Case Report

Left Ventricular-to-right Atrial Shunt (Gerbode Defect) Associated with Interventricular Communication in a 42-year-old Patient

Shunt Ventrículo Esquerdo-Átrio Direito (Defeito de Gerbode) Associado à Comunicação Interventricular em Adulto

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Introduction

The Gerbode defect, a rare ventricular septal anomaly characterized by communication between the left ventricle (LV) and the right atrium (RA), represents approximately 0.08% of intracardiac shunts and less than 1% of congenital heart defects.[1] Although it classically has a congenital etiology, secondary causes of infective endocarditis, myocardial infarction (MI), and iatrogenesis have been reported.[2] The Gerbode defect was first described in 1857, and its first successful surgical treatment was reported by Kirby at the Hospital of the University of Pennsylvania.[3]

Here, we report an unusual case of congenital Gerbode defect in a 42-year-old patient whose diagnosis was challenging due to the presence of an additional shunt at the ventricular level.

Case report

A 42-year-old man from Jaraguá do Sul – Santa Catarina with a 3-month history of dyspnea on moderate effort was referred for a cardiological evaluation. A physical examination revealed jugular venous distension, a grade 4/6 systolic murmur at the left sternal border, and ascites. Electrocardiography demonstrated sinus rhythm and an inverted T-wave in the right precordial leads (V1–V4).

Transesophageal echocardiography and cardiac resonance imaging revealed slight perimembranous interventricular communication (IVC) with a high mean gradient but no clear signs of right chamber overload.

During follow-up, the patient developed right-sided heart failure that was refractory to conventional treatment with massive ascites that required two hospitalizations for paracentesis. Because of the clinical–imaging finding discrepancy, he underwent right heart catheterization, which revealed a significant oxygen saturation step-up in the right ventricle suggestive of a ventricular left-to-right shunt, estimated Qp/Qs of 2.1, moderate pulmonary hypertension, and signs of right ventricular dysfunction. Transthoracic echocardiography demonstrated a 7-mm turbulent jet from the LV outflow tract to the RA compatible with the Gerbode defect (Figure 1).

The defect was repaired via a median sternotomy using cardiopulmonary bypass, and the right chamber was accessed via right atriotomy. A 10-mm LV to RA communication was visualized and characterized as a Gerbode defect (Figure 2) as well as a 3-mm IVC next to the RV outflow tract. A suture was placed to correct both communications. The patient was discharged from the hospital on the seventh postoperative day, and significant symptom improvement was noted during follow-up.

Discussion

Gerbode defects can be defined as abnormal communications between the LV and the RA. This is anatomically possible because the normal tricuspid valve is more apically displaced than the mitral valve. Congenital LV-RA shunt is rare, representing 0.08% of all congenital cardiac anomalies.[1] In contrast, acquired LV-RA communication can be caused by iatrogenic mechanisms such as cardiac surgeries or percutaneous interventions performed close to the membranous RV septum and non-iatrogenic mechanisms such as endocarditis and acute MI.[2,5] In a review by Yuan et al., congenital etiologies represented 26.4% of cases, whereas the acquired form accounted for 72.7%.[2] Nevertheless, it is assumed that acquired forms are more often reported in scientific journals than congenital forms.[6] In our case, the defect was presumably congenital since the patient had no previous history of surgery, MI, or endocarditis. In addition, the presence of an associated interventricular shunt supported the congenital etiology.

Gerbode first categorized this pathology as type I (indirect form) and type II (direct form). In the indirect form, the shunt occurs from the LV to the right ventricle and then flows through a defect in the tricuspid valve into the RA. Thus, the communication occurs below the tricuspid valve. In contrast, in the direct form, the shunt occurs above the tricuspid valve and directly connects the LV to the RA.[1,7] This terminology was later modified to infravalvular and supravalvular to describe shunt location based on its relationship to the tricuspid valve.[8] Finally, Sakakibara and Konno included a third type (intermediate), which has infra- and supravalvular components.[8]

Keywords

Congenital heart defects; Ventricular septal defects; Heart Septal Defects, Ventricular.

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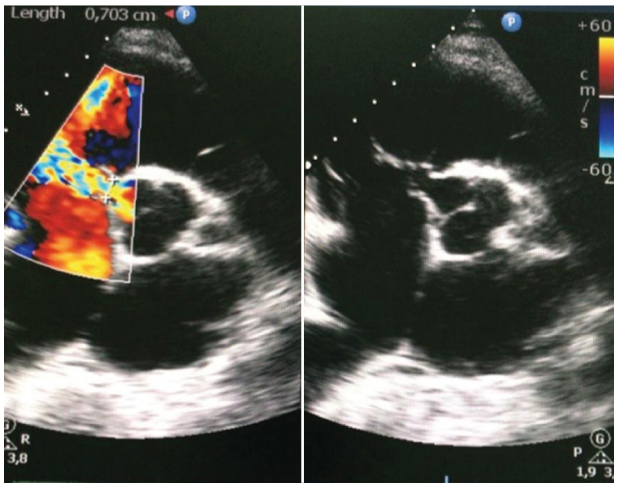

Figure 1 – *Color flow Doppler image from the parasternal transverse heart base view showing flow from the left ventricle to right atrium (left image).*

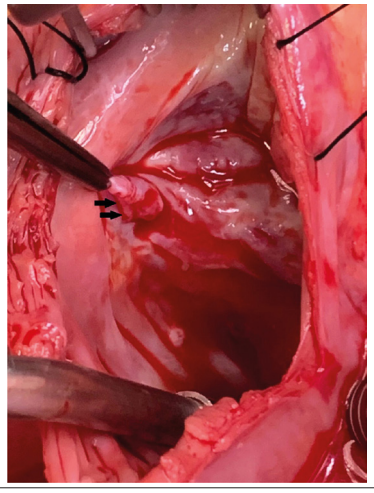

Figure 2 – *Right atrial view during the surgical procedure. The black arrows indicate the Gerbode defect.*

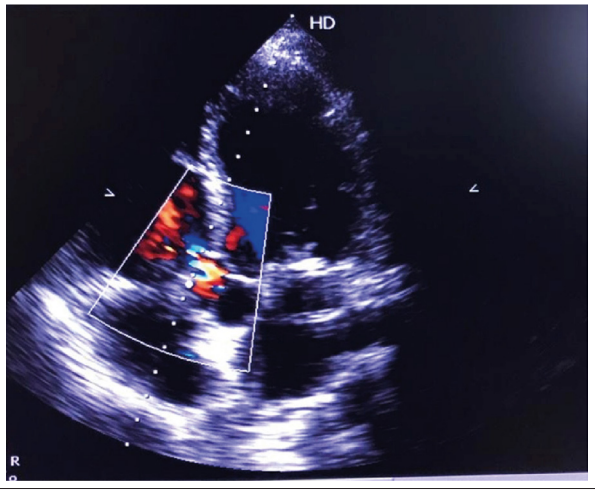

Figure 3 – *Color flow Doppler from 5 chamber apical view showing a perimembranous interventricular communication.*

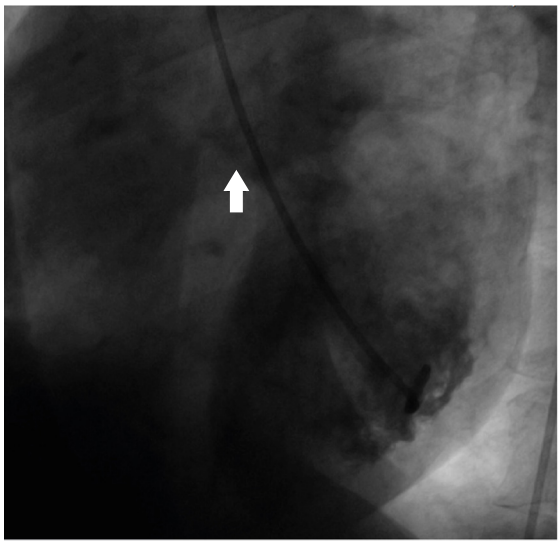

Figure 4 – *Cardiac ventriculography showing a communication at the level of the interventricular septum.*

In our patient, we found two defects that caused a left-to-right shunt: an LV-RA true communication (type II or supravalvular) and an IVC. There were no tricuspid valve abnormalities that could cause LV-RA indirect communication or characterize an infravalvular component. According to the literature, cardiac anomalies related to the Gerbode defect occur in approximately 1/3 of cases; among them, atrial septal defects are the most common.[9] Congenital abnormalities are more often associated with additional cardiac abnormalities than are acquired forms.[2]

Making the LV-RA communication diagnosis is always challenging due to its rarity and the limited diagnostic accuracy of tests such as transthoracic echocardiography.[2,6] However, some echocardiographic findings, such as a normal pulmonary arterial end-diastolic pressure, RA enlargement, leftward bowing of the interatrial septum, an atypical jet direction, and a high Doppler gradient, could suggest the defect.[9,10] The correlation of echocardiographic results among them can also be useful since they can present discrepant findings that are suggestive of Gerbode defect.[10]

The most accurate tests for making the Gerbode defect diagnosis are cardiac catheterization and transesophageal echocardiography.[1,2] In our case, the diagnosis of LV-RA communication was possibly hindered by the coexistence of a concomitant intracardiac shunt. In right heart catheterization, the associated IVC produced an oxygen saturation step-up in the right ventricle that did not help identify the shunt location, although the flow and pressure assessments were useful for confirming the hemodynamic repercussions.

Cardiac magnetic resonance can reveal further anatomical details of the defect and provide information such as accurate left and right heart volume and intracavitary flow measurements.[1] In our case, it failed to discriminate between IVC and LV-RA direct communication.

The need for treatment of Gerbode defect depends on several factors, including symptom severity, shunt magnitude, development time, concomitant anatomical abnormalities, and comorbidities. Small, chronic, and asymptomatic defects, with no signs of circulatory overload, can be managed conservatively, although some authors suggest that all LV-RA defects be repaired to prevent infective endocarditis since surgical treatment demonstrates excellent results.

Conclusion

Gerbode defect is a rare ventricular septal anomaly that results in LV-RA communication. Making this diagnosis is always challenging, and diagnostic delays may lead to worsening of the patient's clinical status. To prevent mistakes, it is important to raise the suspicion of this cardiac defect when faced with echocardiographic findings such as atypical jet direction and normal diastolic pulmonary arterial pressure. In our case, the coexistence of a shunt at the ventricular level and the non-elucidating complementary test results made the diagnosis more difficult, but the defect was repaired successfully and the clinical outcome was favorable.

Authors' contributions

Study concept and design: Takagui ASM; Baggio TC; Obtaining data: Savariz MB; Theilacker G; Baggio TE; data analysis and interpretation: Takagui ASM; Baggio TC; manuscript drafting: Savariz MB; Theilacker G; Baggio TE; and critical review of the manuscript for important intellectual content: Takagui ASM, Baggio TC.

Conflict of interest

The authors have declared that they have no conflict of interest.

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