Pulmonary hypertension
and cardiovascular diseases in interstitial lung disease

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AIMS

- To know that cardiovascular comorbidities are frequent in patients with idiopathic pulmonary fibrosis (IPF)
- To understand the pathophysiology of pulmonary hypertension (PH) associated to IPF
- To know the impact of PH on symptoms and outcome in patients with interstitial lung disease (ILD)
- To know the definition of PH associated to ILD
- To know the guidelines for the diagnosis and management of PH associated to ILD

SUMMARY

PH is frequent in late-stage IPF, and is associated with a shorter survival. It should be suspected in case of dyspnoea or hypoxemia disproportionate with the degree of parenchymal lung disease. It is particularly frequent in patients with the syndrome of combined pulmonary fibrosis and emphysema, and associated with a short survival (median survival less than 1 year). Pulmonary hypertension associated with chronic infiltrative lung diseases can be detected by echocardiography and must be confirmed by right-sided heart catheterization if therapeutic consequences are to be expected, especially to rule out post-capillary pulmonary hypertension frequent in this context [1].

Management is mainly palliative and based on supplemental nasal oxygen. Pulmonary transplantation should be considered in the absence of contra-indication. Therapy specific for pulmonary arterial hypertension has no demonstrated efficacy in PH associated with ILD [2]. One trial using bosentan in patients with idiopathic interstitial pneumonia and associated precapillary PH failed to demonstrate a benefit, either hemodynamic or clinical [3]. Published experience has reported often limited efficacy, and a risk of worsened hypoxemia requiring careful follow-up. However, research is currently ongoing to evaluate whether treatment approved for pulmonary arterial hypertension might be beneficial in some situations, including using newer drugs (e.g. riociguat), or subgroups of patients such as those with severe precapillary PH (mean PAP > 35 mmHg and/or cardiac index < 2.5 L/min/m² not explained by other causes).

REFERENCES


EVALUATION

1. What is the frequency of precapillary pulmonary hypertension at the diagnosis of idiopathic pulmonary fibrosis?
   - a. Less than 5%
   - b. 8-15%
   - c. 30-35%
   - d. More than 50%
   - e. No estimate is available

   Answer B

2. As compared to patients without pulmonary hypertension, those with precapillary hypertension in the setting of IPF have the following characteristics (which of the following are true?)
   - a. Increased mortality
   - b. More pronounced dyspnoea
   - c. Lower DLCO
   - d. Lower PaO2
   - e. Decreased exercise capacity

   Answer A, B, C, D, E

3. About the 2015 guidelines on the diagnosis and management of pulmonary hypertension in patients with chronic lung diseases?
   - a. Echocardiography is recommended for the non-invasive diagnostic assessment of suspected PH in patients with lung disease
   - b. Referral to an expert centre is recommended in patients with echocardiography signs of severe PH and/or right ventricular dysfunction
   - c. The optimal treatment of the underlying lung disease, including long-term O2 therapy in patients with chronic hypoxemia, is recommended in patients with PH due to lung diseases
   - d. Right heart catheterisation is not recommended for suspected PH in patients with lung diseases, unless therapeutic consequences are to be expected
   - e. The use of drugs approved for pulmonary arterial hypertension is not recommended in patients with PH due to lung diseases

   Answer: A, B, C, D, E