

CASE PRESENTATION

Adult type Anomalous Left Coronary Artery from Pulmonary Artery

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Abstract: Anomalous origin left coronary artery from pulmonary artery, also known as ALCAPA syndrome or Bland-White-Garland syndrome, is a rare congenital anomaly. We present the case of a 22-year-old woman, with a history of cardiac pathology since 5-month year-old, that was misinterpreted as endocardial fibroelastosis until a 2 dimensional (2D)-Echocardiography revealed multiple turbulent color flow regions (intercoronary collaterals) in ventricular septum; 2D-Echocardiography was unable to visualize the left coronary artery. A left coronary artery origin anomaly was suspected, so the patient underwent coronary computed tomography (coronary CT), that was inconclusive; thus, she was referred to coronary angiography that revealed typical anatomy of ALCAPA. At present our patient is awaiting surgery.

Keywords: anomalous origin left coronary artery from pulmonary artery, 2D-Echocardiography, intercoronary collaterals, coronary CT, coronary angiography, dual coronary system.

Abstract: Originea anormală a arterei coronare stângi din artera pulmonară, cunoscută ca și sindromul ALCAPA sau sindromul Bland-White-Garland, este o anomalie congenitală rară. Vă prezentăm cazul unei paciente în vârstă de 22 de ani, cu istoric de patologie cardiacă de la vârsta de 5 luni, care a fost interpretat ca fibroelastoză endocardică; la vârsta de 22 de ani ecocardiografia bidimensională a decelat la nivelul septului interventricular trasee vasculare cu flux color turbulent (colaterale intercoronariene); la ecocardiografie nu s-a putut vizualiza artera coronară stângă. S-a ridicat suspiciunea unei anomalii de origine a arterei coronare stângi și s-a realizat coronaro-computer tomografie (coronaro-CT) care a fost neconcludentă; în acest context s-a efectuat angiografia coronariană care a evidențiat modificările tipice ale sindromului ALCAPA. În prezent pacienta a fost programată pentru intervenție chirurgicală.

Cuvinte cheie: originea anormală a arterei coronare stângi din artera pulmonară, ecocardiografia bidimensională, colaterale intercoronariene, coronaro-CT, angiografie coronariană.

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly with an incidence of 1/300.000 live births (0.4% of all congenital heart disease). ALCAPA is the most common anomaly involving a coronary artery arising from the pulmonary trunk^{1,2}. The ALCAPA anomaly may result from 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 arteries⁵. There are two types of ALCAPA syndrome: the infant type and the adult type. Infants experience myocardial infarction and congestive heart failure, and approximately 90% die within the first year

of life²⁻⁴. In a minority of cases (approximately 20%), sufficient myocardial collaterals develop from the normally arising right coronary artery, and survival into adulthood may occur¹; in these situation a left to right shunt into the pulmonary artery is created, leading to a coronary steal phenomenon that results in myocardial ischemia later in life. Few patients survive past childhood without surgical repair, and up to 90% die suddenly at a mean of age of 35 years². Here we describe a case of adult type ALCAPA syndrom.

CASE REPORT

A 22-year-old woman presents with exertional dyspnea on mild effort from approximately 6 months without any other symptoms. There were no pathological fin-

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dings at clinical examination, except tachycardia on cardiac auscultation.

The patient history prior to current observation include the diagnosis of endocardial fibroelastosis and dilated cardiomyopathy since 5-months-age. At the age of 5 months the patient was admitted to pediatric intensive care unit with severely impaired general condition presenting dyspnea with tachypnea, cough, fever, poor feeding and irritability especially during feeding. The clinical examination at that time revealed grade I dystrophy, respiratory failure signs (tachypnea, sharp pulling in of the chest below and between the ribs with each breath, flaring of the nostrils) and cardiac failure signs (tachycardia, poor peripheral perfusion with cyanosis, dilated jugulars, hepatomegaly with liver at 3 cm below the right costal margin, crackles heard in the lung bases). Cardiac auscultation revealed a II/VI ejection systolic murmur at left sternal border. Chest X ray showed global cardiomegaly, electrocardiogram (ECG) revealed sinus rhythm and deep Q waves in leads DI, aVL, V3-V6. Transthoracic echocardiogram showed dilated left ventricle with poor function and dyskinesia of the apex and the left anterior wall. The pathological signs revealed by clinical examination, chest X ray, ECG, transthoracic echocardiography suggested the diagnosis of a possible anomaly of the coronary system; thus the patient was referred to a more specialized center where she underwent cardiac catheterization and nonselective coronary angiography. Cardiac catheterization showed dilated left ventricle with global hypokinesia and severe systolic dysfunction with an ejection fraction (EF) ~26%, small apical lacunar images (possible mural thrombi or endocardial fibroelastosis), mitral regurgitation grade I, tricuspid regurgitation grade I, slight pulmonary hypertension. Nonselective coronary angiography showed both coronary artery apparently in normal limits. At that time the case was interpreted as endocardial fibroelastosis. Until the age of 11 she received positive inotrope agent and coronary dilator agents, when she stopped this treatment because she was symptom free from the age of 8.

Until the age of 22, when she was referred to our unit, she was oligosymptomatic accusing only exertional dyspnea on mild efforts. The ECG on current presentation showed sinus tachycardia with left axis deviation, an anterosuperior block, q waves and T waves inversion in leads DI and aVL, and Q waves in leads V1-V2 (aspect suggesting anterior myocardial infarction sequel), late transition of QRS complex, microvolt in precordial leads (**Figure 1**). Transthoracic echocar-

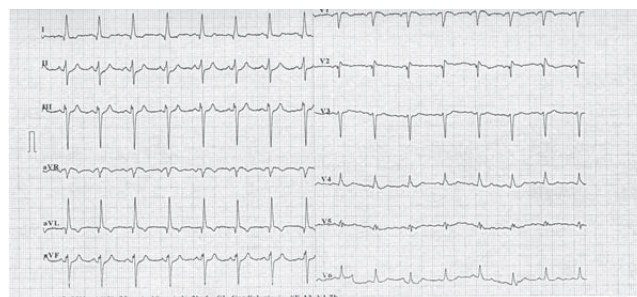


Figure 1. Twelve leads ECG showing sinus tachycardia, left axis deviation, anterosuperior block, q and T wave inversion in DI and aVL leads, Q wave in V1-V2 leads (aspect suggesting anterior myocardial infarction sequel), late transition of QRS complex, microvolt in precordial leads.

diogram showed a mild left ventricle enlargement with apical dyskinesia, with preserved left ventricle contractile function and an global ejection fraction (measured by Simpson method) of 60% (**Figure 2**). 2D-Echocardiography revealed also multiple turbulent color flow regions (intracoronary collaterals) in ventricular septum (**Figure 3**) and was unable to visualize the left coronary artery origin.

In the light of the ECG and 2D-Echocardiography findings a coronary CT was obtained and revealed a left main coronary artery with interarterial course, without showing the exact left main coronary artery origin. At this moment the patient was referred to coronary angiography that showed an enlarged right coronary artery (**Figure 4**) with many collateral branches draining into the pulmonary artery via the left coronary artery (**Figure 5**, **Figure 6**). Coronary angiogram confirmed AL-CAPA syndrome and surgical correction was planned. Before surgery a cardiac magnetic resonance imaging (MRI) was made and no signs of scar fibrosis or infarction were found.

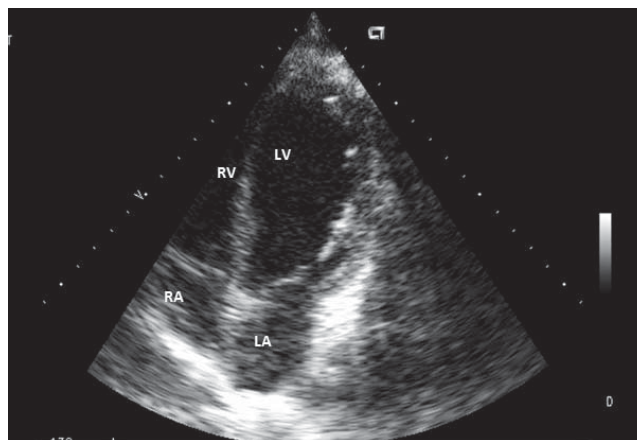


Figure 2. 2D-Echocardiography, 4 chambers view - mild left ventricle enlargement with apical dyskinesia and preserved ejection fraction. LV - left ventricle. RV - right ventricle. LA - left atrium. RA - right atrium.

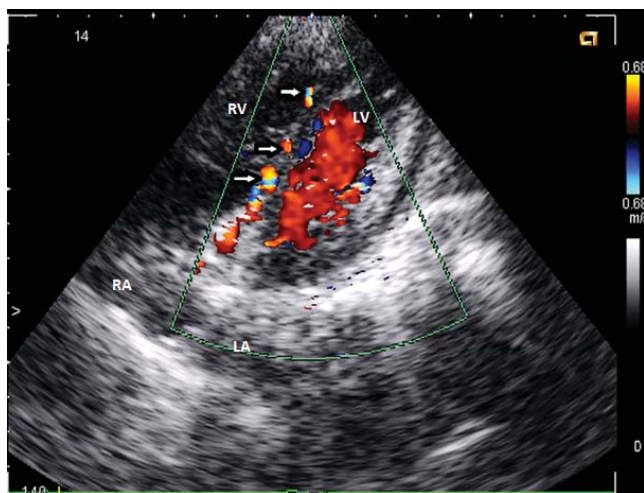


Figure 3. 2D-Echocardiography, 4 chambers view - multiple turbulent color flow regions (intercoronary collaterals) in ventricular septum (white arrows). LV - left ventricle. RV - right ventricle. LA - left atrium. RA - right atrium.

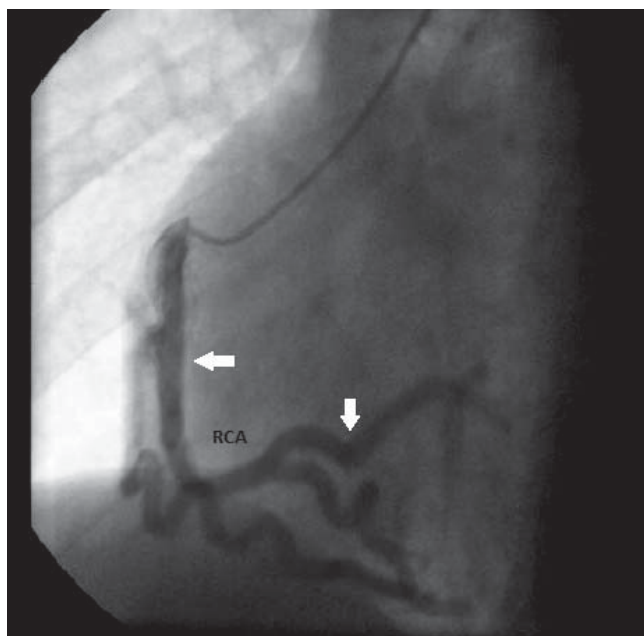


Figure 4. Coronary angiography - catheter inserted into the right coronary artery ostium; enlarged right coronary artery, without atherosclerosis lesions. RCA - right coronary artery.

DISCUSSIONS

The anomalous origin of left coronary artery from pulmonary trunk is a well known, although rare, congenital anomaly in humans. In most cases, it's an isolated anomaly but has occasionally been associated with other congenital heart defects such as patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot and coarctation of the aorta⁶. Our patient presented ALCAPA anomaly without any other congenital heart defects.

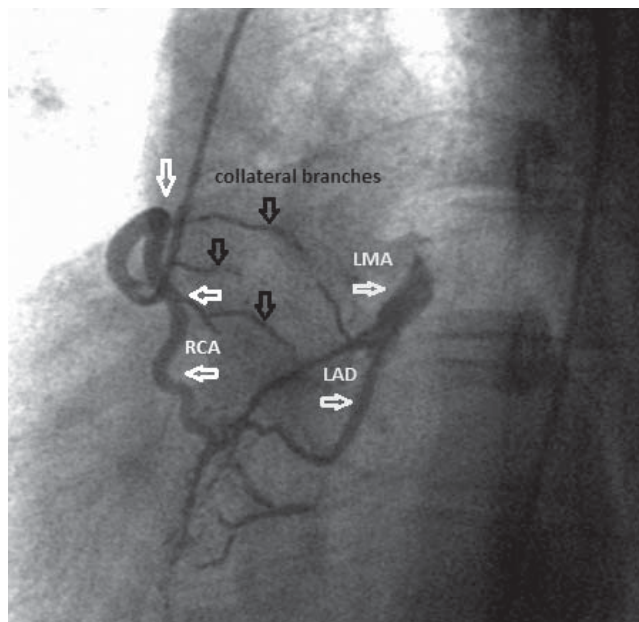


Figure 5. Right coronary angiography in LAO position showing filling of left coronary system through collateral branches (black arrows) originated from right coronary artery. RCA - right coronary artery. LMA - left main coronary artery. LAD - left anterior descending artery.

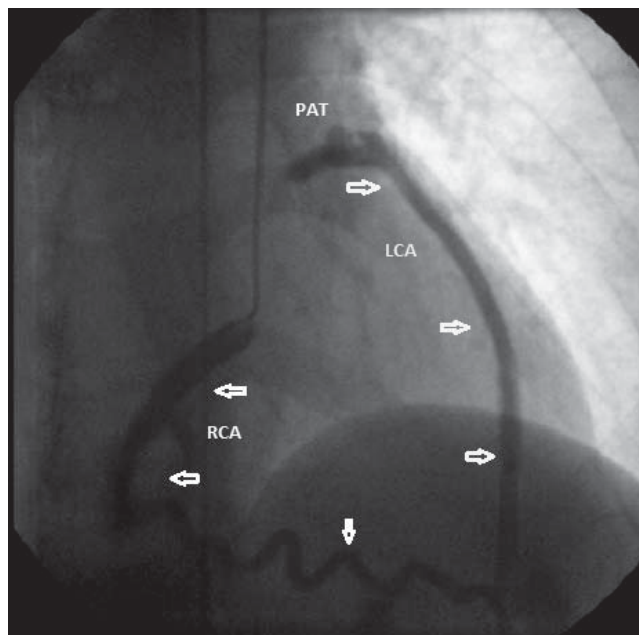


Figure 6. Coronary angiography in RAO position - catheter inserted into right coronary artery ostium; dilated right coronary artery and an abnormal origin of left coronary artery from pulmonary arterial trunk in delayed sequence. RCA - right coronary artery. LCA - left coronary artery. PAT - pulmonary arterial trunk.

The clinical expression of syndrome results from evolving morphological-functional alterations in pulmonary circulation that occur after birth. Soon after birth, resistance of the pulmonary circulation is so high permitting antegrade flow from the pulmonary artery to left coronary artery, which perfuses the left

ventricle. Therefore, occurrence of sudden death is extremely rare in this age group. As pulmonary vascular resistance falls in following weeks, flow from pulmonary artery to left coronary artery stops and left ventricular perfusion totally depends upon collaterals to left coronary artery developed from right coronary artery. Death ensues if collaterals are poorly developed while on the other hand if collaterals enlarge after an initial period of decompensation, improvement and survival into adulthood occurs – so called adult type of ALCAPA⁷. In our case there was an initial cardio-respiratory decompensation at 5 months age, when probably the collaterals from right coronary artery to left coronary artery enlarged causing a progressively increase in left coronary flow and a slow remission of clinical signs.

In patients who survive into adulthood, pulmonary circulation acts as low resistance siphon and collaterals flow uses left coronary artery as a only conduit into pulmonary circulation thus bypassing the left ventricular myocardium. This coronary steal may cause overt ischaemia as well as left ventricular diastolic overload from left to right shunting^{3,7}. In our case, the patient was nearly asymptomatic till date. The presenting symptoms of exertional dyspnea on mild efforts results from myocardial ischaemia due to failure in collateral circulation to ensure a higher flow, and thus more oxygen, to myocardium during effort.

Twelve lead ECG can alert the possibility of ALCAPA if ischemic changes are seen especially in young age group. Broad deep Q waves and associated T wave inversion in leads DI and aVL has been described as being characteristic for ALCAPA⁶. Our patient ECG at 5 months, showed deep Q waves in leads DI and aVL, which is consistent with literature data, and also deep Q waves in leads V3-V6. On ECG signs of infarction are absent in adults⁷, which is not consistent with our findings as electrocardiography at 22 years old showed small q waves with T wave inversion in leads DI, aVL and also deep Q waves in leads V1-V2, aspect suggesting anterior myocardial infarction sequel.

Echocardiography is an important diagnostic tool for the diagnosis of ALCAPA. In infants with suspicion of endocardial fibroelastosis or dilated cardiomyopathy, anomalous origin of coronary artery must be ruled out⁶. Our case was first interpreted as endocardial fibroelastosis based on radiographic (cardiomegaly), echocardiographic (dilated left ventricle with reduced ejection fraction) and angiocoronarographic (nonselective coronary angiography showed both coronary artery apparently in normal limits) findings at

5 months age. This misinterpretation was principally caused by the coronary angiography that showed both coronary artery apparently normal; we must specify that the patient underwent a nonselective coronary angiography that has its limits.

In ALCAPA patients surviving beyond one year without treatment, coronary collaterals have been found to be obvious septal color flow signals echocardiographically. The identification of these septal collaterals is the initial clue for the diagnosis of ALCAPA in patients over one year of age⁶. This echocardiographic aspect was found in our patient at 22 years old; this finding raised the suspicion of a coronary system anomaly. Also, 2D- echocardiography did not reveal mitral regurgitation, which is a common finding in ALCAPA patients. Because the ostium of the left coronary artery was not visualized the patient was submitted to other investigations to confirm the diagnosis.

Additional imaging techniques such as CT scan/ MRI are undertaken only when definitive diagnosis by echocardiography is not possible, or in an effort to exclude other potential diagnoses⁶. In our case coronary CT revealed a left main coronary artery with interarterial course, without showing the exact left main coronary artery origin; therefore we performed selective coronary angiography to confirm the diagnosis.

Three angiographic criteria are established for diagnosing ALCAPA. They are as follows⁷: 1) retrograde filling of left coronary artery, 2) connection of left coronary artery with pulmonary artery and 3) absence of left coronary artery originating from aorta. Since all three criteria are fulfilled in our case, diagnosis of ALCAPA is confirmed.

The adult form of ALCAPA is characterised by exuberance in collateral coronary circulation, which allows survival until adulthood, with case being reported at the age of 72 years⁷.

When coronary artery anomalies like ALCAPA are diagnosed, urgent surgery is often indicated in order to prevent myocardial ischemia, malignant ventricular arrhythmias and sudden cardiac death⁸.

In cases of ALCAPA several surgical treatment options have been proposed⁹. Today surgical procedures are aimed at creating a two-coronary system either via 1) a bypass graft (mammary artery or saphenous vein) in combination with ligation of the anomalous artery, 2) the Takeuchi-procedure where an intrapulmonary tunnel from the aortopulmonary window to the coronary artery is created or 3) translocation of the left coronary artery from the pulmonary trunk to the aortic sinus^{10,11}. The latter depends on the distance between

the origin of the anomalous artery and the aorta, but is possible in the majority of the cases¹². In our case, the patient was proposed for bypass graft with mamary artery.

In infants, most of the patients with corrected ALCAPA show normalization of both ventricular function and mitral valve insufficiency^{13,14}. Estimated long-term survival at 20 years was recently shown to be 94.8%¹⁴.

No long-term studies of large populations of adults with corrected ALCAPA are available, but the prognosis is generally good. The late outcome after revascularization mainly depends on the extend of irreversible impaired left ventricular function and the presence of myocardial scar tissue¹⁵. In our case cardiac MRI was made and no signs of scar fibrosis or infarction were found.

Restoration of a dual coronary system will prevent further ischemia and arrhythmias of acute ischemic origin, but the anatomical substrate for ventricular arrhythmias in patients with old myocardial infarction will not be altered after revascularization. Treatment options include drug therapy, implantable cardiac device (ICD) implantation or catheter ablation. ICD implantation has been shown to be superior to drug treatment in patients with a history of ventricular tachycardia/ventricular fibrillation and previous myocardial infarction (secondary prophylaxis) – especially when there is left ventricular dysfunction¹⁵. With recurrent ventricular tachycardia refractory to these treatment options, direct surgical ablation or resection of the arrhythmogenic focus is an option¹².

ICD implantation may also be considered in patients without a history of arrhythmias, when there is marked left ventricular dysfunction and electrophysiological study shows inducible ventricular tachycardia (primary prophylaxis)¹².

In our patient antiarrhythmic treatment is controversial considering that no arrhythmia was documented in her history, left ventricle sistolic function is preserved and cardiac MRI found no signs of scar fibrosis. In this situation is required long-term clinical and electrocardiographic (ECG and Holter ECG) monitoring.

At this moment there are no clear indications regarding pregnancy in ALCAPA womens. Pregnancy is associated with cardiac overload and may be a trigger for cardiac decompensation in these patients. There are case reports in witch some patients presented cardiac decompensation during pregnancy, but in others, ALCAPA syndrome was diagnosed after some period after birth.

CONCLUSION

Diagnosis of adulthood ALCAPA should be considered not only in adult patients presenting with clear evidence of ischemic heart disease, left ventricular dysfunction, or arrhythmias, but also more importantly in patients with minor symptoms of exercise intolerance or dyspnea that could be easily misinterpreted. ALCAPA syndrome is a congenital anomaly which must be suspected in infants and adults in the presence of ischemic electrocardiographic changes. In adults 2D-echocardiography is a very useful diagnostic tool. Additional imaging investigations are reserved for cases with uncertain echocardiographic diagnosis. Patients prognosis depends on the severity of left ventricular dysfunction and the risk of malignant arrhythmias. ALCAPA syndrome treatment is essentially surgical and should be done as soon as the diagnosis is established. Antiarrhythmic therapy should be individualized according to each patient.

Conflict of interest: none declared.

List of abbreviations:

ALCAPA	Anomalous Left Coronary Artery from Pulmonary Artery
ECG	electrocardiogram
2D- Echocardiography	2 dimentional- Echocardiography
EF	ejection fraction
coronary CT	coronary computed tomography
MRI	magnetic resonance imaging
ICD	implantable cardiac device

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