Case Report

# Adult Survivor of Anomalous Left Coronary Artery Origin from Pulmonary Artery with Cleft Mitral Valve

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**ABSTRACT**

Anomalous origin of Left Coronary Artery from Pulmonary Artery (ALCAPA) is a rare congenital anomaly. Clinical manifestations and survival depend upon formation of collaterals between left coronary artery and right coronary artery after the birth. We report a case of 44 year old female patient diagnosed with ALCAPA and cleft mitral valve that had undergone coronary artery bypass with mitral valve repair.

**Keywords :** ALCAPA; Garland White Syndrome; Cleft mitral valve.

## Introduction -

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly and its association with cleft mitral valve is still rare.[1] Most infants die within the first year of life without treatment. Survival beyond infancy depends on the development of adequate collateral circulation from the right coronary artery (RCA) or from another source to the left coronary artery (LCA). Thus, older patients who survive without surgical intervention are rare. We report the successful surgical treatment of a 44 year old woman with ALCAPA with cleft mitral valve who had undergone CABG with left internal mammary artery graft to LCA (Left Coronary Artery) and mitral valve repair.

## Case Report -

A 44 year old lady presented with angina on exertion of NYHA class II (New York Heart Association.) Since childhood which increased to NYHA class III since last 1 month. Patient was thin built with heart rate of 80/minute, blood pressure of 140/90 mmHg and a soft systolic murmur in left parasternal area. ECG was showing left axis deviation and inverted T wave I, aVL, V1 to V3. Echocardiography was showing left ventricular hypertrophy, left atrial enlargement, anterior mitral leaflet prolapsing into left atrium causing eccentric jet of mitral regurgitation, dilated RCA originating from right coronary sinus, multiple collaterals running across interventricular septum and right ventricular apex and normal left ventricular ejection fraction.

Selective right coronary injection showed hugely dilated RCA filling LCA through collaterals ***(Fig.1).*** LCA was eventually draining into pulmonary artery. Multidetector computed tomography was showing ectatic and tortuous right coronary artery with multiple dilated channels over antero-lateral aspect of left ventricle and LCA arising from left pulmonary artery ***(Fig. 2).***

On confirming ALCAPA patient was posted for surgery. On doing median sternotomy, large tortuous RCAand LAD (Left anterior descending Artery) were seen on cardiac surface. Right and left pulmonary arteries were dissected. Aortic and bicavalcannulations were done and Left Internal Mammary Artery (LIMA) to LAD bypass grafting was done on beating heart. Then pulmonary artery was opened and LAD ostium from pulmonary artery was closed with autologous pericardial patch. Pulmonary artery was then repaired. Left atrium was opened and cleft on posteromedial aspect of mitral valve between A3 and P3 was repaired with prolene and a size 32 mitral annuloplasty ring was implanted to reinforce the mitral repair. Patient recovered well with stable hemodynamics. Now patient is doing well without angina or dyspnoea.

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**Discussion :**

ALCAPA is a rare anomaly in adults because more than 80% of symptomatic infants with this disease die of heart failure within the first year of life if not treated.1In adults, collateral blood flow from the RCA to the LCA system contributes to survival beyond childhood. However, even in the patients who survive to adulthood, sudden death frequently occurs.[1, 2] Therefore, surgical intervention should be performed in all patients, even in asymptomatic adult patients with no objective evidence of ischemia. In the repair of this anomaly, reestablishment of a double-coronary system is desirable. There are several surgical alternatives for the establishment of a double-coronary system, including subclavian-coronary artery anastomosis, ligation of the left coronary artery combined with coronary artery bypass grafting (CABG) with a saphenous vein graft or an internal thoracic artery, intrapulmonary tunnel repair, direct reimplantation of the anomalous left coronary artery to the aorta.

Of these options, coronary button transfer is considered to be the most anatomic correction, 3 and it has excellent long-term results. It is the preferred method of treatment in infants.[4] In adults, the preferred method is ligation of the LCA at its origin from the main pulmonary artery to stop competitive flow combined with CABG placement by using the internal mammary artery or a saphenous vein.[5] So we did closure of LCA origin from pulmonary artery with bypass grafting of left internal mammary artery to LCA.

**Summary :**

We report the successful surgical treatment of a 44 year old woman with ALCAPA with mitral valve cleft presenting with angina on exertion. As sudden deaths are common in adult survivor of ALCAPA we did CABG with left internal mammary artery graft to LCA along with mitral valve repair.

**Conclusion :**

Early recognition and correction of this coronary anomaly is very important to prevent left ventricle dysfunction, mitral regurgitation and unexpected death.

**Conflicts of interest :** None reported by authors

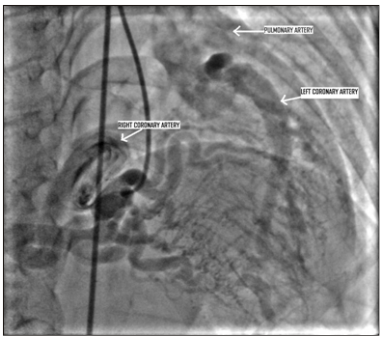


***Fig. 1 :*** **Hugely dilated RCA filling LCA through collaterals**

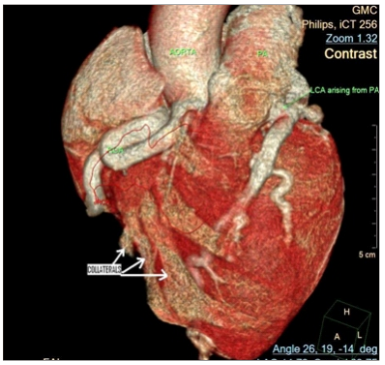


***Fig. 2 :*** **Multidetector CT showing ectatic and tortuous RCA with multiple dilated channels over antero-lateral aspect of left ventricle and LCA arising from left pulmonary artery**

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