Anomalous Left Coronary Artery From the Noncoronary Sinus

Wesam Sourour, MD, Jesus C. Jaile, IV, MD, Stefanie Cheang, MD, Jacquelyn D. Brady, PA-C, and Timothy Pettitt, MD, *New Orleans, Louisiana*

INTRODUCTION

Anomalous aortic origin of coronary arteries (AAOCA) occurs in up to 0.7% of the general population.[1] They can be broadly classified as abnormalities of coronary artery origin, course, destination, and size or number of vessels.[2,3] Despite the low incidence, it is a common cause of sudden cardiac death (SCD) in athletes; however, the absolute risk of SCD in the general population is unknown.[1] As such, there remain differing approaches regarding the optimal means of managing this patient population. We describe a case of a patient with anomalous origin of the left coronary artery (AOLCA) from the noncoronary sinus (NCS), which is an extremely rare condition, who presented with exertional chest pain accompanied with syncope.

CASE PRESENTATION

Our patient was an active 10-year-old with no significant medical history who was referred to cardiology due to syncopal episodes that occur immediately after exertional activity. The patient then reported that they had experienced several episodes of nonexertional syncope that had been occurring for several months. They had also started to experience syncope with strenuous activity when exercising or when playing sports, which was sporadically associated with prodromal symptoms. They described chest pain, dizziness, and lightheadedness that progressed to loss of consciousness despite stopping the activity. The syncopal episodes were brief, typically lasting only a few seconds followed by spontaneous recovery to baseline. There was no incontinence, tonic clonic activity, or a postictal period suggestive of seizures during these episodes. Additionally, the patient denied any associated palpitations in conjunction with these episodes.

Due to initial concerns that the symptoms may be explained by dehydration or vasovagal syncope, a trial of lifestyle modification, increased hydration, and salt intake was advised with close follow-up in 6 months. On reevaluation after 6 months the patient described being symptomatic despite adherence to our recommendations, so fludrocortisone was added for treatment of vasovagal syncope. Lab testing including CBC, CMP, and thyroid function testing was within normal limits for age. Upon further follow-up 3 months later the patient continued to experience exertional dizziness and syncopal episodes on a frequent basis occurring almost once per week. The patient was asymptomatic and without orthostatic variation in vital signs during positional maneuvers.

The electrocardiogram was within normal limits with no evidence of preexcitation, ventricular hypertrophy, strain pattern, or ST- or T-wave abnormalities. Given the concerning history, a transthoracic echocardiogram (TTE) was performed. The TTE demonstrated an AOLCA from the NCS, just rightward of the posterior commissure between the left and noncoronary aortic valve (AV) cusps with an intramural course (Figure 1, Video 1). They otherwise had normal intracardiac anatomy, unobstructed arch, and normal biventricular systolic function.

Cardiac computed tomography (CCT) was obtained, which confirmed the findings of the TTE. The anomalous left coronary artery (LCA) arises from the NCS, takes an acute angle, and courses between the left coronary sinus and NCS. On sagittal views there was an abrupt decrease in the caliber of the lumen of the LCA after its takeoff as it courses leftward along the left aortic sinus, suggestive of a 1.4 cm intramural course (Figure 2).

Given the risk of SCD due to the intramural course, unusual location of this LCA origin, and reported clinical symptoms, in addition to a careful heart team discussion, the patient and family were given options for further management including watchful observation, restriction from strenuous activity, or surgical repair. A stress test was considered, as part of the multidisciplinary discussion, but ultimately deferred. It was felt that AOLCA with an intramural course in a symptomatic patient warranted surgical intervention, and a negative stress test in this case would not alter the decision of proceeding with intervention. Preoperative transesophageal echocardiogram (TEE) again demonstrated the previously discussed findings of the TTE and CCT (Figure 3, Video 2).

Intraoperatively, the left coronary ostium was not easily visualized as it was completely obscured by the superior aspect of the left and noncoronary commissural post. The superior aspect of the commissural post was detached, allowing unroofing of the LCA back into the left sinus (Figure 4). Finally, the AV commissure was resuspended with a pledgeted suture. Postoperative TEE demonstrated normal biventricular systolic function and good antegrade flow into the LCA and no aortic regurgitation (AR; Figure 5, Video 3). The patient returned to the intensive care unit extubated with good hemodynamics. They were eventually discharged on postoperative day 5 after an unremarkable hospital course. Another TTE was performed prior to discharge and the findings were unchanged when compared to the postoperative TTE.

During their postoperative clinic visit 4 weeks later, the patient was asymptomatic from the cardiac standpoint. Their physical exam was significant for a new III/IV diastolic murmur not previously appreciated. A TTE demonstrated prolapse of the noncoronary cusp with

From the Department of Pediatric Cardiology, Children's Hospital New Orleans, Louisiana State University Health Sciences Center (W.S., J.C.J.), Department of Pediatric Cardiology, Children's Hospital New Orleans, Tulane University School of Medicine (S.C.), Department of Pediatric Surgery, Children's Hospital New Orleans (J.D.B.), and Department of Surgery, Children's Hospital New Orleans, Louisiana State University Health Sciences Center, New Orleans, Louisiana (T.P.).

Keywords: Anomalous origin of the left coronary artery, Exertional syncope, Transthoracic echocardiography

Reprint requests: Wesam Sourour, Louisiana State University Health Science Center, Children's Hospital New Orleans, 200 Henry Clay Avenue, New Orleans, LA 70118 (E-mail: *wesam.sourour@lcmchealth.org*).

Published by Elsevier Inc. on behalf of the American Society of Echocardiography. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

2468-6441

https://doi.org/10.1016/j.case.2024.04.009

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal short-axis view, without (*left*) and with (*right*) color-flow Doppler (using a low Nyquist limit 23 cm/sec), demonstrates the LCA arising anomalously from the NCS just rightward of the left and noncoronary commissure.

Video 2: Two-dimensional TEE, midesophageal AV short-axis view, without (*left*) and with (*right*) color-flow Doppler (using a low Nyquist limit 23 cm/sec), demonstrates the LCA arising anomalously from the NCS just rightward of the left and noncoronary commissure.

Video 3: Two-dimensional TEE, midesophageal AV short-axis view, without (*left*) and with (*right*) color-flow Doppler (using a low Nyquist limit; 19.3 cm/sec), demonstrates normal antegrade flow in the proximal LCA, which now originates from the left coronary sinus.

Video 4: Two-dimensional TTE, parasternal long-axis view, without (*left*) and with (*right*) color-flow Doppler, demonstrates prolapse of the noncoronary cusp with resultant moderate AR.

Video 5: Two-dimensional TEE, midesophageal AV long-axis view, without (*left*) and with (*right*) color-flow Doppler, demonstrates prolapse of the noncoronary cusp with resultant moderate AR.

Video 6: Two-dimensional TEE, midesophageal long-axis view with color-flow Doppler, demonstrates normal coaptation of the AV leaflets with no evidence of prolapse of the leaflets and mild AR.

Video 7: Follow-up 10 months postoperative, two-dimensional TTE, parasternal long-axis view with color-flow Doppler, demonstrates normal coaptation of the AV leaflets with mild AR.

View the video content online at www.cvcasejournal.com.

resultant moderate AR with pressure half-time of 245 msec and without holodiastolic flow reversal in the descending aorta (Figure 6A, Video 4). There was normal biventricular systolic function and good flow demonstrated in the proximal coronary arteries bilaterally. The patient was brought back to the operating room for AV repair. Preoperative TEE again confirmed the findings demonstrated on TTE during the clinic visit. (Figure 7, Video 5).

Intraoperatively, the commissural post supporting the left and noncoronary cusps had dehisced (Figure 4C). This was resuspended with additional pledgeted sutures resulting in mild AR on postoperative TEE (Figures 8 and 4D, Video 6). The patient ultimately had an unremarkable hospital course and was discharged on postoperative day 7. Ten months postoperatively, the patient has continued to do well from a cardiovascular standpoint with complete resolution of symptoms and resumption of normal activity and exercise without restrictions. A 7-day Holter monitor demonstrated sinus rhythm without significant arrhythmias, prolonged pauses, or any triggered events. On TTE they continue to have mild AR with pressure half-time of 608 msec by color-flow Doppler with no evidence of stenosis (Figure 6B, Video 7) and good antegrade flow across the unroofed LCA.

DISCUSSION

Anomalous origin of the LCA from the NCS is a rare anomaly with an incidence of 0.0008% to 0.012%.[4,5] Coronary arteries typically arise from the 2 pulmonary facing aortic sinuses. This anomaly has largely been demonstrated in the literature in the form of case reports.

Knowledge of the normal origin and course of coronary arteries is essential to recognize congenital coronary anomalies. Each coronary usually arises from its respective sinus above the AV leaflets. Transthoracic echocardiography is the ideal initial modality given that it is noninvasive, widely available, and can be performed in the clinical setting. Coronary arteries are small and superficial structures and are best imaged using high-frequency transducers that allow optimal penetration and maximal resolution. Coronary arteries are best demonstrated in the parasternal short-axis view by scanning the aortic sinus above the level of the AV. The flow is typically low velocity; thus a low Nyquist limit should be selected, often between 20 and 40 cm/sec.[6] Transthoracic echocardiography may be limited due to the superficial nature of the coronary arteries; interference by lung, ribs, or pleura; rapid heart rate; or poor acoustic windows.

Another imaging modality frequently used in conjunction with echocardiography is cardiovascular magnetic resonance (CMR), which may have been a suitable alternative to the CCT used for this patient. Currently there are well-established guidelines and protocols in place for performing CMR in patients with congenital heart diseases. There is also increasing use of postprocessing techniques and virtual angioscopy to evaluate coronary anatomy, myocardial injury, ischemia, and ventricular dysfunction in the pediatric population.[7] Cardiac computed tomography is a superior diagnostic modality to CMR to evaluate the entirety of the coronary arteries and allows for visualization of an intramural course or stenosis. Even as imaging modalities improve, invasive coronary angiography remains important in the evaluation of coronary anomalies. Invasive coronary angiography also allows for anatomic details from within coronary arteries using intravascular ultrasound and physiologic testing using fractional flow reserve measurements, both of which have been demonstrated to be safe to perform in the pediatric population.[8] There is limited experience in the use of these modalities in the general pediatric population that is largely limited to children with Kawasaki disease and heart transplant recipients.[9,10] Given the lack of experience and data in the use of these modalities for pediatric patients with anomalous coronary arteries, they were not performed on our patient.

Historically this anomaly was thought to be benign; however, there have been associations with SCD and fatal arrhythmias reported in the literature.[11,12] Mechanisms explaining the morbidity associated with this include kinking or compression of the anomalous vessel, intermittent ischemia leading to myocardial injury and subsequent foci for lethal ventricular arrhythmias, intussusception, coronary hypoplasia, lateral compression of the coronary wall by the aorta, and restriction of flow through the relatively noncompliant pericommissural area.[1,13]

A literature review by Bravo-Jaimes *et al.*[13] found a total of 19 studies reporting cases of AOLCA from the NCS. A total of 36 out of 174,262 patients were described across all the studies reviewed. Eighteen patients were symptomatic, 11 of whom experienced SCD; 9 of those 11 cases were diagnosed on autopsy. Three patients were diagnosed with myocardial infarction, and

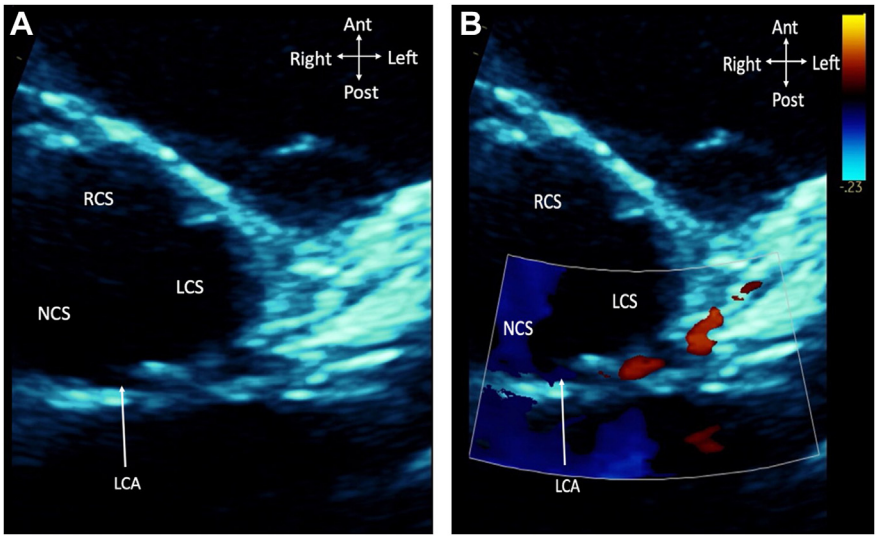

Figure 1 Two-dimensional TTE, parasternal short-axis view, without **(A)** and with **(B)** color-flow Doppler (using a low Nyquist limit 23 cm/sec) during diastole, demonstrates the LCA arising anomalously from the NCS (*arrow*) just rightward of the left and noncommissure with an apparent intramural course. *LCA*, left coronary artery; *LCS*, left coronary sinus; *NCS*, non-coronary sinus; *RCS*, right coronary sinus.

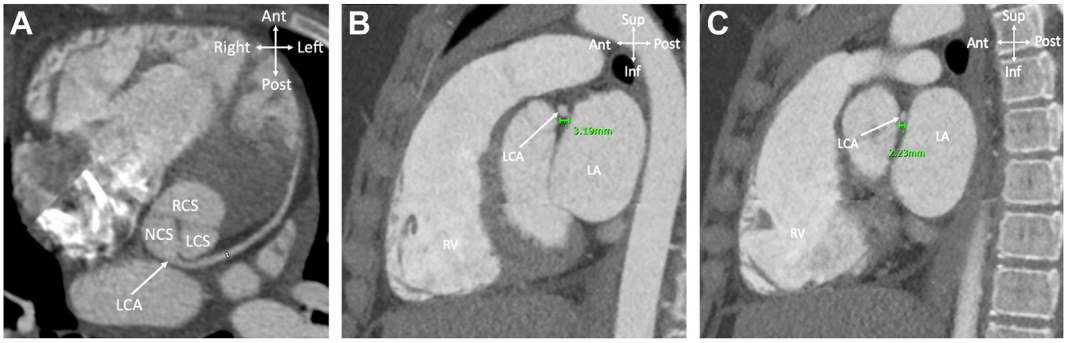

Figure 2 Contrast-enhanced CCT of the coronary arteries in an oblique axial **(A)** and 2 sagittal **(B, C)** multiplanar views demonstrates an AOLCA from the NCS to the right of the commissure between the left and noncoronary cusps. There is an intramural course of the proximal LCA suggested by the variable lumen diameters and nonspherical shape. The distal coronary artery appears to be normal without focal stenosis or aneurysm. *LA*, Left atrium; RV, right ventricle.

5 with angina. Nine patients had high-risk findings, defined as a slit-like orifice, acute angle takeoff, or a proximal intramural segment. Interestingly only 4 patients reportedly were operated on and each with a different surgical technique. A limitation of the studies included in the review is failure to specify the frequency of which high-risk features were demonstrated. Thus, it will be difficult to determine whether the observed overall prognosis is due to the anatomic origin or whether the characteristic of the individual coronary takeoff is a confounding factor. Unfortunately, a comprehensive review of the data pertaining to AOLCA from the NCS in the pediatric population is lacking.

Surgical survival of AAOCA continues to be excellent. It should be cautioned that symptoms and even major cardiac events such as SCD have been observed postoperatively. Thus, families should be counseled about weighing the risks and benefits of undergoing such a repair. Anatomically, the AV commissures are typically located near the intramural segment of the anomalous coronary artery, and commissural takedown often should be performed to achieve complete unroofing if the intramural segment is below the top of the related commissures. Complete unroofing is imperative to achieve an unobstructed neo-ostium and prevention of stenosis postoperatively. The incidence of mild or greater AR is as high as 12% to 17% and may progress, requiring AV replacement.[14]

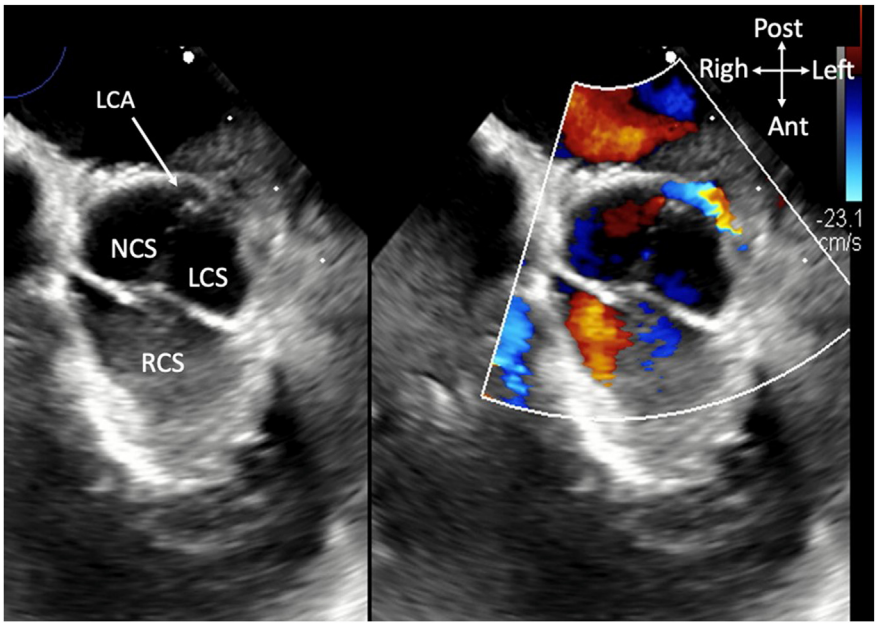

Figure 3 Two-dimensional TEE, midesophageal AV short-axis view, without (*left*) and with (*right*) color-flow Doppler (using a low Nyquist limit; 23 cm/sec) during diastole, demonstrates the LCA arising anomalously from the NCS (*arrow*) with an apparent intramural course.

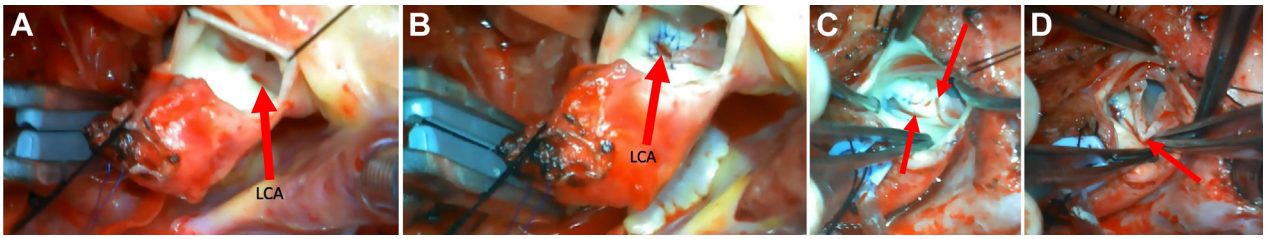

Figure 4 Intraoperative surgical images. Two images from initial unroofing procedure **(A, B)** and 2 images from reintervention on the commissural post (**C**, **D**). (**A**) Aortotomy demonstrating AOLCA from the noncoronary facing sinus (*arrow*). **(B)** Unroofing with marsupialization and resuspension of the superior aspect of the left and noncoronary commissure (*arrow*). **(C)** Failed resuspension and dehiscence of the left and noncoronary commissural post (*arrows*). **(D)** Resuspension of the left and noncoronary commissure (*arrow*).

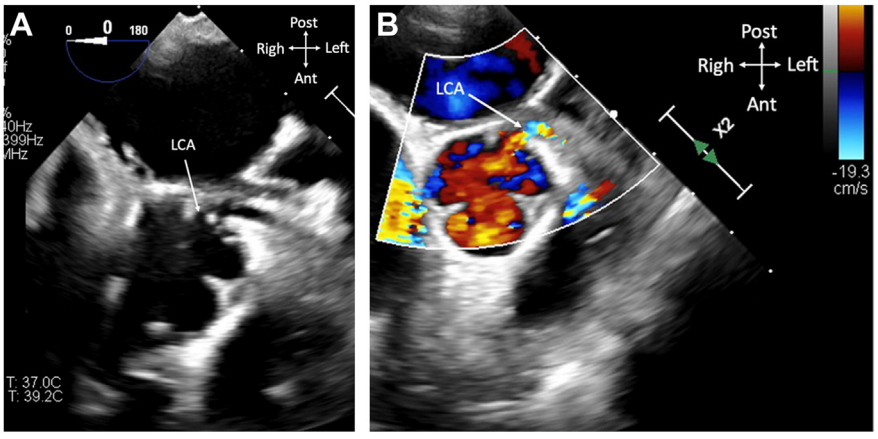

Figure 5 Two-dimensional TEE, midesophageal AV short-axis (0 degree) view, without **(A)** and with **(B)** color-flow Doppler (using a low Nyquist limit; 19.3 cm/sec) during diastole, demonstrates normal antegrade flow in the proximal LCA, which now originates from the left coronary sinus (*arrow*).

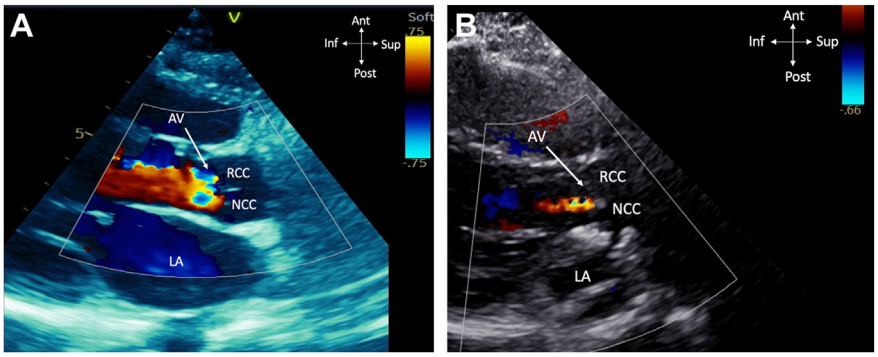

Figure 6 Two-dimensional TTE, parasternal long-axis view, with color-flow Doppler during diastole preoperatively **(A)** demonstrates prolapse of the noncoronary cusp (*arrow*) with resultant moderate AR; 10 months postoperatively **(B)** demonstrates normal coaptation of the AV leaflets and mild AR (*arrow*). *AV*, aortic valve; *NCC*, non-coronary cusp; *RCC*, right coronary cusp.

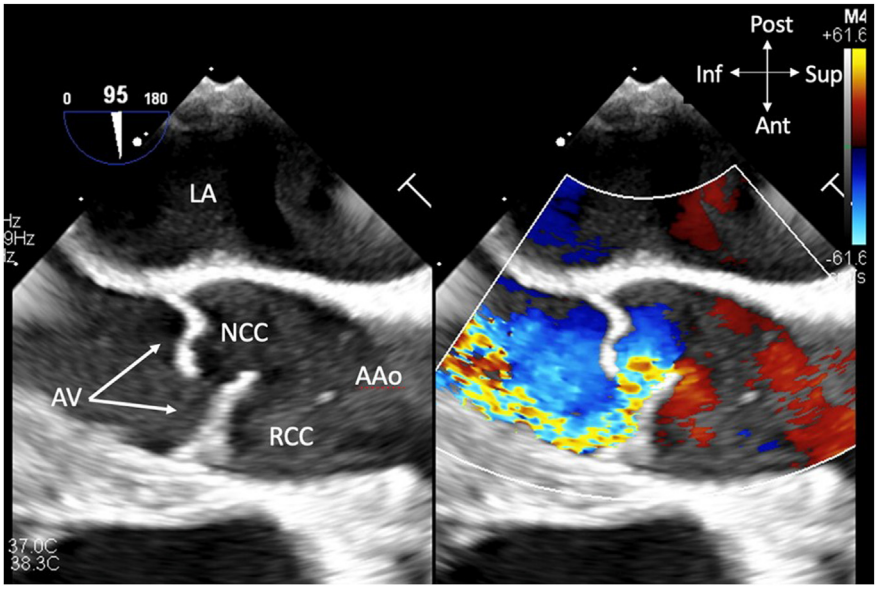

Figure 7 Two-dimensional TEE, midesophageal AV long-axis (95 degree) view, without (*left*) and with (*right*) color-flow Doppler during diastole, demonstrates prolapse of the noncoronary cusp (*arrow*) with resultant moderate AR. *AAo*, Ascending aorta.

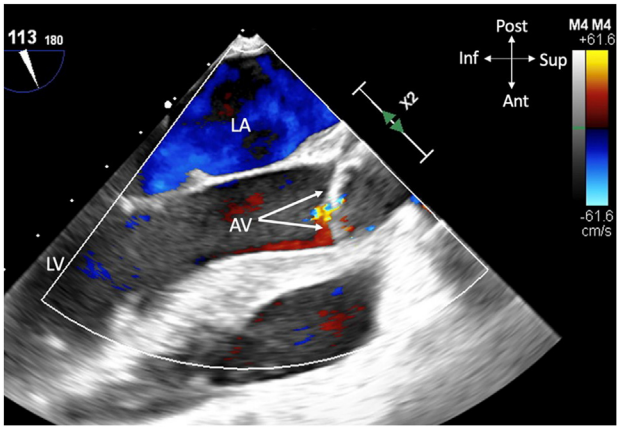

Figure 8 Two-dimensional TEE, midesophageal long-axis (113 degree) view with color-flow Doppler during diastole, demonstrates normal coaptation of the AV leaflets (*arrow*) with no evidence of prolapse of the leaflets. There is mild AR with a vena contracta <0.3 cm. *LV*, left ventricle.

CONCLUSION

Anomalous origin of the LCA from the NCS is a rare condition with limited reports in the literature. Ultimately, due to the risk of SCD, the family opted for surgical management. Our case demonstrates that TTE is a good imaging modality with consistent findings, with the confirmatory CCT in the diagnosis of this rare entity of AOLCA from the NCS with an intramural course. Coronary artery anomalies should be thoroughly interrogated on TTE, specifically the origin and course of the artery with both two-dimensional and color-flow Doppler imaging to rule out AAOCA, especially when there is an index of suspicion with suggestive clinical history.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2024.04.009.

REFERENCES

1. Cheezum MK, Liberthson RR, Shah NR, et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of valsalva. J Am Coll Cardiol 2017;69:1592-608.
2. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation 2007;115:1296-305.
3. Jacobs ML, Mavroudis C. Anomalies of the coronary arteries: nomenclature and classification. Cardiol Young 2010;20:15.
4. Cohen DJ, Kim D, Baim DS. Origin of the left main coronary artery from the "noncoronary" sinus of Valsalva. Cathet Cardiovasc Diagn 1991;22:190-2.
5. Click RL, Holmes DR, Vlietstra RE, et al. Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival: a report from the Coronary Artery Surgery Study. J Am Coll Cardiol 1989;13:531-7.
6. Frommelt P, Lopez L, Dimas VV, et al. Recommendations for Multimodality Assessment of Congenital Coronary Anomalies: A Guide from the American Society of Echocardiography: Developed in Collaboration with the Society for Cardiovascular Angiography and Interventions, Japanese Society of Echocardiography, and Society for Cardiovascular Magnetic Resonance. J Am Soc Echocardiogr 2020;33:259-94.
7. Brothers JA, Kim TS, Fogel MA, et al. Cardiac magnetic resonance imaging characterizes stenosis, perfusion, and fibrosis preoperatively and postoperatively in children with anomalous coronary arteries. J Thorac Cardiovasc Surg 2016;152:205-10.
8. Agrawal H, Molossi S, Alam M, et al. Anomalous coronary arteries and myocardial bridges: risk stratification in children using novel cardiac catheterization techniques. Pediatr Cardiol 2017;38:624-30.
9. Ogawa S, Ohkubo T, Fukazawa R, et al. Estimation of myocardial hemodynamics before and after intervention in children with Kawasaki disease. J Am Coll Cardiol 2004;43:653-61.
10. Kuhn MA, Jutzy KR, Deming DD, et al. The medium-term findings in coronary arteries by intravascular ultrasound in infants and children after heart transplantation. J Am Coll Cardiol 2000;36:250-4.
11. Anwar S, Brook M, Mavroudis C, et al. Anomalous origin of the left coronary artery from the noncoronary cusp: not a benign lesion. Pediatr Cardiol 2012;33:1187-9.
12. Hamamichi Y, Okada E, Ichida F. Anomalous origin of the main stem of the left coronary artery from the non-facing sinus of Valsalva associated with sudden death in a young athlete. Cardiol Young 2000;10:147-1495.
13. Bravo-Jaimes K, Balan P, Garcia-Sayan E. Controversies on the cusp: anomalous origin of the left coronary artery from the NonCoronary cusp. Cureus 2020;12:e7993.
14. Nees SN, Flyer JN, Chelliah A, et al. Patients with anomalous aortic origin of the coronary artery remain at risk after surgical repair. J Thorac Cardiovasc Surg 2018;155:2554-64.