

Percutaneous management of a ruptured sinus of Valsalva aneurysm in an infant

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Brief Report

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Abstract

Ruptured sinus of Valsalva aneurysm is a rare entity in children. To our knowledge, this is the youngest child reported in the literature with a sinus of Valsalva ruptured aneurysm being successfully treated percutaneously using an Amplatzer™ Duct Occluder II Additional Sizes device.

Sinus of Valsalva aneurysms are uncommon congenital heart defects (CHD) accounting for < 1% of all CHD, they more commonly involve right coronary sinus (70%), non-coronary sinus (29%), and rarely involve the left coronary sinus (1%).¹ The malformation arises due to an improper or incomplete fusion of the distal bulbar septum.²

Its rupture usually presents from the second to the fourth decade of life with a wide spectrum of presentation, from asymptomatic continuous murmur to heart failure, cardiogenic shock, and death.¹

Surgery has been the mainstay treatment for these CHDs, however, the morphological features of ruptured sinus of Valsalva aneurysms make them susceptible to percutaneous closure and new techniques are emerging.¹

We highlight this case since there are few reports on this approach with an Amplatzer™ Duct Occluder II Additional Sizes device in children.

Case report

An 18-month-old full-term male with no significant past medical history was referred to our department due to an incidentally detected continuous murmur audible over the precordium. The electrocardiogram was normal. Echocardiography and colour Doppler study on admission showed an intracardiac shunt with a “wind-sock” appearance, arising from the left sinus of Valsalva and creating an inward movement towards the right atrium during systole and diastole (Fig 1a). The aortic valve was trileaflet and there was no regurgitation. The left main coronary artery had normal dimensions (Fig 2b).

The aortogram confirmed the presence of an aneurysmatic fistula of the left sinus of Valsalva (Fig 2a). Since the mean pulmonary artery pressure on cardiac catheterisation was 22 mmHg, on the limit for normality, we decided to close the defect to prevent pulmonary overloading secondary to left-to-right cardiac shunt.

Through a 4-F end-hole catheter, a 0.035-inch exchange guide wire was passed through the fistula into the right atrium, then a continuous arteriovenous loop was created. Over the wire, a Mullins sheath was advanced across the fistula from the aortic side. A 5mm×4mm Amplatzer™ Duct Occluder II Additional Sizes device was advanced and deployed in the fistula. An angiogram using the end-hole catheter was done prior to the release of the device and the final control angiogram showed that the device was in situ with no residual leakage, no significant aortic regurgitation, or any encroachment on coronary arteries (Fig 2b and c). The patient was discharged 24 hours after the procedure with a normal electrocardiogram and completed 6 months of treatment with acetylsalicylic acid. Follow-up 12 months later revealed no residual shunt or aortic regurgitation and a progressively less pronounced dilatation of the left sinus of Valsalva (Fig 1c and d).

Discussion

Since the first report of transcatheter closure of a ruptured sinus of Valsalva aneurysm in 1994, these techniques using different devices are growing in popularity. However, no dedicated closure devices exist yet and open surgical repair remains the mainstay of treatment, mainly when ruptured aneurysms are associated with other intracardiac abnormalities.³

Although the mortality from surgery is low (<2%), some problems limit the application of surgical repair, for instance, haemodynamic instability. In fact, in patients with poor

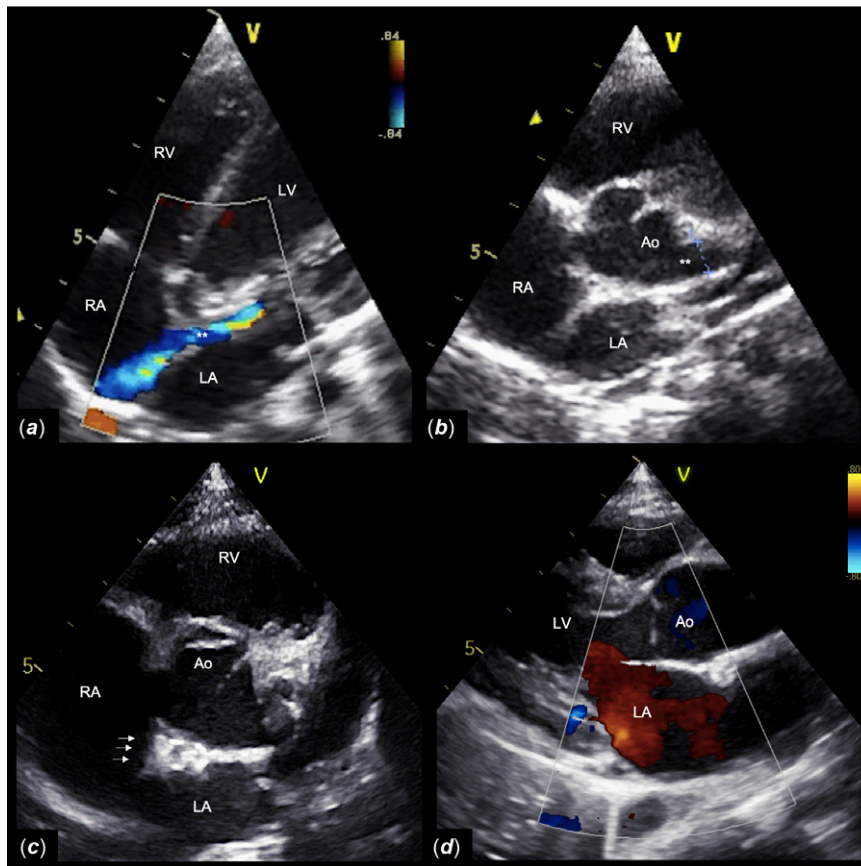


Figure 1. Echocardiogram with colour Doppler study before (**a** and **b**) and 12 months after (**c** and **d**) the transcatheter closure of the ruptured sinus of Valsalva aneurysm. A - 5-chamber view, shows an intracardiac shunt with a “wind-sock” appearance (**). B - Short-axis view, shows an aneurysmatic left sinus of Valsalva (**). C - Short-axis view shows a well-placed device (arrows). D - Long-axis view shows no aortic regurgitation. Ao = Aorta; LA = left atrium; RA = right atrium; RV = right ventricle.

general condition and morbidities, cardiac surgery is high risk, and transcatheter closure can be life-saving. Also, it spared patients from the morbidity related to sternotomy and use of cardiopulmonary bypass.¹

A wide variety of devices have been used to close ruptured sinus of Valsalva aneurysms depending on the anatomy of the lesion and the experience of each centre. The most commonly used in children has been the Amplatzer™ Duct Occluder I, with reports in the literature between the ages of 5 and 17 years.⁴⁻⁷ It is often favoured because its shape is suitable for closing these saccular aneurysms with high occlusion rates and stability through the retention disc and the short waist. Also, it has the distinct advantage of being user-friendly and of easy retrievability.⁴

Amplatzer™ Muscular Ventricular Septal Defect Occluder theoretically has the same advantages as the Amplatzer™ Duct Occluder I with even more stability through the right and left-sided umbrellas, however, there are few paediatric reports concerning the closure of ruptured sinus of Valsalva aneurysms with this device.⁸ Other devices less used in children were the Cocoon Duct Occluder,¹ the Amplatzer Septal Occluder,⁹ and coils, which should be implemented only in cases of minor connections.

Due to the small size, coils were an option for the treatment of the ruptured aneurysm in our patient, however, they have a higher risk of embolisation, therefore, and considering the aforementioned advantages, including our familiarity with this device, we chose to use a Duct Occluder. As the Amplatzer™ Duct Occluder II Additional Sizes has a narrow waist diameter, making it less bulky, we believe that this device is more suitable for closing smaller defects like the one we report. In addition to ours, there is only one case in the literature referring to closure of a paediatric ruptured sinus of Valsalva aneurysm with an Amplatzer™ Duct Occluder II Additional Sizes in a four-year-old child.¹⁰

So far, there is no commonly accepted standard for selecting the size or type of Duct Occluder. The authors chose devices 1–5 mm larger than the maximal diameter of the aortic opening of the aneurysm.^{4,5} In our patient, the opening diameter measured by aortic root angiography was 3 mm, so we used a 5mm×4mm Amplatzer™ Duct Occluder II Additional Sizes.

Although some concerns need to be clarified, we conclude that, in short term, transcatheter closure with the Amplatzer™ Duct Occluder II Additional Sizes was a safe and effective treatment for isolated ruptured sinus of Valsalva aneurysms and could be a cost-effective alternative to surgery in small children.

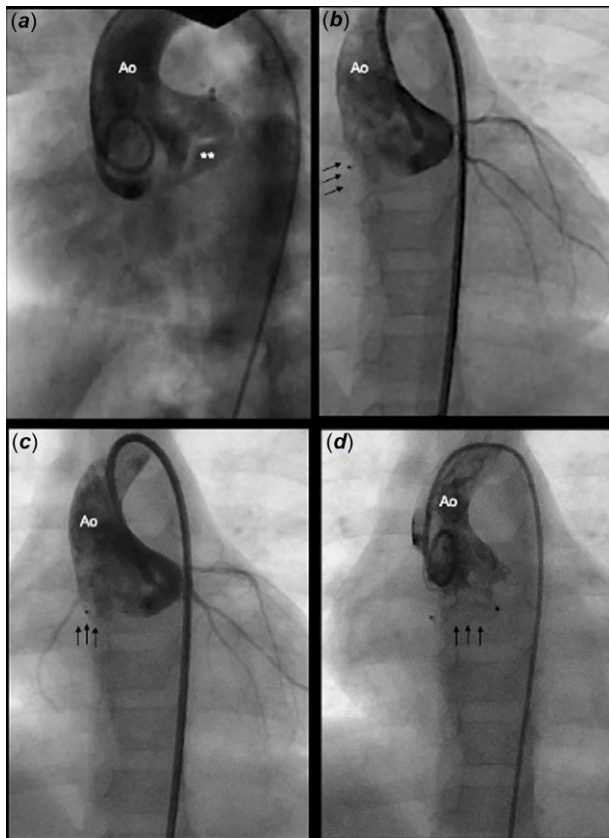


Figure 2. (a) Aortogram, left anterior oblique projection, shows opacification of the right atrium through an aneurysmatic fistula (***) of the left sinus of Valsalva. (b and c) Aortogram, anteroposterior projection, done immediately before the definitive release of the AMPLATZERTM Duct Occluder II Additional Sizes show the device (arrows) inside the fistula with good opacification of the left coronary arteries (b) and the right coronary artery (c), without opacification of the right cavities, revealing no residual leakage. (d) Post-procedure control aortogram, anteroposterior projection, showing the device (arrows) in a good position with no residual shunt.

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Conflicts of interest. None.

Ethical standards. No specific ethical approval from Institutional Reviews Boards are necessary for this type of publication. The authors assure the patients data provided in this case report are anonymized.

References

1. Sinha SC, Sujatha V, Mahapatro AK Percutaneous transcatheter closure of ruptured sinus of valsalva aneurysm: immediate result and long-term follow-up. *Int J Angiol* 2015; 24: 99–104.
2. Kakos GS, Kilman JW, Williams TE, Hosier DM Diagnosis and management of sinus of valsalva aneurysm in children. *Ann Thorac Surg* 1974; 17(5): 474–478.
3. Cullen S, Somerville J, Redington A Transcatheter closure of a ruptured aneurysm of the sinus of Valsalva. *Br Heart J* 1994; 71: 479–480.
4. Kerkar PG, Lanjewar CP, Mishra N et al. Transcatheter closure of ruptured sinus of valsalva aneurysm using the amplatzer duct occluder: immediate results and midterm follow-up. *Eur Heart J* 2010; 31: 2881–2887.
5. Zhao SH, Yan CW, Zhu XY et al. Transcatheter occlusion of the ruptured sinus of valsalva aneurysm with an amplatzer duct occluder. *Int J Cardiol* 2008; 129: 81–85.
6. Srivastava A, Radha A Transcatheter closure of ruptured sinus of valsalva aneurysm into the left ventricle: a retrograde approach. *Pediatr Cardiol* 2012; 33: 347–350.
7. Santoro G, Pacileo G, Bigazzi C et al. Transcatheter closure of a ruptured sinus of valsalva aneurysm causing Fontan circulation failure. *Cardiovasc Med* 2007; 8: 470–472.
8. Schaeffler R, Sarikouch S, Peuster M Transcatheter closure of a ruptured sinus of valsalva aneurysm (RSVA) after aortic valve replacement using the amplatzer muscular VSD occluder. *Clin Res Cardiol* 2007; 96: 904–906.
9. Arora R, Rangasetty U, Thakur A Transcatheter closure of ruptured sinus of valsalva aneurysm. *J Intervent Cardiol* 2004; 17: 53–58.
10. Capogrosso C, Santoro G, Giordano M et al. Transcatheter closure of post-surgical ruptured sinus of valsalva with Amplatzer Duct Occluder II ASTM device. *Ann Pediatr Cardiol* 2018; 11: 86–88.