Anomalous Left Coronary Artery Arising from the Pulmonary Artery (ALCAPA): The Critically Important Role of Color Flow Doppler in Identifying a Rare Intramural Course

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INTRODUCTION

Anomalous left coronary artery (LCA) from the pulmonary artery (ALCAPA) is a rare congenital coronary anomaly that can present with congestive heart failure and ischemic mitral regurgitation in an infant.[1] Coronary imaging by echocardiography in the pediatric population is difficult due to small vessel size, high heart rate, and patient cooperation.[2] This case presents an additional challenge in diagnosis due to a rare variant of ALCAPA with an intramural aortic course that leads to the false impression of normal left coronary origin.[3,4]

CASE PRESENTATION

A 4-month-old ex-full-term male patient presented at 1 month of age with history of progressive labored breathing. On examination the patient was tachypneic, with a respiratory rate of 80 breaths per minute. A low-pitched, holosystolic murmur was heard best at the apex, consistent with mitral regurgitation. Electrocardiogram showed deep Q waves in V5/V6 and ST depressions in the lateral leads (Figure 1). Troponin was normal, and B-type natriuretic peptide was elevated (11,800 pg/mL). Transthoracic echocardiogram showed a severely dilated left ventricle (Z score +7.7; volumes measured by 5/6 area-length product) with moderate systolic dysfunction and ejection fraction of 42% (age adjusted Z score = −4.1). No regional wall motion abnormalities were appreciated. There was severe mitral regurgitation due to both papillary muscle dysfunction and annular dilation. The posteromedial papillary muscle appeared hyperechoic, with restrictive leaflet motion and resultant prolapse of the anterior leaflet, consistent with secondary mitral valve pathology (Video 1). Although the left coronary origin appeared to connect with the aortic root, there was retrograde flow in the LCA, raising concerns for possible ALCAPA (Video 2). The contiguous course of the LCA with the aortic wall made the absolute diagnosis of ALCAPA difficult.

Because of ambiguity on the echocardiogram, the differential diagnosis included a primary congenital mitral valve anomaly, or secondary mitral regurgitation due to ischemia/infarction of the papillary muscles or familial or metabolic causes of dilated cardiomyopathy.

Due to concerns for ALCAPA, a catheterization was undertaken, and a pulmonary artery angiogram demonstrated filling of the LCA (Figure 2A and 2B). The diagnosis of ALCAPA was confirmed, and the patient was transferred to the regional pediatric cardiac surgical referral center for emergent surgery.

Precardiopulmonary bypass transesophageal echo from the midesophageal view demonstrated a section of the LCA immediately adjacent to the aortic wall, giving the two-dimensional (2D) appearance of a normal vessel origin. However, the direction of diastolic retrograde flow *toward* the aortic root, a characteristic echocardiographic finding in ALCAPA, was seen (Video 3, Figure 3). Most notably, simultaneous antegrade blue flow could be demonstrated in the right coronary artery.

Upon intraoperative inspection, the midsection of the coronary artery was intramural and adherent to the aorta in a lengthwise orientation. Given the intramural aortic component, careful dissection into the aortic media was undertaken to better mobilize the coronary artery and then reimplant the ostium onto the aorta as a button (Video 4). Intraoperative inspection and fluid filling of the left ventricle confirmed anterior leaflet prolapse, restricted motion of the posterior leaflet, and immobile retracted papillary muscles, confirming the echocardiographic hypothesis of secondary mitral regurgitation. Due to concerns for postoperative mitral regurgitation, a posterior annuloplasty was performed and the anterior leaflet was supported with artificial chords, with improved valve coaptation after these interventions.

Due to recurrent severe mitral regurgitation and inability to progress off ventilatory support, the patient returned to the operating room on postoperative day 15. Intraoperative inspection was notable for further contraction of the papillary muscles, requiring mitral valve annuloplasty and placement of additional artificial chords. He was ultimately discharged home on postoperative day 55. On most recent follow-up, 2 months after initial repair, left ventricular dilation had resolved and systolic function was normal, with an ejection fraction of 60% (Video 5). In addition, trivial mitral stenosis was present, with a mean gradient of 2-3 mm Hg, as well as qualitatively moderate regurgitation.

DISCUSSION

This case of ALCAPA with intramural aortic course exemplifies the ambiguity in echocardiographic diagnosis. Barbero-Marcial *et al*[3]

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Keywords: Congenital heart disease, Coronary vessel anatomy, Myocardial ischemia, Cardiomyopathy, Echocardiography

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

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2468-6441

https://doi.org/10.1016/j.case.2022.02.001

VIDEO HIGHLIGHTS

Video 1: Sequence of mitral valve images in the parasternal long-axis and apical four-chamber views using color Doppler and 2D/color compare. The mitral valve regurgitation is posteriorly and laterally displaced, consistent with anterior leaflet prolapse. The normal leaflet appearance and the hyperechoic papillary muscles, restrictive motion of the posterior leaflet, and relative prolapse of the anterior leaflet suggest secondary disease related to posterior leaflet ischemia/infarction. Annular dilation secondary to severe left ventricular dilation further contributes to the severity of mitral regurgitation.

Video 2: Transthoracic echocardiogram in a parasternal short-axis view at the level of the aortic root and coronary origins There is subtle retrograde flow within the LCA despite the normal 2D appearance. *Ao*, Aortic root.

Video 3: Transesophageal echocardiogram from the mid-esophageal short-axis view with inverted image at the level of the coronary origins. The opposing flow directions when simultaneously comparing the right coronary artery (*blue*/antegrade and LCA (*red*/retrograde) is readily apparent. *Ao*, Aortic root; *RCA*, right coronary artery.

Video 4: Intraoperative video of surgical repair of ALCAPA. The pulmonary artery is divided to expose the coronary ostium, which is then excised as a button. The intramural left coronary segment runs lengthwise along the ascending aortic wall, which is dissected out and then reimplanted.

Video 5: Preoperative transthoracic echocardiogram of the apical four-chamber view compared with 2-month postoperative study. Note the normalization of left ventricular size and systolic function in the postoperative study. The hyperechoic papillary muscles in the preoperative study are replaced by artificial chords in the postoperative study.

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reported a misdiagnosis of this lesion due to echocardiographic dropout of the thin wall between the coronary and the aorta leading to a falsely normal-appearing ostium on 2D echocardiography.[3] We have captured for video presentation the importance of color Doppler imaging for identifying the abnormal *direction* of coronary flow in ALCAPA with an intramural course.

Anomalous LCA from the pulmonary artery occurs in an estimated one in 300,000 live births. The addition of an intramural aortic course is exceedingly rare, with the literature limited to case reports.[5-8] One center encountered only one case with intramural aortic course out of 42 total ALCAPA repairs over a 35-year experience.[9] Even when ALCAPA was detected on preoperative imaging, failure to recognize the intramural aortic course has led to accidental injury of the LCA.[4]

Due to the intrinsic challenges with diagnosis by noninvasive imaging, it is important to review the physiologic changes that drive the clinical presentation of ALCAPA, along with its associated echocardiographic findings. This will distinguish it from other causes of dilated cardiomyopathy or a congenital mitral valve anomaly. Anomalous LCA from the pulmonary artery most often presents at 2-4 months of age with symptoms of congestive heart failure and poor growth. The timing of presentation is critical, because the physiologic high pulmonary vascular resistance in the fetus and neonate will initially support adequate coronary blood flow. As pulmonary vascular resistance drops in early infancy, a steal phenomenon develops with retrograde flow from the LCA into the pulmonary artery. This coronary blood flow reversal may culminate in myocardial ischemia and possible infarct to the left ventricular myocardium and/or papillary muscles.[1] Mitral valve dysfunction is a consequence of ALCAPA due to ischemic injury to the supporting apparatus and ischemia or infarction of the papillary muscles, often further worsened by left ventricular dilation and dysfunction. This will also result in annular dilation again contributing to anterior leaflet prolapse. On echocardiography the leaflets appear normal; however, the papillary muscles are hyperechoic with tethering and restricted motion of the posterior leaflet with a posteriorly and laterally displaced regurgitant jet, resembling the mechanism of secondary mitral regurgitation following myocardial ischemia/infarction. This appearance of the mitral valve alone should prompt the echocardiographer to pursue a very thorough coronary evaluation.

The widespread usage of color Doppler imaging has become the standard of care in diagnosis of ALCAPA, as reversal of flow in the LCA and egress to the pulmonary artery are diagnostic.[10] In most cases of ALCAPA, the left main coronary artery arises from the left inferolateral aspect of the main pulmonary artery just above the pulmonary valve (Figure 4A).[5] Sometimes the LCA origin from the pulmonary artery cannot be determined, as in this case, because the ALCAPA origin was quite distal at the main pulmonary artery/right pulmonary artery junction (Figure 4B). The proximal LCA then coursed along and was contiguous with the aortic wall in a superior/inferior orientation (directly visualized in Video 3), appearing as if it were in the usual position when viewing the aortic root in cross section.

In general, imaging of the coronary arteries in infants can be challenging due to high heart rate, patient movement, and small vessel size and sometimes requires sedation.[11] On transthoracic imaging, the coronaries are visualized in the parasternal short-axis plane at the level of the aortic root (Video 2). A low Nyquist limit will enhance the low-velocity color Doppler flow in the coronaries, and a narrow sector angle will preserve a higher frame rate to detect subtle directional changes in flow.

CONCLUSION

The 2D echo findings of a contiguous position of the left coronary origin with the aortic wall but associated with abnormal color flow direction should raise suspicion for ALCAPA with intramural aortic course. Knowledge of this rare entity is essential for correct diagnosis, which can allow for prompt lifesaving surgery in the newborn period.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2022.02.001.

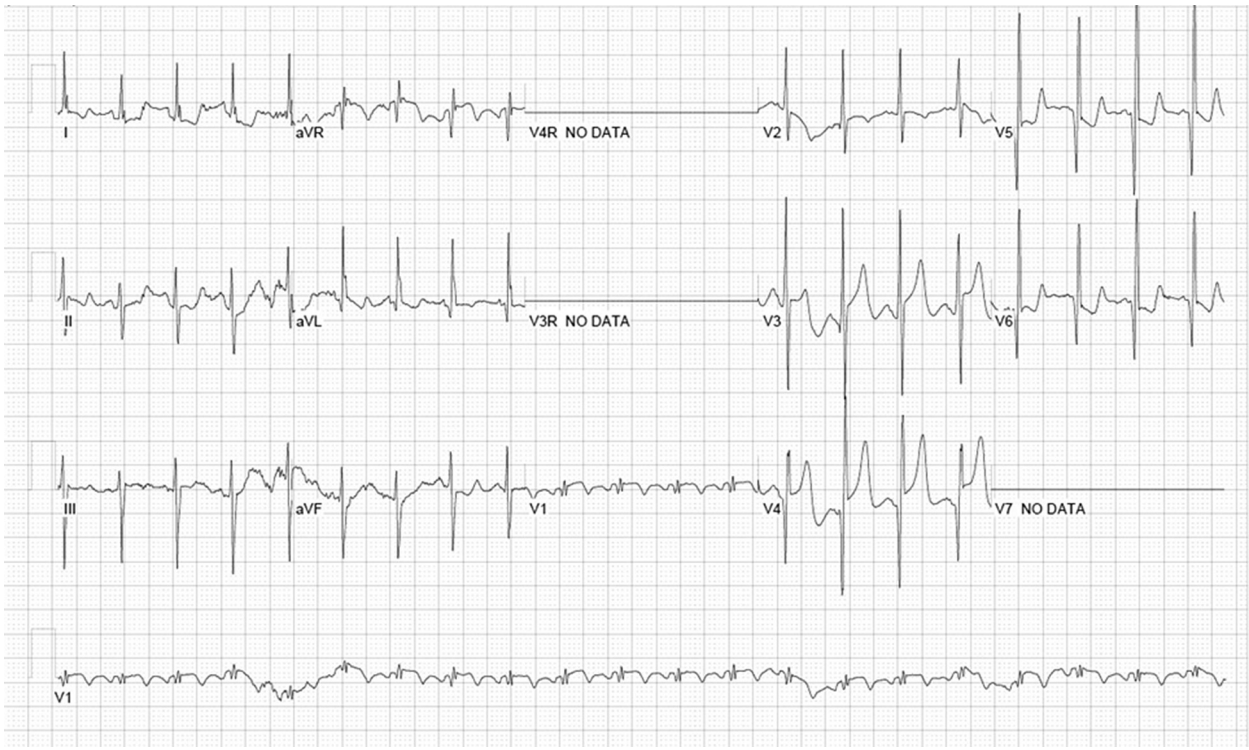

Figure 1 Electrocardiogram on initial presentation showing deep Q waves in V5/V6 and ST depressions in the lateral leads.

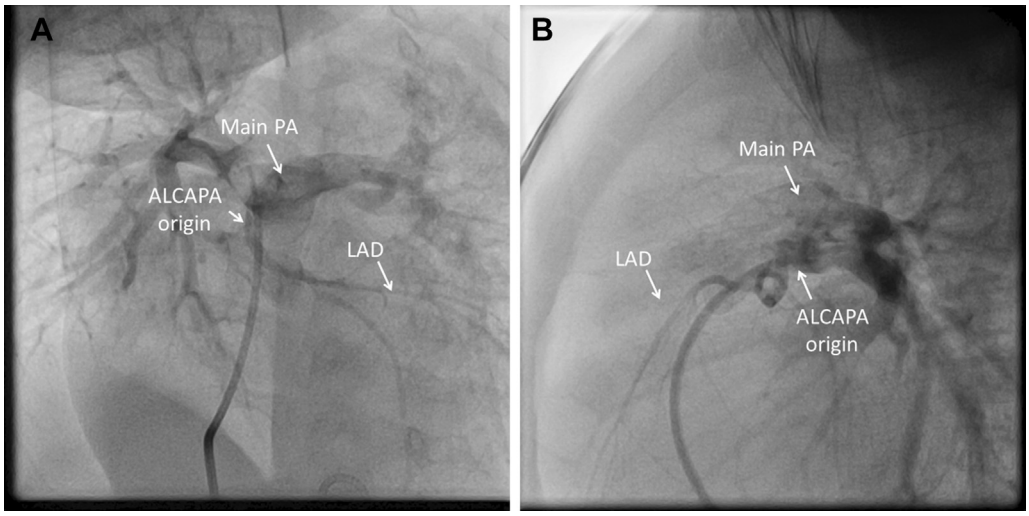

Figure 2 Pulmonary artery contrast injection captured on fluoroscopy from the anteroposterior **(A)** and lateral **(B)** projections demonstrating filling of the left coronary system. The origin of the LCA is near the main pulmonary artery and right pulmonary artery junction. Due to a steal phenomenon, flow in the LCA is primarily retrograde into the pulmonary artery. *LAD*, Left anterior descending; *PA*, pulmonary artery.

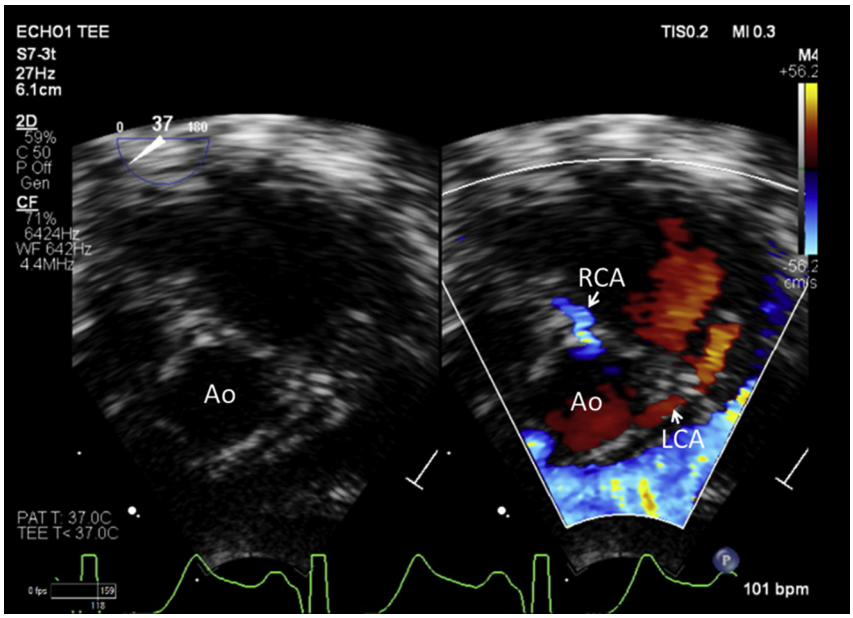

Figure 3 Two-dimensional and color Doppler still image of ALCAPA with intramural course on transesophageal echocardiogram from the midesophageal view. Antegrade (*blue*) flow in the RCA can be seen at the same time as retrograde (*red*) flow in the LCA. *Ao*, Aortic root; *RCA*, right coronary artery.

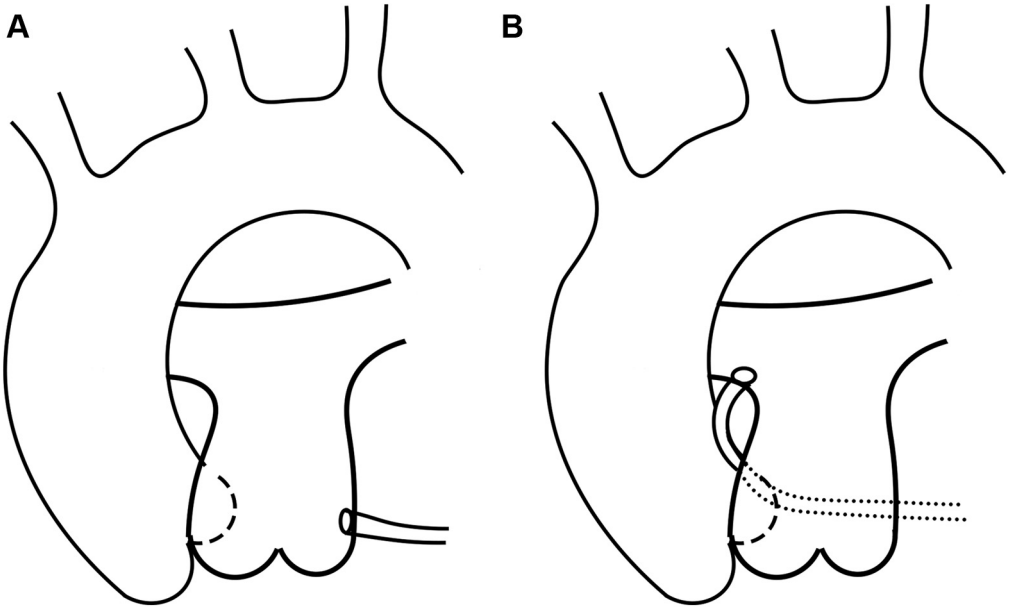

Figure 4 **(A)** Illustration of the most common anatomical configuration of ALCAPA, arising from the proximal main pulmonary artery. **(B)** Illustration of ALCAPA variant with right pulmonary artery origin and intramural aortic course.

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