

Incidental Diagnosis of Anomalous Origin of Right Coronary Artery From the Contralateral (Left) Sinus of Valsalva in a Child: Sonographer and Physician Perspectives



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INTRODUCTION

Congenital coronary arterial anomalies (CAAs) are rare in children, and anomalous origin of the left or right coronary artery (RCA) from the contralateral sinus of Valsalva with an interarterial course is a distinct subset, with patients being at risk for myocardial ischemia, ventricular arrhythmias, and sudden cardiac death (SCD).^{1,2} Symptomatic adolescents, competitive athletes, and young adults undergo further tests toward risk stratification and surgery for relief of symptoms and to prevent SCD. However, optimal management in an asymptomatic child with incidental diagnosis and in whom age precludes risk stratification is a subject of debate. This is relevant in contemporary clinical cardiology practice as incidental detection of CAAs is on the rise.³ We report a case in an infant and highlight the principal concerns faced by the physicians and patients (and their parents) in navigating several management issues related to the diagnosis of CAA.

CASE PRESENTATION

A 1-month-old boy reported for cardiac evaluation for a newly noted heart murmur. On general physical examination, he was normal appearing and well nourished, with length and weight at the 50th percentile, pulse oximetry of 98% on room air, heart rate of 84 beats per minute, symmetric peripheral pulses in all extremities, brisk capillary perfusion, and no brachial femoral delay. His clinical cardiac examination demonstrated normal heart sounds and a grade 2/6 medium-pitched ejection systolic murmur over precordium, and further definition of the murmur was limited secondary to fussiness, tachycardia, and noisy breath sounds. His surface electrocardiogram demonstrated no abnormalities. His transthoracic echocardiogram

demonstrated normal structural cardiac anatomy, normal biventricular systolic function, and a suspicion for anomalous origin of the RCA (AORCA) from the contralateral (left) sinus of Valsalva with an interarterial course (Figures 1 and 2, Video 1). In multiple views, the RCA was arising close to (or straddling) the intercommissural line separating the left and right coronary cusps, precluding definitive diagnosis. Further testing was deferred in light of his age; however, a cardiovascular computed tomography was obtained at 4 years of age, and this study confirmed the diagnosis of AORCA with an interarterial course (Figures 3-6). He continues to be on medical follow-up.

DISCUSSION

The exact prevalence of CAA in children is unknown. A prospective study in schoolchildren using a screening cardiovascular magnetic resonance imaging estimated 0.1% for an anomalous origin of the left coronary artery and 0.6% for the AORCA, and this is much less in pooled studies.^{1,2} Incidental detection of CAA is on the rise, and this is attributed to factors like increased usage of transthoracic echocardiography and greater awareness of these conditions.³ Furthermore, adherence to the imaging protocols by the sonographers and enhanced resolution of the latest echocardiographic machines assisted in the diagnosis.⁴ AORCA with an interarterial course is deemed a high-risk substrate for SCD and attributed to factors like extrinsic compression of the coronary artery from the interarterial course, acute angle of takeoff or kinking of the coronary artery, intramural course, slit-like coronary ostium, and (or) coronary artery spasm. These factors in isolation or in combination are deemed either causal or triggers for myocardial ischemia-induced ventricular arrhythmias and SCD.^{5,6} Tomographic imaging such as cardiovascular computed tomography or magnetic resonance imaging can be combined with a stress test to evaluate for myocardial ischemia and to decide the need for surgery. Similarly, surgery can be a default choice in CAA occurring in conjunction with other congenital heart anomalies. In contrast, decisions related to further imaging and timing of surgery in young children with CAA who are otherwise asymptomatic and in those detected incidentally are difficult. Published studies on SCD report the cumulative incidence with the preponderance of deaths in young and middle-aged adults, and hence, the true incidence of SCD in an exclusively pediatric cohort is unknown. The traditional belief that SCD is a rare occurrence in children is an assumption not supported by robust epidemiological or longitudinal studies.² Consensus guidelines on management of AORCA lend support for surgery to alleviate myocardial ischemia and symptoms and to prevent SCD, but these guidelines are mostly applied to the symptomatic cohort including adolescent, competitive athletes and young adults.^{2,7-9} Rational decisions on benefits of surgery in asymptomatic

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VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiogram in the parasternal short-axis view at the level of aortic valve with color flow mapping showing the origin of the RCA from the left sinus of Valsalva and the interarterial course and antegrade flow into the RCA. Note the pulmonary valve leaflets in this view, a useful landmark to distinguish an interarterial course from the sub-muscular or infundibular course of the proximal RCA.

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

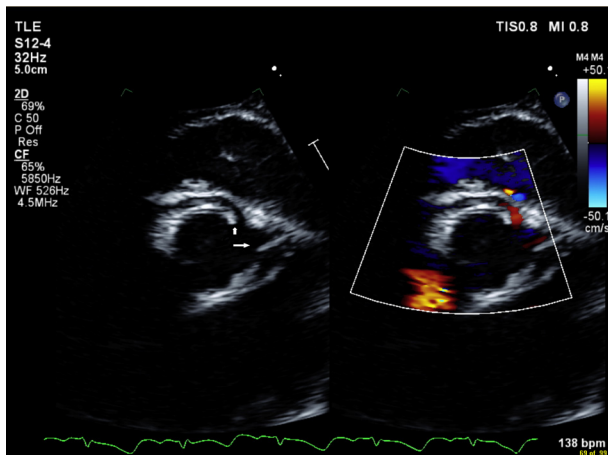


Figure 1 Transthoracic echocardiogram in the parasternal short-axis view showing the aorta with origin of the RCA from the left sinus of Valsalva (*vertical arrow*). The *right panel* in color flow shows the antegrade flow into both coronary arteries.

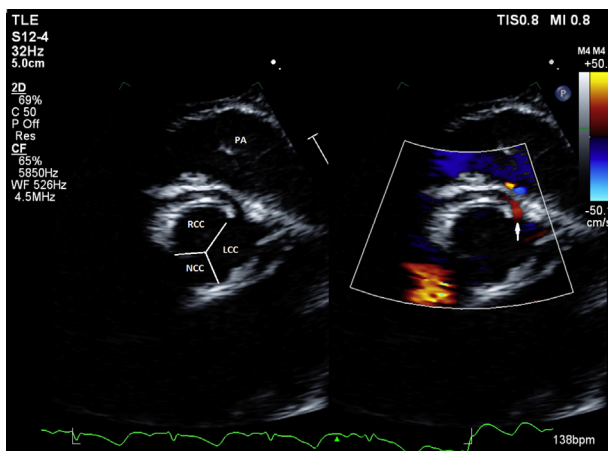


Figure 2 Transthoracic echocardiogram in the parasternal short-axis view with aortic cusps delineated by *solid lines*. The *vertical arrow* pointing upward shows the origin of the RCA. LCC, Left coronary cusp; NCC, noncoronary cusp; RCC, right coronary cusp.

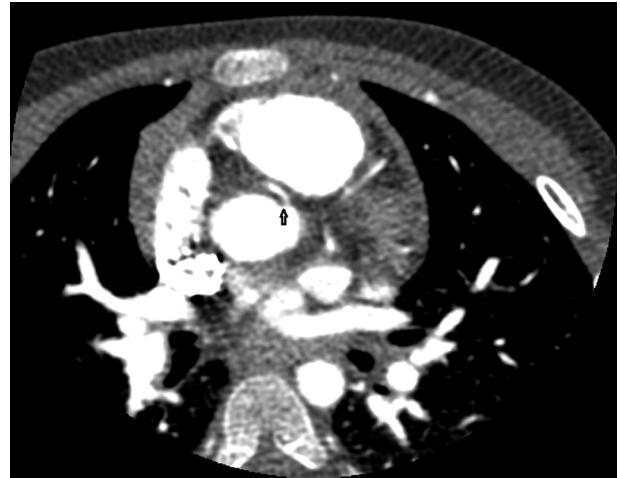


Figure 3 Cardiovascular computed tomography contrast-enhanced image in the axial view showing the origin of the RCA (*vertical arrow*) from the contralateral sinus of Valsalva.

children with incidental diagnosis are difficult to evaluate in the presence of several knowledge gaps and unproven assumptions related to management of AORCA from the contralateral sinus of Valsalva.² Quite often physicians rely on shared decision-making and their anecdotal experience while making decisions related to proceeding to surgery.

Therefore, for the following list of reasons, the incidental diagnosis of AORCA in young children raises several key management dilemmas: (1) there are limited data on an optimal surveillance schedule; (2) the optimal diagnostic testing strategies, their feasibility, and their most reliable parameters are unknown; (3) the reliability of symptoms for risk stratification is poor; (4) the clinical impact and outcome with medical therapy alone is poorly studied; and (5) the timing, the type of surgery, and the clinical impact of surgical intervention are not well defined.

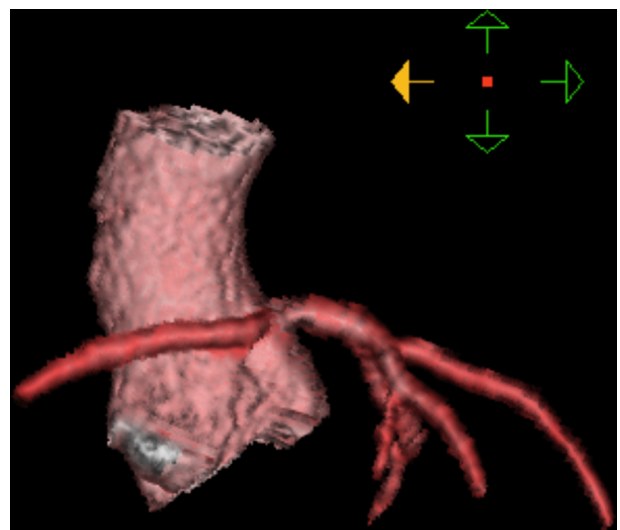


Figure 4 Cardiovascular computed tomography contrast-enhanced three-dimensional volume-rendered display of the aortic root and the ascending aorta showing the RCA from the contralateral sinus of Valsalva.

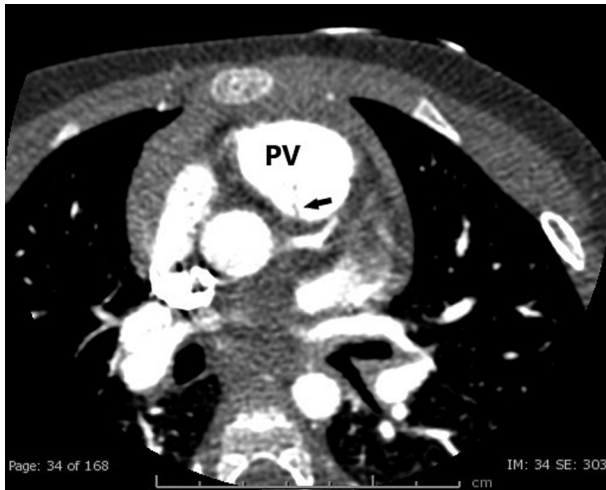


Figure 5 Cardiovascular computed tomography contrast-enhanced image in the axial view to delineate the pulmonary valve (PV) leaflets (*arrow*), which is also a useful landmark to differentiate an interarterial from the subvalvular or muscular course of the anomalous RCA.

The principal rationale for surgical intervention is to alleviate symptoms and to prevent SCD, and studies in children have not addressed these issues conclusively. Limited data on the risk of SCD in a pediatric cohort combined with a widely held presumption that children are at low risk for SCD led to medical follow-up; however, this is more a default choice and not an evidence-based recommendation.^{2,7-9} Hence, can medical follow-up be an acceptable management strategy?

CONCLUSION

Incidental diagnosis of AORCA from the contralateral sinus of Valsalva in children poses numerous management dilemmas, and patients, parents, and their physicians navigate an arduous clinical course that begins with diagnosis, selection of the most desirable imaging modality, a search for the most decisive information from the battery of available tests, prevention of the most dreaded complication of SCD, and deciding whether surgery is the most definitive treatment option. And finally, the most daunting question is whether surgery will prevent SCD? Surgical revascularization data on quality of life, relief of symptoms, and on prevention of SCD in exclusive pediatric cohort with AORCA is too sketchy to assist in definitive management decisions.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.case.2022.05.007>.

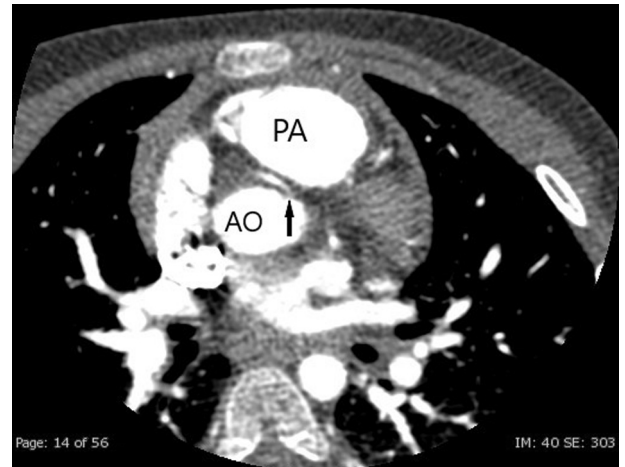


Figure 6 Cardiovascular computed tomography gated contrast-enhanced image in the contiguous axial view showing the anomalous course of the RCA (*arrow*) with an interarterial course. AO, Aorta; PA, pulmonary artery.

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