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Pulmonary Hypertension: When to Refer to Definite Diagnosis

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Introduction

According to the 6th World Symposium on Pulmonary Hypertension in 2018, Pulmonary hypertension (PH) is defined as an increase in mean pulmonary artery pressure (mPAP) >20 mm from direct measurement through the right heart catheterization.

Pulmonary hypertension is a rare disease, but patients with pulmonary hypertension have a poor prognosis because of the high mortality and hospitalization rates. The symptoms of pulmonary hypertension (PH) patients are nor typical, the lack of diagnostic test equipment makes the detection of PH often missed by medical staff and colleagues. It is important here as medical personnel to know PH and when to immediately refer to the PH center for right heart catheterization.

Discussion

Signs and Symptoms of Pulmonary Hypertension (PH)

Early symptoms, such as shortness of breath, fatigue, angina, and syncope, are generally activity-induces and associated with progressive right ventricular dysfunction. In addition, symptoms such as abdominal distension and edema of the legs may be seen in the presence of right-sided heart failure. Other symptoms such as hemoptysis, hoarseness, wheezing, or angina if there is compression of the right coronary artery between the dilated pulmonary artery and the aorta.

Clinical signs of PH include lift in the left parasternal area on the precordial palpation, a loud second heart sound, a third heart sound (gallop) associated with the right ventricle, a pansystolic murmur in tricuspid regurgitation, and a diastolic murmur in pulmonary regurgitation. Elevated jugular venous pressure, hepatomegaly, ascites, and peripheral and cold acral edema may be seen in advanced stages. Cyanosis is common in patients, so it is important to assess for clubbing finger, hepatic and renal dysfunction, and ischemic complications.

Supporting Investigation

On chest X-RAY examination can be found dilatation of the central pulmonary artery accompanied by decreased peripheral vascularization (pruning). Enlargement of the right atrium and ventricle is also present in some cases of advanced PH. Chest X-RAY can help to differentiate arterial and venous PH by depicting an increase or decrease in the arteriovenous ratio, as well as assist in establishing a differential diagnosis of PH if there are signs of pulmonary disease (PH group 3) or pulmonary venous congestion due to left heart disease (PH group 2). As with the ECG, a normal chest X-RAY cannot exclude the presence of PH.

Electrocardiography (ECG) may reveal P pulmonary, right axis deviation, right ventricular hypertrophy, right ventricular strain, right bundle branch block, and QTc prolongation. Strains in the right ventricle have the highest sensitivity rates. Prolongation of the QTc and widening of the QRS complex indicate disease severity. Supraventricular arrhythmias such as atrial flutter and atrial fibrillation can be found at an advanced stage.

Echocardiography examination or mean PAP of more than 25 mmHg indicates the presence of PH. The ESC/ERS guidelines recommend continuous wave Doppler examination by assessing peak tricuspid regurgitation velocity as a determinant factor diagnosis of PH on echocardiography. Coupled with additional parameters such as right ventricular size, interventricular septal function, fluctuations of the inferior vena cava (IVC) against the respiratory cycle, right atrial systolic area, systolic flow velocity pattern against early diastolic pulmonary regurgitant velocity (PVAcct). The tricuspid regurgitant velocity (TR Vmax) value if the value is 2.8 m/s or cannot be measured, then is it probably not PH, but if there are other signs of PH indication, it is necessary to be careful in interpreting these results. If tricuspid regurgitation is between 2.9-3.4 m/s then the possibility of PH can be predicted. If the velocity of the tricuspid regurgitant jet is >3.4 m/s, then the possibility of PH is strongly suspected.

Right heart catheterization is required to confirm the diagnosis of pulmonary hypertension. This examination can assess the degree of hemodynamic disturbances and can be tested for vasoreactivity test was considered positive if there was a decrease in mean PAP of 10 mmHg or more without a decrease in cardiac output.

Cardiac MRI can be used in the diagnosis of PH patients with suspected congenital heart disease if echocardiography is inconclusive. This examination can be used to calculate the intracardiac anatomy to see the type of congenital heart disease, evaluate the size, morphology, and function of the right ventricle, as well as the pulmonary vasculature.

CT imaging examinations can be performed to provide an information picture of vascular, cardiac, mediastinal abnormalities, also assessing the lung parenchyme.

A pulmonary ventilation/perfusion scan (V/Q lung scanning) is used to establish the diagnosis of CTEPH. Normal values from this examination rule out the diagnosis of CTEPH. The V/Q lung scan is a screening method for the presence of CTEPH in PH patients.

Conclusions

Early detection, early diagnosis, and prompt referral to a PH center are important for the management of PH due to various etiologic causes and different therapeutic approaches in PH.

Echocardiography is an important diagnostic modality to screen patients with probable PH so that it can increase the number of referrals to a PH center.

Right heart catheterization is currently able to assess several characteristic parameters compared to a single parameter of mPAP to establish the diagnosis of PH.

A multi-disciplinary PH team is needed to assess and reclassify the PH group.

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