A Simple Guide To Pulmonary Artery Hypertension, Diagnosis, Treatment And Related Conditions (A Simple Guide to Medical Conditions)

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Chapter 1

Pulmonary Artery Hypertension

Pulmonary Arterial Hypertension (PAH) is a rare medical disorder which causes high blood pressure in the arteries to the lungs with no apparent cause.

PAH is also called precapillary pulmonary hypertension or idiopathic pulmonary arterial hypertension (IPAH).

It is a serious condition.

If a person has it, the blood vessels that carry blood from the heart to the lungs become hard and narrow.

In order to pump the blood through, the heart has to work harder.

An overworked heart which becomes weaker over time cannot do its job and the patient can develop heart failure.

IPAH which is not treated will result in right-sided heart failure and death.

Echocardiography in about 30% of patients with pulmonary arterial hypertension (PAH) shows right-to-left shunting across a patent foramen ovale.

There are two main kinds of IPAH.

1. One occurs in families or appears for no known reason (idiopathic).
2. The other kind is linked to another medical disease such as pulmonary and cardiac disease

Signs and symptoms

Non-specific symptoms of IPAH are:

1. Shortness of breath during normal exercise
2. Tiredness
3. Chest pain
4. A racing heartbeat
5. Pain on the upper right side of the abdomen
6. Decreased appetite

7. Recurrent syncope

In patients with PAH cardiovascular findings are:

1. The pulmonary component of the second heart sound is typically raised, which may indicate fixed or paradoxical splitting in the presence of severe poor right ventricular function;

2. Occasionally, the second heart sound may be felt
3. A pulmonary regurgitation (Graham Steell murmur) may be present
4. A murmur of tricuspid regurgitation can be heard, and a right ventricular heave may be seen.
5. Jugular venous pulsations may be raised in the presence of right ventricular failure, volume overload, or both;
6. Large V waves are frequently appear because of the frequently present severe tricuspid regurgitation
7. Right-sided S3 gallop

Other findings are:

1. Hepatomegaly with palpable pulsations of the liver
2. Abnormal abdominal-jugular reflex
3. Ascites - Not rare in untreated patients and in patients with increasing decompensated right heart failure
4. Pitting edema - In the lower limbs
5. Presacral edema - In patients who are unable to get up from bed

The patient may find it difficult to do any physical activities as his IPAH becomes worse.

Diagnosis

Cardiac catheterization

Cardiac catheterization is the main standard test to confirm definitely any form of PAH.

The diagnosis of the patients suspected of IPAH is confirmed by cardiac catheterization.

The exclusion of left-sided heart disease, especially diastolic dysfunction, is very essential in these patients because of major treatment problems.

Catheterization is also done to find out the pulmonary vaso-reactivity which can be useful in prognosis.

Catheterization is also used to assess in the initiation and titration of high-dose calcium channel blocker (CCB) treatment.

Laboratory studies

1. Antinuclear antibody
2. Thyroid function
3. B-type natriuretic peptide

Imaging studies

1. Radiography
2. Echocardiography
3. Computed tomography (CT), magnetic resonance imaging (MRI), and lung scanning
4. Pulmonary angiography

Electrocardiography

The electrocardiography (ECG) results are usually not normal in patients with PAH, showing right atrial enlargement, right axis deviation, right ventricular hypertrophy, and typical ST depression and T-wave inversions in the anterior leads.

An incomplete RBBB may be seen sometimes (typically in patients with atrial septal defects).

Some patients with IPAH will have few or no abnormal electrocardiographic results.

TABLE OF CONTENT

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