

Preserved Left Ventricular Function in Two Infants with Anomalous Left Coronary Artery from the Pulmonary Artery

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Abstract

Anomalous left coronary artery from the pulmonary artery is a rare congenital anomaly that usually presents early in infancy. Typical findings include left ventricular dysfunction and mitral insufficiency. Here we present two cases of infants with ALCAPA with preserved LV function. Their cases illustrate the importance of recognizing the heterogeneity of presentation of ALCAPA in order to facilitate timely diagnosis and repair, and thereby improve associated morbidity and mortality.

Keywords

Anomalous left coronary artery from the pulmonary artery; preserved left ventricular function

Background

Anomalous left coronary artery from the pulmonary artery [ALCAPA] is a rare congenital anomaly that occurs in approximately 1 out of 300,000 births and represents 0.25% of all cardiac congenital abnormalities [1,2].

Typical presentation occurs early in infancy when the pulmonary vascular resistance decreases and flow reverses in the left coronary artery [LCA]. This causes myocardial ischemia and leads to the development of left ventricular systolic dysfunction and mitral insufficiency [1]. Without surgical coronary re-implantation the mortality rate is as high as 90% in the first year of life [2,3]. In spite of this, 10-15% of patients present as adults and are either asymptomatic, have symptoms of ischemia, heart failure or arrhythmias, or sometimes sudden death [4]. In contrast to infants, adult patients typically have good collateralization from the right coronary artery [RCA] to the left coronary circulation, and preserved left ventricular [LV] function [5]. The two cases presented here are uncommon in that they are infants with ALCAPA and preserved LV function. Here we discuss their physiology and diagnostic challenges.

Case 1

A three-week-old previously healthy infant was referred to our outpatient pediatric cardiology clinic for evaluation of a new murmur. Her cardiac exam was notable for a II/VI holosystolic murmur at

the apex with radiation to the axilla and hepatomegaly. An electrocardiogram was obtained that demonstrated non-specific T-wave abnormalities laterally with ST-segment depressions anteriorly and Q-waves in aVL (Fig 1A). An echocardiogram revealed severe mitral regurgitation, moderate tricuspid regurgitation predicting half-systemic right ventricular pressures, normal LV function, and prominence of the anterolateral LV papillary muscle. Importantly, both coronary arteries appeared to arise from the appropriate sinuses by 2D- and color-flow imaging (Fig. 1B). Due to concern for an anomalous left coronary, she underwent diagnostic catheterization, which revealed a dilated RCA and a late-filling left coronary arterial system with retrograde flow of contrast draining into the pulmonary artery (Fig. 1C), consistent with ALCAPA and coronary steal syndrome. On retrospective review of echocardiographic imaging, there was evidence of the LCA arising from the main pulmonary artery with color flow. It would, however, have been difficult to determine by echocardiography the LCA origin without views other than the one seen in Figure 1B. She subsequently underwent surgical coronary artery re-implantation. Her course was uncomplicated and she was discharged home postoperative day 18 with moderate mitral insufficiency, moderate left atrial and LV dilation and good systolic function.

Case 2

A five-month old girl presented to our PICU in severe respiratory distress with cardiomegaly. She had been healthy until about three months of age, at which time she was treated for pneumonia. She never returned to baseline and remained irritable with poor feeding and weight loss. In the days prior to admission, her feeding worsened and she became increasingly inconsolable. On presentation, she was cachectic, and in cardiorespiratory failure. She had a hyperdynamic precordium, a gallop rhythm, and a III/VI holosystolic murmur heard best at the apex. An electrocardiogram showed Q-waves in leads I and V6 and signs of LVH (Fig. 2A). An echocardiogram demonstrated a severely dilated left atrium, severe mitral regurgitation, moderate tricuspid regurgitation estimating at least two-thirds systemic right ventricular pressure, an echo-bright left-ventricular papillary muscle, and preserved LV function. Coronary evaluation suggested anomalous coronary artery origin off the pulmonary artery with antegrade flow (Fig. 2B and 2C). This finding was confirmed by a diagnostic cardiac catheterization (Fig. 2D). She underwent coronary artery re-implantation and mitral valve annuloplasty. She had significant residual mitral insufficiency and underwent repeat surgery for mitral valve plasty. She was discharged home 3 months following initial repair.

Discussion

The typical presentation of infants with ALCAPA are symptoms of ischemia and heart failure and echocardiographic evidence of mitral insufficiency, depressed left ventricular systolic function, RCA dilation, and often endocardial fibroelastosis. The ALCAPA itself is sometimes difficult to visualize, and therefore, cardiac catheterization is often performed to confirm the diagnosis [6]. The two cases presented here are uncommon in that they both had preserved systolic function.

Case 1 presented as an asymptomatic neonate for evaluation of a murmur. She already had EKG changes consistent with ALCAPA, and even though the echocardiogram was not able to make the diagnosis, a cardiac catheterization demonstrated retrograde flow from the LCA into the pulmonary artery. Because ALCAPA was diagnosed and treated promptly, further myocardial ischemia and myocardial dysfunction were prevented. Nine months following the surgical repair, the patient continues

to grow and develop normally with moderate residual mitral insufficiency and normal LV systolic and diastolic function on echo.

Case 2 presented as a five month-old in cardiorespiratory failure. She also had EKG changes suggestive of ALCAPA. Her echocardiogram showed ALCAPA with antegrade perfusion through the pulmonary artery and severe mitral insufficiency with preserved LV systolic function. The diagnosis was confirmed by cardiac catheterization. Preserved LV systolic function is unusual for ALCAPA at this age, but has been described in a few cases who had elevated pulmonary arterial pressure with and without associated congenital heart disease [5,7,8]. In case 2, the diagnosis was delayed. She had been sick for many months and presumably had a lower pulmonary vascular resistance and coronary steal from the LCA into the pulmonary artery prior to presentation. This assumption is founded on echocardiographic signs of subendocardial and mitral papillary muscle ischemia, mitral annular and left ventricular dilation, and severe mitral insufficiency. In adult patients with left heart disease, reactive pre-capillary pulmonary hypertension caused by pulmonary arterial vasoconstriction and vascular remodeling is a well-known, but relatively poorly understood phenomenon[9]. In case 2, this mechanism leads to an increase in pulmonary arterial pressure and reestablishment of antegrade LCA blood flow with systemic venous blood. At presentation, LV function was echocardiographically preserved, but the high mitral regurgitant fraction resulted in diminished cardiac output. We speculate that the patient would have presented earlier and with worse systolic function if reactive pulmonary hypertension had not developed.

Conclusion

In conclusion, these two cases illustrate that clinical presentation and physiology of ALCAPA vary widely. Both infants had ECG changes and significant mitral insufficiency with preserved function. Even with echo imaging, catheterization was necessary to make a definitive diagnosis. Furthermore, the persistence of pulmonary hypertension or development of reactive pulmonary hypertension may be a contributor to preserving LV function. It is important to appreciate this heterogeneity in order to facilitate timely diagnosis and repair, and improve associated morbidity and mortality.

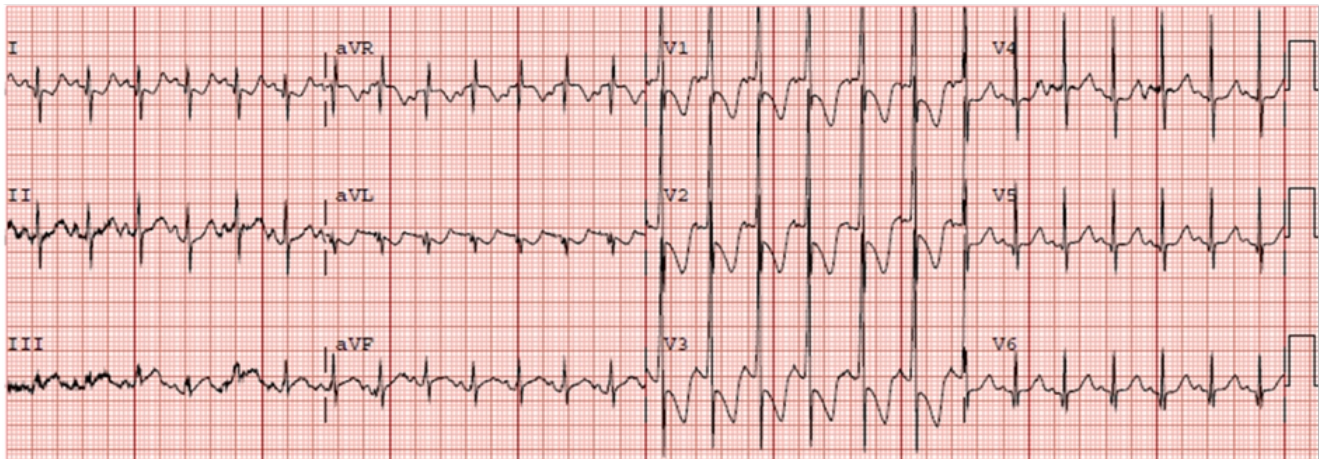
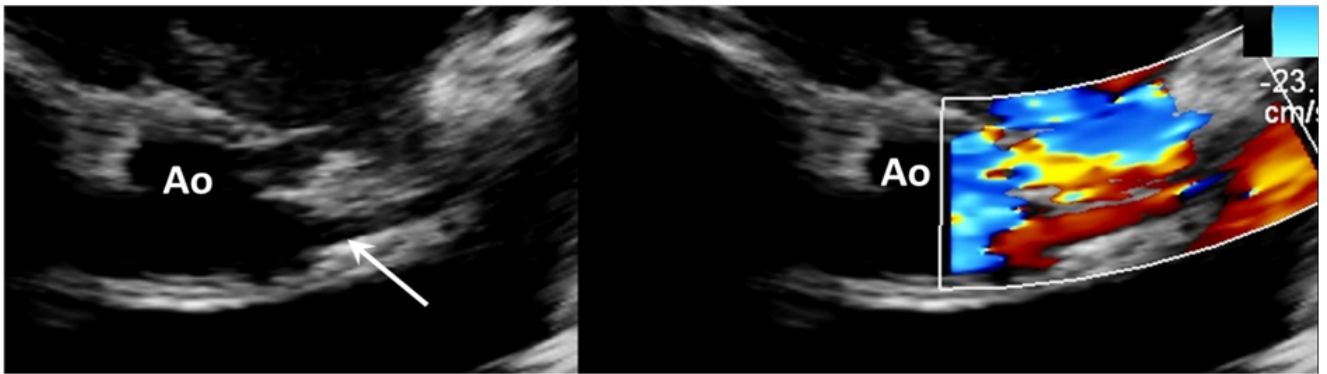
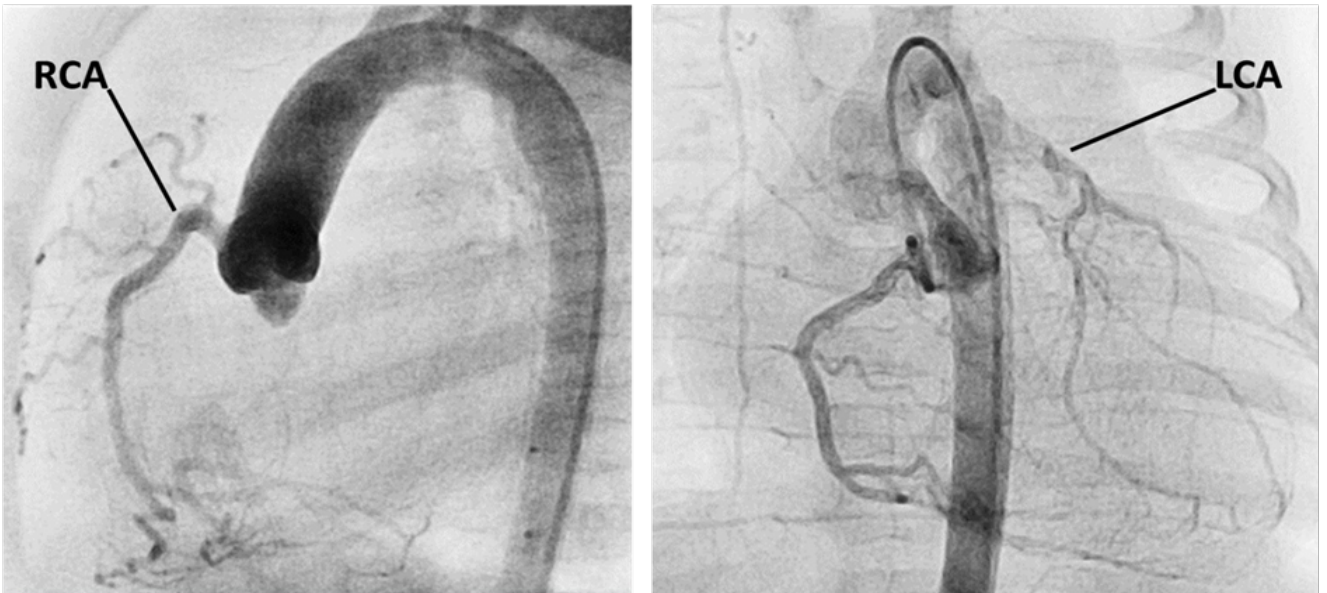
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Figure 1: Diagnostic evaluation of ALCAPA in Case 1.

A. Electrocardiogram demonstrating Q-waves in aVL, non-specific T-wave abnormalities, and ST-segment depression.

B. 2D and color-flow echocardiography in parasternal short axis demonstrating apparent origin of the left coronary [arrow] with antegrade flow from the appropriate sinus of the aortic root [Ao].

C. Cardiac catheterization and aortography demonstrating a dilated right coronary artery [RCA] with late filling of the left coronary artery [LCA] which inserts anterior and superior to the aorta with retrograde flow.

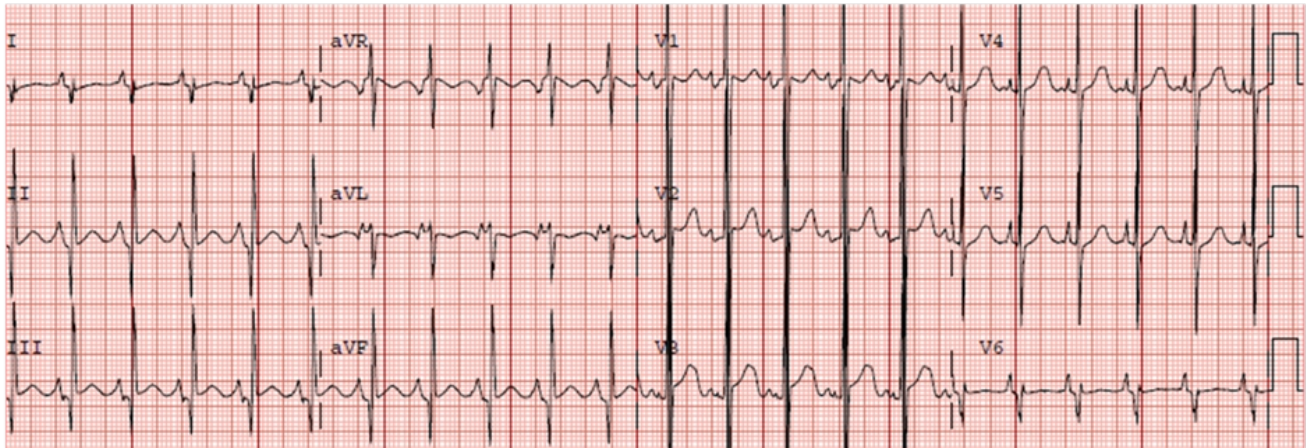
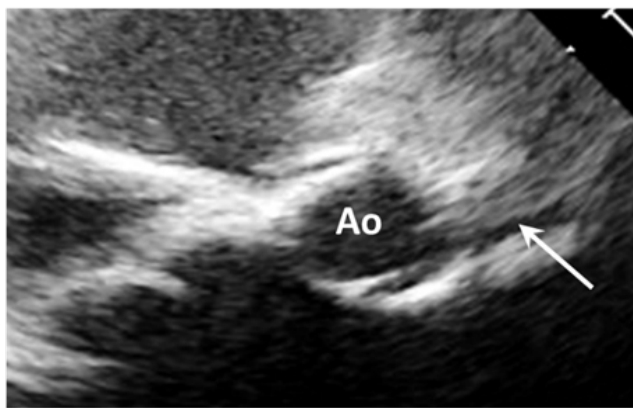
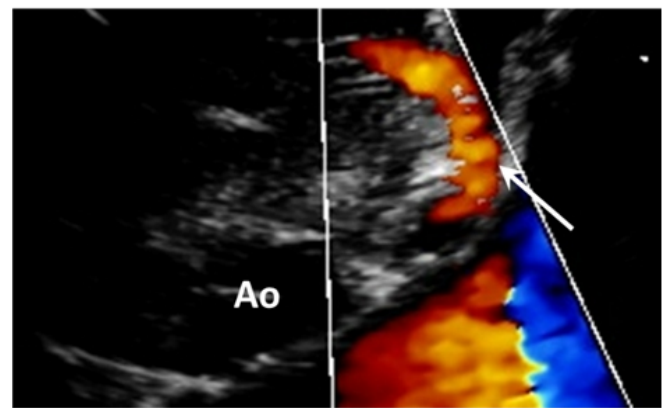
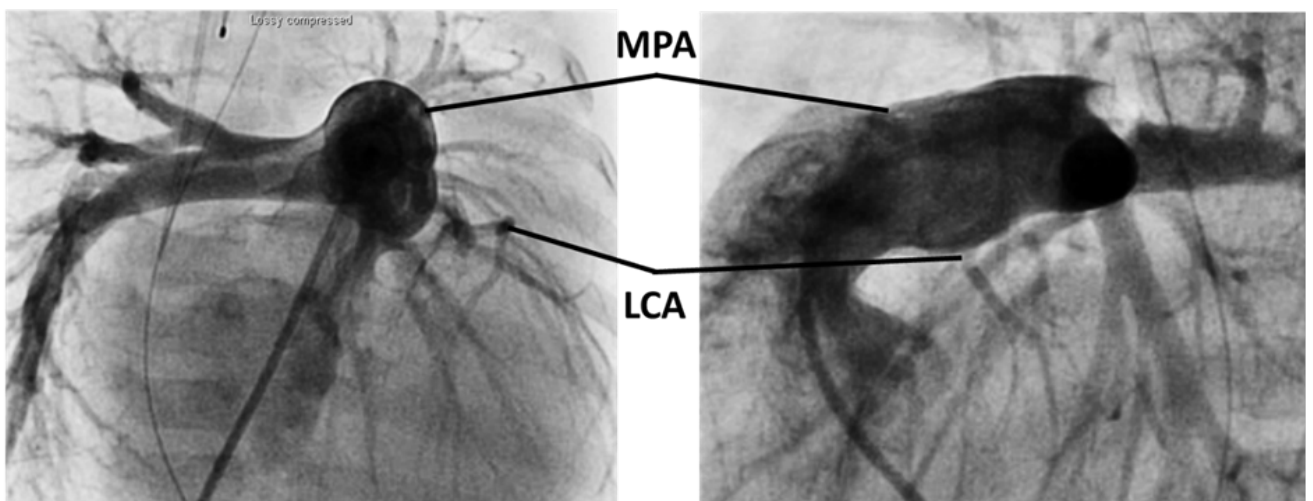
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Figure 2: Diagnostic evaluation of ALCAPA in Case 2.

- A. Electrocardiogram demonstrating Q-waves in lead I and evidence of left atrial enlargement
 B. 2D echocardiogram in parasternal short axis seeming to demonstrate normal origin of the LCA from the aortic root [Ao].
 C. Color-flow echocardiography that seemed to demonstrate antegrade LCA flow from the main pulmonary artery [MPA].
 D. Cardiac catheterization and pulmonary angiography demonstrating ALCAPA with antegrade pulmonary blood flow into the LCA.

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