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Transcatheter Closure of a Ruptured Sinus of Valsalva Aneurysm with the Amplatzer Ductal Occluder II in a 6-Year-Old Girl

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Abstract

We report the successful deployment of a 6 mm Amplatzer Ductal Occluder II via a retrograde approach to treat full occlusion of a type II ruptured right sinus of Valsalva aneurysm in a symptomatic 6-year-old girl with significant left-heart dilation.

Key Words

Occlusion • Sinus of Valsalva • Device

Introduction

Sinus of Valsalva aneurysm is a rare congenital condition accounting for less than 1% of all congenital anomalies, although the prevalence is higher in Asia. It occurs due to deficient elastic fibers in the aortic media, leading to progressive dilatation over time, and should be differentiated from acquired aneurysms caused by infections or connective tissue disorders. The right sinus of Valsalva is affected in up to 85% of cases. A classification system has been reported based on the origin of the aneurysm in relation to the right and noncoronary sinuses [1]. Rupture may be precipitated by an exertional event and usually occurs into the right atrium or ventricle, leading to significant left-to-right shunt and congestive cardiac failure. Once the clinical diagnosis has been confirmed on echocardiography, advanced imaging modalities may be used to clarify the aneurysm morphology because there may be multiple ostia from the aneurysm into the right heart, and this may influence the closure approach.

Case Presentation

A 6-year-old girl was referred with increasing fatigue and was noted to have a harsh grade IV long systolic murmur. She had bounding pulses, and chest x-ray revealed a large heart with increased pulmonary vascular markings. Transthoracic echocardiogram (TTE) demonstrated a dilated left heart with preserved systolic ventricular function and turbulent flow on color Doppler from the aortic root into the pulmonary outflow. This was initially thought to be an aortopulmonary window, but further assessment confirmed that the flow from the aortic root was entering the right ventricular outflow tract (RVOT) beneath the pulmonary valve. The jet measuring 6 mm in diameter originated from the midpoint of the right coronary sinus into the RVOT, with persistence of flow into diastole (although a diastolic component to the murmur was not clearly audible), consistent with a type II ruptured sinus of Valsalva aneurysm according to the original Sakakibara classification [1]. There was an eccentric jet of aortic incompetence graded as mild.

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Catheterization was performed in a single-plane catheterization suite with a C-arm under general anesthesia with transesophageal echocardiography (TEE) guidance. TEE confirmed the presence of a 6 mm ruptured right sinus of Valsalva aneurysm with continuous flow into the RVOT (Figure 1:Panel A and B). These findings were confirmed with ascending aortic angiography (Figure 2:Panel A). The mean pulmonary artery pressure was 26 mmHg, with right ventricular systolic pressure of 42 mmHg and ascending aortic pressures of 91/38. The ratio of total pulmonary to total systemic blood flow (Qp:Qs) measured 4.2:1 but was calculated on 100% oxygen as calculation in room air was not feasible. The defect was easily crossed with a 5-French (Fr) Judkins Right catheter and an exchange-length Glide wire with advancement of the JR4 into the distal right lower pulmonary artery. A 0.035" Amplatzer Extra-Stiff wire (St. Jude Medical, St Paul, MN, USA) was positioned in the right pulmonary artery, and the JR4 was exchanged for a 5Fr guiding sheath (Figure 2:Panel B). A 6:6 Amplatzer Ductal Occluder (ADO) II (St. Jude Medical) was then advanced across the defect, the distal disk was partially deployed, and the sheath was retraced slowly from the main pulmonary artery. As the device crossed the pulmonary valve, the distal disk of the device engaged the right ventricular aspect of the defect, and the waist was deployed within the neck of the aneurysm and finally the proximal disk in the aortic root. TEE confirmed good device positioning (Figure 1:Panel C), but it was unclear if there was a small persistent leak. There was no impingement on the aortic or pulmonary valves. It was decided to release the device because there was some tension from the delivery cable distorting the aortic end of the device away from the root. Following release, the device position was more satisfactory, and the final aortic root angiogram confirmed good position with no residual leak (Figure 2:Panel C).

The patient recovered well from the procedure with no procedural complications. Predischarge echocardiogram the following day demonstrated a good device position, but it initially appeared that there may be a small residual leak. Further assessment revealed that there was a small perimembranous ventricular septal defect (VSD) just beneath the right coronary cusp of the aortic valve that was not initially noticed due to its proximity to the more prominent jet from the ruptured sinus of Valsalva aneurysm. The flow pattern was systolic with no diastolic component and a peak estimated systolic gradient of 82 mmHg. There was no RVOT obstruction or increase in the degree of mild aortic incompetence.

Discussion

Although surgical repair of ruptured sinus of Valsalva aneurysm has been extensively described [2], reports of transcatheter closure are sparse [3-9]. The largest study to date reported 18 successful cases from 20 attempts with a median age of 23 years (17–52 years) [3]. The majority of patients were symptomatic, and few had associated lesions such as VSDs, which were reported in 41% of 725 patients from a large systematic review [2]. In all patients, the defects were closed from the venous side using ADOs 2–4 mm larger than the aortic end of the defect. The ADO sizes ranged from 8/6 to 16/14 mm. Thirteen

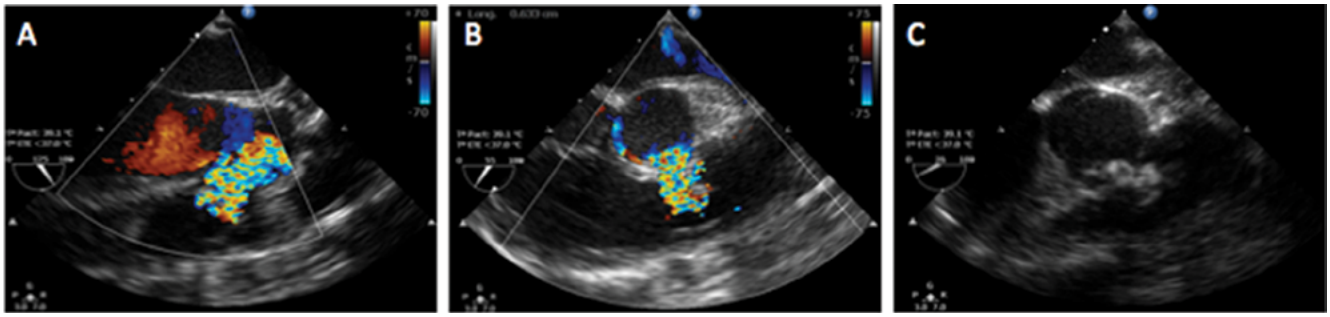

Figure 1. Series of transthoracic echocardiogram images demonstrating the color flow across the ruptured sinus of Valsalva aneurysm from the right coronary sinus into the right ventricular outflow tract in the long (*Panel A*) and short (*Panel B*) axes. (*Panel C*). The Amplatzer Ductal Occluder II is in a good position across the ruptured aneurysm following release.

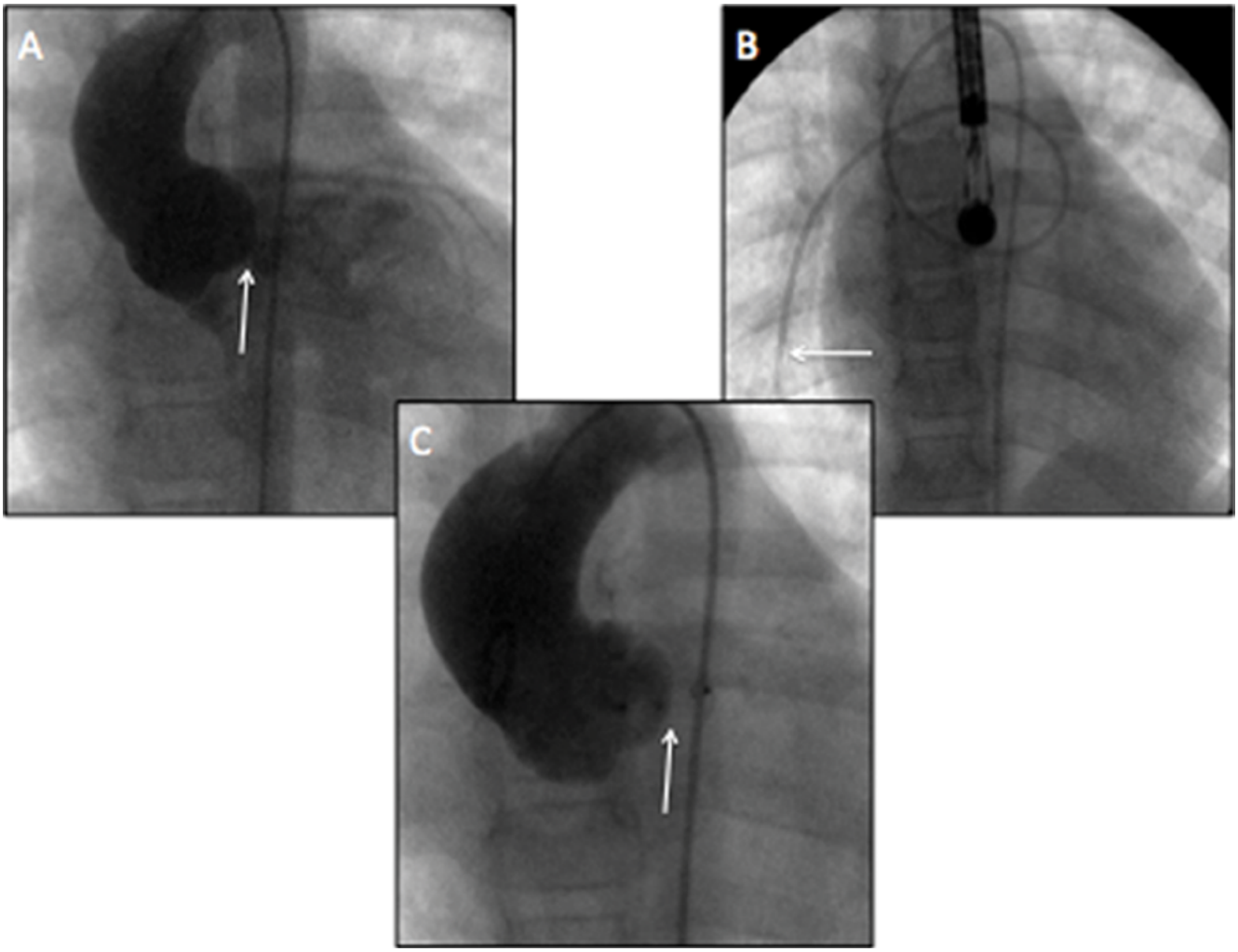

Figure 2. *Panel A.* Initial ascending aortogram demonstrates the sinus of Valsalva aneurysm (white arrow) with rupture into the right ventricular outflow. Mild aortic regurgitation is evident. *Panel B.* Outline of the guiding sheath across the defect from the aorta and positioned distal in the right pulmonary artery. *Panel C.* Final ascending aortogram demonstrates the Amplatzer Ductal Occluder II in a good position (white arrow) with ne residual flow across the ruptured aneurysm.

patients had complete closures at discharge. Five had a residual shunt (four small and one moderate with self-abating hemolysis). Trivial aortic regurgitation (AR) occurred in four. On median follow-up of 24 months (range 1–60 months), 15 patients were in New York Heart Association class I. The residual shunt disappeared in three and was small in two; procedure-related AR vanished in two of four. There was no AR progression, recurrence, infective endocarditis, or device embolization. A variety of other devices have been described in case reports with good success [4-9]. The majority of these closures were in adult patients with delivery from the femoral venous approach following the creation of an arteriovenous loop. In our case, the patient was young, and the low profile of the ADO II allowed us to retrogradely deliver the device from the aorta without the need to create an arteriovenous loop or use a stiff sheath and dilator, which may be more likely to induce hemodynamic instability in a small child. The waist of the device was delivered within the aneurysmal sac toward the aortic end to ensure optimal flow occlusion from the higher pressure aorta. There was no impingement on the aortic or pulmonary valves, and the "soft" nature of the device, which has made it popular for closing perimembranous VSDs [10], was attractive to ensure

minimal distortion of the aortic root in a smaller child. The occlusive nature of the device has been well reported in high flow defects such as VSDs and patent ductus arteriosus [10, 11].

The learning point from the case was to ensure full interrogation of the ventricular septum prior to closure. A small perimembranous VSD may be easily overlooked, especially considering the turbulence seen on color imaging from the right sinus of Valsalva throughout the cardiac cycle. The perimembranous region of the septum is separated from the aortic root only by the thin leaflets of the aortic valve. In retrospect, knowledge of this defect would not have changed our approach. It is arguable that the VSD may have been contributing to the mild AR; however, AR has been reported in up to 71% of patients with ruptured sinus of Valsalva aneurysms [2], which is far greater than seen with a small perimembranous VSD. It is arguable that the device may provide some support to the right sinus and consequently to the support structure of the right coronary cusp to mitigate against further AR.

We elected to use low-dose aspirin following device implantation to limit the risk of thromboembolism. The evidence for such an approach is not strong; however, it has been a widely prescribed practice with device closure where the left-sided discs are proximal to the cerebral circulation origin.

Conclusions

In conclusion, ruptured sinus of Valsalva aneurysms are exceedingly rare in children, but transcatheter closure is feasible, and the ADO II may be an attractive device choice to minimize aortic root distortion.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question

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