A patient with anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA syndrome) and 13 live births

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Abstract
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) or Bland–White–Garland syndrome is a rarely seen congenital anomaly. Adult and infantile types are defined according to the degree of collateral development between the left coronary artery (LCA) and right coronary artery (RCA). If left untreated, ALCAPA has a 90% mortality rate in the first year of life, primarily due to myocardial ischaemia and heart failure. The degree of collateral development and the related LCA perfusion in ALCAPA syndrome determine the occurrence of symptoms. Herein, we present a case of a female patient who had previously, without any symptoms, given live birth to 13 babies. Since our patient had well-developed collaterals to the LCA, she was asymptomatic and able to give birth to the children via the vaginal route without any problems. Having well-formed collateral vessels between the RCA and LCA may prevent patients from developing symptoms, and even stressful conditions such as pregnancy may be tolerable.

Keywords: ALCAPA syndrome, surgical, live birth

Case report
A 56-year-old female patient was admitted to our clinic with exertional angina that was relieved at rest. Her past medical history was normal except for 13 live vaginal deliveries. Electrocardiography was in sinus rhythm without any pathological ST-T abnormalities. The physical examination revealed a 2/6 murmur at the mesocardiac area. Her transthoracic echocardiography (TTE) was normal without any wall-motion abnormality and the ejection fraction was within normal limits. On continuous-wave and colour Doppler echocardiography we detected reverse flow in the pulmonary artery and mild mitral regurgitation. In the exercise stress test, a 3–4-mm ST-segment depression was detected in V3 to V6. In coronary computed tomography angiography (CCTA), the left coronary artery (LCA) was shown to arise from the pulmonary artery and a flow to supply the left ventricle (LV), then preferentially direct it into the lower-pressure pulmonary artery.

Herein, we present a case of a female patient who had previously, without any symptoms, given live birth to 13 children. The patient was treated surgically.
diagnosis of ALCAPA syndrome was made (Fig. 1). The RCA was dilated and tortuous and there were well-developed collaterals between the LCA and RCA.

Corrective surgery was planned. Under cardiopulmonary bypass, the coronary artery was resected, the origin of the LCA was prepared as a button, mobilised 4 cm and implanted directly onto the left side of the ascending aorta. A graft interposition to the main pulmonary artery was performed with a 20-mm PTFE tube. The bypass pump was switched off without any problems.

In the control CCTA one week after surgery, the LCA was noted to be normal (Fig. 2). The patient was discharged on the 10th postoperative day.

Discussion
ALCAPA syndrome is a rare congenital malformation. It occurs in one out of 300 000 live births and constitutes about 0.25–0.5% of all congenital heart defects. The major determinants for symptoms in patients with this condition are the development of collaterals and myocardial ischaemia. The level of symptoms due to ischaemia depends on the degree of development of collaterals between the LCA and RCA.1

Adult-type ALCAPA includes patients with well-developed collaterals, while those without collaterals have the infantile type of this syndrome.2 Unless this infantile type is diagnosed and treated, 90% of children die in their first year of life because of heart failure due to widespread myocardial ischaemia and mitral regurgitation.3 Adult patients can present with angina, dyspnoea, syncope, myocardial infarction or arrhythmias. Sudden cardiac death secondary to malignant ventricular arrhythmias is the most common presentation.4 The few patients who do survive to adulthood without surgery have plentiful, well-formed intercoronary collaterals with adequate perfusion of the LV.2

Chattranukulchai et al. performed corrective surgery on a 79-year-old female patient, who had been admitted to hospital with a three-month history of shortness of breath, after diagnosing her with ALCAPA.2 The abnormal origin of the LCA from the pulmonary artery, the left coronary artery flow via Doppler imaging, the dilated RCA, severe left ventricular dysfunction and mitral regurgitation can be detected by TTE.1,4 In our patient, we detected reverse flow in the pulmonary artery by colour and continuous-wave Doppler on TTE.

CCTA is an important non-invasive diagnostic tool that can be useful to determine the abnormal origin, projection and collaterals of the coronary arteries.1 In our case, we performed CCTA and detected the abnormal origin of the LCA from the pulmonary artery and the collaterals between the LCA and RCA.

In adult patients, especially those with symptoms and large left-to-right shunts, surgical correction is recommended; however, surgery is also recommended by many authors in asymptomatic individuals in order to prevent subendocardial myocardial ischaemia, ventricular arrhythmias and sudden death.1 There are several surgical approaches, including single and double coronary artery repairs. Single coronary artery repair, performed by ligation of the anomalous LCA at its pulmonary origin, has been abandoned due to the high rate of complications, including recanalisation of the ALCAPA, a greater risk of atherosclerosis, severe mitral regurgitation resulting from ischaemic cardiomyopathy and persistent risk of sudden death due to silent ischaemia.1,3

Double coronary artery repairs are preferred nowadays. These include coronary button transfer, the Takeuchi procedure, and placement of a coronary artery bypass graft combined with ligation of the origin of the LCA.4 We performed button transfer of the LCA onto the aorta in our patient.

The degree of collateral development and related LCA perfusion in ALCAPA syndrome determines the occurrence of symptoms. Since our patient had well-developed collaterals to the LCA, she was asymptomatic and able to give birth 13 times via the vaginal route without any problem. We can concluded that very well developed collaterals that effectively supply the LCA territory can prevent cardiac dysfunction and symptoms, even in patients with stressful conditions such as pregnancy and delivery.

Conclusion
In patients with an ALCAPA diagnosis, good LCA perfusion, TTE showing normal left ventricular function, no wall-motion defects and angiographically good collateral blood flow will hide the symptoms and cause delayed diagnosis.

References