Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

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Abstract:
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome is a rare congenital cardiac anomaly. ALCAPA syndrome mostly presents in the first few months of life. Left untreated, the mortality rate in the first year of life is 90% secondary to myocardial ischemia or infarction and mitral valve insufficiency leading to congestive heart failure. Sudden death may occur because of inadequate collateral circulation between the left and right coronary artery systems and/or development of arrhythmia. Currently, the prognosis for patients with ALCAPA syndrome is dramatically improved as a result of both early diagnosis and improvements in surgical techniques, including myocardial preservation. This case report presents a four month old infant with ALCAPA syndrome including clinical presentation of severe cardiomyopathy and left-sided heart failure. Child was operated for reimplantation of ALCAPA to aortic root with satisfactory outcome.

Keywords: ALCAPA, heart failure, infant, treatment

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Introduction
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome is a rare, but serious development anomaly of the coronary arteries. It is accounting for approximately 0.25-0.5% of all congenital heart disease. If left untreated, the mortality rate in the first year of life reaches 90%, secondary to myocardial ischemia or infarction and mitral valve insufficiency leading to heart failure [1].

The first clinical description in conjunction with autopsy findings was described by Bland and colleagues in 1933, so the anomaly is also called Bland-White-Garland syndrome [2-5]. The anomaly arises from either abnormal septation of the conotruncus into the aorta and pulmonary artery or from persistence of the pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries. As a consequence of the decreasing pulmonary artery pressure and resistance shortly after birth, the left ventricular myocardium being perfused by relatively desaturated blood under low pressure, leads to a myocardial ischemia. Collateral circulation between the right and left coronary systems ensues. Decrease in pulmonary arterial pressure results in reversal of flow, as the left coronary artery drains from the right coronary artery through collaterals, into the pulmonary artery. This is known as myocardial steal—hence the nickname of Al Capone of coronary vessels. The steal phenomenon causes ischaemia or infarction of the anterolateral left ventricular wall [6,7]. Consequently, the combination of left ventricular dysfunction and significant mitral valve insufficiency leads to congestive heart failure symptoms in the young infant. Clinical diagnosis of ALCAPA can be challenging. Findings in ALCAPA resemble those of dilated cardiomyopathy and the diagnosis of ALCAPA must be excluded [8], especially in the presence of ischemic ECG findings. ALCAPA
can be treated successfully by several types of operations with good prognosis [8]. In cases of ALCAPA in young children, reestablishment of a dual coronary system through reimplantation of the left coronary artery to the aorta or by bypass grafting and ligation of proximal ALCAPA showed favourable results [9]. Mitral valve surgery is not indicated at initial surgery, except in selected cases with a low potential of recovery [10]. Occasionally, persistent refractory mitral regurgitation will necessitate delayed mitral valve repair or replacement.

In this report we present a four month old infant with ALCAPA syndrome with clinical presentation of severe cardiomyopathy and left-sided heart failure who was operated for reimplantation of ALCAPA to aortic root with satisfactory outcome.

**Case report**

This four months old female patient was admitted to pediatric intensive care unit with history of poor feeding, seizures and irritability for four weeks prior admission. Dyspnea, tachycardia, poor peripheral perfusion, II/VI ejection systolic murmur at left lower sternal border and liver 3 cm below the right costal margin were revealed during examination. Child was hemodynamically compromised and required mechanical ventilation and inotropic support. Chest X-ray (Figure 1) showed cardiomegaly with plethoric lungs and electrocardiography revealed anterolateral ST-segment depression. Echocardiogram showed dilated left ventricle with poor function, fractional shortening of 22%, moderate mitral valve regurgitation, echogenic mid-ventricular septum with akinesia and echogenic left ventricular papillary muscles without visible origin left coronary artery from aortic root on 2D option, while color Doppler revealing blood flow to pulmonary artery in parasternal short axis view. The diagnosis of ALCAPA was strongly suspected, without proved other congenital heart defect. Heart catheterisation (Figure 2) was performed and coronary artery angiogram with 3D reconstruction confirmed the diagnosis of ALCAPA.

Patient was operated for reimplantation of ALCAPA to aortic root. Postoperative echocardiography shows good flow via the newly transpositioned artery. In postoperative management child has developed atelectasis due to obstruction of the left main bronchus, caused by pressure of the enlarged heart. Depressed ST segments could still be observed in V5, V6 leads in ECG, but drop in pro-brain natriuretic peptide (proBNP) values has been noted. Child was, in stable clinical condition, discharged home on digoxin, diuretics and salicylate therapy for further follow up.

**Discussion**

In the described infant, the onset of the disease was typical for the syndrome. After pulmonary arterial pressure had decreased, this child demonstrated distress after feeding and irritability. These were caused by the coronary steal from the left coronary artery into the pulmonary trunk. This coronary steal resulted in diminished perfusion of the left ventricular muscle, extensive myocardial ischemia and finally myocardial infarction.
Early diagnosis and prompt surgical intervention with the aim to restore a two-coronary-artery circulatory system had excellent results and led to gradual myocardial recovery. Inadequate myocardial perfusion likely causes significant chest pain and these symptoms of myocardial ischemia may be misinterpreted as routine infantile colic [11]. In 85% of these patients symptoms appear between 2 to 3 month of age as recurring episodes of distress, marked cardiomegaly and heart failure. In unusual cases, the clinical presentation with symptoms of myocardial ischemia may be delayed into early childhood, or even adulthood. Significant heart murmur is usually absent, with a rare exception of a heart murmur of mitral regurgitation secondary to myocardial infarction. The electrocardiography (ECG) shows an anterolateral myocardial infarction pattern. Cardiac enzyme changes probably occur, but the relatively slow development of myocardial infarction and the uncertainty of the exact time of infarction may make it difficult to interpret laboratory data.

Echocardiography is an important diagnostic tool for the diagnosis of ALCAPA Two dimensional echocardiogram has its limitations to show the exact origin of ALCAPA and color Doppler is much more sensitive. The most common ultrasound findings are mild to moderate mitral valve regurgitation, echogenic left ventricular papillary muscles, some degree of left ventricle dilatation/dysfunction and prominent right coronary artery with collaterals like flow in interventricular septum and then to pulmonary artery. In rare instances, the clinical picture may be atypical and ECG or echocardiogram may not be classical for the diagnosis of ALCAPA. In such atypical cases, cardiac catheterization or computerized axial tomographic scan (CT scan)/magnetic resonance angiography (MRA) may be indicated to confirm the diagnosis [1] and to exclude other potential diagnoses [12]. The most important differential diagnosis in this age group is dilated cardiomyopathy, and others are congenital mitral valve disorder, coronary artery fistula, mitral valve insufficiency and viral myocarditis. Coronary angiography is a simple and quick invasive diagnostic modality in the centers where CT scan/MRA facilities are not fully established. Therefore we performed coronary angiography to confirm the diagnosis prior to surgical intervention.

Initial management of anomalous left coronary artery from the pulmonary artery is both supportive and temporary. Treatment of congestive heart failure includes careful use of diuretics, afterload reduction medications and inotropic drugs. Once the patient is stabilized, the surgical revascularization is performed in order to create a two-coronary artery system including coronary button transfer, the Takeuchi procedure (creation of an aortopulmonary window and an intrapulmonary tunnel extending from the anomalous ostium to the window) and placement of a coronary artery bypass graft combined with ligation of the origin of the left coronary artery. Of these options, coronary button transfer, as preferred method of treatment in infants, is considered to be the best anatomic correction and it has excellent long-term results. Once revascularization to a two-coronary artery system is accomplished, most patients experience normalization of left ventricular systolic function, decreased mitral valve insufficiency and resolution of heart failure symptom, thereby improving long-term survival [13]. In many cases, the classic myocardial infarct pattern on electrocardiography eventually disappears following normalization of left coronary blood flow. The need for simultaneous mitral valve reconstruction, in the presence of severe insufficiency, is controversial because spontaneous improvement of mitral valve function often occurs following surgical revascularization. A study of 23 infants with anomalous left coronary artery from the pulmonary artery, proved aortic reimplantation of the anomalous coronary artery to be effective in improving myocardial function but is a less effective tool for treating severe mitral valve regurgitation [14].

Clinical significance of the presented case is to emphasize the importance of early diagnosis and treatment of ALCAPA syndrome, with awareness of the significant difficulties and possible errors throughout.

Perspectives for research in this field are determination of the long-term outcome after palliation of the syndrome as well as lifelong care in a center experienced with caring for adults with congenital heart disease.

**Conclusion**

We diagnose what we look for and look for what we know. Although ALCAPA syndrome is rare and potentially lethal condition, it should be considered in every clinical findings of severe heart failure, cardiomyopathy and presence of ischemic ECG findings. Early diagnosis and prompt surgical intervention have excellent results, leading to gradual myocardial recovery and better prognosis for these children.

**Declaration of interest**

The authors declare no conflict of interest.
REFERENCES:


