CONDITION

Primary pulmonary hypertension

Primary pulmonary hypertension is high pressure in the lung arteries without an identifiable cause.

WHAT IT IS
High pressure in lung arteries without an identified cause

BODY SYSTEM
Heart, lungs and circulation

MAIN SYMPTOMS
Breathlessness, tiredness, chest discomfort, fainting

WHAT CAUSES IT
Usually unknown; sometimes inherited genetic changes

HOW IT IS DIAGNOSED
Right heart catheterisation after heart and lung testing

MAIN TREATMENT
Pulmonary artery medicines, supportive care and specialist monitoring

CAN IT BE CURED?
No - treatment may control and slow it

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

LONG-TERM OUTLOOK
Serious lifelong condition, often manageable with modern treatment

Overview

Primary pulmonary hypertension is an older name for high blood pressure in the arteries that carry blood from the heart to the lungs, when no underlying cause can be found. Today, specialists usually call this idiopathic pulmonary arterial hypertension , or idiopathic PAH. "Idiopathic" means that the cause is unknown.

This is different from the common type of high blood pressure measured with an arm cuff. In PAH, small arteries inside the lungs become narrowed and stiff. The right side of the heart must work harder to push blood through them.

Over time, this extra workload can enlarge and weaken the right side of the heart. Treatment aims to reduce strain on the heart, improve symptoms and physical ability, and slow progression.

Pulmonary hypertension has several forms. It can result from left-sided heart conditions, lung conditions, blood clots, medicines, or other medical conditions. These forms may look similar but need different treatment. The word "primary" should only be used after a careful assessment has found no other explanation.

Symptoms

Symptoms often develop gradually. Early signs may be mild and can resemble asthma, low fitness, anaemia, or other heart and lung conditions.

Common symptoms include:

Some people notice that activities such as climbing stairs, walking uphill, or carrying groceries become harder. As the condition progresses, breathlessness may occur with lighter activity or at rest.

Symptoms alone cannot confirm primary pulmonary hypertension. Many more common conditions cause the same problems, so testing is needed.

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

Causes and risk factors

In idiopathic PAH, testing does not find a specific cause. Researchers have identified changes in the cells lining the pulmonary arteries, but it is often unclear why these changes begin.

Some cases that were once called primary pulmonary hypertension are now described as heritable PAH . This means an inherited genetic change contributes to the condition. Changes in the `BMPR2` gene are the most commonly identified, but other genes can be involved.

Having a related genetic change does not mean someone will definitely develop PAH. Some people carry a change throughout life without symptoms. A family history of PAH or unexplained early death may lead a specialist to discuss genetic counselling and testing.

Certain medicines, toxins, connective tissue conditions, congenital heart conditions, liver

disease, HIV, and other health problems can cause PAH. When one of these is identified, the condition is not classified as idiopathic PAH, even though the pressure changes may be similar.

Diagnosis

Diagnosis usually starts with a medical history, physical examination, and an echocardiogram . An echocardiogram is an ultrasound scan of the heart. It can estimate pressure in the lung circulation and show whether the right side of the heart is enlarged or under strain.

Other tests help assess the heart and lungs and look for alternative causes. They may include:

An echocardiogram can suggest pulmonary hypertension, but it does not confirm the diagnosis. Confirmation requires right heart catheterisation . 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, blood flow, and resistance in the lung circulation.

During catheterisation, some people have a short test using a medicine that temporarily widens the pulmonary arteries. This is called vasoreactivity testing. Only a small group have the response needed for certain calcium-channel blocker treatment.

Doctors must also rule out other forms of pulmonary hypertension before calling the condition idiopathic. Assessment at a pulmonary hypertension centre can help ensure that the diagnosis and classification are correct.

- An electrocardiogram, or ECG, to record heart rhythm
- Blood tests, including markers of heart strain
- Breathing tests to measure lung function
- Chest imaging, such as a chest X-ray or CT scan
- A ventilation-perfusion scan to look for chronic blood clots
- An exercise or six-minute walking test
- Sleep testing when sleep-related breathing problems are possible

Treatment and management

Treatment is usually planned by a specialist pulmonary hypertension team. The choice depends on symptoms, test results, heart function, other health conditions, and the estimated risk of the condition worsening.

PAH-specific medicines act on pathways that control tightening and abnormal growth in the

pulmonary arteries. The main groups include:

These medicines are taken by mouth, inhaled, injected under the skin, or delivered continuously into a vein, depending on the drug. Many people receive more than one type because the medicines work through different pathways.

Calcium-channel blockers are suitable only for the small number of people who have the required response during vasoreactivity testing. They should not be used for PAH simply because they are also used for ordinary high blood pressure.

Supportive treatment may include diuretics like furosemide or hydrochlorothiazide , sometimes called water tablets, to reduce fluid build-up. Oxygen may be used when blood oxygen levels are low. Anticoagulants like warfarin or apixaban , which reduce blood clotting, are not automatically suitable for everyone with idiopathic PAH; the specialist team weighs possible benefits against bleeding risk.

Supervised exercise or pulmonary rehabilitation may improve physical ability for people whose condition is stable. Activity is generally adjusted to symptoms, without pushing through faintness, chest pain, or severe breathlessness.

Some PAH medicines can harm a developing baby, and pregnancy places major strain on the heart in people with PAH. Pregnancy planning, contraception, and any possible pregnancy need early discussion with a specialist team.

If severe PAH continues to progress despite treatment, lung transplantation or combined heart-lung transplantation may be considered. Eligibility requires a detailed assessment and does not mean that transplantation will be appropriate or available for everyone.

- Endothelin receptor antagonists
- Phosphodiesterase-5 inhibitors
- A soluble guanylate cyclase stimulator
- Prostacyclin-related medicines

Complications

The main complication is right-sided heart failure. This happens when the right side of the heart cannot pump effectively against the high resistance in the pulmonary arteries.

Signs may include increasing breathlessness, swelling, abdominal fullness, rapid weight gain from fluid, reduced appetite, and worsening tiredness.

Other possible complications include abnormal heart rhythms, fainting, low oxygen levels, and coughing up blood. Advanced PAH can reduce blood flow to the body, particularly during

exertion.

Regular assessment is important because test results may show increased heart strain before symptoms change greatly.

Prevention

Idiopathic PAH cannot currently be prevented because its cause is unknown.

Heritable PAH also cannot be prevented by lifestyle changes. Genetic counselling can help families understand inheritance, the limits of testing, and whether screening may be appropriate for relatives.

Avoiding tobacco and unregulated stimulant or weight-loss products supports general heart and lung health, but it does not guarantee prevention of idiopathic PAH. Medicines should not be stopped or changed because of concerns about pulmonary hypertension without medical advice.

Living with it and what to expect

Primary pulmonary hypertension is usually lifelong. Without effective treatment it may worsen, but its pace is not the same for everyone. Modern treatment can improve symptoms, physical ability, and heart function, and some people remain stable for long periods. Others have continuing progression despite several treatments.

Follow-up commonly includes discussion of symptoms and activity, blood tests, walking tests, echocardiograms, and checks for medicine effects. Right heart catheterisation may be repeated when detailed pressure measurements would affect treatment decisions.

Treatment may be adjusted over months or years. A change does not always mean that a medicine has failed; combination treatment is a standard way to target different parts of the condition.

Daily life often involves pacing activities and allowing time for rest. People may need practical adjustments at work, school, or home. Travel and higher altitudes can affect oxygen levels, so plans for flying or visiting high-altitude areas may need to be discussed in advance.

Emotional effects also matter. Uncertainty, limits on activity, and complex treatment can be difficult to manage. Support from family, counselling, rehabilitation teams, or pulmonary hypertension organisations may help.

When to get medical help

Arrange a routine or prompt review if you notice a gradual increase in breathlessness, reduced ability to do normal activities, new ankle swelling, abdominal swelling, rapid weight gain from fluid, or more frequent palpitations. These changes may mean that treatment or fluid management needs reassessment.

Contact the specialist team promptly if medicine side effects are difficult to manage, doses have been missed, or there is a problem with a pump or continuous infusion system. Abrupt interruption of some continuously delivered PAH medicines can be dangerous.

Seek emergency medical help for:

Do not drive yourself if you are faint, severely breathless, or having significant chest pain.

- Severe or rapidly worsening breathlessness, especially at rest
- Fainting or near-fainting
- New or severe chest pain
- Blue or grey lips, confusion, or extreme drowsiness
- Coughing up more than a small streak of blood
- Sudden symptoms suggesting a problem with continuous intravenous treatment

Common questions

Is primary pulmonary hypertension the same as pulmonary arterial hypertension?

Primary pulmonary hypertension is an older term. Most cases are now classified as idiopathic PAH when no cause is found, or heritable PAH when an inherited genetic factor is identified. PAH is one category within the broader group called pulmonary hypertension.

Is it the same as regular high blood pressure?

No. Regular high blood pressure affects arteries throughout the body and is measured with an arm cuff. PAH affects the arteries between the right side of the heart and the lungs. It requires different tests and treatments.

Can primary pulmonary hypertension be cured?

There is no medical cure at present. Treatment can widen or stabilise the lung arteries, reduce strain on the heart, and slow progression. Transplantation may replace severely affected lungs, but it is a major treatment with its own lifelong risks.

Can I exercise with pulmonary hypertension?

Many people can take part in carefully planned, moderate activity when their condition is stable. Supervised rehabilitation may be helpful. Strenuous exertion and activity that causes chest pain, faintness, or severe breathlessness are not considered safe.

Does it run in families?

Sometimes. A proportion of cases involve an inherited genetic change, but carrying such a change does not guarantee that PAH will develop. Genetic counselling can explain testing and what a result may mean for relatives.

How quickly does it get worse?

There is no single timeline. Progression is influenced by heart function, symptoms, response to treatment, and other health conditions. Regular follow-up allows the specialist team to identify changes and adjust treatment.

Sources

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