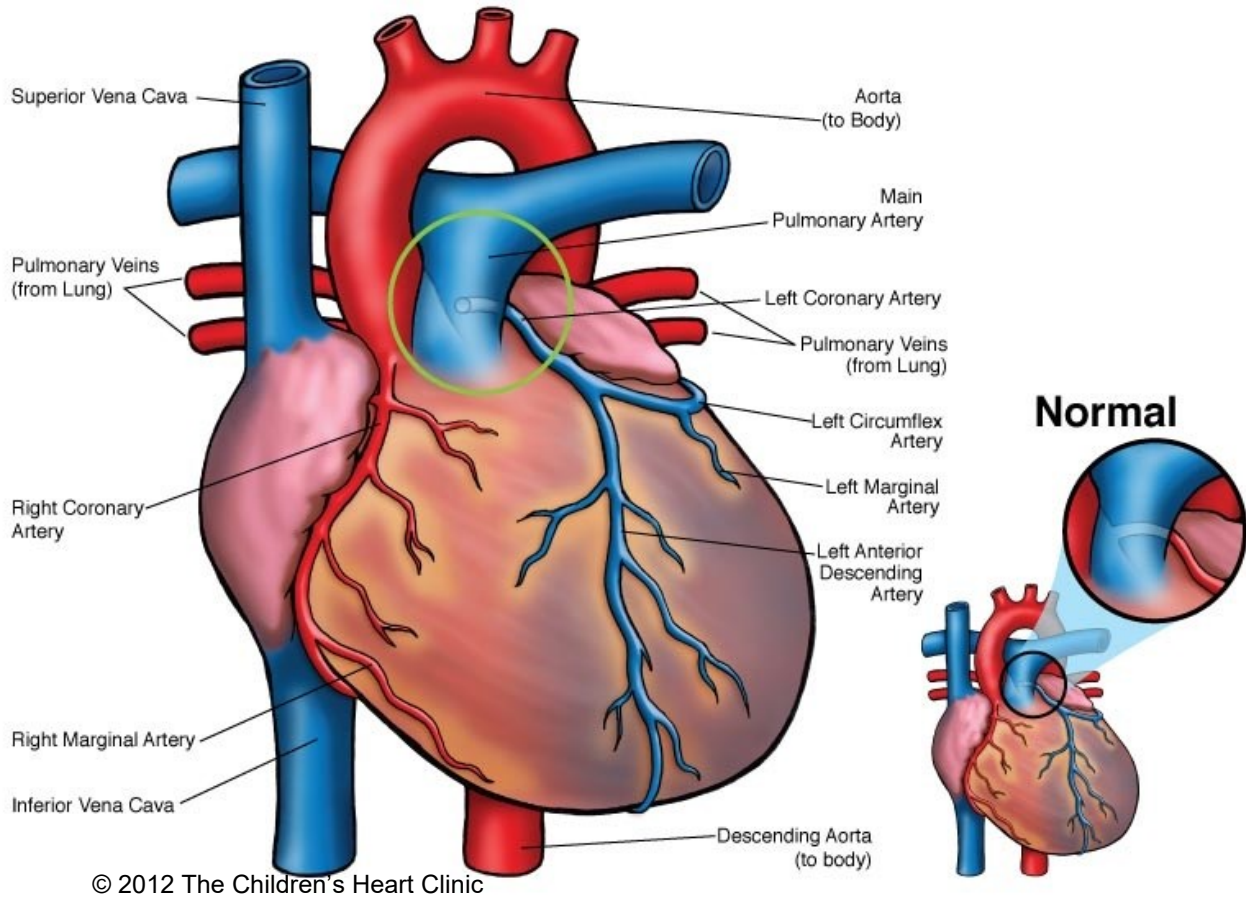


Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA)



Notes:

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In the normal heart, the left coronary artery arises from the aorta, above the left cusp of the aortic valve. This arrangement allows the left coronary artery to provide the left ventricle with oxygenated blood. When the left coronary artery arises abnormally from the pulmonary artery, this is known as ALCAPA. In ALCAPA, blood flow goes from the right coronary artery, through the inter-coronary collaterals to the left coronary artery to the pulmonary artery. This arrangement results in insufficient oxygen supply to the left ventricle, resulting in ischemia (restriction of blood flow) or infarction (severe injury to heart muscle cells). This is an extremely rare congenital heart defect, occurring in 0.5% of individuals with congenital heart disease.

Physical Exam/Symptoms:

- Newborns are asymptomatic.
- Symptoms usually present at about 2-3 months of age, when pulmonary artery pressures fall.
- Cardiomegaly (enlarged heart), episodes of distress/agitation due to anginal pain (severe chest pain due to decreased oxygen supply to the heart muscle) and congestive heart failure (CHF) occur.
- Mitral regurgitation may occur in the setting of left myocardial infarction. In this case, a regurgitant systolic murmur may be present, heard best at the heart apex.

Diagnostics:

- EKG: Deep & wide Q waves, inverted T waves and ST segment changes.
- Coronary angiography: Gold standard for diagnosis.
- Echocardiogram: Aids in diagnoses and evaluation for additional cardiac anomalies.

Medical Management/Treatment:

- 80-100% mortality with medical management alone.
- Surgery to create a two-coronary artery system:
 - Left coronary artery transfer: A “button” or flap around the opening of the left coronary artery is cut and then re-implanted onto the aorta.
 - Coronary artery bypass graft.
- Lifelong cardiology follow up is needed.

Long-term Outcomes:

- If uncorrected, nearly 85% of children with ALCAPA die within the first year of life due to severe left-sided heart failure.
- Early repair is associated with better outcomes/faster recovery time, if left ventricular failure is not severe.
- Presence of mitral regurgitation may require future mitral valve repair/replacement.
- A small number of patients present in adulthood due to development of extensive collateral vessels that supply the left ventricle with adequate oxygenated blood and “stealing” of oxygenated blood from the right coronary circulation.
- In general, with return of adequate heart function post-operatively, life expectancy is normal.