CONDITION

Pulmonary arterial hypertension

Pulmonary arterial hypertension is high blood pressure in narrowed lung arteries that strains the right side of the heart.

WHAT IT IS
High pressure in arteries carrying blood through the lungs

BODY SYSTEM
Heart, lungs and circulation

MAIN SYMPTOMS
Breathlessness, tiredness, chest discomfort, fainting

WHAT CAUSES IT
Often unknown; sometimes genetic, disease-related, or drug-related

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

MAIN TREATMENT
Specialist medicines that lower pressure and support heart function

CAN IT BE CURED?
No, treatments control but do not eliminate it

IS IT AN EMERGENCY?
Sometimes, with fainting or severe breathlessness

LONG-TERM OUTLOOK
Serious but increasingly treatable with lifelong specialist care

Overview

Pulmonary arterial hypertension, or PAH, is a specific type of pulmonary hypertension. Pulmonary hypertension means high blood pressure in the blood vessels of the lungs. In PAH, the small lung arteries become narrowed, thickened or stiff.

These changes make it harder for blood to travel from the right side of the heart through the lungs. The heart must pump against greater resistance. Over time, its right side can enlarge and become weaker.

PAH is not the same as ordinary high blood pressure measured with an arm cuff. It is also different from pulmonary hypertension caused mainly by left-sided heart problems, long-term lung conditions or old blood clots. Correctly identifying the type matters because treatments are different.

PAH is a long-term and potentially serious condition. There is no general cure, but specialist treatments can reduce symptoms, improve physical ability and slow progression. Earlier assessment can allow treatment to begin before the heart is severely strained.

Symptoms

Symptoms often develop gradually. Early signs may be mild or appear only during physical activity, so they can be mistaken for low fitness, asthma or another heart or lung problem.

Possible symptoms include:

PAH does not always cause a cough or wheeze. Some people have few symptoms despite changes in the heart and lung circulation. Symptoms alone cannot confirm PAH.

- Shortness of breath during activity, and sometimes later at rest
- Unusual tiredness or weakness
- Chest pressure, discomfort or pain
- Dizziness, near-fainting or fainting, especially during exertion
- A fast, pounding or irregular heartbeat
- Swelling of the ankles, legs or abdomen
- Reduced ability to walk, climb stairs or complete normal activities

Causes and risk factors

In many people, no cause is found. This is called idiopathic PAH, meaning PAH without an identified cause.

Some cases are heritable. Changes in genes such as BMPR2 can increase the chance of developing PAH, but inheriting a gene change does not mean someone will definitely develop the condition. Genetic counselling can help families understand testing and what a result may mean.

PAH may also be associated with:

Some appetite-suppressing medicines and methamphetamine exposure have been linked with PAH. A healthcare team may ask about current and past medicines and drug exposure without judgement because this information can affect diagnosis and care.

Common heart and lung conditions can cause other forms of pulmonary hypertension rather than PAH. For example, heart valve problems, heart failure, chronic lung conditions and

long-standing blood clots require their own assessment and treatment.

- Connective tissue conditions, particularly systemic sclerosis
- Certain congenital heart conditions present from birth
- HIV infection
- Portal hypertension, which is high pressure in veins serving the liver
- Schistosomiasis, a parasitic infection found mainly in some tropical regions
- Certain medicines, toxins or stimulant drugs

Diagnosis

PAH cannot be diagnosed from symptoms or an echocardiogram alone. Diagnosis usually involves several tests to confirm high pressure, identify the type of pulmonary hypertension and look for an underlying cause.

An echocardiogram uses ultrasound to show the heart's structure and movement. It can estimate pressure in the lung circulation and show whether the right side of the heart is enlarged or strained. It is an important screening test, but its pressure estimate is not exact.

Other tests may include:

Right-heart catheterisation is needed to confirm PAH. A thin tube is passed through a vein into the right side of the heart and lung arteries. It directly measures blood pressure, blood flow and resistance in the lung circulation.

PAH is confirmed when these measurements show high pressure and resistance in the lung arteries without evidence that raised pressure from the left side of the heart is responsible.

During catheterisation, some people with idiopathic, heritable or drug-associated PAH have a vasoreactivity test. This checks whether the lung arteries respond strongly to a short-acting medicine. Only a small, carefully identified group benefits from treatment based on a positive result.

- An electrocardiogram, or ECG, to record the heart's electrical activity
- Blood tests for associated conditions and strain on the heart
- Chest imaging and lung function tests
- An exercise walking test to measure physical ability and oxygen levels
- A ventilation-perfusion scan to look for old blood clots

- Overnight oxygen or sleep testing when appropriate

Treatment and management

PAH should be managed with input from a specialist pulmonary hypertension service. Treatment is selected according to the cause, symptoms, test results, right-heart function and estimated risk of progression.

Most people receive PAH-targeted medicines. These work on different biological pathways involved in narrowing and remodelling of the lung arteries. The main categories include endothelin receptor antagonists, phosphodiesterase-5 inhibitors, a soluble guanylate cyclase stimulator, prostacyclin-pathway medicines and an activin-signalling inhibitor.

More than one medicine may be used because combinations can target different pathways. Some treatments are tablets. Others are inhaled, injected under the skin or given continuously into a vein. Continuous medicines must not be stopped suddenly because this can cause rapid deterioration.

A calcium-channel blocker is appropriate only for the small group with a positive vasoreactivity test and continued specialist monitoring. These medicines should not be used as PAH treatment without that result.

Supportive care may include:

Blood-thinning medicine is not routinely suitable for everyone with PAH. Its possible benefit must be weighed against bleeding risk and the cause of PAH.

Some PAH medicines can cause serious harm during pregnancy. Pregnancy itself places major strain on the heart in PAH and carries substantial risk. Pregnancy planning, reliable contraception and any required pregnancy testing should be discussed with the specialist team. Some medicines can also affect how well hormonal contraceptives work.

If PAH remains severe despite treatment, lung or heart-lung transplantation may be considered. A procedure called atrial septostomy is used only in selected circumstances, usually as a bridge to another treatment.

Medicines commonly used for this include ambrisentan and macitentan . Which one suits you, if any, is a decision for you and your doctor.

- Diuretics like furosemide or hydrochlorothiazide , sometimes called water tablets, for fluid build-up
- Supplemental oxygen when blood oxygen is low

- Supervised exercise or pulmonary rehabilitation when the condition is stable
- Vaccination and prompt care for respiratory infections
- Treatment of an associated condition, where possible

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 high resistance in the lung arteries. It can cause swelling, abdominal discomfort, poor appetite, tiredness and breathlessness.

Other possible complications include:

Regular assessment is intended to detect worsening heart strain before severe complications develop.

- Abnormal heart rhythms
- Fainting because the heart cannot increase blood flow adequately
- Liver or kidney problems related to poor circulation and congestion
- Coughing up blood, which is uncommon but needs prompt assessment

Prevention

Many cases of PAH cannot be prevented. This includes idiopathic PAH and PAH linked to an inherited gene change or another unavoidable condition.

Avoiding methamphetamine and medicines known to increase PAH risk may prevent some exposure-related cases. Do not stop a prescribed medicine because of concern about PAH without discussing it with the prescriber.

Genetic counselling does not prevent PAH, but it can help relatives understand inherited risk and whether testing or clinical screening is appropriate. Treating an associated condition is important, although it may not prevent PAH from developing.

Living with it and what to expect

PAH usually requires lifelong care. Its course is not the same for everyone. Some people remain stable for extended periods with treatment, while others have continuing changes in symptoms or right-heart function.

Follow-up commonly includes discussion of symptoms, a physical examination, blood tests, walking tests and repeat heart imaging. Right-heart catheterisation may be repeated when measurements are needed to guide major treatment decisions.

Specialists regularly assess risk using several findings rather than one test. These include symptoms, fainting, exercise ability, blood results, heart imaging and pressure measurements. Treatment may be added or changed if the condition is not meeting agreed goals.

Many people continue working, travelling and taking part in family life, although they may need to pace activities. Suitable exercise can help maintain muscle strength and confidence. Heavy straining or activity that causes marked breathlessness, chest pain or dizziness should be avoided.

Travel planning may involve arranging enough medicine, checking oxygen needs and identifying specialist support at the destination. People using continuous infusion treatment need a plan for equipment problems and emergency supplies.

Emotional strain is also common. A specialist nurse, counsellor, rehabilitation programme or PAH support group may help with uncertainty, changing abilities and practical adjustments.

When to get medical help

Call emergency services for:

Do not drive yourself if you are severely unwell.

Contact your PAH team promptly for new or steadily worsening breathlessness, swelling, rapid weight gain, repeated dizziness, palpitations or a clear reduction in normal activity. These changes may reflect fluid build-up, an abnormal heart rhythm or increasing strain on the heart.

Routine follow-up is still important when symptoms feel stable. Contact the specialist team before becoming pregnant, if a pregnancy test is positive, before starting a new medicine or if treatment side effects are difficult to manage.

- Severe or rapidly worsening breathlessness, especially at rest
- Fainting, new confusion or difficulty staying awake
- Chest pain that is severe, persistent or accompanied by sweating or nausea
- Blue or grey lips or skin
- Coughing up more than a small streak of blood
- A problem with continuous infusion treatment that interrupts the medicine

Common questions

Is PAH the same as pulmonary hypertension?

No. Pulmonary hypertension is a broad term for high pressure in the lung circulation. PAH is one specific form involving increased resistance in the small lung arteries. Other forms can result from left-heart problems, lung conditions or chronic blood clots.

Can pulmonary arterial hypertension go away?

PAH usually does not go away. Treating an associated problem may help, but the changes in the lung arteries often remain. Treatment aims to control symptoms, protect the right side of the heart and slow progression.

How quickly does PAH progress?

There is no single timeline. Changes may occur slowly or more quickly, even if symptoms initially seem mild. Exercise ability, fainting, blood tests, heart imaging and catheter measurements give more useful information than time since diagnosis alone.

Can I exercise with PAH?

Many people benefit from light or moderate activity when their condition is stable. A specialist-guided or supervised programme is safest when starting. Stop and seek advice if activity causes chest pain, marked dizziness, fainting or severe breathlessness.

Is PAH hereditary?

Some forms are hereditary, but many are not. A person can carry a PAH-related gene change without developing the condition. Genetic counselling is recommended before testing so that you understand possible medical and family implications.

Can I become pregnant if I have PAH?

Pregnancy carries a high risk of serious complications because it greatly increases the work of the heart. Several PAH medicines can also harm a developing baby. Speak with the specialist team before pregnancy or urgently after a positive test; do not stop PAH medicine on your own.

Sources

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