Mechanical hemolysis secondary to percutaneous repair of an iatrogenic Gerbode defect with the Amplatzer™ Septal Occluder

Hemólise mecânica secundária à correção percutânea de um defeito de Gerbode iatrogênico com Amplatzer™ Septal Occluder

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ABSTRACT – The Gerbode defect is defined as an abnormal communication between the left ventricle and the right atrium, and is etiologically classified as congenital or acquired (iatrogenic or not). The typical treatment consists of surgical repair of the shunt, but transcatheter occlusion of this condition has proven to be a safe and effective therapeutic alternative for such patients, especially for those with prior surgeries. The aim of this study was to report a case of transcatheter closure of an acquired Gerbode defect, using the Amplatzer™ Septal Occluder device, in a 58-year-old patient, with two prior mitral valve replacements, and the consequent post-procedure mechanical hemolysis.

Keywords: Heart septal defects, ventricular; Heart ventricles/abnormalities; Heart atria/abnormalities; Septal occluder device; Treatment outcome

RESUMO – O defeito de Gerbode é definido como uma comunicação anormal entre o ventrículo esquerdo e o átrio direito, sendo classificado etiológicamente como congênito ou adquirido (iatrogênico ou não). O tratamento típico consiste na correção cirúrgica do *shunt*, mas a oclusão transcatheter dessa patologia vem se mostrando uma alternativa terapêutica segura e eficaz para tais pacientes, especialmente para aqueles com abordagens cirúrgicas prévias. O objetivo deste estudo foi relatar um caso de fechamento transcatheter de um defeito de Gerbode adquirido utilizando o dispositivo Amplatzer™ Septal Occluder em paciente de 58 anos, com duas trocas valvares mitrais prévias, e a consequente hemólise mecânica pós-procedimento.

Descritores: Comunicação interventricular; Ventriculos do coração/anormalidades; Átrios do coração/anormalidades; Dispositivo para oclusão septal; Resultado do tratamento

INTRODUCTION

The Gerbode defect is defined as an abnormal communication between the left ventricle (LV) and the right atrium (RA), and may be congenital or acquired. When acquired, it is categorized as iatrogenic or non-iatrogenic, the former being an uncommon complication of surgeries performed in the proximity of the atrioventricular (AV) septum.¹ This rare anomaly accounts for only 0.08% of intracardiac communications and <1% of all congenital heart defects;²⁻⁴ however, acquired LV-RA communication has been increasingly reported.^{5,6}

Anatomically, the position of the defect in relation to the tricuspid valve is described, and then classified as supravulvar, infravulvar, or intermediate.^{7,8} The clinical manifestation varies from asymptomatic to severe heart failure and even death, depending on the magnitude of the regurgitant volume, the duration of the LV-RA communication, and the presence of other cardiac anomalies.⁹ Most patients are symptomatic, with dyspnea and fever as the main clinical symptoms.⁵

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Two-dimensional transesophageal echocardiography (TEE) is the most sensitive method to detect LV-RA communication, but it has limitations.⁵ Three-dimensional echocardiography is the most appropriate for anatomical location of the anomaly.^{2,7,10} Other complementary methods may be used, such as magnetic resonance imaging and cardiac catheterization, the latter providing a greater number of preoperative diagnoses.^{2,11} The appropriate treatment depends on the severity of symptoms, and the usual approach is the conventional surgical correction of congenital or acquired LV-RA communications.¹ The surgical technique consists of suturing a patch on the atrial side of the defect, to avoid recurrence and/or complications. However, given the advances in interventional therapies, the percutaneous approach to such defects has become increasingly relevant. The first report of a percutaneous closure of an acquired Gerbode-type defect was performed in 2006, by Trehan et al.,¹² and since then, similar reports have been made,^{5,12} with the experience also expanding to congenital etiology.¹³ So far, percutaneous management has shown favorable results and a lower incidence of complications.

The aim of this study was to report a case of transcatheter closure of an acquired Gerbode defect using the Amplatzer™ Septal Occluder device in a patient with two prior mitral valve replacements, and the consequent post-procedure mechanical hemolysis. This study was evaluated and approved by the Research Ethics Committee of the *Instituto Estadual de Cardiologia Aloysio de Castro*, with protocol 5.053.596, CAAE 52628421.5.0000.5265.

CASE REPORT

A 58-year-old male patient with a history of rheumatic fever and two prior surgeries for mitral valve replacement; the first for placement of a biological prosthesis in 1993, and the second for placement of a metallic prosthesis, in 2012. In addition, a total atrioventricular block and consequent placement of permanent pacemaker, in 2018. He presented with progressive heart failure in New York Heart Association (NYHA) functional class II, refractory to drug treatment. Clinical examination revealed biventricular enlargement and holosystolic murmur 2+/6+ over the left sternal border, irradiating to the dorsum, clavicles, and right carotid artery. The TEE showed a septal defect between the LV and RA with a significant shunt from left to right, dilation of the four heart chambers, preserved LV systolic function, significant dysfunction of the right ventricle with systolic pressure in the pulmonary artery of 45mmHg, in addition to metallic prosthesis; the other functions were normal.

The indication and therapeutic approach were discussed by the multidisciplinary Heart Team, excluding open heart surgery, since it would be a third approach in a patient with limited clinical conditions and moderate surgical risk (EuroSCORE II of 6.08%). A percutaneous approach was chosen for the occlusion of the acquired Gerbode defect, and the procedure was planned by analyzing the TEE images. The procedure was performed under general anesthesia

and guided by TEE. Arterial access was established by left femoral approach with a 6F introducer. With an ARII angiographic catheter and a 0.035"x260cm Roadrunner hydrophilic guidewire (Cook Medical, Bloomington, Indiana, United States), the LV, and then, the RA were retrogradely accessed through the defect. With a 6F introducer positioned in the right femoral vein, a snare was advanced to the RA, and an arteriovenous circuit was established after looping the hydrophilic guide across the defect. Guided by periprocedural TEE images, the Amplatzer™ Septal Occluder (AGA Medical Corporation, Minnesota, United States) device size of 14mm was defined. Through the AV circuit, the LV was accessed from the RA as a delivery system. The prosthesis was released by exteriorizing the left disc (LV) and then releasing the right disc in the membranous septum portion of the RA, achieving occlusion of the Gerbode defect with a small residual transprosthetic shunt (Figure 1 and Videos 1 and 2).

LA: left atrium; LV: left ventricle; RV: right ventricle; RA: right atrium.

Figure 1. Schematic representation demonstrating Gerbode defect, the arteriovenous circuit, and right atrium-left ventricle flow Doppler. (1) Red line: pacemaker electrode previously implanted. (2) Blue line: hydrophilic guide positioned in the anatomical defect, from the left femoral artery, through the aorta, left ventricle, Gerbode defect, right atrium, and then the atrioventricular circuit is completed with the exteriorization of the guide through the right femoral vein. (3) Schematic representation of the metallic valve prosthesis in mitral position. (4) Arrow showing Gerbode defect. (5) Flow through the defect.

After the procedure, the patient was admitted to intensive care and extubated with no complications. During hospitalization, he developed severe hemolytic anemia, requiring blood transfusions, and significant renal dysfunction, requiring dialysis. A control TEE was performed 60 days after the procedure, which showed good positioning of the prosthesis and minimal residual shunt (Figure 2). After 63 days of hospitalization, the patient was discharged, asymptomatic, and in NYHA class I (Video 3).

Video 1. Post-implant fluoroscopy showing residual transprosthetic shunt.

LA: left atrium; LV: left ventricle; RV: right ventricle; RA: right atrium.

Figure 2. Schematic representation demonstrating Gerbode defect occlusion, Amplatzer™ Septal Occluder, and post-procedure right atrium-left ventricle flow Doppler. (1) Red line: previously implanted pacemaker lead. (2) Blue line: pigtail catheter positioned in the left ventricle. (3) Schematic representation of the metallic valve prosthesis in mitral position. (4) Schematic representation of the atrial septal defect prosthesis implanted in the Gerbode defect. (5) Doppler after defect closure.

Video 2. Transesophageal echocardiogram performed 60 days after the procedure, demonstrating reduction of the atrioventricular shunt.

Video 3. Intraoperative transesophageal echocardiogram demonstrating transprosthetic shunt.

DISCUSSION

The Gerbode defect, especially the acquired type, is an extremely rare anomaly.⁹ Acquired non-iatrogenic defects occur secondarily to acute myocardial infarction in right coronary territory, infectious endocarditis, and blunt cardiac trauma, while the iatrogenic types affect patients with previous history of cardiac surgery (mainly aortic or mitral valve replacement, and correction of atrial or ventricular septal defects) or percutaneous cardiac interventions.¹ Thus, as demonstrated by Sinisalo et al.,⁹ and reinforced by the case reported here, mitral valve replacement is one of the main causes of LV-RA communication. The probable mechanism is secondary to some injury to the membranous septum tissue by excessive removal of calcium from the mitral annulus during valve replacement.

Most patients who have the need for correction of iatrogenic LV-RA need to undergo one more cardiac surgery and, therefore, all the risks associated with a reoperation. Thus, the percutaneous approach presents itself as a feasible and safe option, especially in those patients with high surgical risks. However, despite the fact that percutaneous closure of the Gerbode defect has been performed for almost two decades, there are still no formal recommendations or specific devices aimed at the management of these patients. To date, the use of atrial septal occluders, ventricular septal occluders, ductus arteriosus, and Amplatzer™ II vascular plugs have been reported with satisfactory results.^{9,12}

The probable pathophysiological mechanism of hemolysis in these cases is due to the high velocity jets, especially in smaller devices, which provide conditions for cell lysis. Following this rationale, larger devices with more wires in their construction, such as the interventricular communication occluder devices, could provide fewer incidences of shunts and, consequently, less hemolysis. Despite the hemolysis and the transprosthetic flow initially seen, this ceased after a probable endothelialization of the prosthesis, similarly to what occurs in cases of implant complications in other positions. Although massive hemolysis is a high-risk situation with high morbidity, it may only be transient in these cases.

The percutaneous approach to the acquired Gerbode defect is resolvable by adapting various devices already available, and presents itself as a feasible and safe alternative to surgical treatment. Because it is a rare disease and the consequent small number of reports, the task of defining the management and the ideal device becomes challenging. Despite the applicability and use of atrial septal defect occluders have been reported, and the hemolysis presented by the patient was only transient, the use of non-dedicated devices will always be accompanied by a higher probability of therapeutic failure in the medium- to long-term. Thus, it is worth considering the need to define a device that best adapts to this situation or the development of a specific device. Moreover, it should be noted that, due to

the plurality of presentations and etiologies of the Gerbode defect, the transcatheter approach is not always the best management. Percutaneous intervention in patients with more complex anatomies, such as infravalvular defects with multiple openings or larger orifices, may not be favorable, and patient selection and individualization of treatment are essential for the management of this rare disease.

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None.

DECLARATION OF CONFLICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: MJM, EFQ and LH; data collection: LH and VN; data interpretation: LH and VN; text writing: LH; approval of the final version to be published: MJM, EFQ, LH, MOL and AF.

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