CONDITION

Anomalous left coronary artery from the pulmonary artery

Learn how anomalous left coronary artery from the pulmonary artery affects heart blood flow, its symptoms, diagnosis, surgery, and lifelong follow-up.

WHAT IT IS
Congenital abnormal origin of the left coronary artery

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Difficult breathing, feeding-related sweating, poor weight gain, chest discomfort

WHAT CAUSES IT
Unknown developmental cause before birth

HOW IT IS DIAGNOSED
Coronary imaging with CT, MRI, or angiography

MAIN TREATMENT
Surgery

CAN IT BE CURED?
No, lifelong specialist follow-up remains essential

IS IT AN EMERGENCY?
Sometimes, with sudden collapse

LONG-TERM OUTLOOK
Often substantial improvement after surgery, but lifelong risks remain

Overview

Anomalous left coronary artery from the pulmonary artery, often shortened to ALCAPA, is a congenital heart condition, meaning it is present at birth.

Normally, the left coronary artery begins at the aorta and supplies much of the heart muscle with oxygen-rich blood. In ALCAPA, it begins at the pulmonary artery, which carries blood from the heart to the lungs.

Before birth and during the first days of life, pressure in the pulmonary artery is relatively high. After birth, that pressure falls as the lungs begin working. Blood in the pulmonary artery also contains less oxygen than blood in the aorta. The left side of the heart muscle may therefore

receive too little oxygen-rich blood.

Blood may also flow away from the left coronary artery into the lower-pressure pulmonary artery, a process sometimes called coronary steal. This can injure the heart muscle, weaken its pumping ability, and cause leakage through the mitral valve.

Some people develop collateral vessels connecting the right and left coronary arteries. These may allow survival beyond infancy, but they do not restore normal circulation. Heart muscle injury, abnormal rhythms, or sudden cardiac arrest can still occur.

Surgery is generally recommended to create a two-coronary-artery circulation in which both coronary arteries receive oxygen-rich blood from the aorta.

Symptoms

Symptoms depend on age, coronary blood flow, heart function, and whether collateral vessels have developed.

Infants often develop symptoms during the first months of life as pulmonary artery pressure falls. Symptoms may become more noticeable during feeding or crying, when the heart needs more oxygen.

Possible symptoms include:

Feeding-related distress may be mistaken for colic or a feeding problem. A clinician must assess the overall pattern, examination findings, and test results.

Older children and adults may have no obvious symptoms for years. Others develop chest discomfort with activity, shortness of breath, reduced exercise ability, dizziness, fainting, or fast or irregular heartbeats. A murmur, abnormal electrocardiogram, or enlarged heart may lead to diagnosis.

Symptoms alone cannot confirm ALCAPA because many other conditions can cause similar problems.

Seek emergency care for severe or persistent chest pain, fainting during exercise, severe difficulty breathing, blue or gray skin, sudden collapse, or signs of cardiac arrest. An infant needs emergency assessment for marked breathing difficulty, poor responsiveness, or blue or gray coloring.

Contact a clinician promptly about feeding-related sweating or distress, poor growth, unexplained chest discomfort, repeated palpitations, worsening shortness of breath, or declining exercise ability.

- Fast or difficult breathing
- Sweating, pallor, or distress during feeding
- Poor feeding or slow weight gain
- Irritability or intense crying episodes
- Tiring quickly during feeding
- Unusual sleepiness or reduced activity
- Signs of heart failure, such as rapid breathing or swelling

Causes and risk factors

ALCAPA develops while the heart and major blood vessels are forming before birth. The exact developmental event is not fully understood. The opening of the left coronary artery connects to the pulmonary artery instead of the aorta.

The condition is not caused by exercise, diet, stress, or anything a child or adult did. There is usually no preventable cause or established lifestyle risk factor. ALCAPA often occurs alone, although other structural heart differences may occasionally be present.

Most cases do not follow a simple inherited pattern. A cardiology or genetics team may review family history and consider genetic evaluation when other congenital abnormalities or signs of an inherited syndrome are present. Screening recommendations for relatives vary by family.

Diagnosis

Diagnosis usually involves a pediatric or adult congenital cardiologist. Testing determines where the coronary arteries begin, how blood flows, and whether the heart muscle or mitral valve has been affected.

An electrocardiogram (ECG) records electrical activity and may show patterns of heart muscle injury or abnormal rhythms. It cannot define the complete coronary anatomy.

An echocardiogram uses sound waves to create moving images of the heart. It may show the abnormal coronary origin or blood flow and can assess:

A coronary CT angiogram uses X-rays and contrast material to produce detailed images of the coronary arteries. Because it involves radiation, imaging teams use carefully selected techniques, especially for children.

Cardiac MRI can evaluate heart structure, function, and scar tissue without ionizing radiation.

Younger children may need sedation to remain still.

Cardiac catheterization and coronary angiography involve passing a thin tube through a blood vessel to the heart and using contrast dye to outline blood flow. They may be needed when other imaging is inconclusive or clinicians require detailed pressure or anatomy measurements before treatment.

- Heart pumping strength
- Enlargement or abnormal movement of the left ventricle
- Mitral valve leakage
- Blood flow from the coronary artery into the pulmonary artery

Treatment and management

Surgery is the main treatment. Major congenital heart disease guidelines recommend repair because reduced coronary blood flow may cause progressive heart damage or dangerous rhythms even when symptoms are mild or absent.

The goal is to connect both coronary arteries to oxygen-rich blood from the aorta. When possible, surgeons detach the left coronary artery from the pulmonary artery and reconnect it directly to the aorta. This is called coronary reimplantation.

If direct reimplantation is not practical, surgeons may create a tunnel inside the pulmonary artery that directs blood from the aorta to the left coronary artery. This is called a Takeuchi repair. The tunnel and nearby pulmonary artery require long-term monitoring.

In selected people, particularly adults with unusual anatomy, surgeons may close the abnormal connection and perform coronary artery bypass grafting. The approach depends on age, anatomy, previous operations, heart function, and surgical expertise.

Before or after surgery, treatment may be needed for heart failure, fluid buildup, or abnormal rhythms. Medicines are selected according to age, heart function, blood pressure, and other findings. Severe heart failure may temporarily require mechanical circulatory support. Heart transplantation is uncommon but may be considered for irreversible damage that cannot be managed otherwise.

Mitral valve leakage often improves after coronary blood flow and heart function recover. Valve repair during the same operation may be appropriate when leakage is severe or structural valve changes are present.

Many infants and children have substantial improvement after surgery, although recovery may take months. Existing scar tissue may remain. Physical activity and return to sports should be

guided by a congenital cardiologist based on heart function, coronary blood flow, rhythm testing, and scar tissue.

Complications

Without repair, ALCAPA may cause:

Complications can also occur after repair, although surgery greatly improves blood flow. A reimplanted coronary artery may rarely narrow. After a Takeuchi repair, the internal tunnel may narrow or leak, and narrowing can develop in the pulmonary artery.

Some people continue to have reduced heart function, mitral valve leakage, or scar-related rhythms. Additional imaging, rhythm monitoring, catheter procedures, or surgery may be needed.

Emergency diagnosis, infant surgery, and activity restrictions can also cause emotional stress. Families may ask the congenital heart team about counseling, cardiac rehabilitation when appropriate, and age-specific support.

- Heart failure
- Heart muscle injury or heart attack
- Moderate or severe mitral valve leakage
- Ventricular arrhythmias
- Sudden cardiac arrest

Follow up

Lifelong follow-up with a congenital heart disease specialist is essential after ALCAPA repair, even when a person feels well. Monitoring can identify changes before symptoms develop.

The schedule depends on the operation, heart function, mitral valve leakage, coronary anatomy, and rhythm history. Follow-up may include an examination, ECG, echocardiogram, exercise testing, rhythm monitoring, CT, or cardiac MRI.

Patients and families should keep copies of the original anatomy records, operative report, and later imaging. Adults treated during childhood should receive care through an adult congenital heart disease program rather than relying only on general cardiology care.

Anyone with repaired ALCAPA who is considering pregnancy should have a preconception assessment with an adult congenital cardiologist and a pregnancy team experienced in heart

disease.

Contact the cardiology team sooner than scheduled for new palpitations, chest discomfort, fainting, worsening shortness of breath, swelling, or reduced exercise tolerance.

Call emergency services for collapse, severe breathing difficulty, persistent severe chest pain, or signs of cardiac arrest.

Sources

- Stout KK, Daniels CJ, Aboulhosn JA, et al. "2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease." Circulation . 2019;139(14):e698-e800.
- Baumgartner H, De Backer J, Babu-Narayan SV, et al. "2020 ESC Guidelines for the Management of Adult Congenital Heart Disease." European Heart Journal . 2021;42(6):563-645.
- Peña E, Nguyen ET, Merchant N, Dennie C. "ALCAPA Syndrome: Not Just a Pediatric Disease." Radiographics . 2009;29(2):553-565.
- Dodge-Khatami A, Mavroudis C, Backer CL. "Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery: Collective Review of Surgical Therapy." The Annals of Thoracic Surgery . 2002;74(3):946-955.
- Yau JM, Singh R, Halpern EJ, Fischman D. "Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery in Adults: A Comprehensive Review of 151 Adult Cases and a New Diagnosis in a 53-Year-Old Woman." Clinical Cardiology . 2011;34(4):204-210.
- Naimo PS, Fricke TA, d'Udekem Y, et al. "Surgical Intervention for Anomalous Origin of Left Coronary Artery From the Pulmonary Artery in Children: A Long-Term Follow-Up." The Annals of Thoracic Surgery . 2016;101(5):1842-1848.