Case Report

Percutaneous Closure of Multiple Atrial Septal Defects with Hemodynamic Repercussions: Report of Two Cases in First-Degree Related Adult Patients

Fechamento Percutâneo de Comunicação Interatrial Múltipla com Repercussão Hemodinâmica: Relato de Dois Casos em Pacientes Adultos Parentes de Primeiro Grau

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Introduction

Atrial septal defects (ASD) account for 7–10% of congenital heart anomalies.[1,2] ASD are classified as three anatomical types: ostium secundum (OS) (70% of cases), ostium primum (15–20% of cases), and sinus venosus (5–10% of cases) defects. OS involves the fossa ovalis region and results from poor embryological development of septum secundum or excessive absorption of septum primum resulting in the lack of a septum in the fossa ovalis.[1]

Affected patients are usually asymptomatic. Exercise intolerance in the form of dyspnea or fatigue is the most common initial presentation.[1] Echocardiography is a fundamental tool in the diagnosis and management of patients with ASD. Transthoracic echocardiography (TTE) identifies ASD type and size, shunt direction, and the presence of anomalous sinus venous drainage. Three-dimensional transesophageal echocardiography (3DTEE) details the morphology and anatomical relationships of the atrial septum, providing fundamental information for choosing treatment.[1,3]

Based on European guidelines for the diagnosis and treatment of congenital heart diseases, the indications for ASD closure are the presence of atrial and/or right ventricular (RV) dilatation on echocardiography, magnetic resonance imaging (MRI), or computed tomography with no severe pulmonary hypertension and presenting one or more of the following: minimum 10-mm ASD diameter on echocardiography, or Qp/Qs ratio > 1.5:1 measured by echocardiography or contrast-enhanced MRI (class I recommendation; level of evidence, B). Sinus venosus, coronary sinus, or ostium primum ASD must be surgically repaired.[3]

ASD with hemodynamic repercussions should be closed to improve right chamber size, decrease pulmonary arterial pressure, and improve symptoms of reduced functional limitation.[3] Several hospital services implemented the percutaneous closure of OS ASD due to shorter hospitalization time and fewer complications than surgical treatment. Thus, this study aimed to report the presence of OS ASD with hemodynamic repercussions in two first-degree related adults undergoing percutaneous treatment.

The study was approved by the Research Ethics Committee of Hospital de Urgências de Goiânia (CAAE number 85497418.2.0000.0033).

Case 1

RCAS, a 41-year-old man, asymptomatic from the cardiovascular point of view, underwent TTE on February 20, 2018, which showed a patent foramen ovale (PFO) with large passage of microbubbles and a large interatrial septum aneurysm associated with ASD with hemodynamic repercussions (Qp: QS 2 and moderate right chamber enlargement).

On April 6, 2018, a 3DTEE was performed for the morphological and functional assessment of the ASD closure procedure. A multi-fenestrated ASD was visualized in a predominantly posteroinferior location of the septum and PFO (Figures 1A, 1B). The three-dimensional image was fundamental in choosing the device to be implanted, as it showed the reconstructed image of the interatrial septum in which each fenestration and edge was demonstrated in relation to the adjacent structures.

Thus, the multi-fenestrated ASD was occluded with the PFO from the successful implantation of two Amplatzer ASD® cribriform prostheses of 30 and 25 mm without clinical or angiographic complications or residual shunt (Figures 2A, 2B).

The patient's condition progressed without clinical or hemodynamic complications in the immediate postoperative period, and he was discharged on April 8, 2018. He returned to the service on September 1, 2021, for a late follow-up 3DTEE, which showed an intact interatrial and interventricular septum, well-positioned occluding devices in the topography of the interatrial septum, and no residual shunt after an agitated saline injection (microbubble test) (Figures 3A, 3B).

Case 2

RPASC was a 39-year-old woman with dyspnea on minimal exertion and slowly progressing exercise intolerance.

Keywords

Heart Septal Defects, Atrial; Echocardiography; Hemodynamics.

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Manuscript received 9/1/2022; revised 9/30/2022; accepted 10/30/2022

Editor responsible for the review: Daniela do Carmo Rassi Frota

DOI: 10.47593/2675-312X/20223504eabc345i

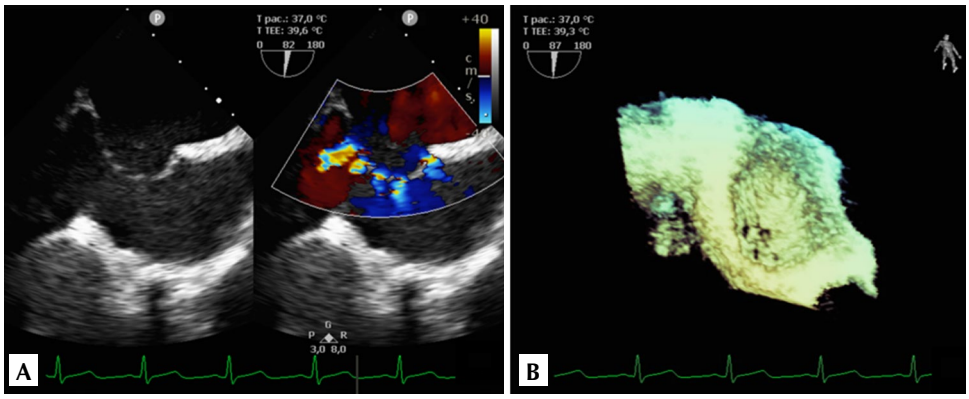


Figure 1 – *Transthoracic echocardiography pre-percutaneous closure procedure performed on April 6, 2018. A. Two-dimensional image of a multi-fenestrated atrial septal defect (ASD; five fenestrations). B. Three-dimensional image of a multi-fenestrated ASD.*

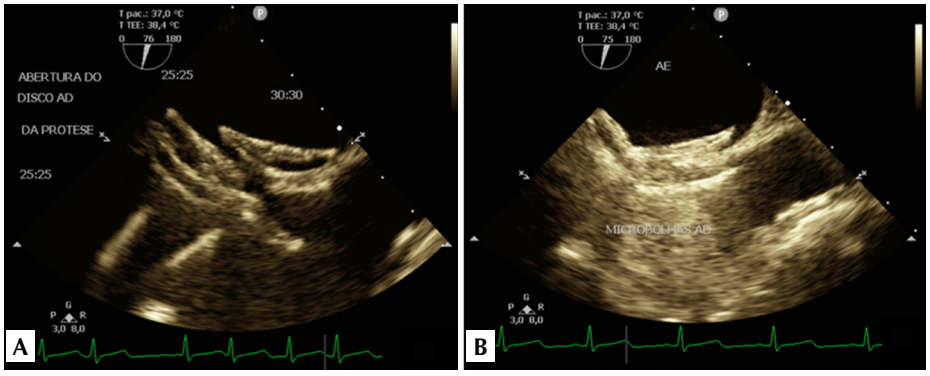


Figure 2 – *Percutaneous procedure for closing a multi-fenestrated atrial septal defect (ASD) with patent foramen ovale on April 6, 2018. A. The implantation of two Amplatzer ASD cribriform prostheses (30 and 25 mm). B. Microbubble test showing no residual shunt.*

On November 10, 2021, a TTE showed slight RV enlargement with preserved systolic function (S', 10 cm/s; tricuspid annular plane systolic excursion, 19 mm; fractional area change, 42%; no indirect signs of pulmonary hypertension pulmonary artery trunk, 21 mm in diameter; inferior vena cava (IVC), 12 mm in diameter; respiratory variability, greater than 50%; and estimated pulmonary arterial systolic pressure, 24 mmHg]), and the presence of two OS ASD, one more anterior measuring 18 × 9 mm and another closer to the IVC measuring 6 × 5 mm, with hemodynamic repercussions. The estimated Qp/Qs was 1.7 with septal borders greater than 5 mm. A 3DTEE performed in another service showed two OS ASD measuring 12 × 14 mm and 4 × 4.3 mm and 12 mm apart with hemodynamic repercussions.

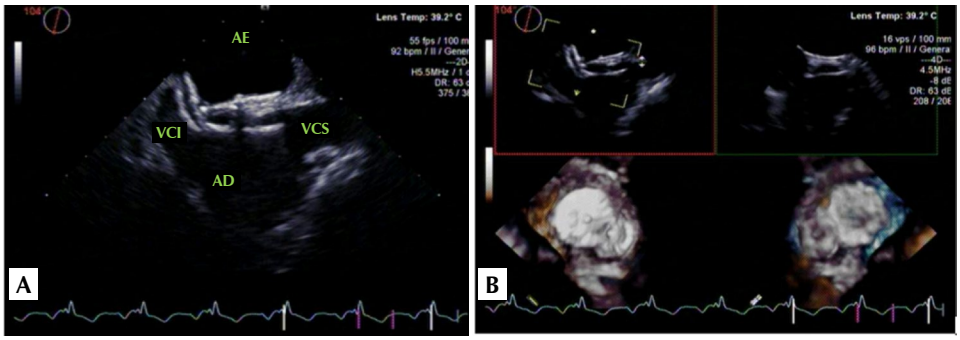
Therefore, percutaneous treatment was indicated due to the presence of an ASD with hemodynamic repercussions (right chamber dilation, Qp/Qs > 1.5).

On November 11, 2021, the patient was taken to the hemodynamics service for intraoperative 3DTEE, which revealed two ASD, the larger one 15 mm and the smaller one 4.3 mm, separated by 14 mm of septal tissue, with moderate hemodynamic repercussion (Qp/Qs, 2) and no other heart diseases (Figures 4A–4C). Thus, the intracardiac ASD was percutaneously closed with two ASD® Amplatzer prostheses (one 24 mm, the other 6 mm) in the major and minor ASD, respectively, guided by 3DTEE without residual shunt or clinical or angiographic complications (Figures 5A, 5B).

The patient's condition progressed with significant clinical improvement in the immediate postoperative period and no hemorrhagic or hemodynamic complications. She returned for a control TTE on December 11, 2021, which showed two occluding devices in the interatrial septum and no residual shunt (Figures 6A, 6B).

Discussion

The decision to repair any type of ASD is based on clinical and echocardiographic information, including signs and symptoms of right heart failure, defect size and location, magnitude and hemodynamic impacts of the left-to-right shunt, and the presence and degree of pulmonary hypertension. Elective closure is recommended for all ASD cases with echocardiographic evidence of RV overload or a clinically significant Qp/Qs ratio greater than 1.5.[4]



AE: left atrial; VCS: superior vena cava; AD: right atrium; VCI: inferior vena cava.

Figure 3 – *Echocardiogram performed on September 1, 2021, in the late postoperative period of percutaneous closure of the multi-fenestrated atrial septal defect. A. Two-dimensional image of the occluding device in the topography of the interatrial septum. B. Three-dimensional image of the prosthesis.*

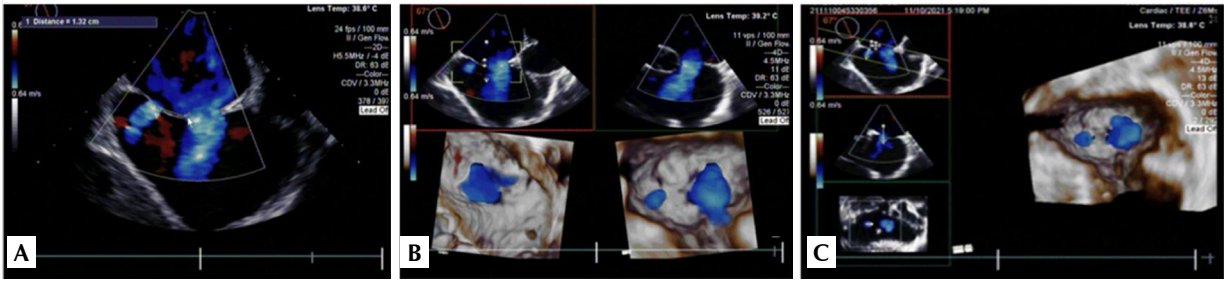


Figure 4 – *Three-dimensional transesophageal echocardiogram taken on November 11, 2021, showing an ostium secundum atrial septal defect (2 fenestrations) with hemodynamic repercussions.*

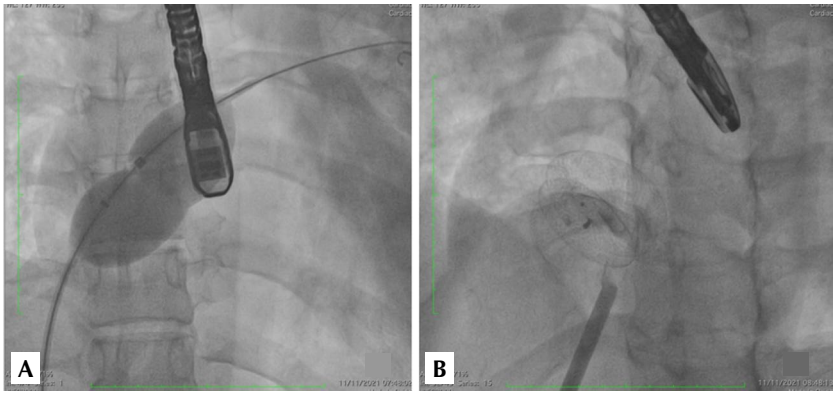
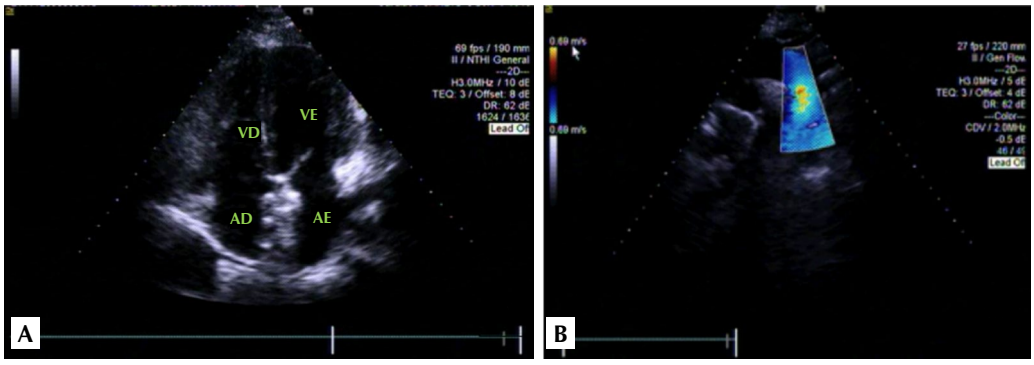


Figure 5 – *Percutaneous closure of two atrial septal defects (ASD) with Amplatzer ASD prostheses on November 11, 2021. A. Balloon sizing. B. Final result.*

Current criteria for the percutaneous treatment of OS ASD include a defect size ≤ 32 mm and a 5-mm rim of atrial septal tissue around the defect, flow through the defect predominantly directed from the left to right atrium, presence of signs of right chamber volumetric overload (increased RV diameter, presence of paradoxical movement in the interventricular septum), no fixed pulmonary hypertension, and no associated defects requiring surgery.[4]

Contraindications to percutaneous treatment include cases in which the defect is small enough to not generate hemodynamic repercussions or severe pulmonary arterial hypertension with indexed pulmonary vascular resistance above 5.0 w.m^2 refractory to pulmonary vasodilator testing.[5]

Interatrial communication has a complex anatomy when presenting the following characteristics: stretched diameter greater than 26 mm; deficient borders measuring less than 4 mm in the anterior, posterior, or inferior region of the septum; two distant fenestrations; multi-fenestrated interatrial



VE: Left ventricle; AE: left atrial; AD: right atrium; VD: right ventricular.

Figure 6 – *Transthoracic echocardiogram taken on November 12, 2021, showing two occluding devices in the interatrial septum (A) and no residual shunt (B).*

septum and interatrial septal aneurysm (redundant and mobile interatrial septum with an excursion greater than 10 mm).[7]

The presence of two or more defects detectable on 3DTEE and distant from one another is amenable to a transcatheter approach since two or more devices can be safely and efficiently implanted simultaneously. In such situations, it is important to individually assess each of the defects. Very small defects (2–3 mm) less than 5 mm away from the largest OS ASD can be indirectly occluded by being covered by the disk of the implanted prosthesis. Multiple small OS ASD (multi-fenestrated septum), provided they are in a region with a diameter of 30–35 mm, can also be percutaneously treated through the implantation of a prosthesis specifically for this purpose.[5]

This prosthesis must have a diameter 20–30% greater than the basal ASD diameter. In cases of multi-fenestrated ASD, the prosthesis must be specific for this type of defect, with two retention disks joined by a thin waist. The device must be implanted through the most central fenestration to allow its disks cover the surrounding fenestrations.[5]

In this context, TTE provides evidence of the shunt through the interatrial septum, cardiac chamber size, RV function, Qp/Qs calculation, and pulmonary pressure estimation, as an insufficient left atrial size can prevent percutaneous ASD closure.[6]

Interestingly, 3DTEE is performed using a sizing catheter to determine the specific anatomical properties of the ASD such as size, location, adequate borders (aortic, mitral, and superior, inferior, and posterior vena cava) and defect relation with adjacent heart structures. In addition, during the procedure, it enables visualizing of the catheters, guiding them through the defect. Color flow Doppler allows defect sizing and helps selecting the proper device size for implantation, confirms the correct device position, evaluates and grades the presence of residual leaks, when possible, checks the need for a second device, and identifies the presence of surgical complications.[6]

The advantages of 3DTEE in the study of ASD include the clear demonstration of the characteristics of the interatrial septum and its defects with special visualization of the extension of the anterosuperior border that established the defect shape and fenestration size. This imaging modality allows an *en face* communication analysis that mimics the surgeon's view with the advantage of being dynamic during the cardiac cycle. After the prosthesis implantation, 3D reconstruction can help identify device position, demonstrate and locate the arms or disks protruding into the right atrium. This method can also be used to assess and measure residual defects.[7]

This procedure presents a low complication rate. Serious complications such as malposition and subsequent surgery occur at a frequency of 1–5%, device embolization in 0.4–4%, stroke in 0.1–0.3%, cardiac tamponade in 0.1%, cardiac perforation in 0.03%, and endocarditis in 0.03%.[4]

An interesting fact in our report was the presence of ASD in non-syndromic siblings. The literature shows reports of patients with OS ASD with a positive family history of this defect or other congenital heart malformations and co-existing heart block. Although an autosomal dominant mode of inheritance for familial ASD was described in some families, the incidence of this defect in these families is many times lower than expected for single-gene defects. Therefore, a multifactorial mode of inheritance has been postulated due to the presence of clinical phenotype diversity combined with generational leaps.[8] According to Benson et al., the familial ASD locus is located in the telomeric region of chromosome 5p. It is a genetically heterogeneous disorder of incomplete penetrance and variable expressivity that results in the significant underestimation of the hereditary nature (multifactorial etiology with complex polygenic interaction and environmental factors) of this condition. The author also suggests that the identification of ASD or other congenital heart defects (such as interatrial septal aneurysm, persistent left superior vena cava, bicuspid aortic valve, which occur in 0.5–1% of cases) in more than one family member should lead to the clinical and genetic evaluation of their relatives. Furthermore, the author emphasized that the identification of other familial genes of ASD should provide new insights about important cardiac morphogenesis steps that lead to atrial septation.[9]

Conclusion

ASD are frequent congenital heart diseases, and a positive family history of this defect should prompt the

investigation of other cardiac anomalies in the index case in addition to the investigation of first-degree relatives. OS ASD occlusion by percutaneous implantation is a safe and effective alternative to traditional surgical closure, but the procedure is not risk-free and must be performed in specialized centers by trained operators. In this context, we highlight the role of echocardiography given the diversity of important data obtained in the pre-, intra-, and postoperative periods of ASD correction as well as in the late follow-up of these patients.

Authors' contributions

Research conception and design: Lopes MP; data collection and manuscript writing: Rocha DFR; critical review of the manuscript for intellectual content: Guimarães HL, Silva JBM and Gardenghi G.

Conflict of interest

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

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