Quadricuspid Aortic Valve With Guarded Left Coronary Artery Ostium in an Infant With Myocardial Infarction

Anne Aurelie Tedga, MD, Frank Pigula, MD, LaTasha Lewis, MD, and Thomas Richard Kimball, MD, *New Orleans, Louisiana*

INTRODUCTION

A quadricupid aortic valve (QAV) is a rare congenital anomaly. Patients with QAV typically present with aortic valve regurgitation (AR) in the fifth and sixth decade of life, often leading to aortic valve repair or replacement.[1] We present the case of an infant diagnosed with QAV and a guarded left coronary ostium in our cardiac intensive care unit (CICU) after a resuscitated cardiac arrest.

CASE PRESENTATION

A previously healthy 2-month-old boy was transferred to our CICU after presenting to an outside emergency department with a cardiac arrest. The patient had an uneventful perinatal history and was exclusively breastfed. On the day of presentation, the patient became increasingly irritable and diaphoretic during feeding and then became unresponsive. After successful resuscitation, the outside transthoracic echocardiogram (TTE) showed severely diminished left ventricular global function with a left ventricular ejection fraction (LVEF) of 28%.

The infant was admitted to the CICU on milrinone and mechanical ventilation. There was profound ST segment depression in the anterolateral leads of the electrocardiogram suggestive of non-ST elevation myocardial infarction (Figure 1). Troponin I levels peaked at 42 ng/mL. A complete TTE demonstrated a QAV with an unusual appearance of the left coronary artery ostium guarded by a leaflet of the valve (Figure 2; Video 1). The left posterior leaflet entrapped the left sinus of Valsalva (Figure 3) with resultant mild AR but no stenosis. The LVEF was 25% (biplane Simpson's). After surgical conference discussion, a cardiac computed tomography scan was performed that demonstrated normal coronary origins, caliber, and course. Given the clearly delineated pathology seen with TTE (Figure 4) and the findings on cardiac computed tomography scan, invasive angiography was deemed unnecessary.

With inotropic support (calcium chloride, dopamine, and epinephrine infusions) and afterload reduction with milrinone, the left ventricular function improved to an LVEF of 43%. However, the patient had 2 consecutive runs of nonsustained pulseless ventricular tachycardia with Torsades de Pointe morphology. These were felt to be secondary to ischemic changes from the myocardial infarction rather than a primary channelopathy. Due to focal seizures (felt to be secondary to the cardiac arrest), a brain magnetic resonance imaging was performed with normal findings, and the patient was cleared for cardiovascular surgery. After the troponin levels stabilized, the patient went to the operating room for aortic valve exploration on day 7 of admission. Intraoperatively, a QAV was confirmed with the left sinus of Valsalva having both a major and a minor component, the minor being adjacent to the noncoronary sinus. The left coronary artery ostium, severely hypoplastic at 1.5 mm in diameter, resided in the rudimentary minor sinus. A left commissuroplasty was performed, and the aortic valve was reconstructed. The surgery was well tolerated, and the postoperative course was uneventful. Light inotropic support was required, and the patient was weaned off of it 2 days postsurgery. The patient remained on mechanical ventilation for 1 day and was extubated to high-flow nasal cannula and subsequently to room air. They were discharged from the cardiology step-down unit 8 days postsurgery. The left ventricular function had normalized to an LVEF of 54%. The echo showed relief of the guarding of the ostium with good flow into the left main coronary artery demonstrated by color Doppler imaging. There was residual moderate AR (initially mild) and still no stenosis. Postop and until this day, there has been no recurrence of ventricular tachycardia.

VIDEO HIGHLIGHTS

Video 1: Parasternal short-axis view demonstrates quadricuspid leaflets with smaller left posterior leaflet *(arrow)* entrapping the left coronary ostium. *A*, Anterior; *AoV*, aortic valve; *L*, left; *LA*, left atrium; *PV*, pulmonary valve.

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

DISCUSSION

The normal aortic valve comprises 3 leaflets: the right, left, and noncoronary sinuses of Valsalva, named after their relationships with corresponding coronary arteries. Anatomical variations of the aortic valve have been described in the literature including the unicuspid, bicuspid, and quadricuspid valves. Among these, the most common anomaly is the bicuspid aortic valve, affecting 1%-2% of the general population.[2] The QAV is a rare malformation and has an incidence of 0.008% on autopsy, 0.043% on echocardiography, and 1% in patients undergoing surgery for isolated AR.[3] Hurwitz and Roberts described 7 anatomic variations of QAV.[4] Type a consists of 4 equal sized coronary cusps. Type b, the most common variation, consists of 3 equal sized cusps and 1 smaller cusp and is consistent with the anatomy of our patient. Type c consists of 2 equal large and 2 equal smaller cusps. Type

From the Division of Cardiology (A.A.T., T.R.K.), Division of Cardiothoracic Surgery (F.P.), and Division of Cardiac Intensive Care Unit (L.L.), Children's Hospital of New Orleans, New Orleans, Louisiana.

Correspondence: Anne Aurelie Tedga, MD, Division of Cardiology, Children's Hospital of New Orleans, 200 Henry Clay Avenue, New Orleans, LA 70118. (E-mail: *Anne.Tedga@lcmchealth.org*).

2468-6441

https://doi.org/10.1016/j.case.2022.10.005

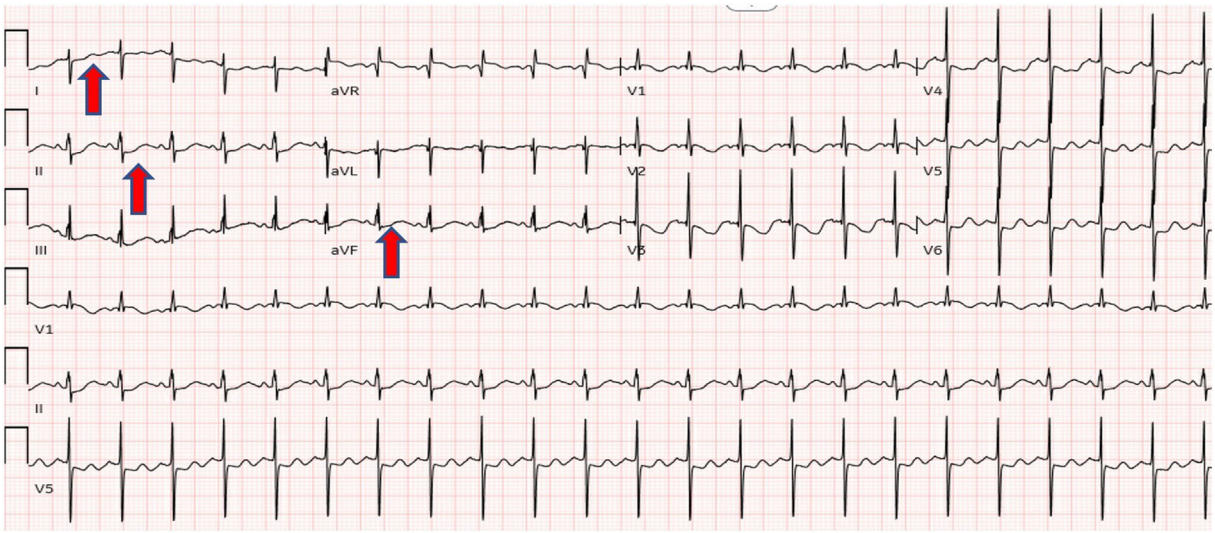

Figure 1 Electrocardiogram of a 2-month-old patient with non-ST elevation myocardial infarction. It demonstrates ST segment depression in leads I, II, and aVF, inferior leads *(arrows)*. It also shows T-wave inversion in the precordial leads.

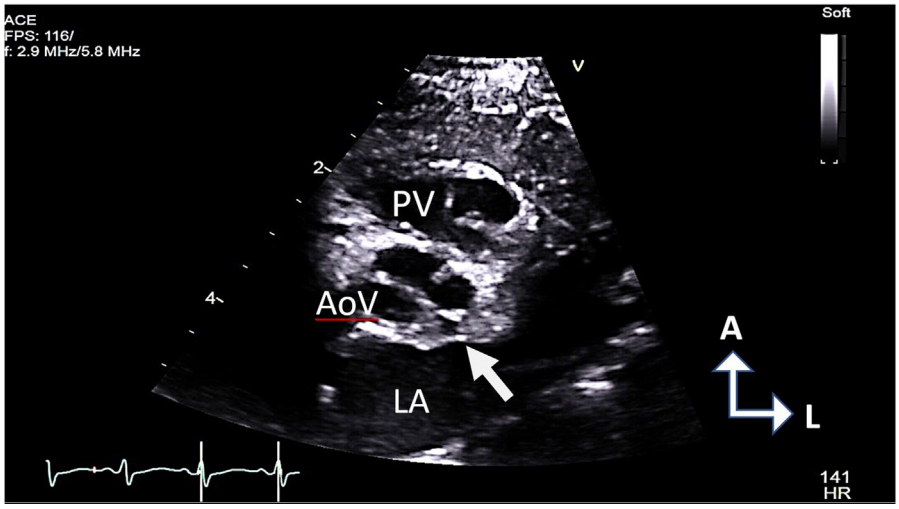

Figure 2 Parasternal short-axis view demonstrates quadricuspid leaflets with 3 equal leaflets and a smaller left posterior leaflet *(arrow)* entrapping the left coronary ostium. *A*, Anterior; *AoV*, aortic valve; *L*, left; *LA*, left atrium; *PV*, pulmonary valve.

d consists of 1 large cusp, 2 intermediate equal cusps, and 1 smaller cusp. Type e consists of 1 large cusp and 3 equal smaller cusps. Type f consists of 2 equal large cusps and 2 small unequal cusps. The last type, type g, consists of 4 unequal cusps.

QAV has a male predilection with a male:female ratio of 1.6:1.[1] QAV is generally an isolated malformation, and adult patients typically present with aortic valve dysfunction such as regurgitation, the most common hemodynamically significant lesion, in their fifth and sixth decade.[2,5] Sometimes it is associated with other congenital malformations such as coronary anomalies, including displacement and dislodgement of coronary ostia, which are reported in up to 10% of patients with a QAV.[1,5] This is important preoperative information for surgeons as it helps them to prevent ostial obstruction during placement of a prosthetic valve. There have only been a handful of pediatric case reports requiring surgery due to a coronary ostial obstruction, one of these occurring in Italy.[6] This child (in Italy) had an anatomically similar left coronary artery ostium, also obstructed by a rudimentary accessory left sinus of Valsalva. However, rather than ostial coronary artery repair, he had Konno surgery with a 19 mm St. Jude mechanical valve placement. Our patient's presentation with a QAV and coronary ostial anomaly in infancy is rare because of associated myocardial infarction leading to cardiac arrest. A 2-month-old boy with the same diagnosis described in Japan by Harada *et al.*[7] had a different presentation. That patient was admitted with poor weight gain and progressive inability to suckle. The ability to diagnose QAV has been greatly enhanced by echocardiography; however, the aortic leaflets may not be well visualized on TTE.[3] Sometimes transesophageal echocardiography may provide additional diagnostic value. Surgical correction was delayed in the infant from Japan because ostial obstruction was not clearly visualized on

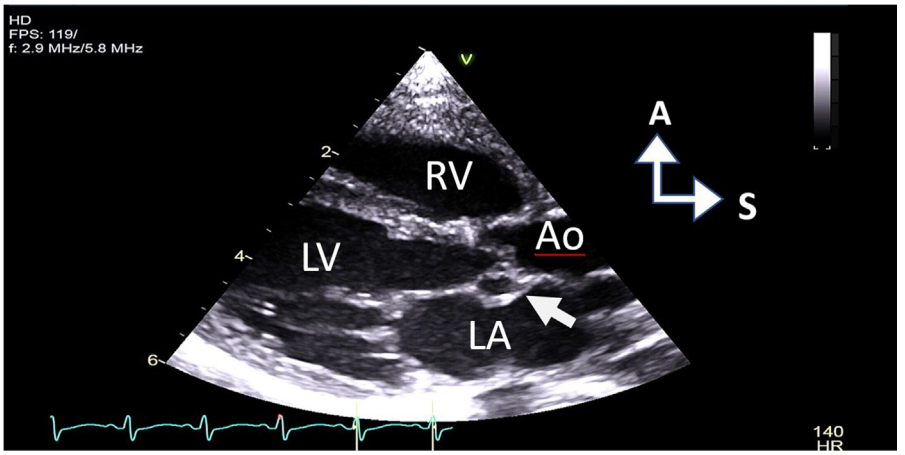

Figure 3 Parasternal long-axis view demonstrates abnormal left posterior leaflet *(arrow)* entrapping left sinus of Valsalva. *A*, Anterior; *Ao*, aortic valve; *LA*, left atrium; *LV*, left ventricle; *RV*, right ventricle; *S*, superior.

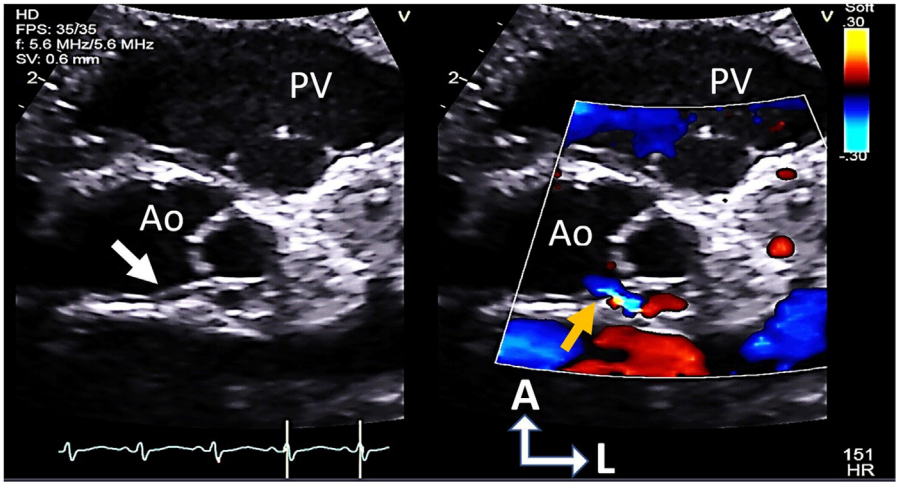

Figure 4 Parasternal short-axis view: two-dimensional image demonstrates quadricuspid leaflets with the smaller left posterior leaflet entrapping the left coronary ostium *(white arrow)*. Color Doppler image demonstrates aliasing of flow as it crosses this rudimentary leaflet coursing to the left coronary artery *(yellow arrow)*. *A*, Anterior; *Ao*, aortic valve; *L*, left; *PV*, pulmonary valve.

both TTE and coronary angiography initially. However, a transesophageal echocardiography and repeat coronary angiography performed 1 month later revealed near occlusion of the left coronary artery ostium by a pouch.[7] In our case, prompt and accurate diagnosis of the aortic valve on initial TTE allowed a valve-sparing repair, correcting the ostial obstruction and stenosis while converting the quadricuspid aortic valve to tricuspid morphology.

CONCLUSION

QAV is a rare congenital heart malformation, which can cause significant AR, requiring surgery in late adulthood. Infrequently, it can present early in life, especially when associated with coronary artery ostial anomalies. Coronary artery anomalies should be ruled out in the setting of aortic valve abnormalities and evidence of myocardial ischemia. The presence of myocardial ischemia is of particular importance because it can predispose to life-threatening cardiac arrest leading to higher mortality and morbidity.

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

The authors declare that written patient consent was not provided for the following reason: Verbal consent was obtained from patient's

mother on October 18, 2022; other than the patient's age and gender, no other identifiers or personal pictures were used.

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.

ACKNOWLEDGMENTS

Special thanks to the following sonographers who helped acquire images: Marilyn Perkins-Capers, RDCS (performed the initial TTE that led to the diagnosis and prompt treatment for the patient); Kim Phuong Nguyen, RDCS; Vanessa Mansey, RDCS; Cristhel Fernandez, RDCS; Ann Cuenca, RDCS; Kristy Reimonenq, RDCS; Nhavan Nguyen, RDCS; Rebecca Kearns, RDCS; Kristie Kussman, RDCS; Baillie Pinion, RDCS; Melinda Tidwell, RDCS; Ally Frye, RDCS.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2022.10.005.

REFERENCES

1. Tutarel O. The quadricuspid aortic valve: a comprehensive review. J Heart Valve Dis 2004;13:534-7.
2. Molinary MC, Martin AB, Asevedo EE, McIlwain EF, Cospolich B, Kerut EK. Normally functioning quadricuspid aortic valve mistaken for bicuspid valve in a septuagenarian. Echocardiography 2021;38:365-7.
3. Timperley J, Milner R, Marshall AJ, Gilbert TJ. Quadricuspid aortic valves. Clin Cardiol 2002;25:548-52.
4. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. Am J Cardiol 1973;31:623-6.
5. Tsang MYC, Abudiab MM, Ammash NM, Naqvi TZ, Edwards WD, Nkomoand VT, et al. Quadricuspid aortic valve characteristics, associated structural cardiovascular abnormalities, and clinical outcomes. Circulation 2016;133:312-9.
6. Benassi F, DiBenedetto G, Labia C, Stefanelli G. Left coronary ostium isolation and aortic valve dysplasia in a young boy: a case report. J Heart Valve Disease 2012;21:135-7.
7. Harada T, Fukae K, Ando Y. Surgical repair of ostial obstruction of the coronary artery for quadricuspid aortic valve in an infant. Interact Cardiovasc Thorac Surg 2017;24:634-5.