Portopulmonary Hypertension: Dx

Portopulmonary hypertension (PPHTN) refers to pulmonary arterial hypertension (PAH) that is associated with portal hypertension; it is a well-recognized complication of chronic liver disease

PAH is defined according to right heart catheterization:

- ●Mean pulmonary artery pressure (mPAP) >25 mmHg at rest
- ●Pulmonary capillary wedge pressure (PCWP) <15 mmHg

Diagnosis of PPHTN is confirmed when:

- ●Pulmonary artery hypertension (PAH) exists, as indicated by an elevated mean pulmonary artery pressure (mPAP), pulmonary vascular resistance (PVR), and transpulmonary gradient during right heart catheterization
- ●Portal hypertension exists, as indicated by an elevated hepatic venous pressure during hepatic vein catheterization
- ●An alternative cause of the PAH cannot be identified

Pearl: Patients with PPHTN are at risk for in situ pulmonary **vascular thrombosis and thromboembolic disease** due to venous stasis, slowed pulmonary blood flow, and right heart enlargement. Patients with PPHTN also tend to have problems related to **volume overload** with **ascites and anasarca**. As a result, anticoagulant and diuretic therapy are often used in patients with PPHTN

Rx: anticoagulants, diuretics, epoprostenol, bosentan, sildenafil, iloprost and transplantation

Question:

A 62 year old male with advanced liver disease presents with dyspnea, increased O2 requirement, ascites and evidence of fluid overload. The following is compatible with portopulmonary hypertension except:

a. Mean pulmonary artery pressure of 30 mmHg
b. Pulmonary capillary wedge pressure of 27 mmHg
c. History of end-stage liver disease
d. Elevated INR

Answer: B. elevated PCWP is compatible with cardiac dysfunction, not PPHTN