






Anomalous origin of coronary arteries from pulmonary artery in adults: a case series

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Background

Anomalous origin of a coronary artery from the pulmonary trunk is a small group of rare congenital anomalies present in up to 1% of the population. These patients, in absence of an adequate collateral supply, may present with congestive heart failure secondary to ischaemia, arrhythmia, or sudden cardiac death in up to 90% of cases within the first months of life.

Case summary

We present four cases diagnosed in adulthood over 10 years in two high-volume centres. The first patient presented with dyspnoea and orthopnoea. The second with chest pain and episodes of non-sustained ventricular tachycardia. The third patient presented during her third pregnancy with chest pain, palpitations, and arrhythmia (non-sustained ventricular tachycardia). The fourth patient presented with sudden cardiac death.

Discussion

In all cases with anomalous origin of coronary arteries, it is recommendable to consider surgical correction to avoid the progression of ischaemia, congestive heart failure, arrhythmia, and sudden death.

Keywords

ARCAPA • ALCAPA • Anomalous origin of coronary artery • Case report • Case series

Learning points

- An anomalous origin of the left coronary artery from the pulmonary artery accounts for 0.25–0.5% of diagnosed congenital heart disease, whilst an anomalous origin of the right coronary artery from the pulmonary artery is rare still accounting for only 0.002%.
- Clinical presentation is variable and can include an incidental finding of a murmur, angina, silent ischaemia, left ventricular dysfunction, or congestive heart failure.
- In all cases of anomalous origin of coronary arteries from pulmonary artery, surgical correction is recommended to avoid a progressive of ischaemia, arrhythmias, or sudden death.

Introduction

Coronary artery anomalies are present in up to 1% of the population. Specifically, the anomalous origin of a coronary artery from the pulmonary trunk is a small group of rare anomalies leading to a high risk of myocardial infarction and cardiac arrest or sudden death.¹

The anomalous origin of the left coronary artery from the pulmonary artery, known as ALCAPA, appears in 1 of 300 000 live births and about 0.25–0.5% of all congenital heart disease. An anomalous origin of the right coronary artery from the pulmonary artery, or ARCAPA, is less frequent, around 0.002% of all congenital heart disease.² We present three cases of ALCAPA and one of ARCAPA diagnosed in adulthood over 10 years (2008–18) in two high-volume university hospitals. In our series (30 341 patients), the overall incidence was 0.01%, the majority were female and underwent cardiac catheterization due to cardiac symptoms.

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Timeline

	Age at presentation	First cardiological evaluation	Presenting symptoms	Anatomical diagnosis	Treatment	Follow-up and outcomes
Case 1	73 years old	January 2010	Dyspnoea and orthopnoea. No chest pain	ARCAPA	Medical therapy (refused surgery)	Died 7 years after diagnosis, in 2017
Case 2	60 years old	February 2014	Chest pain and arrhythmia (non-sustained ventricular tachycardia)	ALCAPA	Surgical (coronary artery bypass grafting, CABG) in May 2014	Alive, with no symptoms and no complications after surgery (date of last follow-up, July 2019)
Case 3	32 years old	May 2015	Chest pain, palpitations, and arrhythmia (non-sustained ventricular tachycardia) during her the third pregnancy	ALCAPA	Medical (refused surgery)	Alive, with dyspnoea and palpitations (date of last follow-up, July 2019)
Case 4	31 years old	November 2018	Out-of-hospital sudden cardiac death treated with defibrillation	ALCAPA	Surgical (CABG) in November 2018	Alive, with no symptoms and no complications after surgery (date of last follow-up, July 2019)

Case presentation

Patient 1

A 73-year-old male patient with no past history of myocardial infarction, presented with dyspnoea and orthopnoea. He did not report any chest pain. On examination, a systolic murmur was audible over the mitral valve area and a diastolic murmur over aortic valve area. Electrocardiogram (ECG) demonstrated a left bundle branch block. Transthoracic echocardiogram (TTE) showed a dilated left ventricle (LV) with global hypokinesis and a left ventricular ejection fraction (LVEF) of 17%. In addition, he had severe mitral regurgitation, moderate to severe aortic regurgitation, and pulmonary systolic artery pressure (PSAP) 67 mmHg (normal reference values for PSAP < 36 mmHg). Coronary angiography revealed an ectatic left anterior descending artery (LAD), which provided collaterals to the right coronary artery (RCA). Retrograde filling of RCA was evident with drainage into the pulmonary artery (PA) (Figure 1, Supplementary material online, S1). A diagnosis of ARCAPA was then made and surgical correction was offered which the patient refused. Medical therapy with betablockers, angiotensin-converting enzyme inhibitors (ACEIs), and diuretic therapy was initiated. The patient did not receive device-based therapy. The patient died 7 years of post-diagnosis.

Patient 2

A 60-year-old female patient presented with chest pain and episodes of arrhythmia (non-sustained ventricular tachycardia) which were treated with oral betablockers. On examination, a systolic murmur was audible over the mitral valve area. Electrocardiogram demonstrated a left bundle branch block. Transthoracic echocardiogram showed normal left ventricular size, an ejection fraction of 55% and anterior, inferior, and posterior segment akinesia at the basal and

mid-levels. In addition, the transoesophageal echocardiography demonstrated severe mitral regurgitation and a PSAP of 21 mmHg. Coronary angiography revealed an ectatic RCA with normal filling which provided collateral circulation to the left circumflex artery (LCx) and LAD. Drainage of left main stem (LMS) into the PA was also evident. Coronary computed tomography angiography confirmed the diagnosis of ALCAPA. The patient underwent surgical mitral valve replacement with a mechanical prosthesis, and coronary artery bypass grafting (CABG): ligation of the left main at the pulmonary level with left internal mammary artery (LIMA) grafting to the LAD. Currently, she remains asymptomatic and has no surgical complications.

Patient 3

A 32-year-old female patient presented during her third pregnancy with chest pain, palpitations, and arrhythmia (non-sustained ventricular tachycardia). One year before the current hospital admission, she underwent cardioversion of non-sustained ventricular tachycardia and was maintained on oral therapy with betablockers. The patient withdrew medical therapy before pregnancy. Cardiac examination was normal. Electrocardiogram demonstrated a left anterior fascicular block. Transthoracic echocardiogram showed a dilated LV with no wall motion abnormalities and an LVEF of 51%. In addition, she had mild mitral regurgitation and normal PSAP of 15 mmHg. Coronary angiography performed after pregnancy revealed an ectatic RCA, with normal filling which provided collateral circulation to the LCx and LAD. Drainage of the LMS into the PA (Figure 2, Supplementary material online, S2). Coronary computed tomography angiography confirmed the diagnosis of ALCAPA. The patient refused undergo to surgery (CABG) and therefore received medical therapy with betablockers and ACEIs. Currently, the patient



Figure 1 Anomalous origin of the right coronary artery from pulmonary artery. Coronary angiography in anteroposterior view displaying an ectatic left coronary artery which gives collateral circulation to an ectatic right coronary artery with final drainage into the pulmonary artery.



Figure 2 Anomalous origin of left coronary artery from pulmonary artery. Coronary angiography in left anterior oblique view which shows an ectatic right coronary artery, with normal filling with left circumflex artery and left anterior descending artery collaterals. The drainage of left coronary artery into the pulmonary artery is depicted.

continues to experience dyspnoea and palpitations but has had no new episodes of ventricular arrhythmia.

Patient 4

A 31-year-old female patient presented with dizziness and loss of consciousness while walking. Prehospital cardiopulmonary resuscitation was initiated due to cardiac arrest and defibrillation with an automatic external defibrillator was performed for ventricular fibrillation, achieving return of spontaneous circulation. On examination, no murmur was audible on cardiac auscultation. Electrocardiogram showed left anterior fascicular block. Transthoracic echocardiogram found a normal LV with apical hypokinesia and an LVEF of 61%. In addition, mild mitral regurgitation and normal PSAP of 17 mmHg were evident on the TTE. Coronary computed tomography angiography was performed, revealing abnormal origin of the left coronary trunk in the posterior third of the PA trunk (pulmonary trunk of normal caliber) (Figure 3). Coronary angiography confirmed an ectatic RCA, with normal filling which provided collateral circulation to the LCx and LAD. Drainage of the LMS into the PA was also noted and a diagnosis of ALCAPA was made. The patient underwent surgical treatment (CABG) with ligation of the LMS at the pulmonary level, right internal mammary artery–LAD graft, and LIMA–LCx graft (Figure 4). The patient remains asymptomatic and has had no complications related to the surgical procedure.

Discussion

An anomalous origin of the coronary artery from the PA is a rare congenital anomaly. Embryologically, coronary artery anomalies

appear as a result of either malrotation of the spiral septum dividing the truncus or malpositioning of the coronary buds themselves.³

An anomalous origin of the left coronary artery from the pulmonary artery can be associated with ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and aortopulmonary window.⁴

The most common cardiac defects reported to be associated with ARCAPA are tetralogy of Fallot and aortopulmonary window anomalies. Other relatively less common anomalies include aortic stenosis and aortic coarctation.⁵

An anomalous origin of the left and right coronary arteries from the pulmonary artery do not cause events prenatally because of the favourable foetal physiology that includes (i) equivalent pressures in the main PA and aorta secondary to a non-restrictive patent ductus arteriosus and (ii) relatively equivalent oxygen concentrations due to parallel circulations. Prior to birth, therefore, there is normal myocardial perfusion and no stimulus for collateral vessel formation.⁶

After birth, the pulmonary vascular resistance drops in the vascular tree, and the pressure also begins to decrease. The ductus arteriosus closes, and flow from the anomalous coronary (left coronary artery, LCA/RCA) reverses, reducing the perfusion pressure of the ectopic artery. As a result, the left ventricular myocardium receives desaturated blood at low pressures.⁷

Generally, the ventricular ischaemia caused by ARCAPA is less prominent than that due to ALCAPA, because right ventricular oxygen demands are lower than that of the LV. Nevertheless, patients with a dominant RCA system do not tolerate the ARCAPA circulation compared with those with a left dominant coronary tree.³

An anomalous origin of the left and right coronary arteries from the pulmonary artery has two main clinical variants, reported in infants and older individuals. These are distinguished by the presence

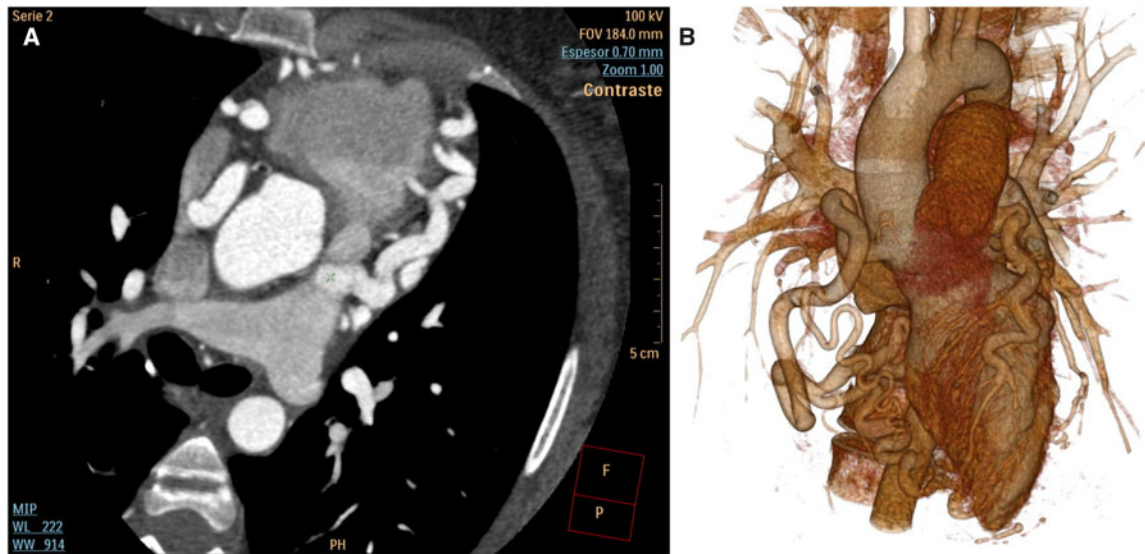


Figure 3 Anomalous origin of left coronary artery from pulmonary artery. (A) Coronary computed tomography angiography (CCTA) displaying the origin of the left main from the pulmonary artery trunk. (B) Three-dimensional reconstruction of CCTA displaying the origin of the right coronary artery from the aortic root and the origin of the left main stem from the pulmonary artery trunk.

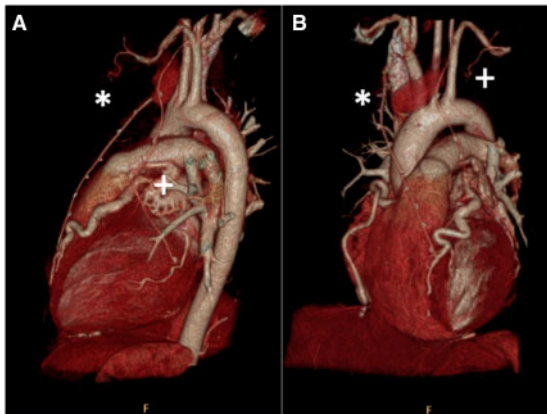


Figure 4 (A and B) Volume rendering images showing arterial bypass graft patency: right internal thoracic artery to left anterior descending artery (*) and left internal thoracic artery to obtuse marginal artery (+). Persistent dilation of the native coronary vessel tree.

or absence of adequate heterocoronary collateral circulation from the RCA. Up to 85% of patients without an adequate collateral supply will present within the first few months of life with congestive heart failure secondary to ischaemia. In these cases, the mortality rate is 90% without early diagnosis and surgical correction. Patients with adequate collateral circulation can remain asymptomatic until adolescence or adulthood. Frequently, the presentation of both ALCAPA and ARCAPA is with a murmur at the left parasternal border as incidental finding. This is usually related to both ischaemic left ventricular

dilatation and ischaemic dysfunction of the papillary muscles. Other presenting features may be angina on exertion, silent, or symptomatic myocardial infarction, left ventricular dysfunction, severe valvular disease, palpitations, or cyanosis. It is also known to be a cause of cardiac arrest or sudden death in adults.^{1,2}

Diagnostic tests

- Electrocardiogram is usually abnormal in patients with ALCAPA syndrome. In this condition, the classic finding is the presence of Q waves in leads I and aVF. Q waves can be found mainly in the anterior and lateral leads together with ST changes. Marked ECG abnormalities could also occur during exercise testing. For ARCAPA, left or right or biventricular enlargement or hypertrophy, ischaemic changes, bundle branch blocks, atrial fibrillation, bradycardia, and deep Q waves in the inferior leads have all been reported.^{4,8}
- Cardiac imaging: on the echocardiogram, ALCAPA patients present with dilatation of the proximal part of the RCA, retrograde flow from LCA into the PA and prominent septal flow. In the ARCAPA cases, usually a retrograde flow in the RCA with diastolic predominance is seen. Wall motion ventricular segmental abnormalities can be frequently delineated with transthoracic echocardiography. Transoesophageal echocardiography can delineate further the anomalous huge and tortuous coronary arteries and characterize their flow patterns. Other advanced imaging modalities such as computed tomography and cardiac magnetic resonance permit the unique visualization of coronary vascular anatomy and course, enhanced with the use of 3D reconstruction of the coronary arterial tree. Myocardial perfusion imaging may also be abnormal.
- Cardiac catheterization and selective coronary angiography may demonstrate the presence and extent of collaterals (Figures 1 and 2), quantify the left to right shunt and provide end-diastolic pressures.^{4,8}

The differential diagnosis for such dilatation of the coronary arteries includes Kawasaki disease, coronary artery–coronary sinus fistula, atherosclerosis-related coronary artery ectasia, vasculitis (polyarteritis nodosa or Takayasu arteritis), scleroderma, Ehlers–Danlos syndrome, hereditary haemorrhagic telangiectasia, and trauma.⁹

Management and prognosis

In all cases of anomalous origin of the coronary arteries from the PA, even in asymptomatic patients, it is recommendable to consider surgical correction to avoid a progressively increasing left to right shunt and poor coronary reserve which will predispose to ischaemia, arrhythmia, and cardiac arrest or sudden death. The aim of surgery is to perform the most physiological procedure to restore normal coronary flow in the anomalous artery, and this approach has shown good long-term results.

In adults, the reimplantation of the coronary artery is more difficult due to the friability and loss of elasticity of the vessel with age. In general, surgical correction has good overall outcomes. Reimplantation of the RCA or LCA to the aorta into the correct aortic sinus is regarded as the treatment of choice.^{1,2} An alternative option is to completely ligate the anomalous RCA or LCA and place a saphenous vein or an internal mammary artery graft.^{9,10} In ARCAPA or ALCAPA, the prognosis after surgery is favourable, the ischaemic yet viable myocardium will recover post-operatively after reperfusion preventing long-term myocardial ischaemia and fibrosis, and the left ventricular function improves substantially. In some cases, the mitral regurgitation also improves after surgery.

Lead author biography



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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 series including image(s) and associated text has been obtained from the patients in line with COPE guidance.

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