Transcatheter closure of postsurgical ruptured sinus of Valsalva with Amplatzer Duct Occluder II AS™ device

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ABSTRACT

Sinus of Valsalva (SV) rupture is a rare, cardiac complication after surgical repair of complex congenital heart disease. This paper reports a 4-year-old male child with double outlet right ventricle (RV) and pulmonary stenosis with superior-inferior arrangement of the ventricles, who was submitted to surgical repair using the "reparation a l'etage ventriculaire" procedure. A few months after an uneventful surgical repair, his clinical condition abruptly worsened because of the rupture of the right SV into the RV outflow tract resulting in large left-to-right shunt and RV functional impairment. To avoid surgical re-do, this late-onset complication was successfully treated by transcatheter implantation of an Amplatzer Duct Occluder Type II Additional Size™ (ADO-IIAS, St. Jude Medical Inc., St. Paul, Minnesota, USA) device.

Keywords: Amplatzer Duct Occluder II AS™ device, double outlet right ventricle, sinus of Valsalva rupture, surgical complication

INTRODUCTION

Sinus of Valsalva (SV) rupture is a rare, cardiac lesion usually occurring in the case of aneurysmal dilatation caused by congenital or acquired wall weakness.[1,2] It can be asymptomatic or more often may result in congestive cardiac failure, arrhythmias, or sudden cardiac death caused by significant and abrupt left-to-right shunt.[3] However, postsurgical fissuration of a morphologically normal SV has so far never been reported in literature as complication of repair of complex congenital heart malformations.

Closure of ruptured SV has been conventionally performed surgically using a cardiopulmonary bypass. Over time, transcatheter closure of ruptured SV has evolved as an acceptable alternative to surgical repair using different off-label devices.[4-7] However, most of them are bulky and require stiff delivery systems. These limitations are all the more important in the case of rupture of a morphologically normal SV due to its proximity to aortic valvular leaflets and coronary artery origin and course. In this setting, using a softer and more low profile device may be safer and crucial for the interventional approach.

This is the first report of percutaneous closure of postsurgical ruptured SV using a novel, softer device of the Amplatzer Occluder family, the Amplatzer Duct Occluder Type II Additional Size™ (ADO-IIAS, St. Jude Medical Inc., St. Paul, Minnesota, USA).

CASE REPORT

A 4-year-old male child with double outlet right ventricle (RV) and pulmonary stenosis with superior-inferior arrangement of the ventricles submitted to right modified systemic pulmonary Blalock–Taussig shunt at the age of 3 months and surgical repair at the age of 2 years showed an abrupt clinical deterioration late after surgery. During the early postoperative

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Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_38_17

How to cite this article: Capogrosso C, Santoro G, Giordano M, Russo MG. Transcatheter closure of postsurgical ruptured sinus of valsalva with amplatzer duct Occluder II AS™ device. Ann Pediatr Card 2018;11:86-8.

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period, neither significant clinical nor instrumental findings were reported. However, about 9 months after a seemingly successful surgical repair, he started to complain progressive worsening of effort tolerance and showed a significant increase of cardiac enlargement as compared to the postsurgical baseline chest X-ray. There were no clinical, hematological or echocardiographic features suggestive of infective endocarditis in the recent past or at the time of admission. In particular, white cell count was within normal values, so blood cultures were not done. The electrocardiogram (EKG) was unremarkable with respect to the early postsurgical picture, showing mild tachycardia and complete right bundle branch block as sole relevant findings. On transthoracic echocardiogram (TTE), a rupture of the right SV into the RV outflow tract, measuring 4 mm, was seen on color Doppler analysis, causing significant left-to-right shunt and resulting in significant RV dilatation with global functional impairment. Based on clinical and anatomic findings, percutaneous treatment of this complication was planned as a lower risk option with respect to surgical repair. After parental informed consent and agreement with the surgical team, the procedure was performed under general anesthesia with TTE guide. At cardiac catheterization, moderate RV hypertension (RV/left ventricular pressure ratio 0.6) and severe RV dilatation were found [Figure 1]. Aortic angiography showed a 3-mm large rupture of the right coronary sinus into the RV outflow, resulting in moderate significant left-to-right shunt (QP/QS 1.6:1) [Figure 2a]. As a cost–benefit approach, device closure from the aortic side was chosen. The ruptured sinus was then negotiated with a coronary guidewire (Abbott Vascular, Santa Clara, CA, USA) and a multipurpose 4 Fr catheter. Over an exchange 0.035" guidewire, the dedicated delivery system (AMPLATZER™ TorqVue™ LP Delivery System, St. Jude Medical Inc., St. Paul, Minnesota, USA) was tracked into the RV outflow. Then, an ADO II-AS™ 5–2 mm (St. Jude Medical Inc., St. Paul, Minnesota, USA) was deployed under fluoroscopic and echocardiographic guide. Angiography was used for the precise deployment of the device. Procedural and fluoroscopic times were 70 and 13 min, respectively. After deployment, aortic root angiogram revealed mild foaming across the device [Figure 2b] without any aortic valve malfunction or right coronary artery compression. The postprocedural course was uneventful, and the child was discharged after 48 h without new-onset EKG anomalies or residual shunt at echocardiographic evaluation. Over a 3-month follow-up, both clinical condition and RV size and function improved significantly.

DISCUSSION

Rupture of morphologically normal SV is so far an unreported complication of surgical repair of congenital heart malformations. Early rupture can result in abrupt clinical deterioration and may be difficult to diagnose in postsurgical patients due to their borderline clinical conditions and poor echocardiographic window. However, late-onset SV rupture in RV outflow tract after a seemingly successful repair may be easily misdiagnosed as chronic pulmonary regurgitation, especially after repair without prosthetic conduit. However, it may cause abrupt clinical deterioration due to sudden RV volume overload as well as decrease of mean aortic pressure and coronary perfusion, so resulting in biventricular failure. Traditional treatment of isolated SV rupture has been surgical repair, with <2% mortality, low rate of recurrent fistula or ventricular septal defect, and as high as 88% long-term survival rate.[8] Development of aortic regurgitation, whether recurrent or *de novo*, is still a risk late after surgical repair.[9] However, several recent reports highlight safety and effectiveness of transcatheter closure of symptomatic ruptured SV.[4-7] This therapeutic option shows potential advantages over surgical repair, mainly in "complex" or unstable patients, namely avoidance of sternotomy and cardiopulmonary bypass. However, the most effective devices often require bulky and relatively stiff delivery systems, thus increasing procedural complexity and risks in younger and more critical children. In addition, different from rupture of SV aneurysm, in the case of rupture of

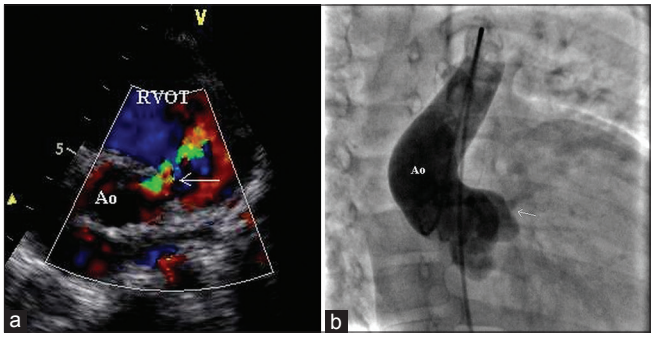

Figure 1: Significant aorta-to-right ventricular outflow tract shunt through the ruptured sinus of Valsalva (arrow) as imaged at echocardiographic color Doppler analysis (a) and aortic angiography (b). Ao: Aorta; RVOT: Right ventricular outflow tract

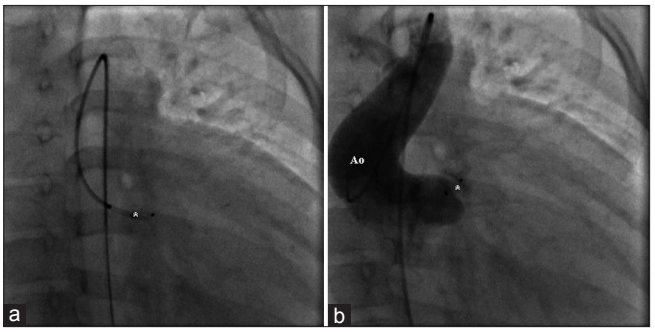

Figure 2: Amplatzer Duct Occluder Type II AS™ device (asterisk) during deployment (a). Mild residual foaming through the implanted device after final release (b). Ao: Aorta

morphologically normal SV, device deployment might potentially cause either distortion and malfunction of the aortic leaflets or occlusion of coronary artery ostium and distortion of proximal course of coronary artery, due to the limited room between fissuration and valvular leaflets or coronary artery origin. In this setting, the newer devices of the Amplatzer Occluder family such as the Amplatzer Vascular Plug or the ADO-II device can be extremely useful, being softer, more trackable, and respectful of local anatomy as compared to older devices.[10] Both devices are made of a fabric free, multilayered, flexible, nitinol wire mesh-shaped into a cylindrical waist with two very low profile retention discs on either end. However, the former is significantly longer and needs of larger delivery systems than the latter one, which is as short as 2 mm and can be tracked by soft 4 Fr dedicated catheters. Thus, the ADO-IIAS needs little room to accommodate and does not interfere with the neighboring structures. In addition, it shows an improved control over placement and release, a high rate of complete occlusion, and a high conformability to local anatomy due to articulating "necks."

To our knowledge, this is the first reported experience of off-label use of this device in the treatment of this uncommon postsurgical complication. Thanks to the high pliability and track ability of the device, the percutaneous approach was very easily performed from the arterial route without the need of arteriovenous loop, resulting in successful and cost-effective treatment of this critical patient.

CONCLUSION

SV rupture is an uncommon, life-threatening postsurgical complication that can be misdiagnosed. Currently, most of these patients are managed using transcatheter intervention. This is the first case in which ADO-II AS™ has been used safely and effectively in a small and sick child to close a small postoperative rupture of SV.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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