

## Adult type anomalous origin of the left coronary artery from the pulmonary artery

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a clinical entity characterized by myocardial necrosis which becomes symptomatic shortly after birth; survival beyond infancy is uncommon because of severe left heart failure.

To our knowledge, it is rare for an ALCAPA patient to survive to adulthood. Here we present a case of a 17-year-old girl with ALCAPA who was referred to our hospital because of palpitation and dyspnea.

**Key words:** anomalous origin of the left coronary artery from the pulmonary artery, adult type, coronary anomalies.

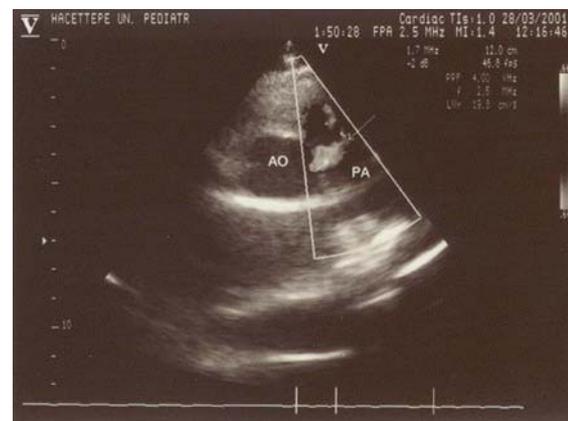
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly occurring in approximately 1 in 300,000 live births<sup>1</sup>. This syndrome can cause severe myocardial ischemia or infarction leading to left ventricular dysfunction, mitral insufficiency, congestive heart failure, and death. Without surgical intervention, about 90% of patients with ALCAPA die within the first year of life<sup>2</sup>. We present a 17-year-old girl with adult type ALCAPA syndrome who survived to her present age without any symptoms except for a six-month history of palpitation and dyspnea.

### Case Report

A 17-year-old girl was admitted to our hospital for attacks of palpitation and dyspnea. Physical examination revealed a grade II-III/VI systolic murmur in the fourth left intercostals space. Her chest radiograph was normal except for pulmonary congestion. Baseline electrocardiogram (ECG) showed inverted T waves and ST depression in leads V4-V6. Holter electrocardiographic recordings revealed sinus tachycardia. Echocardiography revealed a dilated left atrium and left ventricle with hypokinetic anterior septum. Left ventricular function was in normal limits (end-diastolic diameter: 54 mm, end-systolic diameter: 35 mm, ejection fraction: 65%, shortening fraction: 35%). Color

flow mapping and pulsed Doppler investigation demonstrated moderate mitral regurgitation and abnormal flow pattern in the pulmonary artery (Fig. 1). Coronary angiography revealed a tortuous widely dilated right coronary artery communicating through extensive collaterals with the left coronary artery which was filling the pulmonary artery (Fig. 2).

The left main coronary artery was reimplanted in the aorta and she has been free of subjective symptoms for nine months.



**Fig. 1.** Doppler color imaging reveals abnormal flow pattern in the pulmonary artery on transthoracic echocardiographic short axis view. (PA: pulmonary artery, Ao: Aorta).

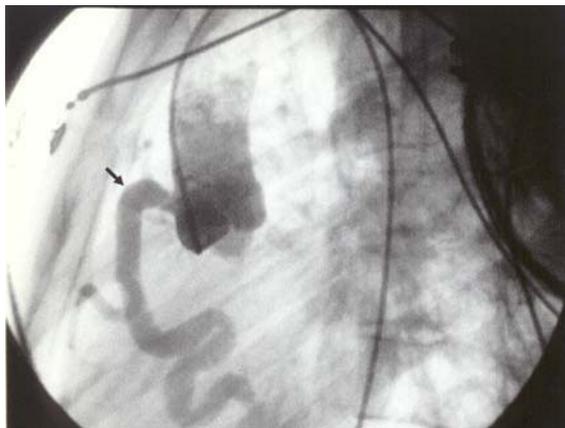


Fig. 2a. Coronary angiography, left anterior oblique view. Catheter is in the aorta. Early phase of angiography reveals a tortuous widely dilated right coronary artery.

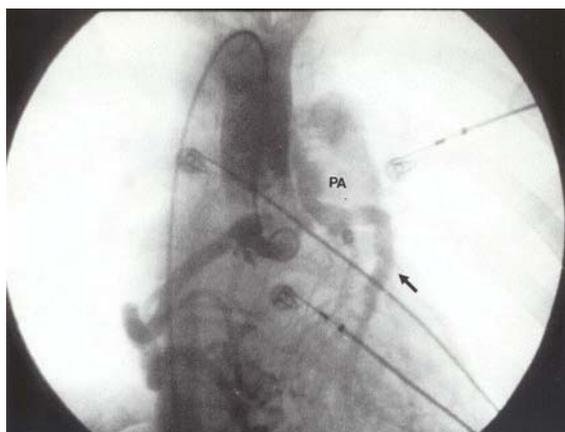


Fig. 2b. Late phase of same angiography reveals extensive collaterals and left coronary artery filling the pulmonary artery. (PA: Pulmonary artery).

## Discussion

Usually, patients with ALCAPA reveal no symptoms in the neonatal period because of physiologic hypertension and the still open ductus arteriosus. A few weeks to months after birth, most infants with this disorder become symptomatic. The balance between speed of closure of the ductus, regulation of the pulmonary hypertension and the speed of development of preexisting collateral between the right and left coronary arteries will determine the extent of myocardial necrosis<sup>3</sup>.

Anomalous origin of the left coronary artery from the pulmonary artery was classified into two groups as an infantile type and an adult

type according to clinical presentation. In general, ALCAPA is often associated with an acutely ill-appearing infant and high mortality, which is classified as infantile type. However, in rare cases, some patients reach adulthood<sup>4-6</sup>, as seen in our case. Our patient survived to this age because of extensive collateralization from the right coronary artery to the left coronary artery as revealed by coronary angiography.

The diagnosis of coronary artery anomalies requires a high index of suspicion during the history and physical examination. Not in infants but in children and adolescents, nonspecific symptoms include shortness of breath, atypical chest pain, fatigue on exertion, and palpitations, as seen in our patient who was admitted for attacks of dyspnea and palpitations. However, sudden death may be the first and only sign of this syndrome. Clues to diagnosis of ALCAPA include a continuous murmur, a murmur of mitral insufficiency, or signs of left ventricular dysfunction. Although not fully satisfactory, echocardiogram may be used for confirmation of this syndrome. In selected cases transesophageal echocardiography may be useful in the diagnosis<sup>7</sup>. Although other techniques may adequately identify the origin of the coronary artery in an anomalous presentation, only coronary arteriography reliably documents the course of the coronary artery<sup>8</sup>. As shown in our case, coronary artery venous fistula was suspected before angiography and the exact anomaly was later confirmed by coronary arteriography.

An aggressive approach to early repair in all children with ALCAPA is warranted, regardless of the degree of the left ventricular dysfunction. By histologic studies it has been shown that, in patients with ALCAPA syndrome, biopsy specimens taken from the region perfused by the anomalous artery showed a variable degree of fibrosis<sup>3</sup>. Corrective coronary artery surgery appears to be the most reasonable choice for patients with ALCAPA. Although the patients with adult type are more asymptomatic, surgical correction should be undertaken as soon as the diagnosis is established.

The severity of preoperative cardiac dysfunction and ventricular dilation is not predictive of outcome, whereas the degree of preoperative mitral regurgitation is<sup>9</sup>. Even some patients who have severe mitral regurgitation may require mitral valve replacement<sup>10</sup>. In this case, although

a moderate degree of mitral regurgitation was demonstrated, this was not considered to be a poor prognostic factor.

After surgical repair to establish blood flow to the left coronary artery from the aorta dramatic improvement in left ventricular function occurs and normalizes within two to three years<sup>2,3,11,12</sup>. However, the extent of recovery of myocardial blood flow reserve and its potential physiologic significance in long-term survivors of surgical repair of ALCAPA are not known<sup>12</sup>.

In conclusion, whenever an adult patient is admitted for dyspnea and palpitation, adult type ALCAPA syndrome should be suspected. Because of marked improvement in morbidity and mortality with surgical correction, it is important to recognize this rare disorder<sup>2,10</sup>.

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