision: April 16 2014

Conflict of interest: Disclosures can be found alongside the online version of this article at erj.ersjournals.com

References

1 Held M, Walthelm J, Baron S, *et al.* Functional impact of pulmonary hypertension due to hypoventilation and changes under noninvasive ventilation. *Eur Respir J* 2014; 43: 156–165.
2 Naeije R. Pulmonary hypertension in hypoventilation syndromes. *Eur Respir J* 2014; 43: 12–15.
3 Humbert M, Sitbon O, Chaouat A, *et al.* Pulmonary arterial hypertension in France: results from a national registry. *Am J Respir Crit Care Med* 2006; 173: 1023–1030.
4 Seeger W, Adir Y, Barbera JA, *et al.* Pulmonary hypertension in chronic lung diseases. *J Am Coll Cardiol* 2013; 62: D109–D116.
5 Zielinski J, Tobiasz M, Hawrylkiewicz I, *et al.* Effects of long-term oxygen therapy on pulmonary hemodynamics in COPD patients: a 6-year prospective study. *Chest* 1998; 113: 65–70.

Eur Respir J 2014; 44: 819–821 | DOI: 10.1183/09031936.00054214 | Copyright ©ERS 2014

From the author:

I would like to thank V. Cottin and co-workers for their interest in our recently published study [1]. Their comments emphasise our conclusion that the treatment of the underlying disease should be the favoured strategy.

Since both patients did not have a reduced cardiac index, when treatment with sildenafil and bosentan was started the lack of a significant improvement was not really surprising. The two patients reported by V. Cottin and co-workers showed different reactions. Patient one deteriorated dramatically in World Health Organization (WHO) functional class and 6-min walk distance (6MWD) "despite" (or due to!) a decrease of pulmonary vascular resistance. This patient showed an excessive increase of cardiac index and a worsening of oxygenation, probably the consequence of increasing shunt perfusion resulting from reversing of vasoconstriction.

Patient two showed an early decrease of pulmonary artery pressure, no change in WHO functional class, and stable oxygenation and cardiac index, but a late improvement in 6MWD. It is questionable whether a short-term follow-up period is appropriate for patients with pulmonary hypertension due to obstructive lung disease. Patients with pulmonary hypertension and chronic obstructive pulmonary disease showed a maximum improvement of 6MWD after 8–9 months, thus, it is probable that these patients need a longer period in order to improve their functional capacity rather than decrease their pulmonary artery pressure [2].

As MEYER *et al.* [3] reported on respiratory muscle dysfunction and respiratory insufficiency in patients with idiopathic pulmonary arterial hypertension, in our daily practice we have to differentiate whether a patient presenting with severe pulmonary hypertension and hypoventilation is a patient with pulmonary

hypertension due to hypoventilation or a patient with late diagnosed pulmonary arterial hypertension and consecutive respiratory failure. The treatment is different.

Another open question is how to treat patients with hypoventilation due to hypoventilation and residual pulmonary hypertension following noninvasive positive-pressure ventilation. Is this a vascular abnormality with a prognostic impact? Perhaps a look at cardiac index might be helpful.

For patients with pulmonary hypertension and lung disease, the current statement of the international conference on pulmonary hypertension recommends to separate patients with a predominantly exhausted circulatory or ventilatory reserve [4]. This is difficult in patients with severe pulmonary hypertension and hypoventilation. As we showed, these patients present with a high minute ventilation/carbon dioxide output slope but low respiratory exchange ratio and increasing end-tidal carbon dioxide tension reflecting a pattern which is different to that seen in patients with pulmonary arterial hypertension and patients with pure ventilatory impairment [1].

V. Cottin and co-workers encourage us to treat the underlying disease as stated in our study [1]. However, we believe that an additional prospective study is needed in order to obtain a clear idea of how to manage these specific patients with residual pulmonary hypertension despite effective noninvasive positive-pressure ventilation.

@ERSpublications
Effects of PH-specific drugs on persistent PH after effective NIPPV are unclear and should be studied prospectively http://ow.ly/xnN4k

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Received: April 21 2014 | Accepted: May 25 2014

Conflict of interest: Disclosures can be found alongside the online version of this article at erj.ersjournals.com

References

1 Held M, Walthelm J, Baron S, *et al.* Functional impact of pulmonary hypertension due to hypoventilation and changes under noninvasive ventilation. *Eur Respir J* 2014; 43: 156–165.
2 Held M, Jany B. Pulmonary hypertension in COPD. *Respir Care* 2013; 58: e 86–e91.
3 Meyer FJ, Lossnitzer D, Kristen AV, *et al.* Respiratory muscle dysfunction in idiopathic pulmonary arterial hypertension. *Eur Respir J* 2005; 25: 125–130.
4 Seeger W, Adir Y, Barbera JA, *et al.* Pulmonary hypertension in chronic lung disease. *J Am Coll Cardiol* 2013; 62: D110–D115.

Eur Respir J 2014; 44: 821–822 | DOI: 10.1183/09031936.00073914 | Copyright ©ERS 2014

The cost of tuberculosis sequelae

To the Editor:

We read with great interest the article by DIEL *et al.* [1], which analysed the total average cost of tuberculosis (TB) per case for the current 27 member states of the European Union (EU-27). The average combined direct and indirect cost of TB in the original EU-15 (Austria, Belgium, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, the Netherlands, Portugal, Spain, Sweden and the UK), plus Cyprus, Malta and Slovenia (EU-18), was calculated to be €10 282 for drug-susceptible TB, €57 213 for multidrug-resistant TB (MDR-TB) and €170 744 for extensively drug-resistant TB (XDR-TB). According to differences in the gross domestic product, the total average cost in the remaining nine countries was extrapolated to be one-third of the mean cost in the EU-18: €3427 for drug-susceptible TB and €24 166 for MDR-TB/XDR-TB. When taking into account the 103 104 disability-adjusted life years caused by TB, the total cost of treating TB patients in the EU-27 in 2012 was estimated to be >€5 billion [1].

As acknowledged by DIEL *et al.* [1], there are various limitations associated with their estimation of the total cost of TB in the EU-27. One of the limitations is that although the disability weight of TB (0.271) might