CONDITION

Pulmonary arteriovenous fistula

A pulmonary arteriovenous fistula is an abnormal connection between an artery and vein in the lung.

WHAT IT IS
Abnormal direct connection between lung arteries and veins

BODY SYSTEM
Lungs, heart and circulation

MAIN SYMPTOMS
Shortness of breath, low oxygen, fatigue, coughing blood

WHAT CAUSES IT
Inherited HHT, sporadic changes, or rarely acquired conditions

HOW IT IS DIAGNOSED
Contrast echocardiography and chest CT

MAIN TREATMENT
Catheter embolization; surgery rarely

CAN IT BE CURED?
Sometimes - treatment can close the abnormal connection

IS IT AN EMERGENCY?
Sometimes - coughing blood, stroke signs, or severe breathlessness

LONG-TERM OUTLOOK
Usually good with treatment and lifelong follow-up

Overview

A pulmonary arteriovenous fistula is an abnormal connection between a pulmonary artery and a pulmonary vein. These blood vessels carry blood through the lungs. The condition is more commonly called a pulmonary arteriovenous malformation, or pulmonary AVM.

Normally, blood passes through tiny lung vessels called capillaries. There it collects oxygen and the capillaries help filter small particles from the circulation. In a pulmonary AVM, some blood bypasses this normal route and flows directly from an artery into a vein.

This bypass is called a right-to-left shunt. It can lower the amount of oxygen in your blood. It can also allow a blood clot, air bubble, or bacteria to pass through the lungs and reach the brain or other organs.

Some pulmonary AVMs are small and cause no noticeable symptoms. Others can affect oxygen levels or lead to complications such as stroke, brain infection, or bleeding. Treatment can often close the abnormal connection and reduce these risks.

A person may have one pulmonary AVM or several. They may occur in one lung or both lungs.

Symptoms

Many people have no symptoms when a pulmonary AVM is first found. It may be discovered during testing for another condition or during screening for hereditary hemorrhagic telangiectasia.

When symptoms occur, they can include:

Clubbing may develop in people with long-term low oxygen. This means the ends of the fingers become wider and the nails curve more than usual.

Symptoms do not always show how important an AVM is. A person who feels well can still have a risk of complications because the lung's normal filtering system is being bypassed.

Stroke, a temporary interruption of blood flow to the brain called a transient ischemic attack, and brain abscess may sometimes be the first sign. A brain abscess is a pocket of infection in the brain.

- Shortness of breath, especially during activity
- Tiredness or reduced exercise tolerance
- Low blood oxygen levels
- Blue or grey lips, skin, or fingertips
- Coughing up blood
- Headaches or migraine
- Dizziness or fainting

Causes and risk factors

Most people with multiple pulmonary AVMs have hereditary hemorrhagic telangiectasia, or HHT. This is an inherited condition affecting blood vessel development. It may also cause frequent nosebleeds, small visible red blood vessels on the skin or inside the mouth, and AVMs in other organs.

HHT can run in families. A parent with HHT can pass the altered gene to a child. However, signs

differ widely, even among relatives. Someone with HHT may have few outward symptoms.

A pulmonary AVM can also occur without HHT. This is called a sporadic pulmonary AVM, meaning that no inherited cause has been identified.

Less commonly, abnormal lung vessel connections may be associated with previous chest surgery or injury, certain liver conditions, or other uncommon medical conditions.

Pregnancy does not cause an inherited pulmonary AVM, but changes in blood volume and blood flow may make an existing AVM enlarge or bleed. This is why screening and treatment planning before pregnancy can be important for someone with HHT or a known pulmonary AVM.

Diagnosis

A clinician may suspect a pulmonary AVM because of low oxygen, an unusual chest scan, symptoms, or a personal or family history of HHT.

A contrast echocardiogram is commonly used to look for abnormal blood flow through the lungs. During this ultrasound scan of the heart, saline containing tiny bubbles is injected into a vein. The timing of bubbles reaching the left side of the heart can show whether blood is bypassing the lung capillaries. This is sometimes called a bubble echocardiogram.

A chest CT scan can confirm whether a visible pulmonary AVM is present. CT provides detailed pictures of its feeding artery, abnormal connection, and draining vein. This information helps determine whether embolization may be suitable.

Not every positive bubble test means that a pulmonary AVM large enough to see or treat will be found on CT. Very small abnormal connections can produce a positive test.

Pulmonary angiography uses a catheter and contrast dye to map the lung blood vessels. It is now usually performed as part of an embolization procedure rather than as the first diagnostic test.

Other assessment may include oxygen measurements, blood tests for anemia, and questions about nosebleeds, family history, skin changes, or AVMs in other organs. Genetic testing may help confirm HHT in some families, but a negative test does not rule out every case.

Treatment and management

The main treatment is transcatheter embolization. A specialist passes a thin tube, called a catheter, through a vein, usually from the groin or neck, and guides it into the blood vessel

supplying the AVM. A plug or coil is then placed to block the abnormal blood flow.

Embolization does not require open-chest surgery. Depending on the number and complexity of the AVMs, treatment may be completed in one procedure or several sessions. Temporary chest discomfort can occur afterward.

Surgery is used much less often. It may be considered when embolization is not possible, has not worked, or urgent control of bleeding is needed. The operation may remove the AVM together with a small section of lung.

Very small AVMs that cannot be treated may be monitored. The decision considers the vessel's anatomy, evidence of abnormal blood flow, previous complications, pregnancy plans, and the person's overall health.

Follow-up imaging is important after embolization. Blood flow can occasionally return through a treated vessel, a nearby artery can begin supplying it, or new AVMs can appear. This is particularly relevant in HHT.

People with a pulmonary AVM are often advised to take specific precautions. These may include:

The exact precautions should be confirmed with the team familiar with your AVM and treatment history. Good dental care is also important because mouth bacteria can enter the blood and, in rare cases, contribute to a brain abscess.

Supplemental oxygen may improve low oxygen symptoms in selected people, but it does not close the abnormal vessel.

- Careful removal of air from intravenous lines
- Antibiotic like amoxicillin or doxycycline prevention before certain dental or other procedures that can place bacteria in the bloodstream
- Avoiding scuba diving because pressure changes can increase the risk from small gas bubbles
- Keeping planned follow-up appointments and scans

Complications

A pulmonary AVM can allow a blood clot to bypass the lung filter and travel to the brain. This may cause a transient ischemic attack or stroke. A stroke can happen even when breathing symptoms are mild or absent.

Bacteria can also cross the abnormal connection and cause a brain abscess. Symptoms may include a persistent or severe headache, fever, vomiting, seizures, confusion, or new weakness.

Bleeding from a pulmonary AVM is uncommon but can be serious. Blood may enter the airways, causing coughing up blood, or collect around the lung. Sudden chest pain, breathlessness, dizziness, or fainting can occur.

Other possible problems include ongoing low blood oxygen, reduced exercise capacity, and high red blood cell levels as the body tries to carry more oxygen.

Risks may increase during pregnancy because circulation changes substantially. With screening, treatment when appropriate, and specialist planning, many people with pulmonary AVMs have successful pregnancies.

Prevention

Inherited pulmonary AVMs cannot currently be prevented. A person does not cause them through diet, exercise, or everyday activity.

Complications can often be reduced by finding and treating suitable AVMs before they cause problems. Screening relatives may be appropriate when HHT is confirmed or strongly suspected.

After diagnosis, prevention focuses on follow-up, dental health, safe intravenous care, and any procedure-related antibiotic advice. Treatment before pregnancy may be recommended when a suitable AVM is found.

Embolization greatly reduces blood flow through a treated AVM, but it does not remove the inherited tendency to form abnormal vessels. Continued monitoring remains important.

Living with it and what to expect

Many people return to usual work, school, exercise, and family life after assessment and treatment. Improvement in oxygen levels or exercise tolerance may occur after embolization, although the amount of change differs.

A treated AVM may remain closed for years. However, blood flow can sometimes return, and untreated small AVMs can grow. If you have HHT, new pulmonary AVMs may also develop over time. Follow-up is therefore usually long term, even when you feel well.

Your monitoring schedule may include contrast echocardiography, chest CT, or both. CT scans are not normally repeated more often than needed because they use radiation.

Tell healthcare professionals that you have or have had a pulmonary AVM, especially before an operation, intravenous treatment, or dental procedure. Keep a record of previous embolization

procedures and implanted coils or plugs.

If you are planning a pregnancy, arrange a review in advance when possible. A team experienced in HHT or pulmonary vascular conditions can check whether screening or treatment is needed and plan care during pregnancy.

Emotional effects are also understandable. Some people worry about stroke, bleeding, or passing HHT to a child. Clear follow-up plans and genetic counselling, when relevant, can make these risks easier to understand.

When to get medical help

Seek emergency medical help for:

These symptoms can have causes other than a pulmonary AVM, but they need urgent assessment.

Contact your usual healthcare team promptly if you develop a small amount of blood in your sputum, worsening shortness of breath, lower exercise tolerance, repeated dizziness, or signs of worsening low oxygen.

Routine contact is appropriate if follow-up imaging is overdue or you are planning pregnancy, dental treatment, surgery, or another procedure that may place bacteria or air into the bloodstream.

- Sudden facial drooping, arm or leg weakness, speech trouble, confusion, or loss of vision
- Coughing up more than a small streak of blood
- Sudden severe breathlessness, chest pain, fainting, or blue-grey skin
- A seizure or loss of consciousness
- Severe headache with fever, vomiting, weakness, or confusion

Common questions

Is a pulmonary arteriovenous fistula the same as a pulmonary AVM?

Usually, yes. Pulmonary arteriovenous malformation is the term used more often. Both describe an abnormal direct connection between an artery and a vein in the lung.

Is a pulmonary AVM cancer?

No. It is an abnormal group or connection of blood vessels, not a cancerous growth. It does not spread to other parts of the body like cancer.

Can a pulmonary AVM close on its own?

Most do not reliably close without treatment. Some remain stable, while others grow or change. Embolization is used to block AVMs that are suitable for treatment.

Is it hereditary?

It can be. Pulmonary AVMs are strongly associated with HHT, an inherited condition. A single AVM can also occur without HHT. Your personal history, family history, examination, and sometimes genetic testing help assess the cause.

Can a pulmonary AVM come back after embolization?

Blood flow can occasionally return through a treated AVM. New feeding vessels or new AVMs can also develop. This is why follow-up scans are needed even after successful treatment.

Can I exercise or fly with a pulmonary AVM?

Many people can exercise and travel by air, but low oxygen levels, recent bleeding, pregnancy, or an untreated large AVM may affect the advice. Scuba diving is generally discouraged because gas bubbles can bypass the lungs through the AVM.

Sources

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