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**CASE REPORT**
*Congenital heart disease*

# Late adult presentation of ALCAPA syndrome: need for a new clinical classification? A case report and literature overview

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**Background** Anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA) is a very uncommon congenital coronary artery anomaly, most commonly presenting in early infancy. Late adult presentation of ALCAPA syndrome is extremely rare.

**Case summary** We present a case of a 76-year-old patient with first presentation of ALCAPA. The coronary anomaly was first diagnosed during elective coronary angiography. The case was discussed at the Heart Team meeting and as the patient was asymptomatic, had good coronary collateral circulation, a medical treatment strategy was chosen and the patient was discharged in a good physical condition. During 3 years of follow-up, the patient has experienced no cardiovascular complications.

**Discussion** We hereby also discuss briefly the clinical presentation, epidemiology, diagnostics and treatment options for adults with newly diagnosed ALCAPA and discuss the need for a new clinical classification. Only a few cases have been published of septuagenarians or octogenarians with first presentation of ALCAPA. To our knowledge, the patient presented in our case was one of the least symptomatic patients during her eight decades of life.

**Keywords** Coronary artery anomaly • ALCAPA • Bland–White–Garland syndrome • Case report

## Learning points

- Congenital heart disease may present at an advanced age.
- Congenital anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA) with late presentation has specific characteristics such as mildness of symptoms and compensatory anatomic changes compared to the infant type ALCAPA.
- First presentation of ALCAPA in the elderly is extremely rare, but these patients are usually asymptomatic or mildly symptomatic.

## Introduction

Anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA) is a very uncommon congenital coronary artery anomaly. Late adult presentation of ALCAPA syndrome is extremely rare. Prior to modern diagnostics and surgical care, most cases were found in infants who died in the first year of life. Symptoms usually develop in infants aged 2–3 months and without surgical treatment, the infantile type is associated with high mortality. However, survival into adult life without surgery is possible. ALCAPA patients have been

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classified into several types based on the clinical presentation, most commonly to infantile and adult types. With modern diagnostics, more asymptomatic or mildly symptomatic elderly patients with ALCAPA are found. In these rare patients, who present with no or mild symptoms, the risk for surgical correction would outweigh the potential benefit.

We present a case of an elderly patient who first presented with ALCAPA in the 8th decade of life. This case was managed conservatively. We also discuss the clinical presentation, epidemiology, diagnostics and treatment options for adults with first diagnosed ALCAPA and review the need for a new clinical classification.

## Timeline

| | |
|---|---|
| Summer 2016 | First symptoms of atrial fibrillation |
| September 2016 | Apical wall abnormalities discovered on bedside echocardiography |
| November 2016 | Underwent coronary angiography for differential diagnosis due to apical wall abnormalities where was diagnosed with anomalous origin of the left main coronary artery from the pulmonary artery |
| November 2016 | Discharged with no further interventions planned due to few symptoms |
| December 2019 | No cardiovascular complications have occurred during 3 years of follow-up |

## Case presentation

In November 2016, a 76-year-old woman was admitted electively to Tartu University Hospital for further investigations due to atrial fibrillation and regional wall motion abnormalities that had been found previously. The past medical history of the patient was insignificant: she had been active in childhood and did not describe any issues with regular physical activities; besides two normal pregnancies and two deliveries, she had never experienced exercise intolerance and recently took part in Nordic walking tours. At presentation, the patient was asymptomatic, but previously, in June 2016, the patient had experienced palpitations and an irregular pulse and therefore underwent electrocardiography (ECG), where atrial fibrillation was found. In September 2016, the patient underwent echocardiography in an outpatient clinic, which demonstrated apical wall motion abnormalities, and therefore was referred to our tertiary care centre for additional investigations. She was started on outpatient rate control and anticoagulation for prevention of thromboembolism and diuretics due to asymptomatic left ventricular dysfunction. She had undergone outpatient Holter monitoring which demonstrated permanent atrial fibrillation and an average heart rate of 73 b.p.m.

Upon presentation to our centre, the patient had been prescribed with metoprolol extended release 50 mg b.i.d., verapamil extended release 120 mg b.i.d., aspirin 75 mg o.d., spironolactone 25 mg o.d., furosemide 20 mg o.d., atorvastatin 20 mg o.d., and warfarin 6/7.5 mg on alternating days. The combination of verapamil and metoprolol had been selected in order to achieve an evidence-based heart rate control (a resting heart rate of <110 b.p.m.). Although neither of the used medications was on the maximum dose and this combination is generally avoided, in this case, it was decided by her prior care providers that this combination was optimal.

Upon presentation, the ECG demonstrated atrial fibrillation with adequate rate control and a resting heart rate of 70 b.p.m. The patient was in a good general condition, no pitting oedema or cyanosis was seen and peripheral pulses were palpable on all arteries. During her hospitalization, she was monitored with telemetry and her resting heart rate was normal at all times. Both systolic and diastolic blood pressures were normal. Inpatient exercise tolerance test demonstrated low exercise capacity (4.6 metabolic equivalents of task) and significant ST-depression without anginal symptoms.

Coronary angiography was undertaken, where no left coronary artery (LCA) was identified with standard techniques. Subsequently, a large right coronary artery (RCA) was found with collateral system to the LCA system (*Figures 1* and *2*). The coronary arteries had minimal atherosclerotic lesions. Coronary computed tomography angiography (CCTA) confirmed the anomalous origin of the LCA from the pulmonary trunk 2.2 cm above the pulmonary valve. Retrograde blood flow to the pulmonary artery was demonstrated also with CCTA (*Figures 3* and *4*).

Echocardiography showed akinesia and apical aneurysm in the left ventricular apex, left ventricular ejection fraction of 41%, slight prolapse of the anterior leaflet of the mitral valve, and moderate mitral regurgitation. The large RCA was echocardiographically visible in the parasternal long (*Figure 5*) and short axis. Retrograde flow from the anomalous LCA into the pulmonary artery was visible on Doppler echocardiography.

The case was discussed at the Heart Team meeting. The patient had no anginal symptoms or dyspnoea following a lenient rate control strategy and there were no signs of congestive heart failure. The

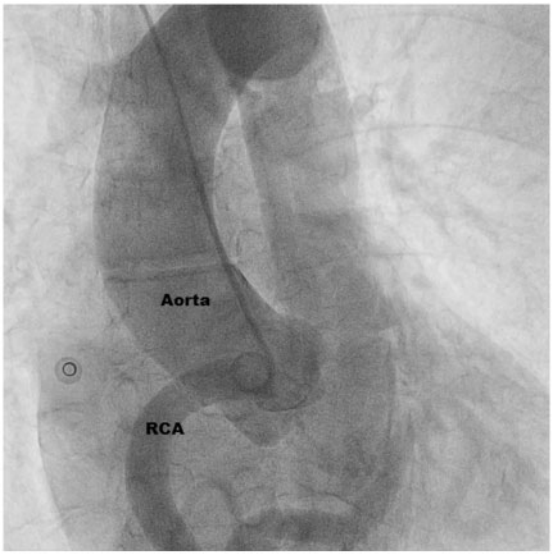



**Figure 1** Aortography with a single coronary artery (right coronary artery) from the aorta.

patient had good coronary collateral circulation, thus it was unclear whether there would be any benefit from either surgical ligation of the LCA or re-implanting the anomalous artery. Therefore, a medical treatment strategy was chosen and the patient was discharged in a good physical condition with a follow-up at her family doctor for rate-control and anticoagulation monitoring. The discharge medications included metoprolol extended release 50 mg b.i.d., verapamil extended release 120 mg o.d., furosemide 20 mg o.d., atorvastatin 20 mg o.d., and warfarin. As the patient was content with her previous medications, the combination treatment for her rate control was not altered, but aspirin was discontinued, as no signs of an acute coronary syndrome were found. The patient has had no cardiovascular rehospitalizations during 3 years of follow-up and is regularly attending follow-up visits at her primary care provider. No additional cardiovascular investigations have been deemed necessary.

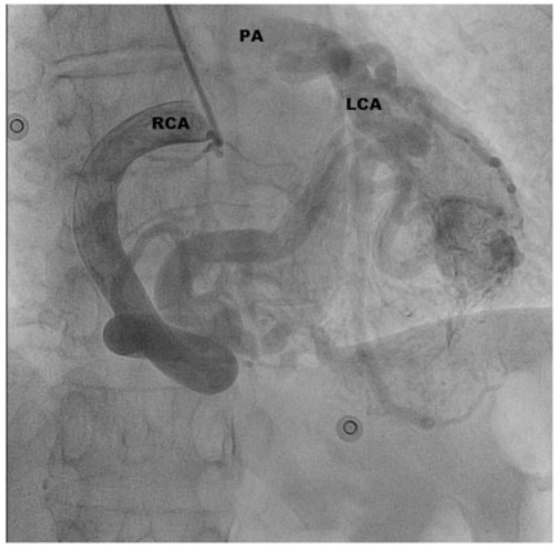



**Figure 2** Coronarography with a severely dilated right coronary artery, collateral flow, and anomalous left coronary artery with retrograde flow into the pulmonary artery.

## Discussion

ALCAPA is an extremely uncommon congenital coronary artery anomaly, most commonly presenting in early infancy. The ALCAPA syndrome is also known eponymously as Bland–White–Garland syndrome after the authors who in 1933 characterized its clinical and pathological data and recorded an electrocardiogram in an infant dying of this condition.[1] Due to the rarity of the condition, its precise

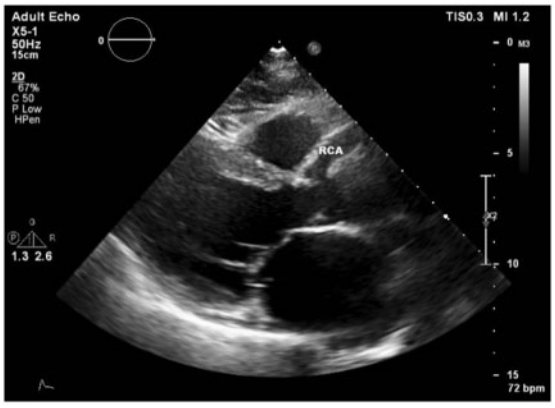



**Figure 4** Parasternal long axis echocardiography. Dilated right coronary artery clearly visible.

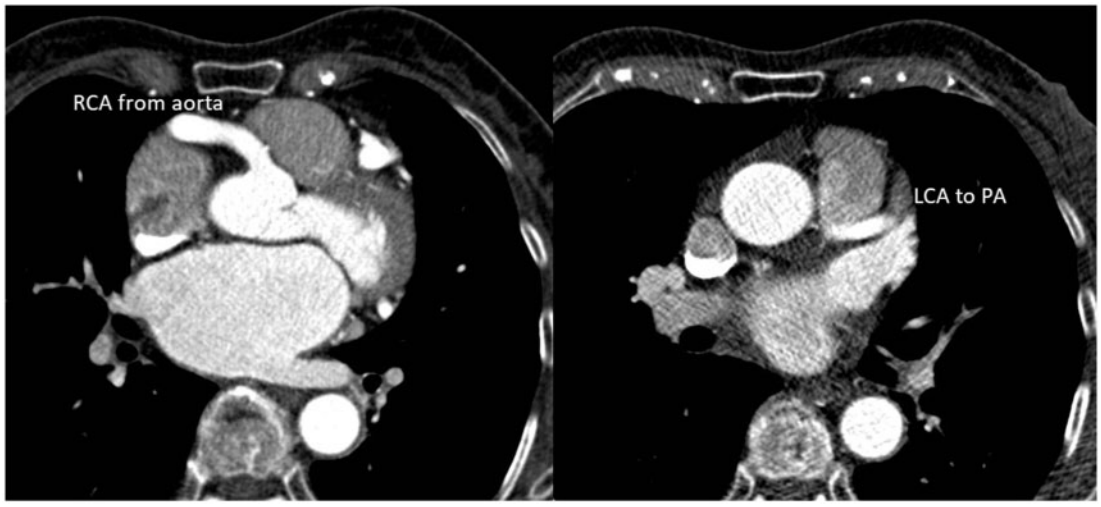



**Figure 3** Coronary computed tomography two-dimensional views of the proximal right coronary artery from the aorta and proximal and anomalous left coronary artery flowing into the pulmonary artery.

incidence is not known but is estimated at one in 300 000 live births.[2] The prevalence in adults in not known. Prior to modern diagnostics and surgical care, most cases were found in infants who died in the first year of life. However, survival into adult life without surgery is possible, and this is the case in approximately 15% of cases.[2] As it predominantly presents in the first year of life, diagnosis in living adults is extremely rare, but owing to modern advances in non-invasive cardiac imaging, the number of adults diagnosed with the syndrome, who have benign clinical outcomes, has increased substantially.[3]

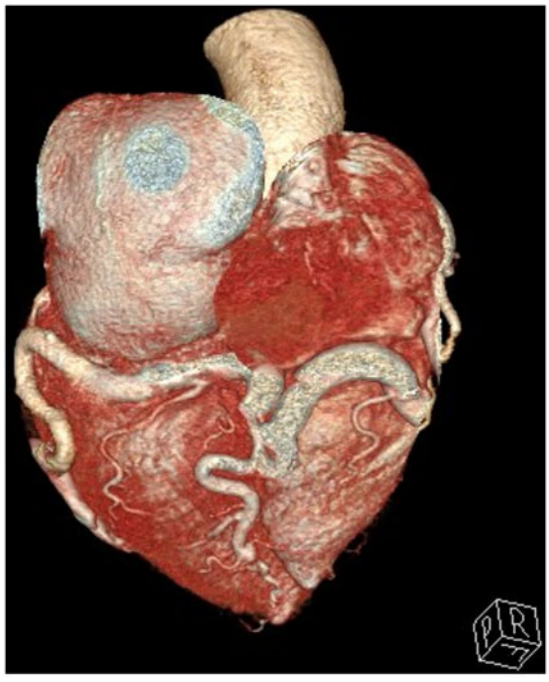


**Figure 5** Coronary computed tomography three-dimensional reconstruction of posterior collaterals from the right coronary artery to the circumflex branch of left coronary artery.

## Clinical types of ALCAPA

ALCAPA patients have been classified into several types based on the clinical presentation, most commonly to infantile and adult types. We hereby argue that a new clinical subtype—the late adult presentation type should be added to the classification (*Table 1*). The types differ in the time of development of symptoms and mortality. Patients with the infantile type present with early myocardial ischaemia, left ventricular dysfunction, and dilatation and mitral regurgitation due to little or no coronary collateral flow. Symptoms usually develop in infants aged 2–3 months and include circumoral pallor, poor weight gain, persistent tachypnoea, and tachycardia.[4] Without surgical treatment, the infantile type is associated with high mortality.

Patients with the adult type ALCAPA experience symptoms that include dyspnoea, angina pectoris, reduced exercise tolerance, and sudden cardiac death. In a comprehensive literature review of 151 published adult cases from 1908 to 2008, Yau *et al.*[3] found that 12% of adult patients were diagnosed at autopsy and the average age of presentation was 40.6 ± 15 years (the oldest being 83 years). At the time of presentation, 66% had symptoms of angina, dyspnoea, palpitations, or fatigue. Seventeen percent presented with life-threatening symptoms (ventricular arrhythmia, syncope, or sudden death).[3]

With modern diagnostics, more asymptomatic or mildly symptomatic elderly patients with ALCAPA are found. In adult patients, the risk of sudden death appears to decline after age 50 years despite less frequent surgical correction in this population.[3] In these rare patients, who present with no or mild symptoms, the risk for surgical correction would outweigh the potential benefit, so these patients should be managed conservatively. Only a few cases have been reported of patients presenting with ALCAPA in the 7th or 8th decade of life.[3,5]

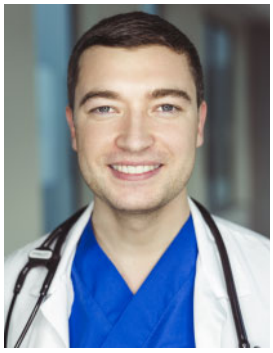
**Table 1 Three distinct types of ALCAPA**

| | Infant type | Symptomatic adult type | Late adult type |
|---|---|---|---|
| Time of presentation | Birth to 3 years of age | Adolescence to 6th decade in life | From 7th decade of life |
| Symptoms | LV infarction, heart failure | Chronic ischaemia, arrhythmias, sudden cardiac death | Mild to no symptoms, no limiting symptoms |
| Echocardiographic findings[6,7] | Dilated LV, anterior, and lateral kinetics disorder | Possible kinetics disorder, mitral regurgitation | Grossly enlarged RCA, visible on regular echocardiography, no dilation of the LV |
| Collateral flow from RCA to LCA[6] | No or very little | Good collaterals with retrograde perfusion of the left ventricle through the RCA | Good collaterals with retrograde perfusion of the left ventricle through the RCA |
| RCA dominant coronary tree | Possible | Usually | Always |
| LCA anatomy | Regular | Possible ostial stenosis | Possible ostial stenosis |
| Systemic blood supply to the LCA | No supply | Dilated bronchial arteries form collaterals to the LCA | Dilated bronchial arteries form collaterals to the LCA |

## Conclusions

ALCAPA is extremely rare in elderly asymptomatic or mildly symptomatic patients, but prognosis in these cases is significantly better than in patients who present in childhood or early adulthood. Only a few cases have been published of septuagenarians or octogenarians with first presentation of ALCAPA. To our knowledge, the patient presented in our case was one of the least symptomatic patients during her eight decades of life. In patients presenting with no or mild symptoms in late adulthood, cardiac surgery is usually not indicated, because the risks would outweigh the benefits. Diagnostics should still be performed until definite diagnosis is established. The type of intervention should be individually tailored and the decision to follow medical treatment should be taken by a Heart Team that includes general cardiologists, interventional cardiologist, cardiac surgeons, and others.

## Lead author biography



Dr Eno-Martin Lotman graduated from the Faculty of Medicine, University of Tartu in 2014. He completed the general cardiology residency in 2019. He is currently working at the North Estonia Medical Centre, Department of Cardiac Intensive Care.

## Supplementary material

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

**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 guidance.

**Conflict of interest:** none declared.

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