

A Rare Cause of Respiratory Distress in the Emergency Department

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 is the most common cause of myocardial ischemia and infarction in children and infants. Most of the affected individuals become symptomatic, owing to dilated cardiomyopathy during the infant period. Because of congestive heart failure, pulmonary congestion and respiratory distress can be seen. Herein, we report a 35-day-old infant with anomalous origin of the left coronary artery from the pulmonary artery who presented with respiratory distress and dilated cardiomyopathy to draw attention to this rare surgically treatable anomaly.

Key Words: respiratory distress, infant, dilated cardiomyopathy, ALCAPA (*Pediatr Emer Care* 2015;31: 357–359)

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) was first described by Brooks in 1886. This rare anomaly is seen in 1 among 300,000 live births.¹ In infants and children, ALCAPA is the most common cause of myocardial ischemia and infarction.² Most of the affected individuals are symptomatic during the infant period characterized by congestive dilated cardiomyopathy, mitral regurgitation, and myocardial infarction or even sudden death. Only 10% of all cases who develop coronary artery collaterals called as adult type have uneventful infancy.³ Because of its poor prognosis, early diagnosis and surgical correction are important. Therefore, infant patients with dilated cardiomyopathy should be evaluated in terms of ALCAPA.

Herein, we reported a 35-day-old infant diagnosed with ALCAPA who presented with respiratory distress and dilated cardiomyopathy. In addition, we wanted to draw attention to this rare anomaly that can be surgically treated.

CASE

A 35-day-old male infant admitted to our emergency department with complaints of cough and respiratory distress. Physical examination was unremarkable except the rales on the bilateral pulmonary areas. Blood pressure was measured as 80/50 mm Hg. Chest x-ray revealed cardiomegaly and pulmonary congestion. Electrocardiographic (ECG) examination showed QR pattern on derivations I, aVL and ST depression on derivations V1 to V4. Echocardiographic examination was done, and severe dilatation and systolic dysfunction of the left ventricle were observed. Fractional shortening was 13%, and abnormal origin of the left coronary artery was determined (Fig. 1). Catheterization and angiographic study were planned urgently. Aortic root and pulmonary artery

angiographies showed dilatation of the right coronary artery, with collaterals between the right and left coronary arteries and ALCAPA (Fig. 2). The left coronary artery was reimplanted to the aorta successfully without any complications. The patient improved quickly after operation and is still being followed up with normal left ventricular systolic function.

DISCUSSION

Anomalous origin of the left coronary artery from the pulmonary artery results from abnormal separation of the aortopulmonary truncus.⁴ During the fetal life, because of high pulmonary resistance and pressure, adequate blood flow can be maintained to the left coronary artery. After birth, the left coronary artery blood flow directs toward the pulmonary artery (coronary steal) as a consequence of decreasing pulmonary pressure and vascular resistance. For the maintenance of the left ventricular perfusion, collaterals from the right coronary artery are necessary. Because of the underdevelopment of these collaterals, myocardial infarction and dilated cardiomyopathy may occur in infancy.³

The initial presentation of ALCAPA can be similar with more common clinical conditions such as infantile colic, respiratory infections, and food intolerance during infancy.⁵ The most prevalent presenting symptoms are attacks of intense crying, a sign of angina pectoris, and symptoms of congestive heart failure such as dyspnea, respiratory distress, diaphoresis, poor feeding, and failure to thrive.^{2,6} Chest radiography always shows cardiomegaly and pulmonary congestion.⁶ Anomalous origin of the left coronary artery from the pulmonary artery is an important cause of congestive dilated cardiomyopathy in infancy and childhood. Once diagnosed, it can be treated with surgery; however, clinical diagnosis of ALCAPA and differentiation from other causes of dilated cardiomyopathy may not be always possible even by the pediatric cardiologists.³ Electrocardiography, which is a cheap and easily accessible diagnostic tool, can provide valuable clues in suspected patients. Pathological Q waves and T inversions on ECG derivations I and aVL in conjunction with the clinical findings and cardiomegaly on chest x-ray may be warning for ALCAPA.⁷

A 2-dimensional echocardiography is commonly used to visualize the origin of the coronary arteries and to differentiate ALCAPA from idiopathic dilated cardiomyopathy. However, because of the lack of lateral resolution of echocardiography, less than 50% of the patients with ALCAPA can be diagnosed by 2-dimensional echocardiography.⁸ Therefore, cardiac catheterization with aortic root angiography should be performed to determine accurate anatomic origins of the coronary arteries and also to show collaterals in patients with uncertain diagnosis.⁸

In infancy, most of the patients with ALCAPA present with findings of decreased left ventricular function and mitral regurgitation secondary to myocardial ischemia and papillary muscle damage, respectively.⁹ Immediate coronary reimplantation and early improvement of the left ventricular function are essential

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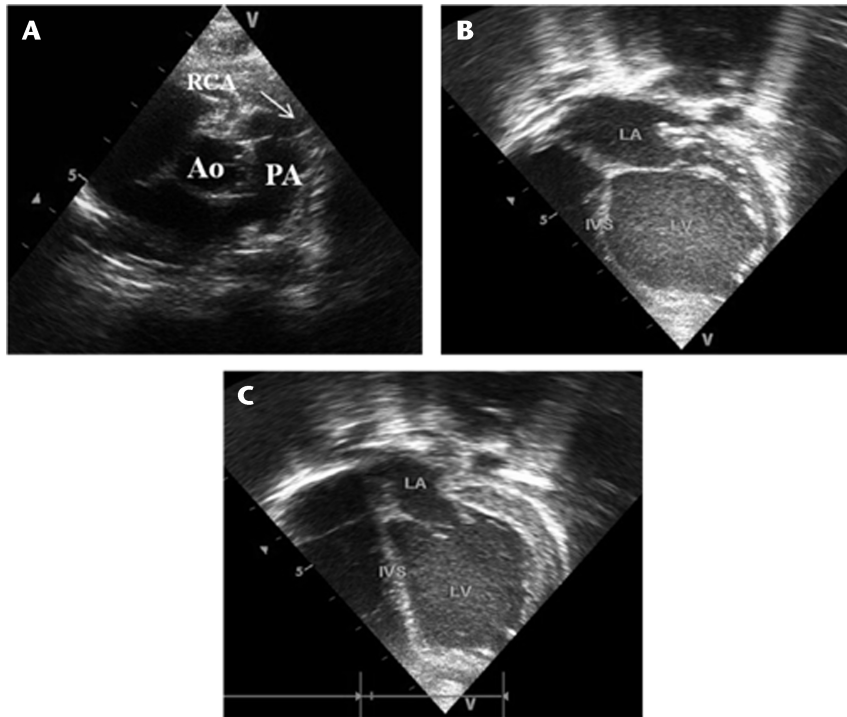


FIGURE 1. Echocardiographic images of the patient with ALCAPA. A, Abnormal left coronary artery from the pulmonary artery (arrow). B, Preoperative dilatation of the left ventricle. C, Postoperative improvement of the left ventricular dilatation. Ao indicates aorta; PA, pulmonary artery; RCA, right coronary artery; LA, left atrium; LV, left ventricle; IVS, interventricular septum.

for recovery. Surgical repair outcomes are quite excellent in most patients. After successful repair, ventricular function, dilatation, and mitral regurgitation improve gradually.¹⁰ Similar with these data, our case was presented with mitral regurgitation and dilated cardiomyopathy. Anomalous origin of the left coronary artery from the pulmonary artery was diagnosed at the beginning of the second postnatal month, and after immediate surgical correction, dilatation of the left ventricle and mitral regurgitation completely disappeared.

In some cases with severe heart failure, preoperative mechanical circulatory support by extracorporeal membrane oxygenation (ECMO) may be necessary.¹¹ Extracorporeal membrane oxygenation may also be required in patients whose ventricular functions do not improve in early postoperative period, despite provision of adequate coronary blood flow. In recent reported series, necessity of ECMO in early postoperative period varies from 0% to 43%. Nevertheless, Lange et al¹⁰ reported 95% survival rate in their surgically treated ALCAPA patients, none of whom needed postoperative ECMO support. Another study denoted postoperative ECMO requirement rate of 19% in patients with higher risk of reoperation.¹¹

In conclusion, differential diagnosis of all previously healthy infants presenting with respiratory distress, dilated cardiomyopathy, and classical ECG findings should focus on ALCAPA. The coronary arteries should be carefully examined by echocardiography to prevent morbidity and mortality in those cases.

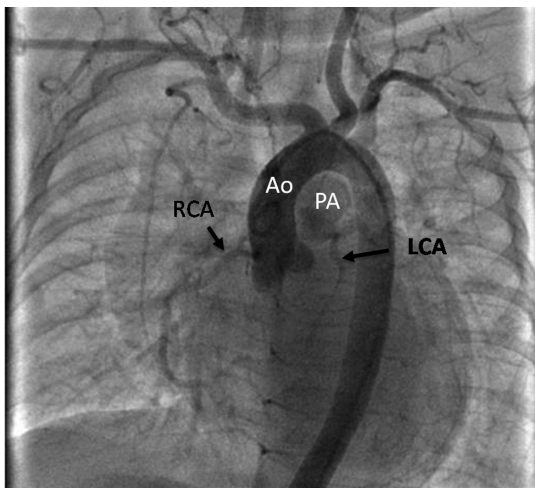


FIGURE 2. Aortic root angiography shows dilated right coronary artery and ALCAPA that was filled by collaterals from the right coronary artery. Ao indicates aorta; PA, pulmonary artery; RCA, right coronary artery; LCA, left coronary artery.

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