

Supracristal Ventricular Septal Defect Complicated by Formation of an Aorto-Right Ventricular Outflow Tract Fistula: A Rare Cause of Biventricular Enlargement

Amer Muhyieddeen, MD; Ashwini Sadhale, MD; Siri Kunchakarra, MD; Ankit Rathod, MD

UNIVERSITY OF CALIFORNIA, SAN FRANCISCO-FRESNO, FRESNO, CALIFORNIA

ABSTRACT: Aorto-right ventricular outflow tract fistulas typically occur secondary to trauma, infective endocarditis, and sinus of Valsalva aneurysm rupture. We describe an unusual case of a spontaneous aorto-right ventricular outflow tract fistula in the absence of such findings, instead forming secondary to a complicating supracristal ventricular septal defect and leading to dilated cardiomyopathy.

HISTORY

A 41-year-old male presented to the emergency department with progressively worsening exertional dyspnea and symptoms of heart failure. The patient had been previously followed by a pediatric cardiologist for a congenital ventricular septal defect (VSD) but was lost to follow-up. On physical examination, vital signs were stable. The patient had elevated jugular venous distension, regular heart rate and rhythm with split S2 and 2/6 pansystolic murmur, diminished breath sounds on bilateral lung fields, a distended abdomen, and edematous bilateral lower extremities. His medical history was significant for hypertension and perimembranous VSD.

INVESTIGATION

An electrocardiogram (ECG) showed normal sinus rhythm and right axis deviation with no suggestion of ischemia. Labs were normal with no indication of anemia, electrolyte derangements, or metabolic abnormalities. Troponins were also normal. A transthoracic echocardiogram showed a severely dilated left ventricular chamber with global hypokinesis and an ejection fraction of 25%. A transesophageal echocardiogram (TEE) confirmed the severely dilated left ventricle, diffuse global hypokinesis, and estimated ejection fraction of 20% to 25%. A VSD with predominant left-to-right shunting was observed on color Doppler TEE (Figures 1, 2). A chest x-ray showed cardiomegaly with a prominent cardiac contour.

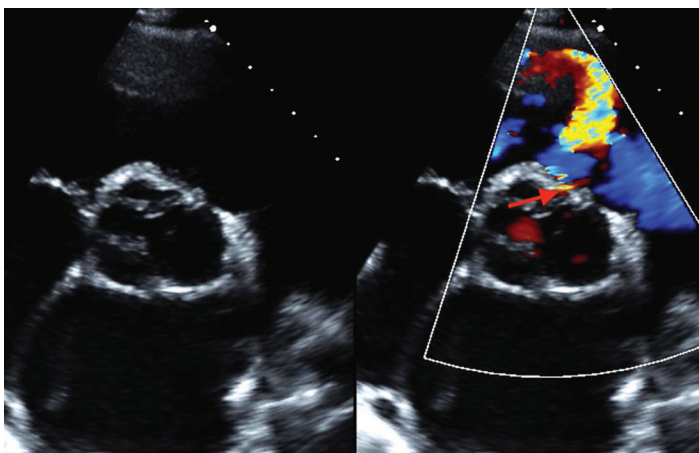


Figure 1.
Transesophageal echocardiography showing ventricular septal defect jet between the 12:00 and 2:00 positions (red arrow).

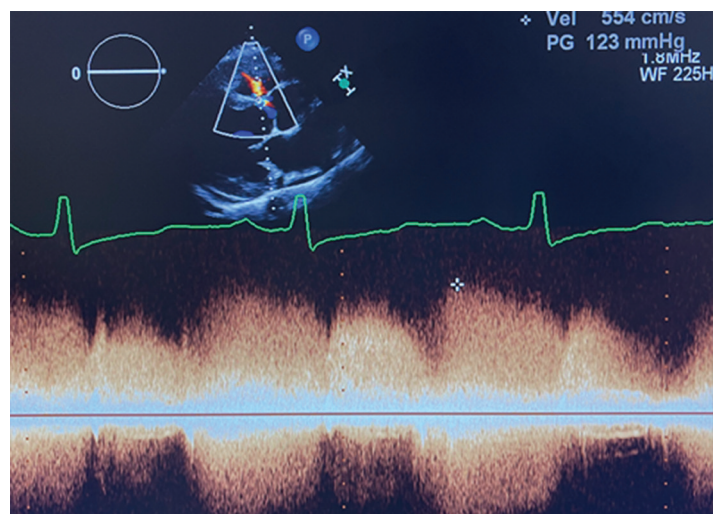


Figure 2.
Doppler wave form on transesophageal echocardiogram.

Given the patient's history of a congenital VSD, cardiac magnetic resonance imaging (CMR) was used to evaluate the morphology and possible associated complications. CMR showed severe biventricular dilation and evidence of a supracristal VSD with an 8.5-mm defect adjacent to the

right sinus of Valsalva (Figures 3, 4). The VSD was further complicated by development of an aorto-right ventricular outflow tract (RVOT) fistula with evidence of continuous left-to-right shunt across the VSD from the left side into the RVOT in the subpulmonic region (Figures 5, 6). The calculated pulmonary-to-systemic flow ratio ($Q_p:Q_s$)

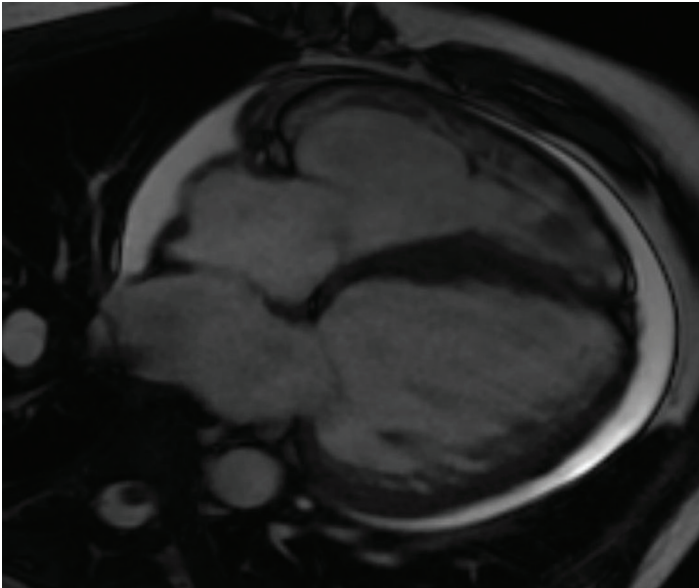


Figure 3.
Severe biventricular dilation seen on cardiac magnetic resonance imaging.

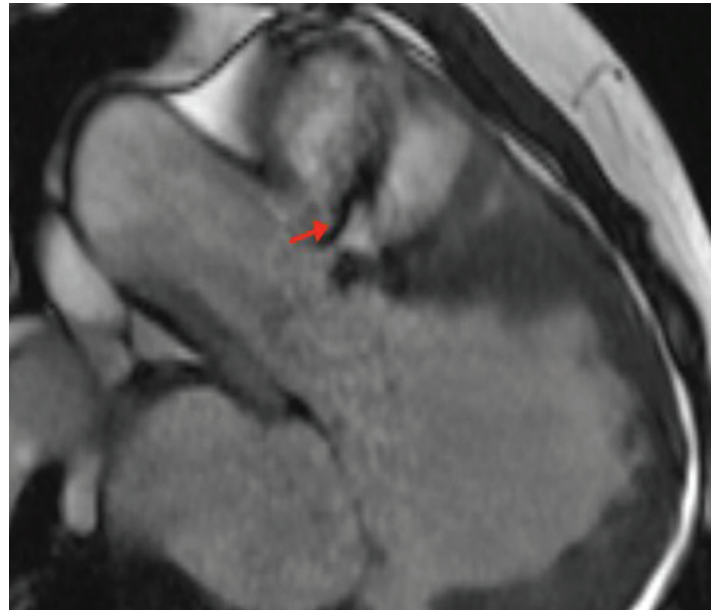


Figure 5.
Aorta-right ventricular outflow tract fistula demonstrated by the presence of systolic flow jet (red arrow).

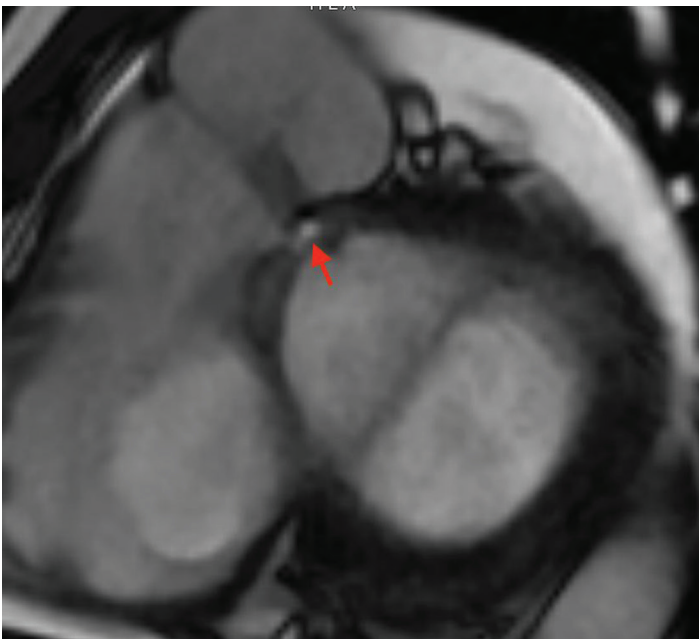


Figure 4.
Supracristal ventricular septal defect (red arrow) with an 8.5-mm defect adjacent to the right sinus of Valsalva.

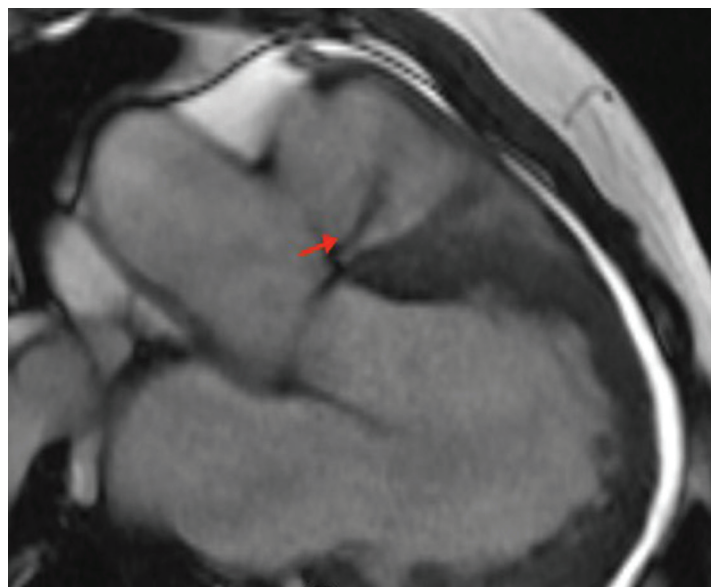


Figure 6.
Aorta-right ventricular outflow tract fistula demonstrated by the presence of diastolic flow jet (red arrow).

was 2:1, suggesting significant left-to-right shunt. The right ventricle was significantly dilated, with features of pulmonary hypertension suggestive of some chronicity to the development of the fistulous connection.

The patient subsequently underwent a left and right heart catheterization with coronary and aortic root angiography. Left ventricular systolic pressure was 115 mm Hg, and left ventricular end diastolic pressure was 15 mm Hg. Right heart catheterization showed a mean pulmonary arterial pressure of 28 mm Hg, pulmonary arterial saturation of 81%, right ventricular saturation of 73%, right atrial saturation of 66%, inferior vena cava saturation of 66%, superior vena cava saturation of 66%, and arterial saturation of 94%. The Qp:Qs ratio was 2.2:1. Coronary artery angiography revealed nonobstructive coronary arterial disease, and aortic root angiography showed 2+ aortic regurgitation.

MANAGEMENT

The cardiothoracic surgery team evaluated the patient for VSD closure and closed the supracristal VSD with a pliable collagen bioscaffold patch, which was also used to repair an aortic valve perforation in the area of the right coronary cusp. The patient experienced no immediate complications during or after surgery.

FOLLOW-UP

At 1-month follow-up, the patient was doing well and reported near-complete resolution of his symptoms. At 6-month follow-

up, a repeat CMR showed no sign of a fistulous connection across the right cusp of the aorta and RVOT. The left ventricular systolic function was improved to between 50% and 55%. The left ventricle was noted to be mildly dilated, with an end diastolic diameter of 5.5 cm. The right ventricular size was noted to be grossly normal (Figure 7).

DISCUSSION

Ventricular septal defects are the most common form of congenital heart disease diagnosed in children, with some studies showing an estimated incidence as high as 5% of all births.¹ Historically, echocardiography has been used to evaluate patients with VSDs and quantify the severity of shunt morphology. Although echocardiography is highly accurate in evaluating shunt morphology, cardiac output measurements strongly depend on the assumed flow area and sampling site.² On the other hand, CMR can evaluate cardiac morphology and function independent of body habitus and proper probe positioning. CMR has also been useful in quantifying the magnitude of shunting. In one study, use of contrast during CMR showed that it was more accurate than even invasive cardiac catheterization in quantifying flow and shunt size.³ Thus, CMR enables noninvasive visualization of cardiac anatomy and function with superior temporal and spatial resolution, facilitating disease diagnosis, management, and treatment.

Through the use of CMR, we were able to elucidate and characterize the morphology of our patient's VSD and

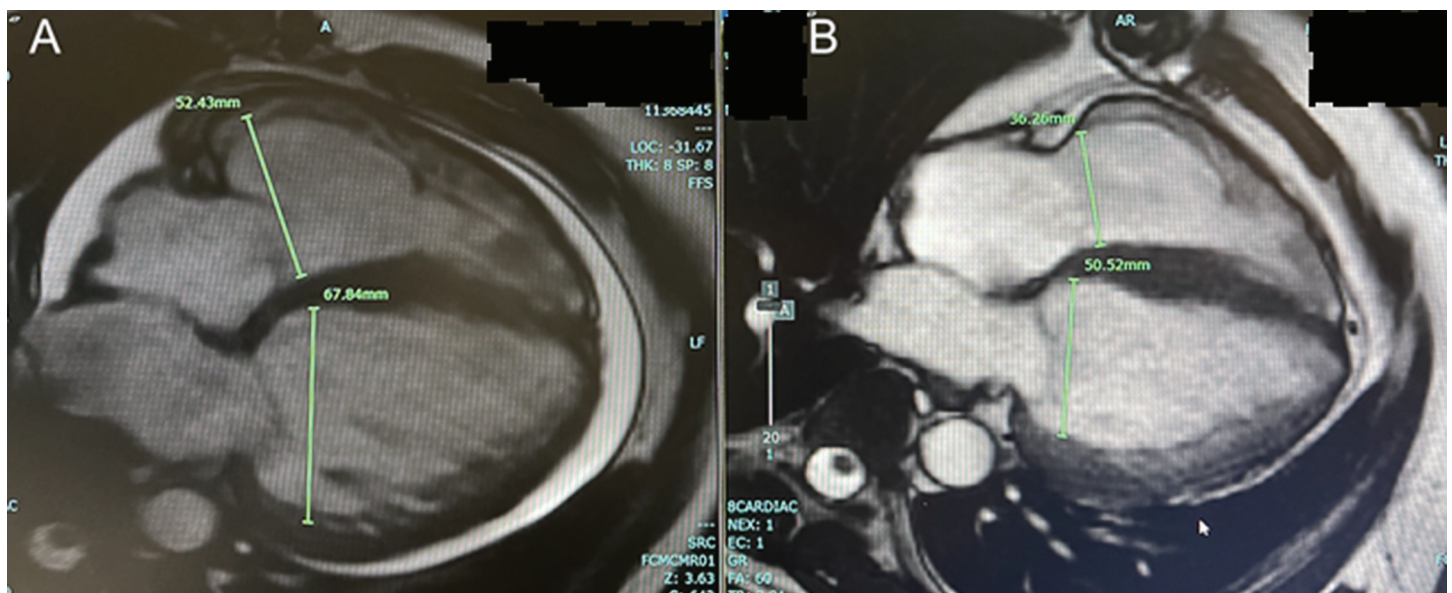


Figure 7.

(A) Biventricular dilation prior to surgery. (B) After surgery. At 6-month follow-up, there was marked improvement in end diastolic diameters of both the left and right ventricles.

determine that it had been complicated by fistula formation. Our patient had a type of VSD known as supracristal or subarterial. Supracristal VSD only accounts for about 5% of known VSDs in Western populations but has been seen as high as 20% in Asian populations.^{4,5} These VSDs have been associated with several other congenital heart defects, including sinus of Valsalva aneurysms (SOVAs), aortic valve prolapses, and aortic valve regurgitation. Observational studies have also shown that up to 61% of patients over 20 years of age with a supracristal VSD had a coexisting SOVA, and within this group, 91% experienced rupture.⁶ Supracristal VSDs have also been found to lead to a substantially higher risk of aortic valve prolapse and aortic regurgitation.⁴ Thus, characterizing this subtype of VSD at a young age is extremely important due to the detrimental consequences it may bring later in life.

The defining feature of our patient was the presence of a supracristal VSD complicated by the formation of an aorto-right ventricular fistula. Aorto-cardiac fistulas are extremely rare and, when found, are usually a complication of trauma or infective endocarditis.⁷ Some observational studies have found that up to 76% of aorto-cardiac fistulas were due to ruptured SOVAs.⁸ With the advent and increased use of transcatheter aortic valve replacement (TAVR) for aortic stenosis, aorto-cardiac fistula formation is recognized as a major complication.⁹ Our patient was notable for lacking the typical inciting events that are usually known to cause these aorto-cardiac fistulas; without a history of trauma or infection, the fistula's mechanism of formation was unclear. There was no obvious valvular endocarditis-like vegetation or obvious SOVA visualized by the CMR. Additionally, the presence of right ventricular enlargement is an uncommon finding with VSD. Our patient was thought to have biventricular enlargement from the fistula, causing chronic volume overload.

CONCLUSION

This case highlights the increasingly indispensable role CMR plays in diagnosing aorto-cardiac fistulas. An aorto-cardiac fistula is an exceedingly rare and serious complication that typically results from an inciting incident. In our patient, it led to the development of symptomatic heart failure with reduced ejection fraction and biventricular dilation. Most medical literature has described the formation of such fistulas as secondary to trauma, TAVR, infective endocarditis, or SOVAs. In our patient, none of these pre-existing conditions were present. Given CMR's excellent imaging resolution and noninvasiveness, it is an excellent choice for evaluating such patients, especially young pediatric populations where ionizing radiation is a concern. Future strategies may involve a broader application and earlier use of CMR in patients

with suspected congenital heart defects. Timely intervention for clinically significant VSDs may prevent potential harmful sequelae and reduce associated morbidities.

Corresponding Author:

Amer.Muhyieddeen@ucsf.edu

Conflict of Interest Disclosure:

The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

Keywords:

congenital ventricular septal defect, cardiac magnetic resonance imaging, congestive heart failure, congenital cardiac disease, cardiology

REFERENCES

1. Sands AJ, Casey FA, Craig BG, Dornan JC, Rogers J, Mulholland HC. Incidence and risk factors for ventricular septal defect in "low risk" neonates. *Arch Dis Child Fetal Neonatal Ed.* 1999;81(1):F61-63. doi:10.1136/fn.81.1.f61.
2. Dittmann H, Voelker W, Karsch K-R, Seipel L. Influence of sampling site and flow area on cardiac output measurements by Doppler echocardiography. *J Am Coll Cardiol.* 1987 Oct;10(4):818-823. doi:10.1016/S0735-1097(87)80275-9.
3. Muthurangu V, Taylor A, Andriantsimivona R, et al. Novel method of quantifying pulmonary vascular resistance by use of simultaneous invasive pressure monitoring and phase-contrast magnetic resonance flow. *Circulation.* 2004 Aug 17;110(7):826-34. doi:10.1161/01.CIR.0000138741.72946.84.
4. Layangool T, Kirawittaya T, Sangtawesin C, et al. Natural aortic valve complications of ventricular septal defect: a prospective cohort study. *J Med Assoc Thai.* 2008 Oct;91 Suppl 3:S53-9.
5. Ammash NM, Warnes CA. Ventricular septal defects in adults. *Ann Intern Med.* 2001 Nov 6;135(9):812-24. doi:10.7326/0003-4819-135-9-200111060-00011.
6. Momma K, Toyama K, Takao A, et al. Natural history of subarterial infundibular ventricular septal defect. *Am Heart J.* 1984 Nov;108(5):1312-7. doi:10.1016/0002-8703(84)90759-2.
7. Samuels LE, Kaufman MS, Rodriguez-Vega J, Morris RJ, Brockman SK. Diagnosis and management of traumatic aorto-right ventricular fistulas. *Ann Thorac Surg.* 1998 Jan;65(1):288-92. doi:10.1016/s0003-4975(97)01084-9.
8. Rustad DG, Hopeman AR, Murr PC, Van Way CW. Aortocardiac fistula with aortic valve injury from penetrating trauma. *J Trauma.* 1986 Mar 1;26(3):266-270. doi:10.1097/00005373-198603000-00010.
9. Konda MK, Kalavakunta JK, Pratt JW, Martin D, Gupta V. Aorto-right Ventricular Fistula Following Percutaneous Transcatheter Aortic Valve Replacement: Case Report and Literature Review. *Heart Views.* Oct-Dec 2017;18(4):133-136. doi:10.4103/HEARTVIEWS.HEARTVIEWS_115_16.