

A Case of Gerbode Ventricular Septal Defect Endocarditis



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INTRODUCTION

Communication between the left ventricle and right atrium, termed a Gerbode ventricular septal defect (VSD), was first described in 1838 and later explained with varying etiologies, including congenital and acquired forms.¹⁻³ Among these etiologies, infective endocarditis is a rare cause, and echocardiography is a mainstay of its diagnosis and clinical management. Here we describe the case of a patient with bacterial endocarditis as a cause of a Gerbode VSD, with subsequent echocardiographic diagnosis and surgical repair.

CASE PRESENTATION

A 65-year-old man with a history of bacterial infective endocarditis in 1985, with no known prior treatment and subsequent resolution, who was being followed for aortic valve stenosis presented with worsening exertional dyspnea. Preoperative transthoracic echocardiography revealed a left ventricular–right atrial communication with a measured peak velocity of 5.3 m/sec (Video 1, time index 00:01), gradient of 113 mm Hg, and critical aortic stenosis with an aortic valve area of 0.4 cm² and a mean gradient of 88 mm Hg. The patient was subsequently taken to the operating room for aortic valve replacement and VSD closure. Intraoperative transesophageal echocardiography (TEE) confirmed the preoperative diagnosis of bicuspid aortic valve, with severe aortic valve stenosis and a left ventricular–right atrial communication (Video 1, time index 00:15), with a presumed etiology of the prior infective endocarditis. The patient underwent aortic valve replacement with a 21-mm Regent mechanical valve (St. Jude Medical, St. Paul, MN) and primary closure of a 2 × 2 mm septal defect using pledgeted sutures. The patient's postoperative course was uneventful, and he was discharged home on postoperative day 8.

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DISCUSSION

Communication between the left ventricle and right atrium was first described by Thurman¹ in 1838 and, later, with the successful surgical repair of five cases, by Gerbode *et al.*² in 1957. The congenital form of left ventricular–right atrial shunt has historically been more common, but acquired forms of this shunt have been increasingly reported. Acquired noniatrogenic etiologies include trauma, infective endocarditis, and inferior myocardial infarction. Iatrogenic etiologies include valvular surgery, atrial septal defect and VSD repair, percutaneous cardiac interventions such as atrioventricular node ablation, and endomyocardial biopsy.^{3,4}

Two classifications of left ventricular–right atrial communication have been described: infravalvular and supravulvular. This distinction depends on the supra- or infravalvular location of the membranous defect with respect to the tricuspid valve.⁵ Because the septal leaflet of the tricuspid valve will divide the membranous septum into inter-ventricular and atrioventricular portions, its insertion point in part determines the classification of these two subtypes.⁶ The congenital type originates in the interventricular membranous septum with a shunt existing between the left ventricle and the right ventricle and subsequently into the right atrium as a result of a defect in the septal leaflet of the tricuspid valve (type I, indirect; Figure 1B).⁷ In acquired forms, as in those resulting from infective endocarditis or iatrogenic causes, the communication exists between the left ventricle and right atrium superior to the intact septal leaflet of the tricuspid valve (type II, direct; Figure 1A).^{8,9}

TEE has been demonstrated to be superior to transthoracic echocardiography in the recognition of endocarditis vegetations and associated complications such as fistula and abscess formation.¹⁰ As such, every case of Gerbode VSD deemed or suspected to result from endocarditis as a primary etiology should warrant thorough TEE, examining all atrioventricular and semilunar valves in multiple views for potential vegetations. As was the case with our patient, no valvular pathology was noted outside of the previously mentioned aortic valve stenosis, which resulted from a congenital bicuspid aortic valve.

Care must also be taken to identify the defect with regard to the location of the communication, which can often be difficult to precisely determine. Three-dimensional (3D) color echocardiography through full-volume gated acquisition, which was used in our case (Video 1, time index 00:34), may provide significant aid in the accurate localization of these defects. Indeed, cases have been reported of the misinterpretation of Gerbode VSDs as pulmonary arterial hypertension in the setting of a high-velocity jet in the right atrium coupled with tricuspid regurgitation.¹¹ The direction of the jet and calculation of mean and diastolic pulmonary artery pressures from a pulmonary regurgitation jet can aid in distinguishing tricuspid regurgitation with pulmonary hypertension from a Gerbode VSD. A high continuous-wave Doppler gradient between the left ventricle and right atrium on echocardiogram is also one of the hallmarks of the Gerbode defect

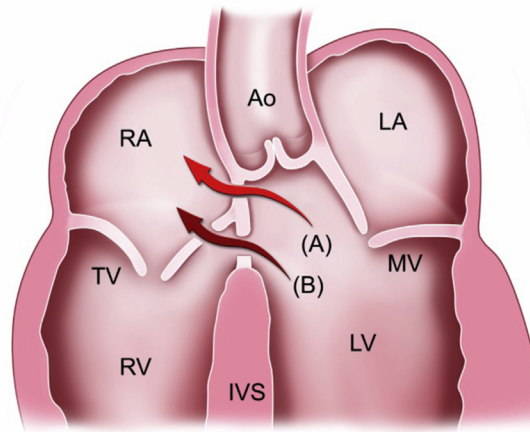


Figure 1 (A) Acquired type with the left ventricular and right atrial communication superior to the intact septal leaflet of the tricuspid valve (TV; type II, direct). (B) Congenital type with a communication existing between the left ventricle (LV) and right ventricle (RV) and a defect in the septal leaflet of the TV (type I, indirect). Ao, Aorta; IVS, interventricular septum; MV, mitral valve; RA, right atrium.

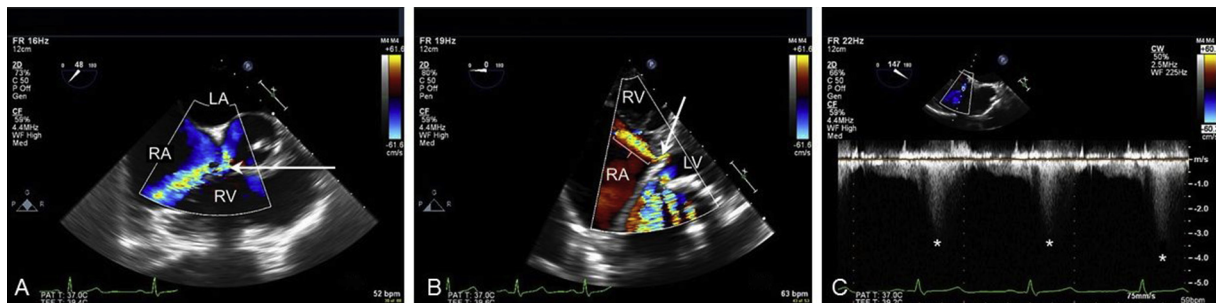


Figure 2 (A) Midesophageal right ventricular inflow-outflow view demonstrating turbulent flow from the left ventricular outflow tract into the right atrium (RA). The *arrow* denotes the origin of the Gerbode VSD. (B) Deep transgastric five-chamber view demonstrating flow originating from the left ventricle (LV) into the RA (above the tricuspid valve). The *arrow* denotes the origin of the Gerbode VSD. (C) Continuous-wave Doppler interrogation of the preoperative transthoracic subcostal four-chamber view demonstrating the velocity of the jet (*asterisk*) through the ventricular-atrial defect. LA, Left atrium.

(Figure 2).^{4,12,13} An additional distinguishing echocardiographic element to better delineate the Gerbode VSD is the occurrence of an enlarged right atrium with systolic expansion,² resulting from the shunt primarily occurring during systole because of the large pressure gradient favoring flow from the left ventricle to the right atrium. This is in contrast to a communication between the aorta and right atrium, which would have flow in both systole and diastole, peaking at end-systole.¹⁴ Perimembranous VSDs, which also result from a communication between the left and right ventricles, as with a type I Gerbode VSD, lack the right atrial communication and would not demonstrate this directional flow on color flow Doppler.

We recommend visualizing the ventricular-atrial shunt in multiple views, including the midesophageal right ventricular inflow-outflow view (60°), midesophageal four-chamber view (0°–30°), and deep transgastric long-axis view (0°–30°). TEE can further help characterize the defect through 3D planimetric measurement of the defect using postprocessing after 3D image acquisition. In our case, 3D imaging was used to help characterize the defect both with and without color (Video 1, time index 00:39), but planimetry was excluded because of the small size of the defect.

The magnitude of the shunt is the main predictor of long-term outcomes in this patient population¹⁵ and can be quantified through echocardiographic measurement and calculation of shunt

fraction (Qp-Qs) through determination of the ratio of pulmonary to systemic flow. In our case, Qp-Qs was not calculated, because of an inability to accurately measure the right ventricular stroke volume because of a lack of a parallel intercept angle while taking spectral Doppler measurements at the right ventricular outflow tract.

CONCLUSION

Gerbode VSDs are a rare defect, accounting for 0.08% of all congenital cardiac defects. Acquired Gerbode VSDs are increasing in incidence because of the increase in the number of cardiac surgical and percutaneous cardiac procedures as well as cases of infective endocarditis. Echocardiographic evaluation for this defect requires a high level of suspicion and careful interrogation and is aided by the advanced technique of 3D echocardiography.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2018.03.005>.

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