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Anatomic correction of ALCAPA in an adult presenting with sudden cardiac death

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ABSTRACT
We report on a young adult with ALCAPA, who was successfully resuscitated after collapsing in ventricular fibrillation while playing football. This was followed by anatomical correction of the anomaly with a smooth recovery and return to his daily activities. The advantages of this approach are discussed in this brief report.

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BACKGROUND

Anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA) is a rare but serious congenital anomaly that occurs in 0.5% of children with congenital heart disease. It may result from abnormal formation of the conotruncus or from persistence of pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries[1].

It predominantly presents in the first year of life, resulting in left ventricular ischemia and dysfunction. If uncorrected, the condition carries a very poor prognosis. Owing to the development of extensive collateral circulation, only a very small percentage of patients survive to adulthood, where various manifestations of myocardial ischemia (such as left ventricular dysfunction and mitral regurgitation) are usually present. However, sudden cardiac death may be the first presentation in this group[2].

The management and longer-term outcomes have not been adequately defined, but anatomical surgical correction remains the best treatment modality for this subset of patients[3].

PATIENT AND METHODS

A 17 year-old male patient presented with sudden cardiac death (SCD) while playing football and was successfully resuscitated. There was no prior history of chest pain, shortness of breath or palpitations. He was then referred to our unit for further investigations and management.

Electrocardiography revealed sinus rhythm and signs of left ventricular hypertrophy, without pathological Q waves. Two-dimensional color flow Doppler echocardiography revealed mildly dilated left ventricular dimensions with borderline global systolic function (ejection fraction = 50%) and mild mitral regurgitation (MR).

The left main coronary artery (LMCA) was arising from the non-facing sinus of the pulmonary artery with flow reversal. Multi-slice computed tomography (MSCT) confirmed the diagnosis and the exact location of the orifice and proximal course of the anomalous coronary artery (Fig. 1a, 2a & 3a).

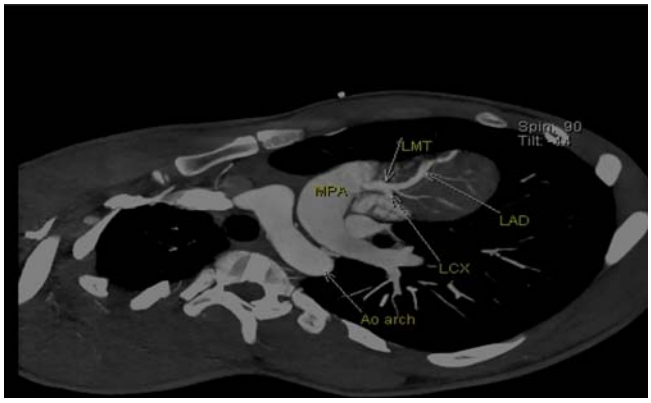

Figure 1 (a). Preoperative axial CT scan showing the origin of LMCA from pulmonary artery.

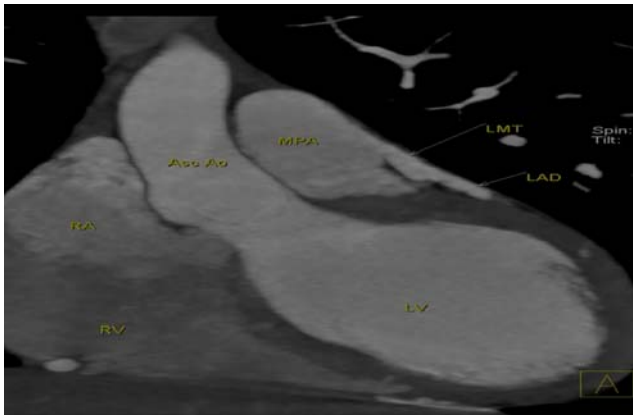

Figure 2 (a). Preoperative coronal section of CT scan showing the origin of LMCA from pulmonary artery.

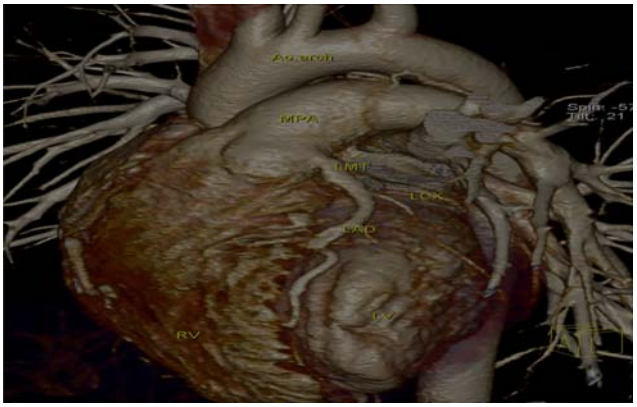

Figure 3 (a). **Preoperative reconstructive 3D MSCT angiography showing the origin of LMCA from pulmonary artery.**

The condition was surgically corrected by harvesting the button of anomalous left main coronary ostium with the surrounding wall of the non-facing sinus of the pulmonary artery, mobilizing the button with the left main coronary artery and reimplanting them in the anatomical position within the left sinus of Valsalva. Care was taken to perform the coronary transfer without tension, torsion or kinking.

The postoperative course was smooth and the patient was discharged on the fifth postoperative day. Pre-discharge MSCT coronary angiography revealed a smooth takeoff of the LMCA from the aorta with no obstruction (Fig. 1b, 2b & 3b).

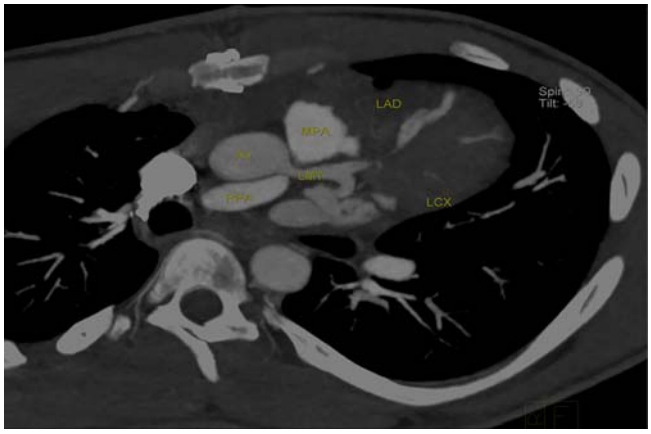

Figure 1 (b). **Postoperative axial CT scan showing the origin of LMCA from the aorta after it's re-implantation.**

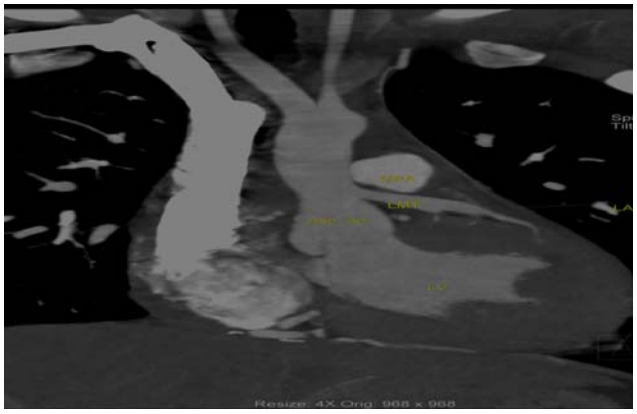

Figure 2 (b). **Postoperative coronal section of CT scan showing the origin of LMCA from the aorta after it's re-implantation.**

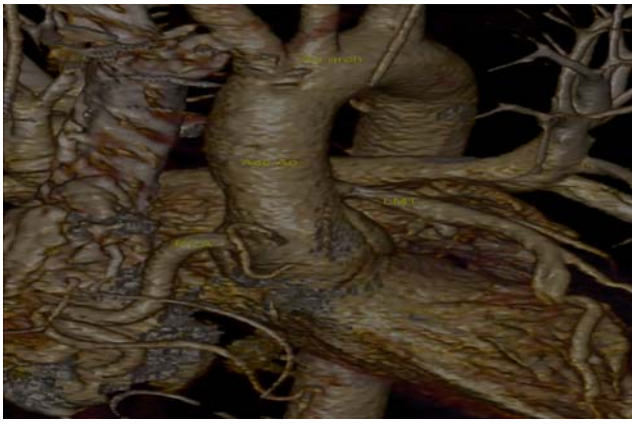

Figure 3 (b). Postoperative reconstructive 3D MSCT angiography subtracting the pulmonary artery showing the origin of LMCA from the aorta after it's re-implantation.

Six months later, the patient was completely asymptomatic and had returned to his normal daily activities. Follow up echocardiography showed significant reverse remodeling with normal LV dimensions and ejection fraction, and no mitral regurgitation. No resting regional wall motion abnormalities were detected. A dobutamine stress echocardiogram was negative. Cardiac magnetic resonance imaging revealed mild subendocardial fibrosis in the basal and mid anteroseptal wall.

WHAT HAVE WE LEARNED?

While cardiomyopathies and channelopathies are responsible for the vast majority of SCD in young adults, coronary artery anomalies including ALCAPA should always be included in the differential diagnosis and workup of survivors. Albeit rare, it is important to remember that SCD may be the first presentation in the latter group. In the absence of widespread myocardial scarring, anatomic correction helps restore left ventricular function and myocardial perfusion, thus eliminating the substrate for further arrhythmias/SCD.

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