

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery: Late Pediatric Presentation of a Rare Disease

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare form of congenital heart disease that typically presents in infancy with signs of severe heart failure. We present 3 cases of ALCAPA that presented in late childhood or adolescence without significant cardiac symptomatology and evidence of minimal to no cardiac dysfunction. These 3 cases demonstrated significant coronary collateral vasculature, either by echocardiography, cardiac catheterization or direct surgical inspection. Earlier and more robust development of coronary collaterals, potentially in-utero, is the most likely reason for late presentation, absence of symptoms, and preserved cardiac function in this rare subset of patients. Given the high morbidity and mortality of this disease and potential for vague symptoms and a normal physical exam in late childhood or adolescence, an extremely high index of suspicion must be maintained to facilitate early diagnosis and treatment.

Keywords

anomalous coronary artery; congenital heart disease; heart failure; coronary collaterals

Abbreviations

LCA: Left Coronary Artery; ALCAPA: Anomalous Left Coronary Artery from the Pulmonary Artery; ECG: Electrocardiogram; EFE: Endocardial Fibroelastosis; LAD: Left Anterior Descending; MPA: Main Pulmonary Artery; RCA: Right Coronary Artery; MR: Mitral Regurgitation; AP: Aortopulmonary

Background

Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is a rare anomaly that commonly presents in infancy, which if untreated has up to 85% mortality, with most deaths occurring before 1 year of age from complications related to intractable heart failure [1]. Some patients remain asymptomatic likely secondary to an extensive network of collateral vessels between the right and left coronary system, with potential additional benefit of a restrictive opening between the origin of the LCA and pulmonary trunk [2]. The small minority of patients who survive to adulthood with ALCAPA will ultimately develop symptoms, or present with arrhythmia, syncope or sudden cardiac death [3]. We present the clinical symptoms and imaging findings of 3 patients with ALCAPA diagnosed in late childhood or adolescence. In addition, we searched our echocardiographic database (1994 – 2015) identifying 20 additional patients all with the typical presentation in infancy (under 2 years of

age). With review of the presentation of these 3 patients, we hoped to identify unifying diagnostic features in patients with a later presentation and less severe clinical course.

Case Series

Case 1

A 14 year old previously healthy female presented to our outpatient cardiology clinic for evaluation after a syncopal event during a prolonged indoor soccer game. Her physical exam and electrocardiogram (ECG) were normal. Echocardiogram showed normal cardiac chamber size and normal biventricular function without mitral or tricuspid regurgitation. There was no evidence of endocardial fibroelastosis (EFE), myocardial injury, or regional wall motion abnormalities. Antegrade flow in the LCA could not be proven. The left anterior descending (LAD) and circumflex coronary arteries were dilated with the appearance of reversal of flow. Color flow Doppler in the main pulmonary artery (MPA) showed that the LCA arose from the pulmonary artery trunk (Figure 1a). The right coronary artery (RCA) was significantly dilated (Figure 1b) with increased flow in the posterior descending artery and along the posterior wall and interventricular septum. Evidence of significant collateral vasculature was noted on the 4-chamber view with Doppler interrogation (Figure 1c and 1d).

Cardiac catheterization demonstrated a dilated RCA arising from the aorta with multiple large inter-arterial coronary collaterals. The LAD and circumflex coronary arteries filled retrograde from the coronary collaterals with retrograde flow seen into the MPA. Both the LAD and circumflex coronary arteries were significantly dilated and tortuous. The left ventricle had normal systolic and diastolic function. She underwent repair with an uncomplicated post-operative course and was discharged home on postoperative day #4 on no cardiac medications. At her 2 month outpatient visit, exercise stress testing revealed no ischemia or arrhythmias. She has resumed normal physical activity.

Case 2

A 12 year old previously healthy female presented to the outpatient cardiology clinic for evaluation of fatigue, murmur and decreased physical endurance. She denied any history of chest pain, syncope, or dizziness at rest or related to exercise. Her physical examination revealed a grade 1 systolic ejection murmur heard at the left lower sternal border with radiation to the apex. ECG showed normal sinus rhythm, left axis deviation, voltage criteria for left ventricular hypertrophy (LVH), and Q-waves in leads I and AVL.

Echocardiogram demonstrated a dilated LCA arising from the pulmonary artery. There was collateral flow noted from a dilated RCA to the left coronary artery system. The left ventricle was mildly dilated with preserved systolic and diastolic function. There was mild EFE of the left ventricle papillary muscle without any mitral regurgitation (MR). A cardiac CT confirmed the echocardiographic findings. She underwent uneventful repair and was noted to have large coronary collaterals on intra-operative inspection. She was discharged home on postoperative day #4 on furosemide and aspirin. At her 1 week follow-up visit, she was asymptomatic with echocardiographic evidence of normal left ventricular function.

Case 3

A 3 year old female presented to her pediatrician in the setting of an asthma exacerbation and

was found to have a murmur. There was no past history of limited exercise, syncope, or chest pain. Her physical exam revealed a grade 2 systolic murmur best heard at the apex. ECG showed normal sinus rhythm, LVH, and evidence of non-specific ST segment and T wave changes.

Echocardiogram showed an anomalous origin of the LCA from the pulmonary artery with multiple coronary collaterals. She had moderate mitral regurgitation (MR), moderate left ventricular dilation and left atrial enlargement. She had preserved left ventricular systolic function without any evidence of wall-motion abnormalities or EFE. Cardiac catheterization demonstrated a dilated RCA, delayed filling of the left coronary artery system and diffuse coronary collaterals (Figure 2). There was moderate MR and normal left ventricular systolic and diastolic function. She underwent repair with mitral valve commissural annuloplasty. She had an uneventful post-operative course and was discharged home on a diuretic and beta-blocker. At her most recent clinic visit, approximately 12 years after repair, she had mild MR and normal left ventricular systolic and diastolic function.

Discussion

The earliest reports of “anomalous arteries” arising from the pulmonary artery are from autopsy accounts described in 1865 [4] and 1885 [5]. In 1906 the details of the anatomy of ALCAPA was defined in a 2 day old infant, and subsequently in 1911 in an autopsy of a 5-month-old child with ALCAPA and a left ventricular aneurysm [6]. In 1933, the landmark paper of Bland, Garland, and White was the first to correlate a clinical syndrome with autopsy findings of ALCAPA [7].

Patients with ALCAPA are usually asymptomatic at birth, however within the first month of life, the pulmonary vascular resistance falls, leading to decreased antegrade flow from the pulmonary artery to the anomalous left coronary. This results in progressive coronary insufficiency and ischemia leading to left ventricular dilation and dysfunction. Patients with ALCAPA usually present in early infancy with failure to thrive, irritability with feeding likely due to “exercise induced” angina, and symptoms of heart failure, including respiratory distress, diaphoresis and fatigue with feeding [7]. Early reports have quoted an incidence ranging from 0.01 to 0.02% with 72% found in early infancy [8]. A more recent case series reported that 84% of their cases presented under 1 year of age [9]. This is similar to our rate of 83% (19/23) presenting under 1 year of age with 1 additional patient presenting at 16 months with significant symptoms and severely decreased left ventricular function and EFE.

Patients with ALCAPA who survive beyond infancy have evidence of collateral vessels on direct inspection or on cardiac imaging with angiography or CT scan. The early formation of extensive inter-arterial coronary collateral vessels is believed to allow for blood flow with a higher perfusion pressure into the left coronary artery system. Unfortunately, since this flow is retrograde and at still often at a lower perfusion pressure compared with normal coronaries, these patients often develop a state of chronic hypo-perfusion in a left coronary artery distribution pattern. Additional left to right shunting through the collateral system imposes a further volume overload to the ischemic left ventricle potentially leading to left ventricular dilation, dysfunction, and MR in adulthood.

The timing and extent of collateral development may affect the clinical presentation and age at diagnosis in patients with ALCAPA. Our 3 patients had either minimal or no evidence of myocardial injury, though the youngest patient had moderate MR with left heart enlargement which resolved after coronary reimplantation. The current body of literature has described late ALCAPA presentation and the possible

effect of coronary collateral vasculature, but the patients often had either significant symptoms or evidence of ventricular dysfunction [9-11]. Additionally, the literature does not provide direct comparison of collateral burden between infant and late presentation of ALCAPA either by catheterization or other imaging modalities. Both adolescent patients (Case 1 and Case 2) had intra-operative evidence of significant coronary collateral vasculature. In addition, the angiogram performed in Case 3 (Figure 2) demonstrated a significant collateral network supplying the left ventricle. We believe that the collaterals documented in this patient are more numerous and robust compared to the “typical” appearance of the collateral vessels seen in infants presenting with the disease (see Figure 3 as a representative case). This lends more credence to the hypothesis that earlier and more pronounced collateral vasculature is responsible for the absence of symptoms and preservation of left ventricular function. Furthermore, we speculate that the development of coronary collaterals in those surviving to late childhood or adolescence may actually begin in utero. Further study including assessment of the fetal heart and the coronary circulation in the diagnosis of ALCAPA will be instrumental in proving this hypothesis.

The most common types of surgical repair for ALCAPA are direct implantation and the Takeuchi repair. Direct implantation involves the creation of aortic and main pulmonary artery flaps with transfer of the left main coronary artery, while the Takeuchi repair is performed by the creation of an aorto-pulmonary window and intra-pulmonary tunnel. The Takeuchi repair is preferred if the LCA arises from the non-facing sinus of the pulmonary artery or too distant from the aortic root [12]. Hoashi et al followed 19 patients with repaired ALCAPA post-operatively and showed increased survival in the Takeuchi group (mean follow-up 7.4 years) compared with the translocation group (mean follow-up 9.3 years), but increased re-intervention rate. The most common reason for re-intervention was Takeuchi baffle leak or stenosis and supra-valvar pulmonary stenosis in the translocation group [12]. The direct implantation procedure is preferred at our institution, though Case 2 did undergo a Takeuchi repair.

Our patients suggest that in this age group, ECG and physical examination findings that are often diagnostic, may be equivocal. Two of our patients presented with a murmur, one with additional symptoms of fatigue, and the other during an asthma exacerbation. Others have reported that a heart murmur is a common presentation of ALCAPA after infancy [9-13]. In a large review, 66% of adult patients with ALCAPA had symptoms of angina, dyspnea, palpitations or fatigue. Seventeen percent presented with arrhythmia, syncope or sudden cardiac death and only 14% were asymptomatic. Two of our patients had abnormal Q waves in leads I and avL, suggestive of myocardial injury, however of note is that one had a normal ECG. This suggests that a normal ECG does not completely rule-out the presence of ALCAPA.

Conclusion

ALCAPA is a rare form of congenital heart disease that can lead to significant morbidity and mortality. Amongst the subset of patients with ALCAPA, there are individuals who survive beyond infancy. This less severe phenotype most likely occurs secondary to earlier, more extensive and more robust formation of collateral coronary vasculature that allows for retrograde perfusion of the left coronary artery system and minimal left ventricular injury. These patients can present with vague symptoms and sometimes have a normal physical examination and ECG. Older age at presentation,

particularly in adulthood, increases the risk of morbidity. Due to the high mortality of this disorder, a high clinical suspicion for ALCAPA particularly out of the typical presentation window in infancy must be maintained in order to facilitate rapid diagnosis and definitive surgical repair.

Figures

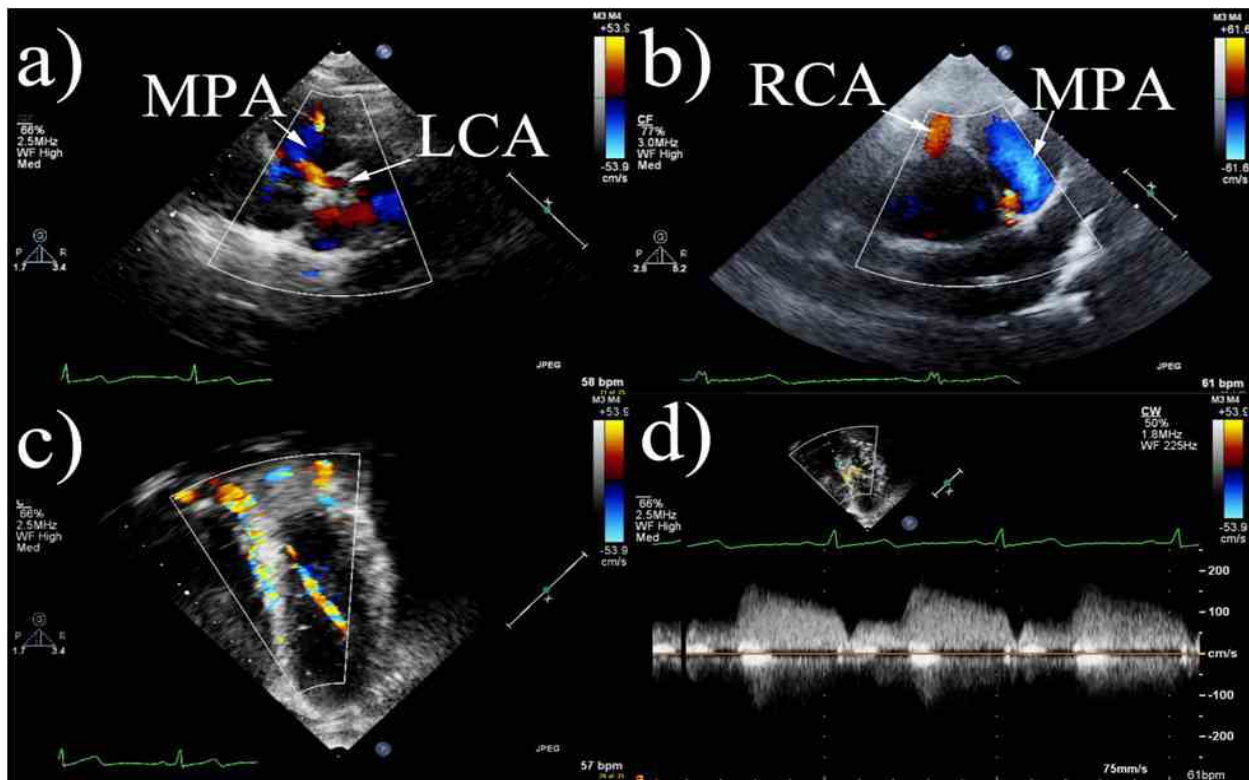


Figure 1: Echocardiographic demonstration of ALCAPA: echocardiographic imaging **a)** modified parasternal short axis view demonstrating retrograde left coronary artery (LCA) flow into the main pulmonary artery (MPA) **b)** parasternal short axis view with increased blood flow into a dilated right coronary artery (RCA) **c)** multiple coronary artery collateral vessels along the posterior aspect of the heart in the apical 4 chamber view **d)** Doppler flow pattern of the posterior coronary collaterals.

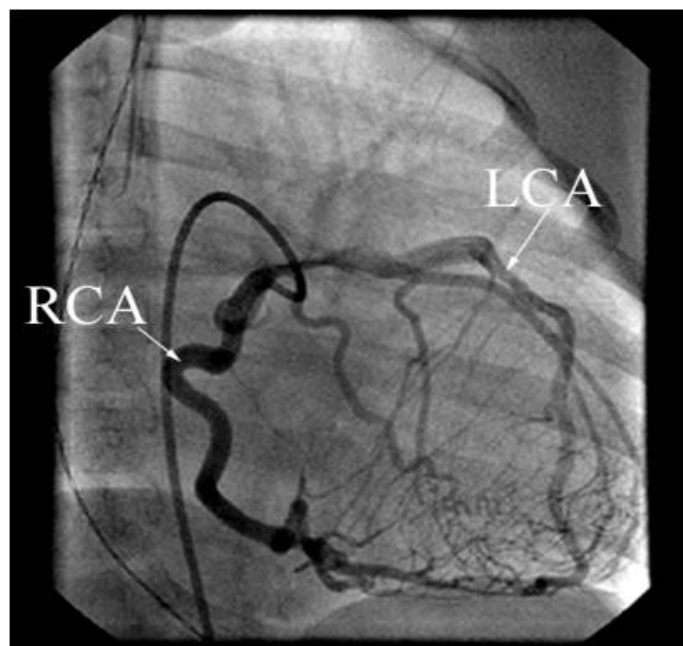


Figure 2: Cardiac catheterization in an adolescent patient (Case 3) with ALCAPA: right coronary artery angiogram in the right anterior oblique projection, demonstrating a dilated right coronary artery with a fine inter-coronary collateral network, with retrograde filling of the left anterior descending, circumflex, and left coronary artery. The left coronary artery fills the main pulmonary artery in a retrograde fashion.

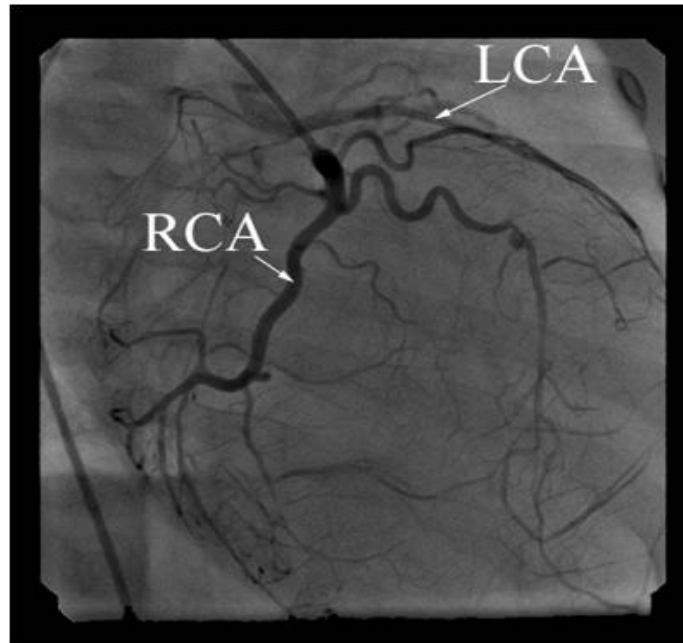


Figure 3: Cardiac catheterization in a typical infant presentation of ALCAPA: right coronary artery angiogram in the right anterior oblique projection, performed on a patient presenting with heart failure in early infancy demonstrating a less prominent inter-coronary network when compared to Figure 3. Again the left coronary artery is seen entering the main pulmonary artery in a retrograde fashion.

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