

A Case Report of an Adolescent with Anomalous Origin of the Left Coronary Artery from the Pulmonary Trunk (ALCAPA)

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Abstract:

Anomalous origin of the left coronary artery from the pulmonary artery is a serious congenital cardiac defect associated with high mortality rates in infancy. It is undoubtedly a rare defect but its diagnosis during life is possible in most cases, and successful surgery appears within reach. The object of this paper is to report our experience with a fourteen years old girl with this rare anomaly. The presenting symptoms of our patient during infancy were not that are considered classic for this anomaly. Though the child had excessive sweating and symptoms of heart failure during infancy. The girl was suspected to have ALCAPA clinically and echocardiographically finally selective right coronary arteriography established the diagnosis.

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Keywords:

Congenital heart disease,
ALCAPA.

Introduction:

The congenital anomaly in which the left coronary artery arises from the main pulmonary artery is rare, but lethal, and since it can be alleviated surgically, its recognition and hemodynamic consequences are important. It was first described by Brooks¹ in two adults at necropsy in 1886. In 1911 Abrikosoff² reported on an autopsy on a 5 month old baby. Cases were reported sporadically in the next twenty years. But clinical interest was not aroused until Bland, White, and Garland, in 1933, integrated the clinical and pathological data and recorded an electrocardiogram in an infant dying of this condition.³

Most instances of this defect have been found in babies who have died in the first year of life. However, survival into adult life is possible, and this appears to occur in approximately 15 per cent of cases.⁴ Hadwig Wesselhoeft,⁵ on the basis of 140 reported cases and seven further cases, divided patient with this lesion into four groups: (1) in infancy with angina-like symptoms or as cardiomyopathy, and later (2) as mitral insufficiency, (3) continuous murmur or (4) by sudden death. The object of this paper is to report our experience with an adolescent girl with this rare anomaly.

Case summary:

A fourteen years old very bright girl presented with poor appetite, weight loss, nausea & vomiting, abdominal pain, palpitation and leg swelling on and off for last one and half year. She is the only child of a middle class nonconsanguineous parents. Her birth history was uncomplicated. She was delivered by normal vaginal delivery at term. Her birth weight was 2.5 kg. Her mother noticed increased precordial activity, mild chest indrawing at about twenty days of age, excessive sweating around one and half months of age. For which she was brought to a physician who advised a chest X ray (figure 1) which showed cardiac enlargement and was referred to a cardiologist. An ECG was done which showed Deep q wave in II, aVf, V5, V6; T inversion I, I, III, aVf, V5, V6. An echocardiogram was performed and was diagnosed as Viral myocarditis/ dilated cardiomyopathy as the patient had global hypokinesia, LV dysfunction, LVEF:48%. Since then she was on digoxin, frusemide, captopril and was relatively well though she was thriving poorly. Thereafter she underwent multiple echocardiographic examinations which showed gradual improvement of left ventricular ejection fraction though she developed significant mitral regurgitation subsequently with left ventricular enlargement (table-I). She developed jaundice three years back and diagnosed to have Gilbert syndrome.

Table-I

Serial echocardiogram showing Ejection fraction(EF) of left ventricle and left ventricular end diastolic diameter(LVIDd)

Age	EF(%)	LVIDd (mm)
3 months	48	37
7 months	47	44
1 .5 years	42	46
2.3 years	48	43
3.5 years	60	40
5 years	74	40
7 years	78	46
9 years	74	45
10	63	52
13	68	61
14	72	59

On examination she was mildly pale and icteric. Her weight (32.5 kg) and height (138 cm) falls below 3rd centile. Her oxygen saturation was 96% with pulse oximeter. She was tachycardic (heart rate 105 per minute), blood pressure was 90/50 mm Hg, respiratory rate was 18 breath per minute. 1st heart sound was soft, 2nd heart sound was mildly accentuated. There was a pansystolic murmur of grade 3/6 at the apex radiating to axilla. Liver was soft and enlarged 3 cm from right costal margin along midclavicular line. Other systems were unremarkable.

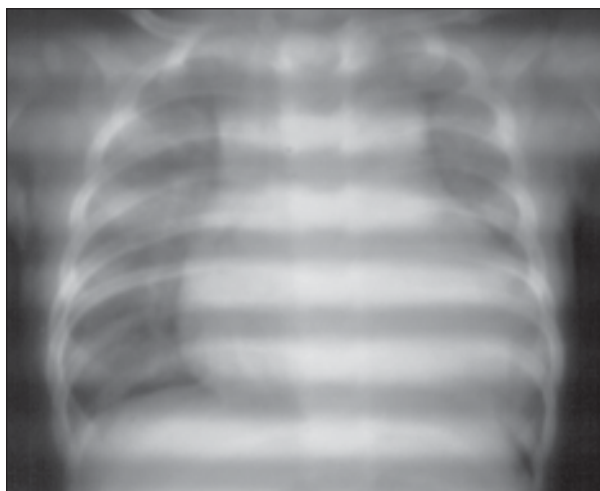


Fig.-1: CXR at 3 months showed cardiomegaly,

Her lab investigation shows CBC, Renal function tests, RBS and serum lipid profile were normal. Chest X ray showed cardiomegaly with prominent pulmonary vasculature, suggesting pulmonary venous congestion; electrocardiogram showed atrial fibrillation. Her echocardiogram showed Mitral valve prolapse with moderate MR, hugely dilated left atrium and ventricle, moderate pulmonary hypertension, Small ASD shunting left to right, dilated right coronary artery, left coronary artery was also dilated but its origin could not be defined as well as flow reversal seen in both LAD (blue)& LCX(red)(figure 2,3,4). So she was provisionally diagnosed as anomalous origin of left coronary artery from pulmonary artery(ALCAPA) and she was taken for cardiac catheterization. In right sided cardiac catheterization (table 2, 3) there was an increase in oxygen saturation in the pulmonary artery (9% step up) confirmed left to right shunt at pulmonary artery level. The right ventricular and pulmonary artery systolic pressures were moderately elevated (60 mm Hg). Selective ascending aortography there was evidence of a single coronary artery arising from the aorta in the usual position of the right coronary artery. Selective right coronary arteriography showed retrograde opacification of the anomalous left coronary artery and of the pulmonary trunk from right coronary artery, well developed collaterals (figure 5,6).

Table-II

Oximetry table obtained from cardiac catheter

Site	SpO2(%)
SVC(H)	77
SVC(L)	74
RA(H)	75
RA(M)	80
RA(L)	81
PA	83
RV	74
LA	83

Table-III

Pressure table obtained from cardiac catheterization

Site	systole	diastole	mean
RA	08	02	05
PA	60	30	45
LV	100	10	
AO	100	80	96
RV	60	05	32
LA	08	03	04

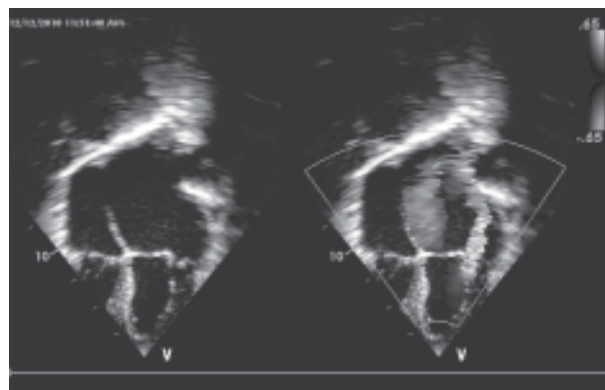


Fig.-2 : Dilated Left atrium and ventricle and moderate mitral regurgitation.

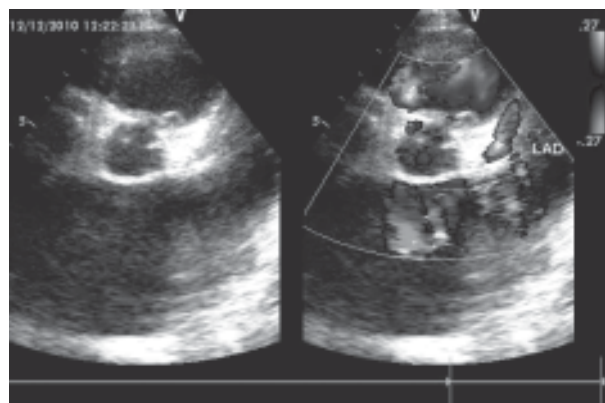


Fig.-3 : Dilated LAD with flow reversal(blue)

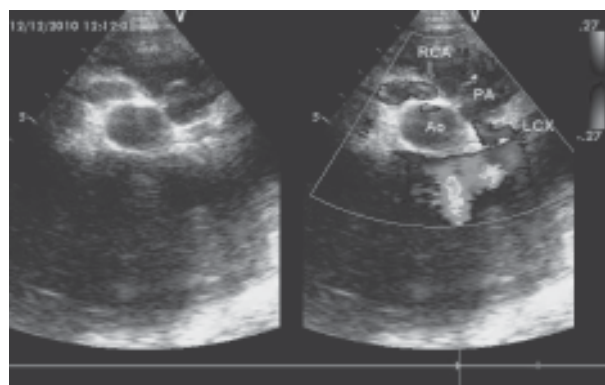


Fig.-4:Dilated RCA and flow reversal in LCX(red)

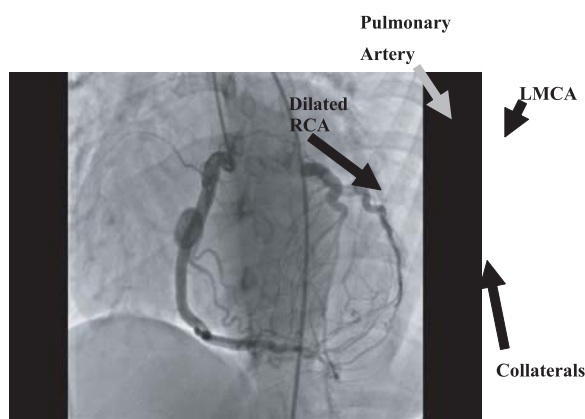


Fig.-5: Selective right coronary arteriography showed retrograde opacification of the anomalous left coronary artery and of the pulmonary trunk from right coronary artery, well developed collaterals.

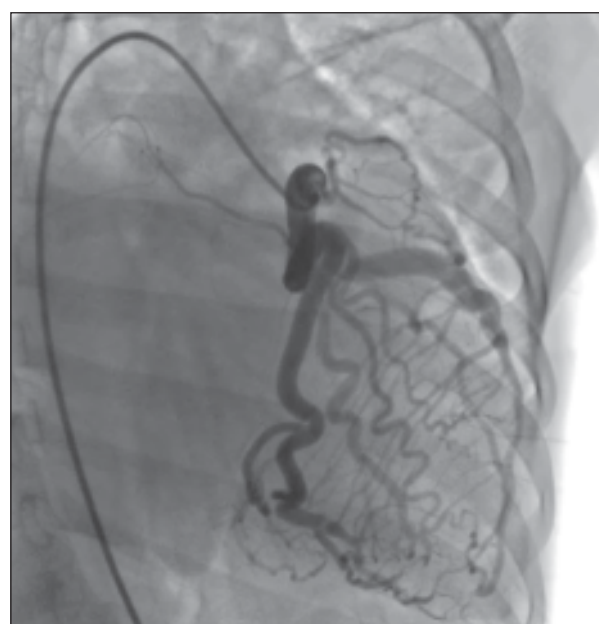


Fig.-6: Well developed collaterals between RCA and LCA

Discussion

Edwards⁶ proposed that in every patient there is a gradually changing functional pattern of the coronary flow. In the first phase (newborn period) the anomalous left coronary artery is supplied by the relatively high pulmonary artery pressure. In the second phase, after establishment of collateral vessels, the anomalous left coronary artery carries blood into the pulmonary artery (adult type). He assumed that during the transitional phase (beyond the neonatal period), corresponding to the time of onset of symptoms of myocardial ischemia,

the flow of blood into the part of the myocardium supplied through the anomalous left coronary might be at an all time low. Survival would then depend upon the extent of collateral vessel development and the predominance of the right coronary artery. In a fourth phase, anastomosis⁷ no longer function as “feeding” vessels but rather act as a “steal” for the drainage system. Interestingly, Brooks¹ in 1886 had considered that the “pulmonary coronary branch acted as the channel by which the blood was drained away.

The presenting symptoms of our patient during infancy were not that are considered classic for this anomaly, taking the form of episodes of pallor, dyspnea, and perspiration⁸. Though the child had excessive sweating and symptoms of heart failure during infancy and response to antifailure was effective.

The early clinical course and the subsequent improvement, however, suggests that this cardiac malformation may give rise to a pathophysiologic spectrum,⁹⁻¹¹ in our case. In this case, the patient was asymptomatic during the first few weeks of life, but each developed symptoms during infancy. Then improved and had an apparent “remission” from the disease. Despite this apparent improvement, there was evidence of residual myocardial damage. This was manifested by persistent cardiomegaly, mitral insufficiency.

Of the 50 electrocardiograms reviewed from reports in the literature, Wesselhoeft and associates⁵ found anomalous left coronary artery from the pulmonary artery to be associated with myocardial infarction in 80%. Our patient had evidence of infarction, although one had only loss of R-wave voltage when first seen. T-wave changes in the precordial leads were noted in cases in the literature as in our case. S-T segment elevation was present in 40% of the cases in the literature but not in our case.

From the radiologic point of view,⁸ the most definitive diagnostic procedure is selective right coronary arteriography. If aortography is performed, it is important to inject the contrast material at the root of the aorta and to make exposures in rapid succession, since the transit of contrast material from the right to the left coronary artery may be very rapid. Opacification of the pulmonary trunk may be evident if the reversal of blood flow through the left coronary artery is well established. The best demonstration of delayed filling of the left coronary artery with

reversed blood flow and secondary opacification of the pulmonary trunk is accomplished by selective right coronary arteriography.

Conclusion:

Anomalous origin of the left coronary artery from the pulmonary artery is a rare and serious congenital cardiac defect. Infants less than six months of age presenting with heart failure without any apparent cause should be investigated for ALCAPA by electrocardiogram and color doppler echocardiography. Echocardiography can identify ALCAPA but selective ascending aortography or selective right coronary arteriography is necessary to establish the diagnosis.

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