

Euro Heart Congress 2020: Anomalous Left Coronary Artery from the Right Pulmonary Artery

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Abstract

Introduction:

One of the rarest but highly serious congenital cardiac anomalies can be distinguished as Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). Anomalous left coronary artery from the pulmonary artery (ALCAPA) is also a kind of heart defect. The artery that carries the blood to the heart muscles i.e., left coronary artery (LCA), actually begins from the pulmonary artery instead of the aorta.

History: The term ALCAPA was first described in 1866. Bland and colleagues in the year 1933 described the first clinical description in conjunction with the autopsy findings, and hence this particular anomaly is named as Bland-White-Garland syndrome. In the year 1962, a series of 58 post-mortem specimens that demonstrated that most patients had died at a young age was reported by cardiologists Fontana and Edwards.

The Description of the abnormality is given as follows: Both coronary arteries should arise (branch) from the aorta in case of a normal heart. However in the case of an anomalous left coronary artery from the pulmonary artery (ALCAPA), while the heart of an infant is forming in the mother's womb something goes wrong, and the left coronary artery arises from the right pulmonary artery instead of arising from the aorta. And the medical personalities have named this type of heart defect as "anomalous origin of the left coronary artery from the pulmonary artery." The word Anomalous means irregular. 1 in every 300 000 live births has been recorded with this heart defect of Anomalous origins of the left coronary artery arising from the pulmonary artery (ALCAPA).

The most common origin of the abnormal LCA is from the pulmonary truncal sinuses. In the most common form of ALCAPA, the abnormal coronary artery arises from the adjacent pulmonary valvar sinus, rather than the pulmonary trunk. In this case, the Anomalous Left Coronary Artery originated the Right Pulmonary Artery. Such case has an incident of 1 in 2,000,000 live births. This is the first reported case in a tertiary cardiovascular referral center. In fetal life, this has no detrimental effect since pressures and saturations are similar in the aorta and pulmonary artery. After birth, however, the pulmonary artery contains de-saturated blood at pressures that rapidly fall below systemic pressures. The left ventricle is perfused with de-saturated blood

at low pressures leading to infarction with ventricular dysfunction. Coronary translocation and Lecompton maneuver was done which provided relief for the patient's condition. In children suffering from with ALCAPA heart defect, the left coronary artery arises from the pulmonary artery and it carries blood without oxygen to the left side of the heart. In this situation, the heart muscles can weaken or die which is similar to having a heart attack because the heart doesn't get enough oxygen. Hence the damaged heart muscle cannot pump effectively, which becomes the route cause for cardiomyopathy and heart failure. In infants, ALCAPA is a very rare and dangerous health condition that may cause dangerously poor cardiac function. Performing Surgery is a must in order to correct the defect. Without intervention, most babies don't survive their first year, but with timely surgery, most babies do well and live a normal life.

The rarest form of ALCAPA presents with anomalous left coronary artery arising from the right pulmonary artery. This is a case of 1 month old female presenting with Dyspnea, 2D echocardiography revealed ALCAPA. Intraoperatively, the Left Coronary Artery was found to be originating from the Right Pulmonary Artery. The patient underwent coronary implantation and LeCompte procedure.

Causes:

ALCAPA is a heart defect or a problem that is most likely to occur early in the pregnancy when the baby's heart is still developing. This issue arises when the developing blood vessel does not attach to the heart muscle correctly.

The LCA originates from the pulmonary artery in children suffering with the rarest disease of ALCAPA. The pulmonary artery which acts as the major blood vessel that pumps oxygen and poor blood from the heart to the lungs to pick up oxygen.

Hence the blood that is lacking or insufficient in oxygen is carried to the heart muscle on the left side of the heart when this defect occurs. And hence the heart muscle does not get enough of oxygen. As a result of which, the tissue begins to die due to lack of oxygen. This can become the root cause a heart attack in the baby.

Another deadliest condition which is known as "coronary steal" evokes the further damages the heart in babies with ALCAPA. The blood from the abnormally connected LCA is flowed back toward the pulmonary artery instead of toward the heart muscle because of the low blood pressure in the

pulmonary artery. This backward flow causes and results in less blood and oxygen transferred to the heart muscle. This problem can also lead to a heart attack in a baby. If the condition is not treated as early as possible in babies with ALCAPA, Coronary steal develops over time.

Treatment: Surgery is needed to treat anomalous left coronary artery from the pulmonary artery (ALCAPA). There are mainly two options for repair which are performed commonly, they are 1. Detaching the anomalous left coronary artery from the pulmonary artery and moving it over to the aorta directly (translocation) 2. With the help of a Takeuchi repair method that includes creating a natural tunnel from its abnormal location to the aorta. If the mitral valve is leaking, it can be repaired at the same time. However in some extreme cases where the heart is severely damaged the support of mechanical circulatory/ventricular assist device may be required and a heart transplant may be needed.

Though ALCAPA is a serious issue with the help of a timely therapy, most children do very well once after the surgery is performed and can be expected to live a normal and healthy life. However, there should be routine checkup and life-long follow-up with a heart-failure specialist will be necessary to ensure recovery of the heart muscle and good function of the mitral valve. An increased risk for heart-rhythm problems later in life time can be seen in children born with ALCAPA. However, adults have been reported to survive better owing to the development of collateral perfusion from right coronary artery. In these cases, however, revascularization is necessary to prevent sudden cardiac death.

The Takeuchi procedure for treating ALCAPA refers to performing a direct anastomosis of the anomalous left coronary artery from the pulmonary artery directly to the aorta was described in the 1970s and currently remains the best procedure of choice.

An intrapulmonary aorta-coronary tunnel or baffle was performed by Takeuchi prior to the above procedure. If the above procedure is not feasible due to coronary anatomy or due to lack of surgical experience this surgery is performed even today.

Medication:

In order to build up the strength necessary for the procedure, some children may need medications prior to surgery. These medications for building up the strength may include:

- Diuretics: "water pills"
- Inotropic agents: medications that can help the heart muscles to pump harder the beta-blockers,
- ACE inhibitors: medications that help in lower the work load on the heart.
- In addition, some of these medications may also be needed after surgery.

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