CONDITION

Aortopulmonary window

Learn how an aortopulmonary window affects blood flow, the symptoms it may cause, and how specialists diagnose, repair, and monitor this rare congenital heart defect.

WHAT IT IS
Rare congenital heart defect

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Fast or difficult breathing, feeding fatigue, slow weight gain, fast heartbeat

WHAT CAUSES IT
Unknown developmental change

HOW IT IS DIAGNOSED
Echocardiogram

MAIN TREATMENT
Surgical or catheter-based closure

CAN IT BE CURED?
Sometimes, closure definitively treats the opening

IS IT AN EMERGENCY?
Sometimes, with severe trouble breathing

LONG-TERM OUTLOOK
Good before permanent lung vessel damage

Overview

An aortopulmonary window is a rare congenital heart defect. Congenital means that it is present at birth. In this condition, there is an abnormal opening between two major blood vessels leaving the heart: the aorta and the pulmonary artery.

The aorta carries oxygen-rich blood from the heart to the body. The pulmonary artery carries oxygen-poor blood from the heart to the lungs. Normally, these vessels are completely separate.

An aortopulmonary window allows blood to pass directly from the aorta into the pulmonary artery. Because pressure is usually higher in the aorta, a large amount of blood may flow back toward the lungs. This is called a left-to-right shunt.

The extra blood flow makes the heart and lungs work harder. A large opening can lead to heart failure during infancy and can damage the blood vessels in the lungs. Over time, that damage may cause pulmonary hypertension, which means unusually high pressure in the lung arteries.

An aortopulmonary window is different from truncus arteriosus, another congenital heart defect. In truncus arteriosus, one large blood vessel leaves the heart instead of a separate aorta and pulmonary artery. With an aortopulmonary window, the two vessels and their valves are separate but have an opening between them.

The opening can vary in size and location. Some extend close to the heart, while others are farther along the vessels. An aortopulmonary window may occur by itself or with another heart or blood vessel abnormality, such as an interrupted or narrowed aortic arch.

Because ongoing excess blood flow can permanently affect the lung arteries, specialists usually recommend closing the opening after diagnosis if it can be done safely.

Symptoms

Symptoms depend mainly on the size of the opening and whether other heart defects are present. A large opening can cause symptoms within the first days or weeks of life. A small opening may cause few symptoms and may be found during an examination for another reason.

Possible symptoms in a baby include:

These symptoms can occur with many childhood conditions and do not confirm an aortopulmonary window.

During an examination, a clinician may hear a heart murmur, which is an extra sound caused by turbulent blood flow. The pulses may feel unusually strong or "bounding." Some children have signs of an enlarged or overworked heart.

Blue or gray skin is not usually an early feature of an isolated aortopulmonary window because oxygen-rich blood initially flows toward the lungs. Blue coloring may occur if there is another heart defect or if severe pulmonary hypertension eventually reverses the direction of blood flow.

- Fast or difficult breathing
- Tiring, sweating, or breathing harder during feeding
- Taking a long time to feed or being unable to finish feeds
- Slow weight gain

- Irritability or unusual sleepiness
- Frequent respiratory illnesses
- A fast heartbeat
- Swelling, although this is less common in young infants

Causes and risk factors

An aortopulmonary window forms early in pregnancy while the heart and major blood vessels are developing. Tissue that should completely separate the aorta from the pulmonary artery does not form normally, leaving an opening.

In most cases, the exact reason for this developmental change is unknown. It is not considered the result of something a parent did or did not do during pregnancy.

There are no well-established lifestyle risk factors specific to an aortopulmonary window. Some affected children have other congenital heart abnormalities, including:

The presence of another abnormality can change symptoms, treatment planning, and the timing of surgery. A genetics evaluation may be offered when a baby has several birth differences, developmental concerns, or a family history suggesting an inherited condition. An isolated aortopulmonary window does not by itself establish a genetic disorder.

- An interrupted aortic arch, in which part of the aorta does not connect normally
- Coarctation, or narrowing, of the aorta
- Abnormalities involving the pulmonary arteries
- A ventricular septal defect, which is an opening between the heart's lower chambers
- Tetralogy of Fallot or other complex congenital heart conditions
- Unusual origins of the coronary arteries, which supply blood to the heart muscle

Diagnosis

An aortopulmonary window may be suspected because of symptoms, a heart murmur, strong pulses, or signs of heart failure. It can occasionally be seen before birth during a fetal echocardiogram, but prenatal diagnosis may be difficult.

An echocardiogram is the main diagnostic test. This painless ultrasound examination shows the heart's structure, blood flow, and pumping function. It can identify the opening, estimate

how much blood is passing through it, and look for associated heart defects.

Other tests may include:

Cardiac catheterization is not necessary for every child. It may be especially helpful when diagnosis has been delayed, pulmonary hypertension is suspected, or a catheter-based closure is being considered.

The care team must also distinguish an aortopulmonary window from conditions that can produce similar blood-flow patterns, such as a patent ductus arteriosus, truncus arteriosus, or certain abnormal artery connections.

- Pulse oximetry: A small sensor measures the amount of oxygen in the blood.
- Electrocardiogram: Sensors placed on the skin record the heart's electrical activity and may show strain or enlargement of the heart chambers.
- Chest X-ray: This may show an enlarged heart or increased blood flow through the lungs.
- Cardiac CT or MRI: These scans provide detailed images when the opening or nearby blood vessels cannot be fully assessed by echocardiography.
- Cardiac catheterization: A specialist passes a thin tube through a blood vessel to the heart. This can measure pressures, calculate blood flow, and assess resistance in the lung arteries.

Treatment and management

Closing the abnormal opening is the definitive treatment. The timing and method depend on the child's age, symptoms, anatomy, associated heart defects, and pressure in the lung arteries.

For most infants with a large aortopulmonary window, surgical repair is recommended early because excess lung blood flow can quickly lead to heart failure and pulmonary vascular disease. During surgery, the surgeon closes the opening, often with a patch, while preserving both major vessels. Other heart or aortic abnormalities may be repaired during the same operation.

Some small, well-positioned openings may be closed during cardiac catheterization. A specialist guides a closure device through a blood vessel and places it across the opening. This approach is only suitable when there is enough tissue to hold the device safely and when it will not block the aorta, pulmonary arteries, heart valves, or coronary arteries. Surgical repair remains necessary for many patients.

Before repair, medicines may be used to manage heart failure symptoms. These can help the

body remove excess fluid and reduce the heart's workload. Some babies also need higher-calorie feeding, feeding-tube support, or respiratory support. These measures can stabilize a baby and improve growth, but they do not close the opening.

If pulmonary hypertension is advanced, the team may perform additional testing before recommending closure. When lung blood vessel damage has become severe and irreversible, closing the opening can be dangerous because the right side of the heart may no longer be able to pump effectively against the high pressure. These decisions require evaluation at a center experienced in congenital heart disease and pulmonary hypertension.

After surgery or catheter treatment, care may include pain relief, monitoring of heart rhythm and blood pressure, and repeat imaging. Recovery depends partly on whether the child had heart failure, pulmonary hypertension, or another heart defect before treatment.

Complications

Without treatment, a substantial aortopulmonary window can cause complications from prolonged excess blood flow to the lungs. These may include poor growth, enlargement of the heart, heart failure, and repeated respiratory problems.

Pulmonary hypertension is one of the most important concerns. Persistent high blood flow can thicken and narrow the small arteries in the lungs. If this becomes irreversible, blood may eventually flow from the pulmonary artery toward the aorta. This advanced condition can lower oxygen levels and cause blue or gray coloring, fatigue, and reduced exercise ability.

Possible complications after repair include:

Serious complications are not inevitable. Children treated before permanent lung vessel damage develops may have good heart function and normal or near-normal activity. Individual outcomes depend on the anatomy, the timing of treatment, and associated conditions.

Infective endocarditis is an infection involving the inner lining of the heart or nearby material used in a repair. Antibiotics like amoxicillin or doxycycline before dental procedures are recommended only for certain higher-risk heart conditions and time periods. A cardiologist or dentist can explain whether preventive antibiotics apply to a specific patient.

- A small residual opening or leak
- Narrowing of the aorta or pulmonary artery near the repair
- Persistent pulmonary hypertension
- Heart rhythm problems

- Problems involving a closure device, if one was used
- The need for another catheter procedure or operation

Follow up

Regular congenital cardiology follow-up remains important after a successful repair. The cardiologist will check growth, blood pressure, heart sounds, exercise tolerance, and oxygen level. Echocardiograms may be used to look for a residual opening, narrowing near the repair, pressure in the lung arteries, or changes in heart function.

The schedule varies according to the repair and the presence of other heart defects. Some people need only periodic routine visits after a complete repair, while those with residual problems or pulmonary hypertension need closer monitoring. Children should transition to an adult congenital heart disease specialist as they reach adulthood rather than relying only on general adult cardiac care.

The care team can advise about physical activity. Many people with a fully repaired defect and normal heart and lung pressures can participate in ordinary activities. Restrictions may be needed when there is pulmonary hypertension, vessel narrowing, abnormal heart function, or another heart condition.

Contact the clinical team promptly for worsening feeding, new sweating with feeds, poor weight gain, increasing breathlessness, swelling, fewer wet diapers, unusual sleepiness, fever after a recent procedure, or redness and drainage from a surgical wound. These concerns need timely assessment but are not always emergencies.

Seek emergency care for severe trouble breathing, blue or gray lips or skin, collapse, inability to wake normally, or a sudden major decline in responsiveness. Follow any emergency plan provided by the congenital heart team.

Sources

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