



University Hospital
Southampton
NHS Foundation Trust

Eisenmenger syndrome

Long term care and information for patients



Eisenmenger syndrome is a form of congenital heart disease in which the pressure in the pulmonary (lung) arteries is high, causing an increased resistance to blood flow in the lungs. Congenital means present from birth.

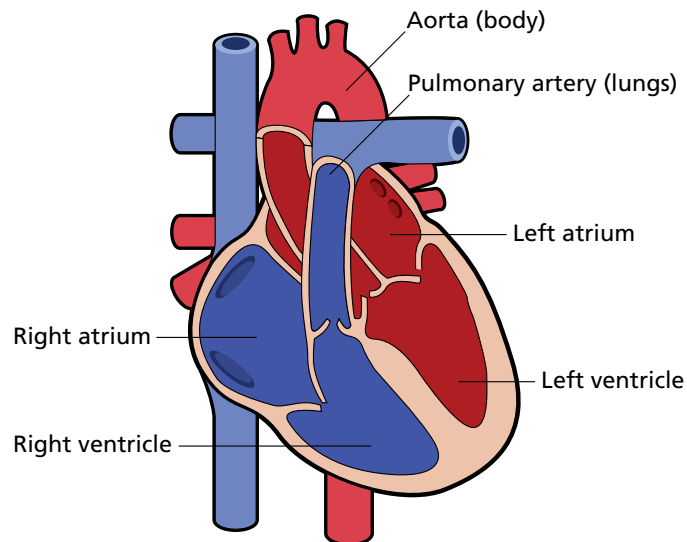
Understanding how the heart works

In order to understand what Eisenmenger syndrome is, it helps to know how the normal heart works (see diagram below).

The heart is a pump. It has four parts: two separate collecting chambers (atria) and two separate pumping chambers (ventricles).

The right side of the heart pumps oxygen-poor (blue) blood to the lungs where oxygen is added. The left heart receives oxygen-rich (red) blood from the lungs and pumps this blood out to the rest of the body.

The blood pressure in the left side of the heart is at least three times higher than in the right side of the heart.



What is Eisenmenger syndrome?

Patients with Eisenmenger syndrome have the following:

- A large hole/communication from birth (congenital), either on the inside or outside of the heart.
 - If the hole/communication is inside the heart, it allows the right and left sided pumping or collecting chambers to connect directly to each other (see diagrams 1 to 3 over the page).
 - If the hole/communication is outside of the heart, it allows the artery going to the lungs (pulmonary artery) to connect directly to the aorta, which is the artery going to the body (see diagram 4 over the page).
- High blood pressure in the lungs.
- A “blue” appearance, known as cyanosis, due to lower than normal oxygen levels.

Causes of Eisenmenger syndrome

When there is a large hole/communication between the left and the right side of the heart, the higher pressure on the left side pushes oxygen-rich (red) blood through the hole/communication into the right side of the heart.

This causes the blood pressure in the right side of the heart to become high, like the left side of the heart. This is known as pulmonary hypertension.

If the hole/communication is not repaired, the high blood pressure on the right side of the heart damages the arteries within the lungs as the person gets older.

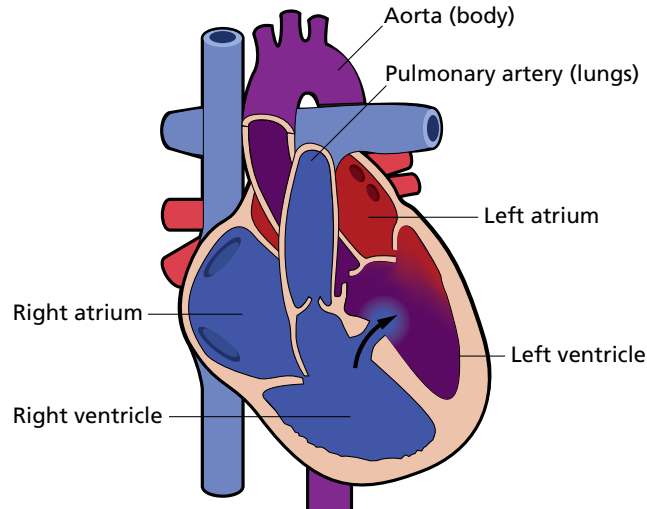
Eventually the damaged lung arteries are under such high pressure that they begin to force oxygen-poor (blue) blood backwards, reversing the direction of blood flow through the hole (at this point the condition is called Eisenmenger syndrome). This results in blood passing from the right side directly to the left side of the heart and around the body, causing the person to appear cyanosed.

Why do I have Eisenmenger syndrome?

You were born with one, or a combination, of the conditions listed below.

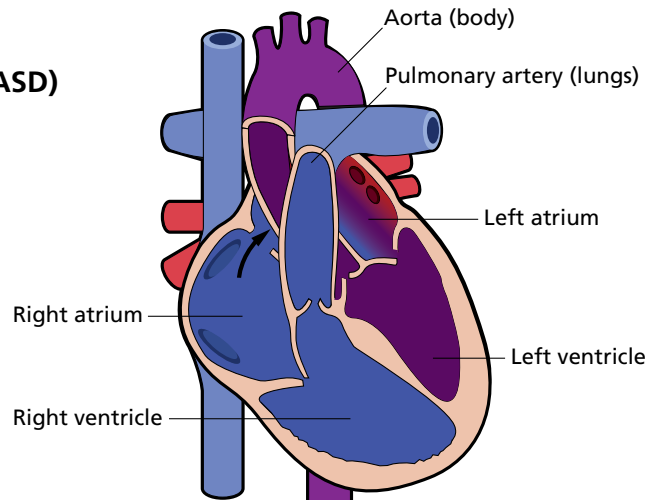
1. Ventricular septal defect (VSD)

A hole between the ventricles of the heart.



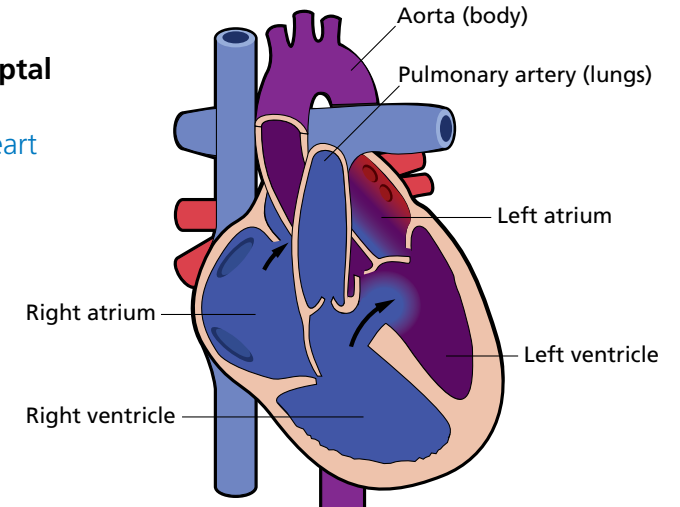
2. Atrial septal defect (ASD)

A hole between the atria of the heart.



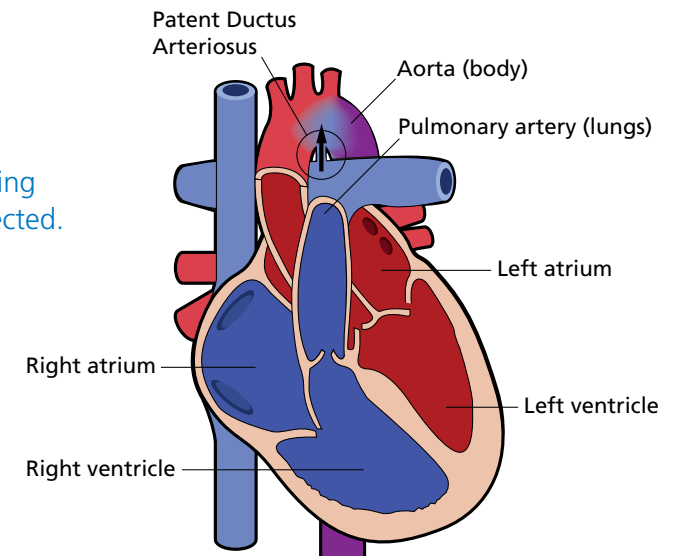
3. Atrioventricular septal defect (AVSD)

A central hole in the heart involving all chambers.



4. Patent ductus arteriosus (PDA)

Two major arteries leaving the heart remain connected.



5. Complex congenital heart conditions

A variety of heart conditions giving rise to higher blood flow to the lungs.

All these conditions mean that there is a hole somewhere in your heart or communication between the arteries leading away from your heart. This causes mixing of red and blue blood as mentioned above which causes damage to your lung arteries.

What are the signs and symptoms of the condition?

You may have one or more of the following:

- “blue” appearance, known as cyanosis, caused by the mixing of blue blood with red blood
- breathlessness on exertion or sometimes even when at rest
- increased tiredness
- headaches
- dizziness or fainting
- heart palpitations (fast or irregular heartbeats)
- chest pain or chest tightness
- numbness and/or tingling of fingers and toes
- blurred vision

How is the condition diagnosed?

Your doctor will use the following tests to make a diagnosis, and to better understand how severe your condition is:

- **Blood tests** – to find out how your body is responding to the lower oxygen levels.
- **Oxygen saturations** – a finger probe to measure how much oxygen is in your blood.
- **6 minute walk test** – to assess your tolerance to a mild level of exercise.
- **Chest x-ray**
- **Electrocardiogram (ECG)** – a tracing of your heartbeat and rhythm.
- **Echocardiogram (echo)** – to look at the way your heart is formed. Also to measure which way blood flows to assess pressures within your heart and to look at the pumping function.
- **Cardiac catheterisation** – a small tube is placed into a blood vessel leading to the inside of your heart. This is used to measure the blood pressure directly in your lungs and to find out how much mixing there is between the red and blue blood.
- **MRI (magnetic resonance imaging)** – a specialised scan to look at the anatomy and function of your heart, aorta and pulmonary arteries.
- **CT scan (computerised tomography)** – a detailed x-ray scan to look at your lung arteries.

What treatments are available?

Treatment for Eisenmenger syndrome is aimed at lowering the blood pressure in your lung arteries. This treatment can help to give you better oxygen levels, greater energy and more stamina when you are active. Your condition should be treated and monitored by a cardiologist who specialises in adult congenital heart disease and pulmonary hypertension.

You may require any or all of the following as treatment for your condition:

- diuretics such as frusemide (water tablets)
- medicines to stabilise your heart rate, rhythm and function
- overnight oxygen (although this is not usually required)
- specific pulmonary hypertension medicines (such as sildenafil, bosentan, ambrisentan and prostacyclines) which work by helping to lower the blood pressure in the lungs by relaxing the small arteries
- when Eisenmenger syndrome is very severe, transplantation may be considered

What are the potential complications?

- **Raised blood thickness** due to increased red blood cell production as the natural response to increase circulating oxygen levels. This may cause additional symptoms, particularly headaches.
- **Iron deficiency anaemia** – with Eisenmenger syndrome your body produces more red blood cells but they are of poorer quality and this may lead to low blood iron levels, causing the anaemia.
- **Blood clotting abnormalities** which may lead to either being more prone to bleeding or excessive clotting. Any damage that results from clots will depend on the location and size of the clot.
- **Reduced heart pumping function** which can lead to heart failure.
- **Kidney and liver abnormalities** can sometimes occur.
- **Gout** – this is when there are very painful, swollen and red joints. Usually only one joint is affected at a time. Gout commonly affects the joints of the toes and feet.
- **Gallstones** – stones in the gallbladder are common. They do not usually generate problems. Occasionally they may cause infection or pain if blockages occur in the gallbladder.

- **Brain abscess** – this is very rare but can occur in people with Eisenmenger syndrome. This may present as a high temperature, headaches, nausea, disorientation or weakness of an arm and/or leg on one side of your body.

What symptoms or conditions should I be worried about?

It is very important to seek help for any of the following. If you are being cared for by your local doctor/hospital please ask for your specialist adult congenital heart disease (ACHD) and pulmonary hypertension doctor/nurse to be notified.

- **Coughing up blood** – go to your closest emergency department for treatment.
- **Any sudden weakness in your arms or legs** – this may mean that you are suffering a stroke. Report to your closest emergency department.
- **Any sudden loss in your vision** – this may mean that you are suffering a stroke. Report to your closest emergency department.
- **Infections or unexplained temperatures/fever** – if serious go to your closest emergency department. However if symptoms are less serious, contact your ACHD/pulmonary hypertension doctor to see where you need to be assessed.
- **If you need to have any operation** – however minor it may seem, please discuss this with your ACHD and pulmonary hypertension doctor to help formulate a safe plan for you. Your heart may be very vulnerable and not cope well with an operation/anaesthetic, so decisions will need to be made carefully.
- **If you need a general anaesthetic** – it is crucial to discuss this with your ACHD/pulmonary hypertension doctor. Anaesthetics, particularly general anaesthetics where you are put to sleep, may cause your heart to become unstable. Therefore it is generally best to perform such procedures at your specialist centre.
- **Feeling faint or passing out** – go to your local emergency department, and ensure your ACHD/pulmonary hypertension doctor or nurse is aware.
- **Heart palpitations** – discuss with your ACHD/pulmonary hypertension doctor or nurse but if symptoms are severe or you feel lightheaded go to your local emergency department.

- **Swelling of your legs and or abdomen** – go to your GP, but also inform your ACHD/pulmonary hypertension doctor or nurse.

Pregnancy

Due to the demands pregnancy places on a mother's heart and lungs, women who have Eisenmenger syndrome should not become pregnant.

Pregnancy for a woman who has Eisenmenger syndrome poses a high risk of death for both the mother and baby, and as a result you will be strongly advised against becoming pregnant. Effective contraception is therefore vital; we recommend progesterone-only preparations, and this will be discussed with you during your visits to clinic.

However, if you do become pregnant, it is essential for your ACHD and pulmonary hypertension doctor to see you as soon as possible to discuss further management. Please make contact through the specialist nursing service (details on the next page).

Important advice if you are admitted to hospital

- Take a supply of pulmonary hypertension medications with you
- Do not miss any doses of your pulmonary hypertension medications
- Take your last heart clinic letter with you
- Ask doctors to contact your specialist pulmonary hypertension centre to inform them of your admission

Other things to be aware of

- Avoid missing a dose of your pulmonary hypertension medications
- Only take medication or herbal remedies after checking with your GP and/or ACHD/pulmonary hypertension doctor or nurse
- Avoid dehydration
- Avoid smoking/passive smoking

- Contact your ACHD/pulmonary hypertension doctor if you need any surgery or other invasive procedures such as endoscopy or dental extractions, particularly if this includes a general anaesthetic.
- You may need to take antibiotics for any invasive dentistry, or for operations or procedures agreed by your ACHD/pulmonary hypertension doctor.
- Look out for signs of infection such as a high temperature and seek help early.
- Avoid excessive physical activity, but keep active within your own capabilities.
- Ensure you are up to date with your annual flu and pneumonia vaccinations.
- Avoid high altitudes in excess of 1500 to 2000 meters above sea level. If this is unavoidable then supplemental oxygen should be used.
- Commercial flights can be undertaken safely as long as your condition is stable and you are not breathless at rest or with very minimal exertion prior to flying. Check with your ACHD/pulmonary hypertension doctor first.
- Ensure that you take with you written information about your condition and contact numbers of your ACHD/pulmonary hypertension doctor/nurse. Then if you need medical attention whilst overseas the doctors managing your situation can access medical information.
- You may find talking to others who have been through similar situations to be helpful, encouraging and reassuring. Relevant support groups are detailed on the back page.

If you have any cause to worry about your condition, or have noticed any changes it is advisable to discuss this with the ACHD/pulmonary hypertension team. Please feel free to contact the specialist pulmonary hypertension service for adult patients with congenital heart disease.

Pulmonary hypertension clinical nurse specialist

Mobile **07717 714724** or

Extension: **023 8120 4739**

Bleep: **023 8077 7222** and bleep **1479**

For urgent medical enquiries out of hours please contact Ward E2 via University Hospital Southampton switchboard – **023 8077 7222**

Description of terms

Aorta – This is the large blood vessel leading from the heart to the rest of the body. Under normal circumstances this carries oxygen-rich (red) blood. However in Eisenmenger Syndrome there is a mixture of oxygen-rich and oxygen-poor blood which accounts for the bluer than normal appearance.

Atria – These are the collecting chambers situated at the back of your heart. On the right side it is known as the right atrium and on the left side it is known as the left atrium.

Cyanosis – Blue tinged skin, lips and finger nails resulting from a lower oxygen level in the blood.

Deoxygenated blood – Blue blood that has not yet passed to the lungs to receive oxygen, and therefore has a low oxygen level (oxygen-poor blood).

Finger clubbing – A thickening of the fingertips that gives a rounded appearance. This is commonly associated with heart, lung and gastrointestinal disorders. The exact cause of finger clubbing is not fully understood, but it is believed to be associated with reduced oxygen circulating in the blood.

Oxygenated blood – Red blood that has passed through the lung vessels and received oxygen into the blood, and therefore has a higher oxygen level (oxygen-rich blood).

Pulmonary artery – This is the large blood vessel leading from the heart to the lungs, carrying oxygen-poor blood to the lungs for the uptake of oxygen.

Pulmonary hypertension – High blood pressure within the blood vessels in the lungs.

Ventricle – This is the part of your heart that pumps blood either into the lungs (the right ventricle) or around your body (the left ventricle).

Information and support groups

The following charities may be useful for providing additional information and support:

Pulmonary Hypertension Association

Email: office@phauk.org

Telephone: **01709 761450**

Website: www.phauk.org

The Somerville Foundation Patient Association

Email: helpline@thesf.org.uk

Telephone: **0800 854 759**

Website: www.thesf.org.uk

Downs Syndrome Association

Email: info@downs-syndrome.org.uk

Telephone: **0333 1212 300**

Website: www.downs-syndrome.org.uk

British Heart Foundation

Heart Helpline: **0300 330 3311**

Website: www.bhf.org.uk

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For help preparing for your visit, arranging an interpreter or accessing the hospital, please visit www.uhs.nhs.uk/additionalneeds

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