CONDITION

Pulmonary hypertension

Pulmonary hypertension is high blood pressure in the blood vessels carrying blood from the heart to the lungs.

WHAT IT IS
High pressure in the lung arteries

BODY SYSTEM
Heart, circulation, and lungs

MAIN SYMPTOMS
Breathlessness, tiredness, chest discomfort, fainting

WHAT CAUSES IT
Heart, lung, clotting, inherited, or unknown causes

HOW IT IS DIAGNOSED
Right heart catheterization after screening tests

MAIN TREATMENT
Cause-specific care and pulmonary hypertension medicines

CAN IT BE CURED?
Sometimes - when the underlying cause can be corrected

IS IT AN EMERGENCY?
Sometimes - fainting, chest pain, or severe breathlessness

LONG-TERM OUTLOOK
Often chronic; outlook improves with early, cause-specific care

Overview

Pulmonary hypertension means the pressure is too high in the blood vessels that carry blood from the heart to the lungs. These vessels are called the pulmonary arteries.

This is different from the blood pressure measured with a cuff on your arm. In pulmonary hypertension, the right side of the heart must work harder to pump blood through the lungs. Over time, that extra work can strain and weaken the heart.

Pulmonary hypertension is a broad condition with several causes. Pulmonary arterial hypertension, often shortened to PAH, is one specific type. In PAH, the small arteries in the lungs become narrowed, thickened, or stiff.

Some forms improve when their cause is treated. Others are long-term conditions that need

specialist monitoring and treatment. Finding the type matters because a treatment that helps one type may not help another.

Symptoms

Symptoms often develop slowly and may resemble those of heart, lung, or fitness problems. Early symptoms may only appear during exercise.

Common symptoms include:

As the condition progresses, breathlessness may occur with lighter activity or at rest. The lips or skin can sometimes look blue or grey because the blood is carrying less oxygen.

These symptoms do not always mean pulmonary hypertension. An assessment is needed to find their cause.

- Shortness of breath during activity
- Unusual tiredness or weakness
- Chest pressure, discomfort, or pain
- Dizziness or fainting
- A fast, pounding, or irregular heartbeat
- Swelling in the ankles, legs, or abdomen

Causes and risk factors

Doctors divide pulmonary hypertension into groups based on what is causing the high pressure.

Pulmonary arterial hypertension may have no identified cause. It can also be inherited or linked to connective tissue conditions such as systemic sclerosis, congenital heart problems, liver disease with portal hypertension, HIV, or certain drugs and toxins.

Other major causes include:

Having a risk factor does not mean you have pulmonary hypertension. In some people, no clear cause is found even after detailed testing.

- Left-sided heart conditions: Heart valve problems or weakness and stiffness of the left side of the heart can make pressure back up toward the lungs.

- Lung conditions or low oxygen: Chronic obstructive pulmonary disease, interstitial lung disease, sleep-related breathing disorders, and long-term exposure to low oxygen can raise lung artery pressure.
- Chronic blood clots: Clots that do not fully clear can obstruct the pulmonary arteries. This is called chronic thromboembolic pulmonary hypertension, or CTEPH.
- Mixed or unclear causes: Some blood, inflammatory, metabolic, and other conditions can affect the lung vessels in several ways.

Diagnosis

Diagnosis usually begins with a discussion of your symptoms, health history, medicines, family history, and activity level. A physical examination may identify changes in heart sounds, swelling, or signs of another condition.

An echocardiogram, which is an ultrasound scan of the heart, can estimate pressure in the pulmonary arteries. It also shows how well the right and left sides of the heart are working. An echocardiogram can suggest pulmonary hypertension, but it does not confirm the diagnosis by itself.

Other tests may include:

Pulmonary hypertension is confirmed with right heart catheterization. During this procedure, a thin tube is passed through a vein into the right side of the heart and pulmonary artery. It directly measures pressures and blood flow. These results help establish the type and guide treatment.

Some people with suspected PAH have an additional test during catheterization to see whether their lung arteries respond to a short-acting medicine.

- An electrocardiogram to record the heart's electrical activity
- Blood tests, including tests for heart strain and possible causes
- Chest imaging and lung function tests
- A walking test to measure exercise ability and oxygen levels
- A sleep study if sleep-related breathing problems are suspected
- A ventilation-perfusion scan to look for chronic blood clots

Treatment and management

Treatment is based on the cause, severity, symptoms, and effect on the right side of the heart. Care is often coordinated by a pulmonary hypertension specialist team.

Treating an underlying heart or lung condition may lower pulmonary pressure or prevent it from worsening. This can include care for heart valve disease, heart failure, lung disease, low oxygen, or sleep-related breathing problems.

Supportive treatments may include oxygen when blood oxygen is low, medicines called diuretics to reduce fluid buildup, and supervised exercise or rehabilitation. Anticoagulants like warfarin or apixaban , which reduce blood clotting, are important for chronic clot-related pulmonary hypertension but are not suitable for every type.

PAH can be treated with medicines that act on the narrowed lung arteries. These include endothelin receptor antagonists, phosphodiesterase-5 inhibitors, soluble guanylate cyclase stimulators, and medicines that act on the prostacyclin pathway. Some are tablets, while others are inhaled, injected, or given continuously through a pump. More than one medicine may be used.

PAH medicines should not be assumed to work for pulmonary hypertension caused by left heart or lung conditions. In those groups, treatment usually focuses on the underlying problem.

For CTEPH, surgery to remove organized clots may offer major improvement and can sometimes be curative. Balloon pulmonary angioplasty or targeted medicine may be considered when surgery is not possible or pressure remains high afterward.

Lung or heart-lung transplantation may be considered for a small number of people with severe disease despite other treatments.

Pregnancy places major stress on the heart and circulation in pulmonary hypertension. If pregnancy is possible or being considered, specialist advice about risks, contraception, and planning is important.

Complications

The main complication is right-sided heart failure. This happens when the right side of the heart can no longer pump effectively against the raised pressure. It can cause increasing breathlessness, tiredness, and fluid buildup.

Other possible complications include abnormal heart rhythms, fainting, reduced oxygen levels, and worsening limits on physical activity. Severe disease can affect the liver, kidneys, and other organs because blood flow and oxygen delivery are reduced.

Regular monitoring aims to identify these changes early.

Prevention

Pulmonary hypertension cannot always be prevented. Inherited PAH and cases with no known cause have no established prevention method.

Managing heart and lung conditions, avoiding smoking, and treating low oxygen or sleep-related breathing problems may reduce risk in some situations. Following advice about blood clot prevention after surgery, illness, or long periods without movement may help reduce the chance of pulmonary embolism.

Avoiding illegal stimulant drugs and unapproved weight-loss drugs also removes known risk factors for some forms of PAH. These steps cannot guarantee prevention.

Living with it and what to expect

The course of pulmonary hypertension differs according to its cause and how the heart responds to treatment. Some forms improve when an underlying problem is corrected. Many forms are chronic and require treatment and follow-up over years.

Follow-up may include blood tests, echocardiograms, walking tests, oxygen checks, and reviews of symptoms and daily activity. Right heart catheterization may be repeated when more precise information is needed.

Many people can remain active, work, and take part in family life, although they may need to pace activities. Gentle or moderate exercise may help when it is planned with the specialist team. Heavy lifting or activity that causes chest pain, marked breathlessness, or dizziness may be unsafe.

Take prescribed medicines consistently and do not stop them suddenly without medical guidance. Some continuous PAH treatments can cause serious problems if interrupted.

Air travel and time at high altitude may lower oxygen levels. People with low oxygen or more advanced pulmonary hypertension may need testing, supplemental oxygen, or other planning before travel.

The outlook cannot be predicted from the diagnosis alone. The underlying group, right-heart function, symptoms, other health conditions, and response to treatment all matter.

When to get medical help

Seek emergency medical help for:

These symptoms can indicate severe strain on the heart or another urgent problem. Do not drive yourself if you feel faint or severely short of breath.

Contact your healthcare team promptly for increasing ankle or abdominal swelling, rapid unexplained weight gain, new palpitations, reduced ability to manage usual activities, or worsening breathlessness. These changes need timely assessment but are not always emergencies.

Routine appointments remain important even when symptoms are stable. They allow treatment to be adjusted before problems become more noticeable.

- Severe or rapidly worsening breathlessness
- Fainting, especially during activity
- New or persistent chest pain
- Blue or grey lips or skin
- Confusion or difficulty staying awake
- Coughing up more than a small streak of blood

Common questions

Is pulmonary hypertension the same as pulmonary arterial hypertension?

No. Pulmonary hypertension is the broad term for high pressure in the lung circulation. Pulmonary arterial hypertension is one subgroup caused by changes in the small pulmonary arteries. Other types are linked to heart disease, lung disease, chronic clots, or mixed causes.

Can pulmonary hypertension go away?

Sometimes. Pressure may improve if a reversible cause is treated, such as certain heart problems or operable chronic blood clots. Many forms, including much of PAH, are long-term conditions that can be controlled but not cured with current medicines.

Is pulmonary hypertension fatal?

It is a serious condition, but the outlook is not the same for everyone. Cause-specific treatments can improve symptoms, heart function, and long-term outcomes. Risk is higher when the right side of the heart is severely affected or the condition does not respond to treatment.

Can I exercise with pulmonary hypertension?

Many people can do carefully planned activity. Walking and supervised rehabilitation may improve strength and confidence. The safe level depends on symptoms, oxygen levels, heart

function, and treatment. Stop and seek advice if activity causes chest pain, faintness, or severe breathlessness.

Can I fly with pulmonary hypertension?

Many people can fly, but aircraft cabins have lower oxygen levels than ground level. You may need an assessment or oxygen during the flight, especially if you already use oxygen, have low oxygen levels, or have more advanced symptoms. Plan well before travelling.

Why do I need a right heart catheterization?

An echocardiogram can estimate pressure, but it cannot make the final diagnosis or fully classify the condition. Right heart catheterization directly measures pressure and blood flow. This information helps prevent the wrong type of treatment from being used.

Sources

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