What does one need to survive with Anomalous origin of left coronary artery from pulmonary trunk (ALCAPA) beyond infancy.

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What does one need to survive with Anomalous origin of left coronary artery from pulmonary trunk (ALCAPA) beyond infancy

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Abstract

A 53-year old man presented with sustained monomorphic ventricular tachycardia which was restored with 200 joules of direct current cardioversion. Complete haemogram, liver, and kidney function tests, and serum electrolytes were within normal limit. His past history was insignificant except exertional dyspnoea NYHA class-I for last one year. Transathoracic echocardiography revealed dilated left ventricle and atrium with severe systolic dysfunction (EF = 28%). There was continuous flow entering the pulmonary trunk. Origin of right coronary artery was dilated and left coronary artery was originating from main pulmonary artery suggestive of anomalous origin of left coronary artery from pulmonary trunk (ALCAPA). Coronary angiogram showed dilated and tortuous right coronary artery (RCA) with absence of a left coronary ostium in aortic sinus. During the delayed filling phase, abundant intercoronary anastomoses (Rentrop grade 3) were seen, communicating with left coronary artery which was later opacifying the main pulmonary artery thus establishing diagnosis of ALCAPA. Multidetector computed tomography angiography confirmed its diagnosis. Implantable cardioverter defibrillator was implanted. ALCAPA is a rare congenital cardiovascular anomaly which occurs 1 in 300,000 live births with rare survival to adulthood as 90% succumbs by third decade. Those with this syndrome who manages to survive beyond childhood are often incapacitated by myocardial ischemia, arrhythmias and severe ischemic cardiomyopathy. Survival beyond fourth or fifth decade with ALCAPA without surgical intervention is exceedingly rare as low pulmonary artery pressure results in coronary steal.
phenomenon. Rarely some remain asymptomatic until adulthood due to superdominant RCA, although they generally become symptomatic in early adulthood due to fixed myocardial ischemia and coronary steal phenomenon. In utero, as the systemic and pulmonary arterial pressures are equal, there is anterograde flow in both the left and the right coronary. In the neonatal period as the pulmonary artery pressure falls and ductus arteriosus closes, the flow in the left coronary artery reverses. The more collateral channels exist between RCA and anomalous left coronary system (ALCA), the longer the patient may survive. Another contributing factor may be the area supplied by the ALCA as the smaller the area supplied by the ALCA, the less extensive the myocardial ischemia is likely to be. In few adult cases, RCA supplies not only the posterior wall of the left ventricle but also its lateral wall. May be this reduction in the area of the myocardium supplied by the ALCA would favour survival. However, even extensive collateral circulation may not totally prevent ischemic damage though can delay it as low pulmonary artery pressure may still result in coronary steal phenomenon, which may cause sudden deaths. We believe right coronary artery may have “supernormal” flow reserve to account for his survival.

Surgical correction such as direct transfer of the left coronary artery from pulmonary artery (like in arterial switch operation), Takeuchi repair (creating aorto-pulmonary window), and bypass graft when these two are not possible, it is one of the coronary anomalies not compatible to life. In our case, it was decided not to intervene surgically as risk of cardiac surgery was outweighing the benefit.

**Key words:** ALCAPA syndrome, coronary steal phenomenon, NYHA Functional Class, superdominant RCA, ventricular tachycardia

A 53-year old man presented with sustained monomorphic ventricular tachycardia which was restored with 200 joules of direct current cardioversion. Complete haemogram, liver, and kidney function tests, and serum electrolytes were within normal limit. His past history was insignificant except exertional dyspnoea NYHA class-I for last one year. Transrthoracic echocardiography revealed dilated left ventricle and atrium with severe systolic dysfunction (EF = 28 %). There was continuous flow entering the pulmonary trunk. Origin of right coronary artery was dilated and left coronary artery was originating from main pulmonary artery suggestive of anomalous origin of left coronary artery from pulmonary trunk (ALCAPA). Coronary angiogram showed dilated and tortuous right coronary artery (RCA)
with absence of a left coronary ostium in aortic sinus (Fig. 1A,C). During the delayed filling phase, abundant intercoronary anastomoses (Rentrop grade 3) were seen, communicating with left coronary artery (Fig. 1B,D) which was later opacifying the main pulmonary artery thus establishing diagnosis of ALCAPA (Fig. 1D). Multidetector computed tomography angiography confirmed its diagnosis (Fig. 2). Implantable cardioverter defibrillator was implanted. ALCAPA is a rare congenital cardiovascular anomaly which occurs 1 in 300,000 live births with rare survival to adulthood as 90% succumbs by third decade.[1] Those with this syndrome who manages to survive beyond childhood are often incapacitated by myocardial ischemia, arrhythmias and severe ischemic cardiomyopathy. Survival beyond fourth or fifth decade with ALCAPA without surgical intervention is exceedingly rare as low pulmonary artery pressure results in coronary steal phenomenon. Rarely some remain asymptomatic until adulthood due to superdominant RCA, although they generally become symptomatic in early adulthood due to fixed myocardial ischemia and coronary steal phenomenon.[2] In utero, as the systemic and pulmonary arterial pressures are equal, there is anterograde flow in both the left and the right coronary. In the neonatal period as the pulmonary artery pressure falls and ductus arteriosus closes, the flow in the left coronary artery reverses. The more collateral channels exist between RCA and anomalous left coronary system (ALCA), the longer the patient may survive. Another contributing factor may be the area supplied by the ALCA as the smaller the area supplied by the ALCA, the less extensive the myocardial ischemia is likely to be. In few adult cases, RCA supplies not only the posterior wall of the left ventricle but also its lateral wall. May be this reduction in the area of the myocardium supplied by the ALCA would favour survival.[1] However, even extensive collateral circulation may not totally prevent ischemic damage though can delay it as low pulmonary artery pressure may still result in coronary steal phenomenon, which may cause sudden deaths. We believe right coronary artery may have “supernormal” flow reserve to account for his survival.

Surgical correction such as direct transfer of the left coronary artery from pulmonary artery (like in arterial switch operation), Takeuchi repair (creating aorto-pulmonary window),[3] and bypass graft when these two are not possible, it is one of the coronary anomalies not compatible to life. In our case, it was decided not to intervene surgically as risk of cardiac surgery was outweighing the benefit.
Key words - ALCAPA syndrome; Coronary steal phenomenon; NYHA Functional Class; Superdominant RCA; Ventricular tachycardia.

References


Figure legends

Figure 1. Tortuous and dilated superdominant right coronary artery (A); During the delayed filling phase, abundant intercoronary anastomoses (white arrow) were seen communicating with left coronary artery (B); Absence of a left coronary ostium in aortic sinus (C); Left coronary artery opacifying the main pulmonary artery (red arrowhead; D)

Figure 2. Dilated RCA with left main arising from pulmonary trunk (RCA — right coronary artery; Ao — aorta; PT — pulmonary trunk; LMCA — left main coronary artery).