CONDITION

Eisenmenger syndrome

Eisenmenger syndrome is severe lung artery disease caused by a congenital heart defect that reverses blood flow and lowers blood oxygen.

WHAT IT IS
Advanced lung artery disease from a congenital heart defect

BODY SYSTEM
Heart, lungs and circulation

MAIN SYMPTOMS
Breathlessness, tiredness, blue-grey skin, fainting

WHAT CAUSES IT
Congenital heart defect with long-term abnormal blood flow

HOW IT IS DIAGNOSED
Echocardiogram, oxygen measurements and cardiac catheterisation

MAIN TREATMENT
Pulmonary hypertension medicines and specialist supportive care

CAN IT BE CURED?
Sometimes - transplantation can provide a cure

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

LONG-TERM OUTLOOK
Serious lifelong condition; treatment can improve symptoms and survival

Overview

Eisenmenger syndrome is a serious, long-term complication of certain congenital heart defects, meaning structural differences in the heart that are present from birth.

Some defects create an abnormal opening or connection between the heart's chambers or major blood vessels. At first, this usually sends extra blood from the left side of the heart to the lungs. Over time, the increased flow can permanently damage and narrow the small lung arteries.

This causes pulmonary arterial hypertension: very high pressure in the arteries carrying blood from the heart to the lungs. Eventually, pressure on the right side of the heart can become high enough to reverse blood flow through the abnormal connection. Blood that has not collected enough oxygen then enters the body's circulation.

Low blood oxygen can make the skin, lips or nails look blue or grey, called cyanosis. It can also affect other organs. Treatment may reduce symptoms and improve survival, but it does not usually restore the lung arteries to normal. Care should be coordinated through a specialist adult congenital heart disease and pulmonary hypertension service.

Symptoms

Symptoms may develop gradually, often years after birth. They can include:

The body responds to low oxygen by making more red blood cells, called secondary erythrocytosis. This helps carry oxygen, but very high levels may contribute to headaches and dizziness. Dehydration and iron deficiency can cause similar symptoms, so blood tests need specialist interpretation.

New symptoms, symptoms at rest or noticeable worsening need medical review. Sudden severe symptoms may require emergency care.

- Shortness of breath, especially during activity
- Tiredness or reduced ability to exercise
- Blue or grey lips, skin, fingers or toes
- Dizziness or fainting
- Chest discomfort or a racing, pounding or irregular heartbeat
- Headaches, poor concentration or blurred vision
- Coughing up blood
- Swelling of the ankles, legs or abdomen
- Clubbing, where the fingertips widen and the nails curve around them

Causes and risk factors

Eisenmenger syndrome develops when a congenital heart defect allows too much blood to flow into the lungs for a long time. The pressure and stress progressively damage the lung arteries.

Associated defects include:

Not everyone with one of these defects develops Eisenmenger syndrome. Risk is higher when the connection is large and is not repaired before permanent lung artery damage occurs.

The original defect often has no identifiable cause. Some congenital heart defects are linked to inherited or chromosome-related conditions, while many occur without a known family cause. Eisenmenger syndrome is not caused by infection or lifestyle choices.

- Ventricular septal defect, an opening between the lower heart chambers
- Atrial septal defect, an opening between the upper chambers
- Patent ductus arteriosus, a blood vessel that remains open after birth
- Atrioventricular septal defect, involving the centre of the heart
- More complex defects or abnormal connections between major blood vessels

Diagnosis

A specialist will review your symptoms, medical history and heart anatomy. Examination may identify cyanosis, clubbing, abnormal heart sounds or signs of heart strain.

Tests may include:

Cardiac catheterisation provides the most direct pressure measurements and may be needed when the diagnosis or treatment plan is uncertain. It should be performed by a team experienced in congenital heart disease and pulmonary hypertension.

- Pulse oximetry: Measures blood oxygen using a finger sensor.
- Blood tests: Check red blood cells, iron, kidney and liver function, and other causes of symptoms.
- Electrocardiogram: Records heart rhythm and may show strain.
- Echocardiogram: Shows heart structure, blood flow and pumping function.
- Chest imaging: An X-ray, CT scan or MRI examines the heart, lungs and blood vessels.
- Exercise testing: Measures the response of the heart, lungs and oxygen level to controlled activity.
- Cardiac catheterisation: Uses a thin tube passed through a blood vessel to measure pressures and blood flow.

Treatment and management

Treatment aims to improve symptoms, protect organs and reduce complications. The plan depends on heart anatomy, oxygen levels, symptoms and test results.

Pulmonary arterial hypertension medicines relax or alter the lung arteries. They include endothelin receptor antagonists, phosphodiesterase-5 inhibitors and medicines acting on the prostacyclin pathway. Some people need more than one medicine. Specialist monitoring is essential.

Oxygen may help selected people, particularly when oxygen levels fall during sleep, illness, activity or air travel. It does not repair the circulation and is not helpful for everyone.

Other management may include:

Routine removal of blood, called phlebotomy or venesection, is generally avoided. The increased red blood cell level helps compensate for low oxygen. Unnecessary removal can cause iron deficiency and reduce oxygen delivery. It may occasionally be considered for significant hyperviscosity symptoms, but only under specialist supervision after excluding other causes.

Blood thinners like warfarin or apixaban are not routinely used because Eisenmenger syndrome can increase both clotting and bleeding risks. They may be prescribed for another reason, such as a particular abnormal heart rhythm.

Closing the original heart defect is usually unsafe once Eisenmenger syndrome is established. Removing this pressure-release pathway can trigger severe right-sided heart failure. Closure should only be considered after detailed expert assessment in unusual circumstances.

For advanced illness that does not respond to treatment, heart-lung transplantation or lung transplantation with repair of the defect may be considered. Transplantation can provide a cure but requires major surgery, lifelong anti-rejection treatment and a suitable donor.

- Treating confirmed iron deficiency
- Managing abnormal heart rhythms
- Using diuretics like furosemide or hydrochlorothiazide for fluid build-up in selected people
- Keeping vaccinations up to date, particularly against respiratory infections
- Planning dental procedures, surgery and anaesthesia with the specialist team
- Reviewing contraception, pregnancy risks and medicines before any pregnancy

Complications

Long-term low oxygen and high lung artery pressure can cause:

Small air bubbles may cross the heart defect instead of being filtered by the lungs, so hospital teams may use special precautions with intravenous lines.

Pregnancy places major additional strain on the circulation. It carries a very high risk of severe complications or death for the pregnant person, as well as miscarriage, premature birth and poor fetal growth.

- Right-sided heart failure
- Abnormal heart rhythms
- Blood clots in the lungs or elsewhere
- Stroke or temporary interruption of brain blood flow
- Bleeding, including coughing up blood
- Iron deficiency or symptoms from very high red blood cell levels
- Kidney or liver problems
- Gout
- Infective endocarditis, an infection of the heart's inner lining
- Sudden deterioration during infection, dehydration, anaesthesia or surgery

Prevention

The congenital heart defect itself often cannot be prevented. However, Eisenmenger syndrome can sometimes be prevented by identifying and repairing a significant defect before permanent lung artery disease develops. Timing depends on the anatomy, blood flow and pressure measurements.

Once irreversible damage and reversed blood flow are established, closing the defect generally cannot prevent or correct the syndrome.

Good dental care helps reduce bloodstream infections that could affect the heart. Some people with unrepaired cyanotic congenital heart disease need preventive antibiotics like amoxicillin or doxycycline before certain dental procedures. Ask the congenital heart team whether this applies to you.

Living with it and what to expect

Eisenmenger syndrome is lifelong unless transplantation is possible. Symptoms and daily ability may improve with pulmonary hypertension treatment, although the abnormal circulation usually remains.

Regular specialist follow-up is essential even when you feel stable. It may include symptom reviews, blood tests, oxygen measurements, heart imaging and exercise assessment.

Regular movement is often beneficial, but intense exertion may cause dizziness, fainting or excessive strain. Stay within limits agreed with your congenital heart team. Stop and rest if you develop unusual breathlessness, dizziness, faintness or chest pain.

Avoid dehydration because fluid loss can make the blood more concentrated and impair circulation. Seek earlier advice during vomiting, diarrhoea, fever or very hot conditions.

Air travel and high altitude can lower oxygen levels. Some people need extra oxygen or should avoid particular journeys. Arrange a pre-travel assessment, especially if your symptoms have changed.

Pregnancy is strongly discouraged because of the high risk. Contraception needs individual planning because some methods increase clotting risk or interact with pulmonary hypertension medicines. Seek specialist advice before considering pregnancy.

Many people continue with work, education, relationships and adapted activities. Emotional support, peer groups and practical adjustments may help with energy limits, appointments and uncertainty.

When to get medical help

Attend all routine specialist appointments. Contact your care team promptly for sustained changes such as increasing breathlessness, reduced activity, new swelling, more frequent palpitations or worsening headaches and dizziness.

Seek prompt medical advice for:

Call emergency services for:

Tell emergency staff that you have Eisenmenger syndrome or cyanotic congenital heart disease. If possible, carry a medical summary listing your heart anatomy, usual oxygen level, medicines and specialist contact details.

- Fever or persistent signs of infection
- Vomiting or diarrhoea that could cause dehydration
- A small amount of blood when coughing
- New or repeated palpitations
- New swelling or rapid weight gain from fluid

- Possible pregnancy or plans to become pregnant
- Severe or sudden difficulty breathing
- Fainting, especially during activity
- Severe or persistent chest pain
- Coughing up more than a small streak of blood
- Signs of stroke, such as facial drooping, arm weakness or speech difficulty
- Blue-grey colouring that is suddenly much worse than usual
- Confusion, collapse or inability to wake normally

Common questions

Is Eisenmenger syndrome the same as pulmonary hypertension?

It is a specific type of pulmonary arterial hypertension caused by long-standing abnormal blood flow through a congenital heart defect. It also involves reversed blood flow and low body oxygen.

Can the hole in my heart be closed?

Usually not after Eisenmenger syndrome is established. Closing it may dangerously increase pressure on the right side of the heart. Assessment requires detailed testing at an expert congenital heart centre.

What is the life expectancy with Eisenmenger syndrome?

Life expectancy is generally shorter than in the wider population, but there is no single figure for an individual. Heart anatomy, heart function, complications and response to treatment all affect outlook. Modern specialist treatment has improved outcomes.

Can I exercise?

Many people can do light or moderate activity within personal limits. Strenuous exercise, heavy lifting and activities that cause chest pain, dizziness or fainting may be unsafe. Ask your specialist team for individual guidance.

Can I fly with Eisenmenger syndrome?

Some people can fly, but lower cabin oxygen may worsen symptoms. A pre-travel assessment can determine whether you need extra oxygen, medicine planning or other precautions.

Why is pregnancy so dangerous?

Pregnancy increases blood volume and makes the heart work harder. Eisenmenger circulation

may not tolerate these changes, and life-threatening complications can occur during pregnancy, delivery or the following weeks. Specialist contraception and pregnancy counselling are essential.

Sources

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