

Anomalous left coronary artery from pulmonary artery (ALCAPA): A cause of myocardial ischemia in children

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Abstract

ALCAPA is a rare congenital anomaly of the coronary system of the heart in which the left coronary artery, which normally arises from the aorta, takes its origin from the pulmonary artery. It is a cause of myocardial ischemia in infancy and causes up to 90% mortality in the first year of life if left untreated. Diagnosis has become easy with the advent of 2D echocardiography. Surgery is the mainstay of therapy with good short and long term results.

Keywords: ALCAPA, Myocardial ischemia, Pulmonary artery, Coronary artery.

ALCAPA or Bland White Garland syndrome is a rare congenital anomaly of the heart in which the left coronary artery arises from the pulmonary artery, instead of aorta. Literature reports the incidence to be 1 in 3 lakh live births. The portion of the left ventricle which is supplied by this anomalously originating artery is thin, scarred and dilated and is a cause of myocardial ischemia in children. It is usually an isolated congenital anomaly although it might sometimes be associated with patent ductus arteriosus, ventricular septal defect, tetralogy of fallot or coarctation of aorta.

The myocardial ischemia in ALCAPA is not due to the perfusion by deoxygenated blood or due to a single normally functioning coronary artery (right coronary artery), but due to the direction of blood flow through the coronary bed. In the fetal and early neonatal life, owing to increased pulmonary arterial pressures, there is ante grade flow into anomalous coronary artery from the pulmonary artery. Subsequently, the neonatal pulmonary vascular resistance decreases and there is a parallel fall in blood flow in anomalous left coronary artery. At this time the perfusion of cardiac myocardium depends entirely on right coronary artery. Hence, ischemia is unavoidable unless there is adequate circulation from right to left coronary artery via inter coronary anastomosis. Initially this ischemia is transient which

occurs mainly during feeding, crying and exertion. Even if collaterals are adequate, left coronary artery is connected to a low pressure pulmonary artery, so this collateral flow tends to pass into pulmonary artery rather than into higher pressure myocardial blood vessels (pulmonary coronary steal with left to right shunt).

There can be three types of presentation of these children. Firstly, they can present with serious symptoms in early infancy. The classical history is that of acute episodes of irritability and pallor which occurs secondary to myocardial ischemia, typically during feeding and distress. Without treatment 80-85% of these children will not survive their infancy. They have poor growth and development due to congestive heart failure. The second group of children can present with early symptoms initially, followed by gradual attenuation of symptoms. Another 10% of children will have absence of early symptoms with asymptomatic survival till adulthood. These adults can present with mitral regurgitation, angina, myocardial infarction, congestive cardiac failure, atrial fibrillation, ventricular tachyarrhythmia. Approximately one third of these patients will have sudden cardiac death.

On examination of these children they are found to have rapid labored breathing, weak cry, cough and diaphoresis. Pallor accompanies episodes of

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symptomatic myocardial ischemia. Features of congestive heart failure are present with raised jugular venous pressure. Pulmonary arterial hypertension with loud second heart sound and right ventricular enlargement is a feature in ALCAPA. Pan-systolic murmur due to mitral regurgitation and continuous murmur due to inter coronary anastomosis are present.

Deep narrowed q waves in leads I, aVL, V4 and V5 with q wave width of >30 msec in lead I and a depth of > 3 mm in lead aVL with a QR pattern in aVL is significantly associated with ALCAPA. Left ventricular hypertrophy due to selective replication of postero-basal left ventricle by immature cardiocytes in response to hypoxia is seen. Left atrial dilatation due to left superior direction of major depolarization vector is also a feature. Abnormal R waves and R wave progression in left precordial leads with ST elevation and T wave inversion are seen. Even in asymptomatic adults, resting ECG is often found to be abnormal.

Chest x-ray shows massive cardiomegaly with features of pulmonary venous congestion. With the advent of 2D echocardiography, the diagnosis has become rapid and much simplified. 2D Echocardiography can identify the pulmonary origin of left coronary artery. Doppler flow patterns show continuous or diastolic flow entering pulmonary trunk just distal to pulmonary valve and adhering to medial wall of pulmonary trunk. Left ventricle is dilated with echogenic antero-lateral papillary muscle due to ischemic scarring. Mitral regurgitation is seen due to annular dilatation, papillary muscle shortening and even papillary muscle rupture. Ejection fraction is severely decreased. Right coronary artery is dilated with increased flow. In adults with normal left ventricular global function, strain and stress echo can detect regional myocardial abnormalities.

In adolescents and adults with compromised echo windows, CT and CMR have a useful role to play. CT has a better spatial resolution and rapid acquisition time and high resolution but it has radiation hazard. On the other hand, CMR is free of radiation hazard and guides in functional assessment of left ventricle and myocardium viability (late gadolinium enhancement). Cardiac angiogram remains the gold standard for diagnosis in which selective right

coronary arterial angiogram is diagnostic with dilated RCA and left to right shunt at pulmonary arterial level.

Surgery is the main modality of treatment, but the child should be stabilized medically before it. Initial surgical techniques aimed at decreasing the pulmonary steal by pulmonary arterial banding and ligation of the origin of left coronary artery. Subsequently, along with the ligation of the origin of the left coronary artery, revascularization via internal mammary artery or saphenous venous grafts was started. The standard surgical therapy which is presently employed across the globe is direct reimplantation of left coronary artery origin into the aorta with a pulmonary arterial button around it. An alternative to this is Takeuchi procedure in which an aorta-pulmonary window is created with a tunnel fashioned directing the blood from aorta to left coronary osmium. The surgical mortality has reduced from 75-80% in early 1980s to 0-23% now. The risk factors for early mortality included early age at operation and decreased pre operative left ventricular function. Degree of mitral regurgitation is not found consistently to influence the mortality and in fact it is know to improve with time.

Long term follow up of these children revealed that left ventricular function normalizes by 1-2 years. Late coronary stenosis, graft occlusion presents as symptoms of ischemia, decreased left ventricular function warrants assessment of coronary patency.

Conclusion

Though a rare anomaly, ALCAPA has a very high mortality in infancy. 2D echocardiography has made the diagnosis very easy and feasible. Surgery is the mainstay of therapy and the surgical results have tremendously improved in the last 4 decades. Overall long term prognosis of these patients is good.

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Conflict of Interest

None.

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