ESSENTIAL MESSAGES FROM ESC GUIDELINES

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European Respiratory Society

EUROPEAN SOCIETY OF CARDIOLOGY®

PULMONARY HYPERTENSION

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ESC ESSENTIAL MESSAGES

ESC GUIDELINES FOR THE DIAGNOSIS AND TREATMENT OF PULMONARY HYPERTENSION*

The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT) and by the Association for European Paediatric Cardiology (AEPC)

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European Heart Journal 2009;30:2493–2537
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Take home messages

1. Pulmonary Hypertension (PH), a haemodynamic and pathophysiological condition, should not be confused with Pulmonary Arterial Hypertension (PAH) a clinical group of rare diseases.

2. The current clinical classification of PH includes 37 clinical conditions which are classified into six groups according to similar pathological, pathophysiological and therapeutic characteristics.

3. Doppler-echocardiography does not measure pulmonary arterial pressure but gives only an estimate of it.

4. Right heart catheterization is mandatory for the confirmation of the diagnosis of PAH and may be required also in other clinical groups.

5. The correct clinical diagnosis in a patient with demonstrated PH requires the application of an appropriate diagnostic algorithm.

6. The prognostic assessment and the definition of clinical status of PAH patients is multidimensional and it requires the combination of different procedures assessing symptoms (e.g. WHO functional class), exercise capacity (e.g. 6-minute walk test) and right ventricular function (e.g. right heart catheterization).

7. The evidence-based treatment algorithm is appropriate only in patients with PAH (Clinical group 1). It is not recommended in the other clinical groups.

8. Acute vasoreactivity test preferably with inhaled nitric oxide is strongly recommended in particular in idiopathic PAH.

9. The appropriate prospective and objective evaluation of the clinical response to PAH therapy is recommended and should be performed with a structured follow-up.

10. Goal-oriented treatment strategy and sequential combination therapy are an appropriate approach to optimize the effects of currently available PAH specific drugs. Patients with PAH should be followed in "expert centers" objectively defined and prospectively audited. Self-election is not acceptable.

11. Despite recent progress, the current treatment strategy for PAH remains inadequate because the mortality rate continues to be high and the functional and haemodynamic impairments are still extensive in many patients. Interventional procedures such as lung transplantation are often required in particular in young patients.

Take home messages

12. The optimal treatment of the underlying left heart disease is recommended in patients with PH due to left heart disease. The specific PAH drugs are not recommended.

13. The optimal treatment of the underlying lung disease including long-term O_2 therapy in patients with chronic hypoxaemia is recommended in patients with PH due to lung diseases. The specific PAH drugs are not recommended.

14. Surgical pulmonary endarterectomy is the recommended treatment for patients with chronic thromboembolic pulmonary hypertension.

Major gaps in evidence

1. The definition of normal haemodynamic response to exercise and the definition of PH on exercise is needed.

2. The outcome of patients with PAH and mean pulmonary arterial pressure between 21 and 24 mmHg has to be established.

3. The diagnosis of post-capillary PH in left ventricular diastolic dysfunction (with "borderline high pulmonary wedge pressure") requires validation. The value and standardization of exercise, fluid challenge and diuretic challenge is required.

4. The prospective validation of the arbitrary echocardiographic jet velocity criteria for the definition of PH likelihood and the prospective validation of the "Probability of PAH diagnosis" is needed.

5. The derivation and validation of prognostic formulas/scores/calculators (multidimensional risk assessment) for PAH patients is required.

6. The identification of methods able to assess the disease activity in the distal pulmonary vessels is needed.

7. Appropriate comparisons between goal-oriented treatment strategy as compared with empiric therapy and between initial (upfront) drugs combination therapy as compared to sequential combination therapy is required.

8. The definitions of clinical status and clinical response to treatments require validation in adults and children.

9. The precise indications for balloon atrial septostomy and for listing for lung transplantation are required.

10. The appropriate definition of "out of proportion" PH in left heart disease and in lung diseases is required as well as randomized controlled studies with PAH approved drugs in these patients.

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