

# Ventricular Fibrillation in a Teenager as First Manifestation of Anomalous Origin of Left Coronary Artery from Pulmonary Artery

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The anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome or ALCAPA) is a rare cause of sudden cardiac death in adults. We describe the case of a teenager with ventricular fibrillation as the first manifestation of this syndrome. The patient was successfully resuscitated and remains asymptomatic over one year, following surgical reimplantation of the left coronary artery to the aorta.

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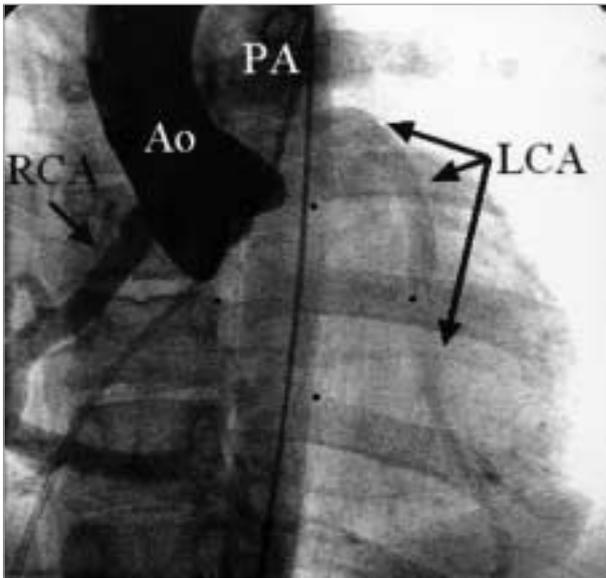
**A**nomalous left coronary artery (LCA) originating from the pulmonary artery (ALCAPA) is a rare congenital anomaly with significant mortality during childhood<sup>1,2</sup>. In the rare patient who survives to adulthood, disease manifestations range from non-specific symptoms of angina and fatigue during exertion to sudden death in the previously asymptomatic patient<sup>3-6</sup>. The diagnosis of the latter condition is stated postmortem, as very few patients survive cardiac arrest due to malignant ventricular arrhythmias.

As far as we can determine from the literature, we present the youngest formerly asymptomatic patient with ALCAPA, who survived resuscitation of ventricular fibrillation and remains asymptomatic over a one year period following successful surgery.

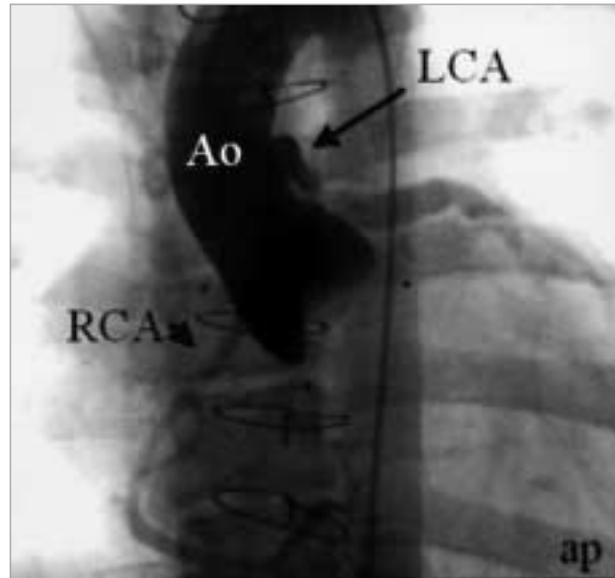
## Case report

A 15 year old teenager suffered an episode of cardiac arrest accompanied with generalized seizures. Cardiopulmonary resuscitation was conducted immediately. Upon arrival of the emergency helicopter, ventricular fibrillation was documented and converted to sinus rhythm by

defibrillation (240 J). The patient was intubated and transported to the regional general hospital. Upon admission, pulmonary congestion was clinically detected and effectively treated with furosemide. The electrocardiogram showed repolarisation abnormalities in the anteroseptal leads and the blood tests revealed increased cardiac enzymes (CK-MB max 1166 / 117 U/l, Troponin I 99,8 µg/L). Eight hours later an echocardiogram showed normal left ventricular contractility and normal cardiac chamber dimensions with no mitral valve regurgitation. Colour-Doppler imaging showed a continuous systolic-diastolic flow into the pulmonary artery, which led to the suspected diagnosis of ALCAPA. The patient was transported to the University Hospital of Freiburg, where a coronary angiography confirmed the diagnosis (Figure 1). Thallium-201 myocardial imaging revealed a reduced perfusion of the anterior wall without a complete transmural defect. Corrective surgery was performed 8 days after the cardiac arrest. Following medial sternotomy and cardiopulmonary bypass, the markedly dilated left coronary artery was mobilised and reimplanted into the ascending aorta, using an interponate of auto-



**Figure 1.** Preoperative aortic root (Ao) angiography: visualization of the dilated right coronary artery (RCA), and of the retrograde filling of the left coronary artery (LCA) which is not arising from the aorta. Contrast medium slightly visualized the pulmonary artery (PA, marked by the pigtail catheter), demonstrating the pulmonary steal phenomenon.



**Figure 2.** Postoperative aortic root (Ao) angiography: Both coronary arteries are visualised, the left coronary artery (LCA) has been surgically reimplanted to aorta. The right coronary artery (RCA) has been reduced to normal size one year after surgery.

logous pericardium to complete the anterior aspect of the anastomosis. The pulmonary artery was closed using a pericardial patch. Aortic cross clamp and cardiopulmonary bypass times were 68 min and 157 min, respectively. Mechanical ventilation was discontinued 2 hours later and the patient was discharged from the hospital 7 days later. One year after surgery his medical history was negative for clinical signs of myocardial ischemia or congestive heart failure. In the one-year follow-up examination remaining negative T waves at the anteroseptal leads were noted at the resting ECG. The exercise test and cardiac SPECT scintigraphy were negative for ischemia. Coronary angiography showed normal flow through the reimplanted left coronary artery (Figure 2).

## Discussion

Coronary artery anomalies are rare (0,2% to 1,2%) in the general population and some forms are associated with a significantly increased risk of sudden death<sup>1-4</sup>. Anomalous origin of the LCA from the pulmonary artery was first described in 1885 by Brooks<sup>5</sup> and Bland. White and Garland in 1933 observed the association of this anomaly with the clinical syn-

drome of angina, myocardial ischemia and death in early infancy<sup>6</sup>. This rare syndrome (incidence of 1 / 300000 live births) manifests itself in early infancy with congestive heart failure, often mimicking dilated cardiomyopathy with severe mitral valve regurgitation and is associated with a high mortality<sup>7</sup>. After the first weeks of life, following the normal decline in the pulmonary blood pressure, the left ventricle is perfused with desaturated blood at low pressures. The affected infants manifest paroxysmal attacks of acute myocardial ischemia precipitated by exertion during feeding or crying. During these attacks infants are severely distressed with marked pallor and a shock-like appearance. Severe heart failure leads to death in 65% to 85 before 1 year of age, usually 2 months after birth<sup>8</sup>. Only about 15% of patients reach adulthood, thanks to adequate collateral vessel development between the normal right and abnormal left coronary artery, which allows retrograde perfusion of the left coronary artery<sup>8,9</sup>. However, these patients have steal phenomenon from the left coronary artery into the pulmonary artery and associated ischemia of the left coronary artery perfusion area<sup>10</sup>. Adult patients, due to adequate myocardial perfusion, can be asymptomatic<sup>11</sup>, or may present dyspnea and angina at effort<sup>12,13</sup>. The

first symptom can be a cardiac arrest or sudden cardiac death due to malignant arrhythmia<sup>3,14</sup>. On physical examination there are signs of congestive heart failure in infants whereas in asymptomatic adults a murmur of mitral regurgitation or a soft continuous murmur at the upper left sternal border may be present.

Our patient was asymptomatic until the episode of cardiac arrest due to ventricular fibrillation. Patients with ALCAPA who were successfully resuscitated of ventricular fibrillation have been rarely reported. Weiss et al reported a previously asymptomatic 31 year old man, who was successfully resuscitated<sup>15</sup>. While another report the cases of two women, 47 and 26 year old, who were also successfully resuscitated of ventricular fibrillation<sup>16,17</sup>. Both patients were previously asymptomatic, although a heart murmur had been existed since childhood. In contrast, Askenazi et al reported failure to resuscitate a 18-year-old girl with ALCAPA, who had previously been symptomatic<sup>13</sup>. Our 15 year old patient is the youngest known previously asymptomatic patient with ALCAPA, who survived ventricular fibrillation<sup>15-17</sup>. All of the reported survivors of ventricular fibrillation were previously asymptomatic and operations using internal mammary artery graft<sup>16,17</sup> or direct reimplantation of left coronary artery to the aorta<sup>15</sup> were successful. Our patient was successfully treated using the latter technique. Both surgical approaches are superior to the simple ligation of the origin of the left coronary artery, the first described surgical treatment, since the latter cannot prevent late sudden death<sup>18</sup>. Grafts using the internal mammary artery may have a longer survival than saphenous vein grafts<sup>19</sup>. An alternative surgical approach described by Takeuchi et al, consists of establishing the connection of the anomalous left coronary artery with the aorta through a surgically created aortopulmonary window and an internal tunnel created in the main pulmonary trunk<sup>20</sup>. Although the best surgical approach is still controversial, patients with ALCAPA can be safely operated on irrespective of their age. Previously asymptomatic adult patients with ALCAPA presenting ventricular fibrillation as a first symptom of their disease, if resuscitated successfully, may have a good postoperative prognosis. A detailed evaluation to rule out ALCAPA is necessary in cases of cardiac arrest in young previously asymptomatic adults.

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