Spontaneous Closure of Perimembranous Ventricular Septal Defects: A Janus-Faced Condition

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INTRODUCTION

A ventricular septal defect (VSD) is a communication between the left and right ventricles resulting in shunt flow and is the most common isolated congenital heart defect.[1] Depending on localization, size, and shunt flow, VSD closure may be necessary. Spontaneous closure of muscular VSDs is frequently seen in early childhood, whereas spontaneous closure of a perimembranous VSD is more rare, especially when >4 mm.[2] Nevertheless, in some cases even large defects may close spontaneously by an aneurysm, most frequently originating from tissue of the tricuspid valve's septal leaflet. This is usually a pleasant finding from a pediatric cardiologist's point of view, as imminent surgery will fortunately no longer be necessary.

Recently, there has been much discussion of whether patients with VSDs require ongoing observation during adulthood, if the shunt is closed during childhood (regardless of whether surgically, interventionally, or spontaneously), when there is no residual shunt or history of arrhythmia.[3,4] This case presentation underlines the fact that these patients cannot be regarded as cured but rather need lifelong observation.

CASE PRESENTATION

We present the case of a 29-year-old woman of Moroccan origin who migrated to Germany 1 year before presentation. She was referred to our center with a recent history of exertional dyspnea, fatigue, and vertigo and a newly diagnosed systolic murmur. Previously her medical history had been unremarkable. She had no documented history of congenital heart disease, and she had given birth to three healthy children. Physical examination revealed a 5/6 hissing systolic murmur. She was acyanotic and in good overall condition. Laboratory values, including N-terminal pro–brain natriuretic peptide, were normal. Electrocardiography showed pathologic right-axis deviation.

Transthoracic echocardiography revealed a large perimembranous VSD that was fully covered by an interventricular septal aneurysm (Figures 1A and 1B, Videos 1-3). There was no detectable residual left-to-right shunt. Doppler echocardiography within the right ventricular outflow tract revealed relevant flow acceleration (Figure 2).

Significant obstruction of the right ventricular outflow tract induced by the septal aneurysm was confirmed by transesophageal echocardiography (Videos 4 and 5) and cardiac magnetic resonance imaging (Figure 3, Videos 6 and 7).

In addition, cardiac catheterization was performed to rule out significant pulmonary hypertension and coronary heart disease. On catheterization, the invasively measured intraventricular peak-to-peak gradient was 76 mm Hg. Absence of residual shunt flow was confirmed by angiography.

Subsequently the patient underwent cardiac surgery. Using cardiopulmonary bypass, the right atrium of the cardioplegic heart was opened. Through the tricuspid valve, the aneurysm was

VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiography showing the large VSD and the septal aneurysm bulging into the right ventricular outflow tract in the parasternal long-axis view.
Video 2: Transthoracic echocardiography showing the large VSD and the septal aneurysm bulging into the right ventricular outflow tract in the parasternal short-axis view.
Video 3: Color flow Doppler reveals severe right ventricular outflow tract obstruction due to the aneurysm.
Video 4: Transesophageal echocardiography (modified transgastric long-axis view) revealing severe right ventricular outflow tract obstruction due to the aneurysm.
Video 5: Color Doppler of right ventricular outflow tract obstruction due to the aneurysm.
Video 6: Cine magnetic resonance imaging (MRI; balanced steady-state free precision [SSFP] sequence) with sweep from tricuspid valve plane to the right ventricular (RV) apex.
Video 7: Cine steady-state free precession magnetic resonance imaging with focus on the right ventricular outflow tract obstruction due to the septal aneurysm.

View the video content online at www.cvcasejournal.com.

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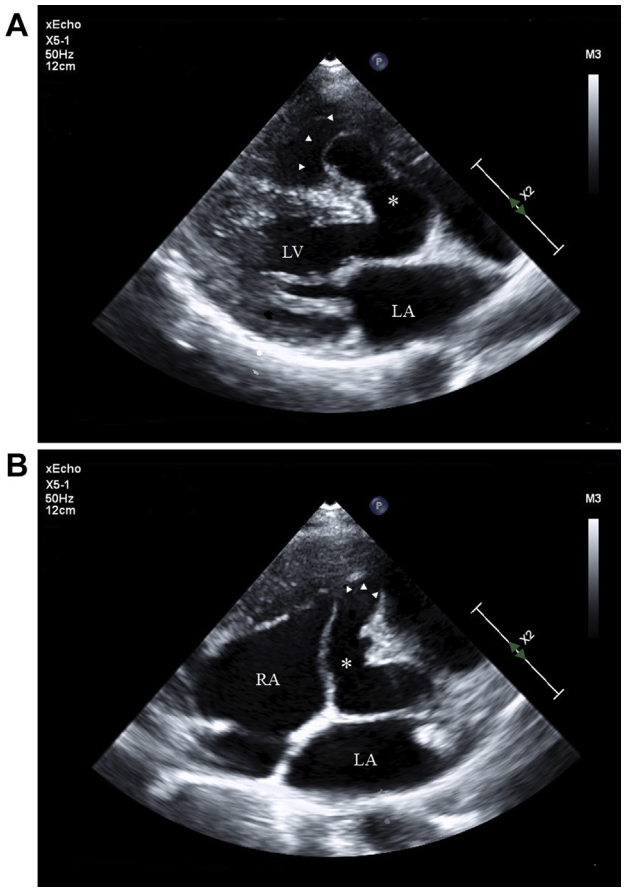

Figure 1 Transthoracic echocardiography showing the large VSD (*asterisk*) and the septal aneurysm (*arrowheads*) bulging into the right ventricular outflow tract in parasternal long-axis **(A)** and parasternal short-axis **(B)** views. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium.

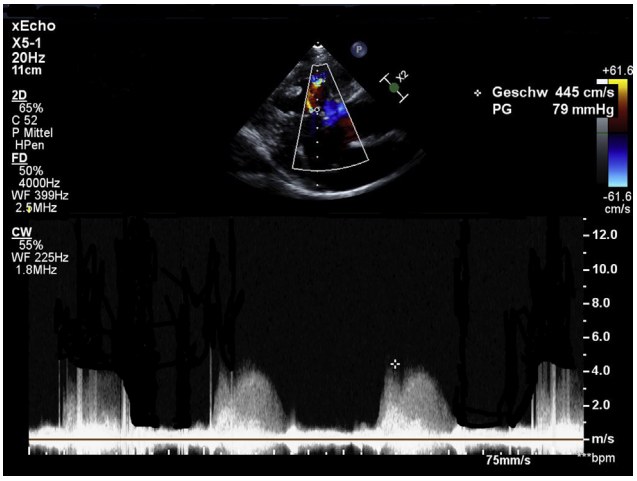

Figure 2 Doppler echocardiography within the right ventricular outflow tract revealing relevant flow acceleration (estimated peak gradient 79 mm Hg).

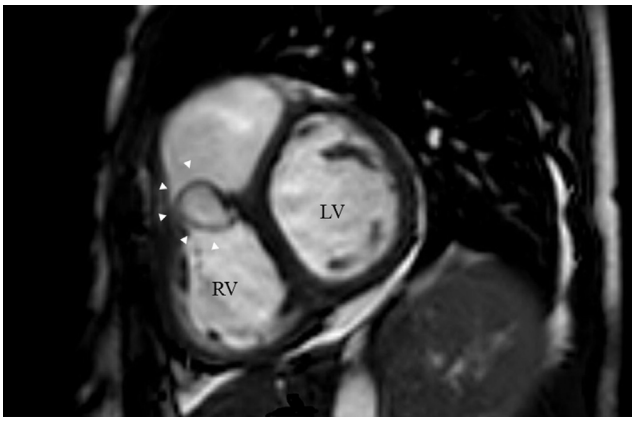

Figure 3 Cardiac magnetic resonance imaging (short-axis view) demonstrating the septal aneurysm (*arrowheads*) almost reaching the free wall of the right ventricle (RV). *LV*, Left ventricle.

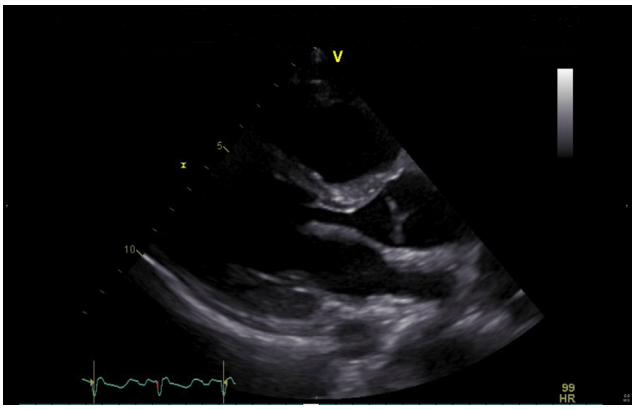

Figure 4 Postoperative echocardiography (parasternal long-axis view) showing good result of VSD patch closure.

visualized and revealed to originate from the septal leaflet of the tricuspid valve. The diameter of the aneurysm was 17 × 20 mm, and severe obstruction of the right ventricular outflow tract was detected. The base of the aneurysm was formed by the perimembranous VSD, with a diameter of 10 mm. The VSD was closed using a Gore-Tex patch. The aneurysm was then resected at the level of the coaptation to the tricuspid valve. Because of dilation, the tricuspid valve showed a relevant central insufficiency during intraoperative transesophageal echocardiography, making a De Vega annuloplasty necessary. The postoperative course was uneventful, and the patient was discharged 11 days after surgery. Postoperative echocardiography showed good result of VSD patch closure, without any residual shunt (Figure 4).

DISCUSSION

Aneurysms originating from tissue of the tricuspid valve's septal leaflet are usually considered beneficial because of their effects on the reduction of shunt size of VSDs or even spontaneous VSD closure, but it has previously been shown that even in patients with complete

spontaneous VSD closure, complications may occur during follow-up induced by the septal aneurysm, which include tricuspid insufficiency, aortic valve prolapse, bacterial endocarditis, rupture, and right ventricular outflow tract obstruction.[5-8]

To prevent further enlargement of the aneurysm and hence avoid the risk for complications, some authors have recommended complete aneurysm resection and defect patch closure, even if there are no cardiac symptoms.[9] Clearly, this approach can be controversially discussed, as the occurrence of clinical relevant complications is rather uncommon. A more conservative approach would include at least regular routine follow-up echocardiographic assessment pursued during adulthood, as right ventricular outflow tract obstruction and other complications may occur late because of the growth of the aneurysm.

According to our experience, regular follow-up at specialized centers for adult patients with congenital heart disease is an adequate scenario. The use of multimodality imaging allows accurate diagnosis of this rare finding; in particular, transesophageal echocardiography enabled an excellent visualization of the spatial relationship of the aneurysm and intraventricular obstruction, allowing optimal preoperative planning.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.08.005.

REFERENCES

1. Schwedler G, Lindinger A, Lange PE, Sax U, Olchvary J, Peters B, et al. Frequency and spectrum of congenital heart defects among live births in Germany: a study of the Competence Network for Congenital Heart Defects. Clin Res Cardiol 2011;100:1111-7.
2. Zhao QM, Niu C, Liu F, Wu L, Ma XJ, Huang GY. Spontaneous closure rates of ventricular septal defects (6,750 consecutive neonates). Am J Cardiol 2019;124:613-7.
3. Dittrich S, Ewert P, Lê TP, Schirmer KR, Hörer J. Isolated ventricular septal defect (VSD) in children and adolescents. Guidelines for the management of congenital heart diseases in childhood and adolescence. Cardiol Young 2017;27(suppl 3):S8-10.
4. Kaemmerer H, Apitz C, Brockmeier K, Eicken A, Gorenflo M, Hager A, et al. Pulmonary hypertension in adults with congenital heart disease: updated recommendations from the Cologne Consensus Conference 2018. Int J Cardiol 2018;272S:79-88.
5. Privitera F, Monte IP, Indelicato A, Tamburino C. Membranous septal aneurysm causing right ventricular outflow tract obstruction in adult. J Cardiovasc Echogr 2017;27:145-8.
6. Mahesh SK, Kumar S, Satheesh S, Jayaraman B. Real time three-dimensional transesophageal echocardiography demonstration of membranous septal aneurysm causing severe right ventricular outflow tract obstruction. Echocardiography 2015;32:184-6.
7. Altunbas G, Gokaslan G, Ercan S, Davutoglu V, Deniz H. Contribution of giant aneurysm of a membranous ventricular septal defect to right ventricular outflow tract obstruction. Thorac Cardiovasc Surg 2013;61:320-2.
8. Santos de Soto J, Lillo Jiménez T, López Haldon J, González Barrero A, Alvárez Madrid A, Descalzo Señorans A. Subpulmonary obstruction caused by an aneurysm of the membranous ventricular septum. A study of 2 cases. Rev Esp Cardiol 1999;52:351-4.
9. Yilmaz AT, Ozal E, Arslan M, Tatar H, Oztürk OY. Aneurysm of the membranous septum in adult patients with perimembranous ventricular septal defect. Eur J Cardiothorac Surg 1997;11:307-11.