

Pulmonary hypertension: Rx

Definition: Pulmonary hypertension (PH) is defined by a mean pulmonary arterial pressure (mPAP) ≥ 25 mmHg at rest usually confirmed by right heart catheterization [1]. Normal pulmonary arterial systolic pressure ranges from 15 to 30 mmHg, diastolic pressure from 4 to 12 mmHg and normal mPAP is ≤ 20 mmHg

Classification:

Group 1 – PAH (idiopathic)

Group 2 – PH due to left heart disease

Group 3 – PH due to chronic lung disease and/or hypoxemia

Group 4 – Chronic thromboembolic pulmonary hypertension (CTEPH)

Group 5 – PH due to unclear multifactorial mechanisms

Rx: Early treatment of pulmonary hypertension (PH) is generally suggested because advanced disease may be less responsive to therapy. Treatment options include **oxygen, anticoagulation, diuretics, treatment of the underlying cause, digoxin (+/-), exercise, vaccination, and for some cases advanced PAH-specific therapy (epoprostenol, prostanoid, an endothelin receptor antagonist, or a phosphodiesterase-5 inhibitor). Advanced therapy often referred to as "pulmonary vasodilator therapy" is directed at the pulmonary hypertension (PH) itself, rather than the underlying cause of the PH. It includes treatment with prostacyclin pathway agonists, endothelin receptor antagonists, nitric oxide (NO)-cGMP enhancers, or rarely, certain calcium channel blockers.** The treatment of patients with group 1 pulmonary arterial hypertension (PAH) is different from that of the other groups who are, in general, less amenable to advanced PAH-specific medical therapy. Some patients in group 4 are candidates for pulmonary thromboendarterectomy. Occasionally, patients with severe disease unresponsive to therapy may require lung or heart-lung transplantation or right to left shunts.

Question:

Treatment for a patient with mean pulmonary pressure of 40 is best described as:

- a. Not required
- b. Directed at possible cause avoiding hypoxia, acidosis and hypercarbia
- c. Responsive to NO
- d. Based on clinical requirements

Answer: B – see keyword