Case report of asymptomatic very late presentation of ALCAPA syndrome: review of the literature since pathophysiology until treatment

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Background
Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is an unusual congenital heart defect which affects approximately 1 in 300 000 live births and accounts for 0.5% of all congenital heart disease. Without surgical intervention, most patients die in infancy (nearly 90%).

Case summary
We present a rare case of an asymptomatic 67-year-old female. Transthoracic echocardiography demonstrated a dilated right coronary artery (RCA) and multiple collaterals. ALCAPA was confirmed by multidetector computed tomography. The left main artery was seen originating from the pulmonary artery and well-developed collaterals were visualized between the RCA and LCA. No areas of myocardial infarction were identified on cardiac magnetic resonance. Stress studies showed no inducible ischaemia.

Discussion
Our clinical case of an ALCAPA patient who survived and remained asymptomatic to their late 60’s, highlights the importance of well-collateralized and pressurized coronary system to maintain adequate myocardial perfusion. Physicians should be aware of this congenital anomaly as appropriate early diagnosis is crucial to prevent irreversible myocardial damage, acute ischaemia, and arrhythmias, and can improve patient outcomes. Surgical treatment is suggested irrespective of symptomatology or the presence of inducible myocardial ischaemia.

Keywords
Case report • Congenital coronary artery anomaly • Adult-type ALCAPA • Epicardial collaterals • No inducible myocardial ischaemia • Cardiac multidetector computed tomography

Learning points
• ALCAPA patients with well-developed collaterals network between the right and left coronary arteries that provided adequate oxygenated blood to the myocardium, can survive to advanced adulthood without symptoms.
• Stress imaging modalities might not detect myocardial ischaemia due to the possibility of a balanced ischaemia and/or hibernated myocardium.

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**Introduction**

Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) or Bland–White–Garland syndrome, is an uncommon congenital abnormality that affects 1 of every 300 000 live births and accounts for 0.25–0.5% of all congenital heart conditions. In 1885, Brooks et al. reported the first case of ALCAPA and in 1933, Bland, White, and Garland published the first comprehensive clinical presentation of this syndrome. Its origin is the mal-implantation of the vascular network into the pulmonary artery (PA) portion of the truncus arteriosus instead of the aortic side of the primitive truncus arteriosus. The patients are classified on the basis of their survival patterns into infantile or adult type.

Although ALCAPA is commonly an isolated entity, this syndrome could be associated with other congenital anomalies, including atrial and ventricular septal defect, patent arterial duct, aortic coarctation or another aortic arch abnormality, and conotruncal heart malformations such as tetralogy of Fallot or common arterial trunk.

Without treatment, the mortality of this anomaly is very high, and nearly 90% of untreated patients die in the first year of life (infancy type). Only patients with adequate collateral circulation between the right coronary artery (RCA) to the left coronary artery (LCA) may survive beyond infancy (adult type). We present an uncommon case of ALCAPA. Our patient is one of the oldest asymptomatic ALCAPA patients reported, with normal ejection fraction and no myocardial ischaemia or myocardial infarction.

**Timeline**

| October 2011 | The patient was referred to our outpatient clinic for atypical chest pain. |
| November 2011 | Transthoracic echocardiogram (TTE) showed left main artery arising from the pulmonary artery. |
| November 2011 | Cardiac multidetector computed tomography confirmed the origin of the left main coronary artery from the pulmonary artery. |
| Between November 2011 and January 2012 | Exercise stress test, stress TTE, and exercise stress myocardial perfusion scintigraphy were negative for inducible ischaemia. Cardiac magnetic resonance showed mildly dilated left ventricle with normal ejection fraction. Late gadolinium enhancement sequences displayed subtle mid-wall myocardial fibrosis at basal septum. Qp:Qs = 1.8:1. |
| From 2012 to 2019 | The patient has been followed-up in our outpatient clinic and she remains asymptomatic from a cardiovascular perspective with good exercise capacity. |

**Case presentation**

A 67-year-old female was referred to our centre for atypical chest pain of mechanical characteristics. She had no previous medical history, and she was an ex-smoker. Blood pressure on the clinic was 130/67 mmHg and heart rate was 70 b.p.m. Physical examination revealed a 3/6 holosystolic murmur in the precordium. The electrocardiogram showed sinus rhythm (SR), normal QRS duration, and no ST-segment or T wave abnormalities. Routine blood tests were normal. A transthoracic echocardiogram (TTE) showed surprisingly a dilated RCA arising from the right sinus of Valsalva and an aneurysmal left main artery (LMA) arising from the dilated PA (Video 1). Doppler colour imaging showed diastolic flow in the RCA filing the left coronary arteries via epicardial collaterals (Video 2). Biventricular ejection fraction was normal. The mitral valve was myxomatous with no papillary muscle abnormalities and mild to moderate regurgitation. A cardiac magnetic resonance (CMR) showed mildly dilated LV with normal ejection fraction (LVEF: 60%). Late gadolinium enhancement sequences demonstrated very subtle mid-wall myocardial fibrosis at...
the basal septum, with no areas of myocardial infarction (Figure 1). The pulmonary to systemic blood flow ratio (Qp:Qs ratio) was 1.8.

A cardiac multidetector computed tomography confirmed the anomalous origin of the LMA from the PA. The RCA was arising from the aorta running a normal course. The RCA was markedly dilated throughout its extent with large collaterals across the anterior right ventricle free wall and septum terminating to the LCA system. The coronary arteries were free of atherosclerosis or other congenital anomalies (Figure 2). Exercise stress echocardiography showed no regional wall motion abnormalities and no deterioration in the degree of mitral regurgitation at peak stress. Exercise stress myocardial perfusion scintigraphy showed no perfusion defect. A 24-h electrocardiogram tape showed SR with no arrhythmic events.

Although surgical correction is the recommended treatment of ALCAPA, medical treatment was decided for our patient. The decision was based on patient’s desire. A beta-blocker and aspirin were commenced. The patient has been followed for 7 years, and she remains asymptomatic from a cardiovascular perspective with good exercise capacity. The exercise echocardiogram was repeated 2 years ago and was still negative for ischaemia.

**Discussion**

ALCAPA presentation in adults is rare, and around 90% of adults may suffer from sudden death at a mean age of 35. It is very uncommon patients to exceed the age of 50 and at this stage they may present with symptoms of new-onset exertional angina, myocardial infarction, dyspnoea, heart failure, syncope, or sudden death. A well-collateralized and pressurized system would prevent significant hypoperfusion and will allow survival to adulthood. However, the left to right shunt will cause a chronic increase in LV preload resulting to gradual dilatation of the LV and deterioration of LV function.

The onset of symptoms and the degree of myocardial dysfunction depends on the timing of ductus arteriosus closure, the pressure difference between the pulmonary artery and coronary arteries, and the development of collaterals from the RCA to the LCA. A well-collateralized and pressurized system would prevent significant hypoperfusion and will allow survival to adulthood. However, the left to right shunt will cause a chronic increase in LV preload resulting to gradual dilatation of the LV and deterioration of LV function.

The echocardiographic findings of ALCAPA include a dilated RCA arising from the aorta, diastolic blood flow detected with colour Doppler in the interventricular septum, and an LCA arising from the PA. Multidetector computed tomography is a useful tool to confirm anomalous coronary arteries origin, and course, and CMR allows for imaging of the coronary arteries and surrounding structures.
assessment of ventricular function, myocardial ischaemia, and also proximal coronary arteries course. However, imaging modalities might not detect myocardial ischaemia due to the possibility of hibernated myocardium and/or balanced ischaemia.

When ALCAPA is diagnosed, even in asymptomatic patients, surgical treatment is suggested in order to prevent irreversible myocardial damage, malignant ventricular arrhythmias, and sudden cardiac death, regardless of the presence of myocardial ischaemia in functional tests. In young children, good results have been reported after re-implantation of the LCA into the aorta or after bypass grafting. If the above techniques are not feasible, due to anatomical limitations or significant co-morbidities, closure of the anomalous artery with surgical ligation or percutaneously may lead to clinical improvement. In patients with significant mitral regurgitation, mitral valve repair may also be necessary. However, despite a successful anatomical repair, left ventricular function and/or myocardial blood flow may be remained impaired. So, early surgical correction of this condition at the time of diagnosis with the aim of creating a two-coronary system is highly recommended in order to improve patient’s outcomes.

Conclusions
Our case report highlights the possibility that ALCAPA may undergo undiagnosed for decades and patients can survive with no symptoms.
to their late 60’s. This can be attributed to the presence of well-collateralized and/or pressurized coronary network system from RCA to LCA. Surgical treatment is suggested irrespective of symptoms and the presence of inducible myocardial ischaemia.

**Lead author biography**

Dr Esther Cambronero-Cortinas was born in Cuenca, Spain, and studied Medicine in Albacete, Spain at University of Castilla-La Mancha, where she was qualified in 2007. She then came to University Hospital of Albacete for doing her specialist register in Cardiology. In 2013, she was qualified as Cardiology Specialist. After this, she started her specialization in Cardiovascular Imaging and Congenital Heart Disease with training in ‘La Paz’ Hospital, Royal Brompton Hospital, Barts Hospital, Hammersmith Hospital, and Kings College Hospital. She is currently Cardiovascular Consultant in Cardiovascular Imaging and Congenital Heart Disease at University Hospital of Valladolid.

**Supplementary material**

**Supplementary material** is available at *European Heart Journal - Case Reports* online.

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidelines.

**Conflict of interest:** none declared.

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