Closure of insufficient, native right ventricular outflow tract with AMPLATZER™ muscular ventricular septal defect occluder in a patient with tetralogy of Fallot post-Melody® valve

Neha Bansal, Sanjeev Aggarwal, Daniel R Turner

The Carmen and Ann Adams Department of Pediatrics, Division of Cardiology, The Children's Hospital of Michigan, Wayne State University School of Medicine, Detroit, MI, USA

ABSTRACT

AMPLATZER™ muscular ventricular septal defect occluder is used commonly for off-label purposes. We describe an unusual case of a patient with tetralogy of Fallot who underwent repair with a right ventricle to pulmonary artery homograft due to abnormal coronary artery pattern. During the initial surgery, the native right ventricular outflow tract was left open. At 30 years of age, he was symptomatic due to severe native right ventricular outflow tract insufficiency. Cardiac MRI confirmed a dilated right ventricle and pulmonary insufficiency through the native right ventricular outflow tract, despite no significant homograft insufficiency due to previous Melody® valve placement. The right ventricular outflow tract was closed successfully using an 18 mm AMPLATZER™ muscular ventricular septal defect occluder. At 5-year follow-up, there is no native right ventricular outflow tract insufficiency and no additional arrhythmia. We suggest that percutaneous closure of the insufficient; native right ventricular outflow tract using a septal occluder is an alternative to surgical management.

Keywords: AMPLATZER™ ventricular septal defect occluder, native outflow tract, tetralogy of Fallot

INTRODUCTION

The AMPLATZER™ septal occluder, AMPLATZER™ muscular ventricular septal defect (VSD) occluder, and AMPLATZER™ duct occluder devices are used extensively for closure of septal defects in congenital heart disease and more recently for nonconventional and off-label uses.[1-5] The AMPLATZER™ muscular VSD occluder has been used for patent ductus arteriosus occlusion,[6] perforated sinus of Valsalva aneurysm closure,[7] and closure of pulmonary artery (PA) to left atrium fistulae.[8,9] There are two case reports of muscular VSD occluders used for closure of native right ventricular outflow tract (RVOT), one in a patient with bidirectional cavopulmonary anastomosis and the other in a patient with repaired tetralogy of Fallot (TOF) with limited follow-up.[10,11] We report the case of a 38-year-old male with TOF, status-post repair with a right ventricle (RV) to PA homograft due to an abnormal coronary artery pattern. He recently underwent Melody® valve placement for conduit insufficiency, but remained symptomatic due to a persistently dilated RV and a significantly regurgitant native RVOT. He had a successful percutaneous closure of the native RVOT using an AMPLATZER™ muscular VSD occluder with persistent closure at the 5-year follow-up.

CASE REPORT

Our patient is a 38-year-old male born with TOF, right aortic arch with mirror image branching, left superior vena cava draining to the coronary sinus, and a large accessory left anterior descending coronary artery

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_76_18

How to cite this article: Bansal N, Aggarwal S, Turner DR. Closure of insufficient, native right ventricular outflow tract with AMPLATZER™ muscular ventricular septal defect occluder in a patient with tetralogy of Fallot post-Melody® valve. Ann Pediatr Card 2019;12:159-62.

Address for correspondence: Dr. Neha Bansal, The Carmen and Ann Adams Department of Pediatrics, Division of Cardiology, The Children's Hospital of Michigan, Wayne State University School of Medicine, Detroit, MI, USA. E-mail: nehban@gmail.com

originating from the right coronary artery and crossing the RVOT to the ventricular septum. He initially underwent placement of a left Blalock–Taussig shunt in infancy followed by a complete repair at 6 months of age with the placement of a RV to PA homograft due to the coronary artery abnormality. His homograft was subsequently repaired at 13 years of age, with the removal of the valve and placement of a patch over the anterior surface of the homograft. Over the years, he developed worsening homograft stenosis and regurgitation. Cardiac magnetic resonance imaging showed mild RV dilation with an end-diastolic volume index of 139.2 ml/m^2, RV end-systolic volume index of 70.6 ml/m^2, and RV ejection fraction of 49.3%. There was dual outflow from the RV to the PAs via the homograft and the native RVOT, and the combined regurgitant fraction was 63%. He developed nonsustained ventricular tachycardia (VT) thought secondary to increased right ventricular systolic pressure (RVSP) and volume overload. At 31 years of age, he underwent Implantable Cardioverter-Defibrillator (ICD) placement for the VT as well as placement of a Melody® valve for the homograft stenosis and insufficiency. Prestenting was not performed due to the relatively mild degree of stenosis. The native outflow tract remained open and continued to be a source of moderate-to-severe regurgitation. He continued to complain of significant fatigue and inability to keep up with activities of daily living despite successful elimination of the homograft insufficiency by Melody® valve placement. Transthoracic echocardiography showed the RV to be moderately dilated with mild stenosis at the Melody® valve. Doppler interrogation of the native RVOT revealed moderate-to-severe insufficiency [Figure 1]. Due to the symptoms, persistent RV dilation, and severe regurgitation from the native RVOT, we decided to close the native RVOT by percutaneous intervention.

Cardiac catheterization was performed under general anesthesia. RVSP was moderately elevated (49% systemic), and there was a 10 mmHg peak gradient across the Melody® valve. The pulmonary capillary wedge pressure was mildly elevated (16 mmHg). There was free pulmonary insufficiency through the native RVOT [Figure 2]. A 24-mm AMPLATZER™ Sizing Balloon was used for test occlusion of the native RVOT, with no change in the RVSP. The patient was hemodynamically stable. There was no ST segment or T-wave changes noted on the continuously monitored electrocardiography. However, specific coronary angiographies were not performed before the device placement. The narrowest diameter of the native RVOT was 13–15 mm at its mid-portion. An 18-mm AMPLATZER™ muscular VSD occluder was delivered with the center waist at the narrowest portion of the RVOT. The device remained in satisfactory position after release [Figure 2]. After device closure, there was no significant change in the RVSP (52% systemic). There were no complications.

The patient tolerated the procedure well and was discharged home the next day on aspirin. On follow-up 3 years after the device placement, there was no regurgitation of the native pulmonary tract and the RVSP was stable (40 mmHg). A computed tomography scan showed satisfactory position of the device [Figure 3]. However, at 5-year follow-up, echocardiography showed increased RV pressure at 76 mmHg due to type II fractures of the Melody® valve with loss of stent integrity. At this time, the homograft was replaced using a 25-mm porcine-valved conduit.

DISCUSSION

Percutaneous devices are utilized increasingly for imaginative off-label use to decrease the number of surgical interventions in patients with unrepaired or repaired congenital heart defects. TOF survivors with the need for RV-to-PA homograft often require multiple surgical revisions due to outgrowing the size of the homograft or due to conduit valve dysfunction. In patients who require the initial repair with a conduit due to an abnormal coronary artery pattern, the native pulmonary outflow tract is often left open to allow for a second pathway for blood to exit the RV, in the event of conduit

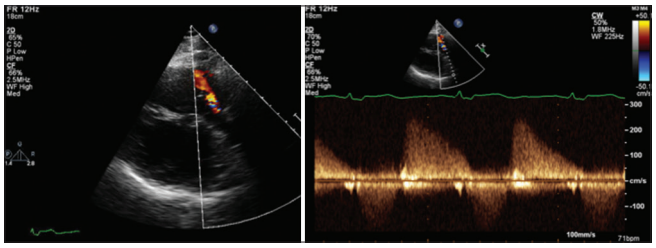

Figure 1: Parasternal short axis view (left) and the Doppler interrogation (right) of the native pulmonary tract demonstrating the moderate-to-severe regurgitation by color echocardiography

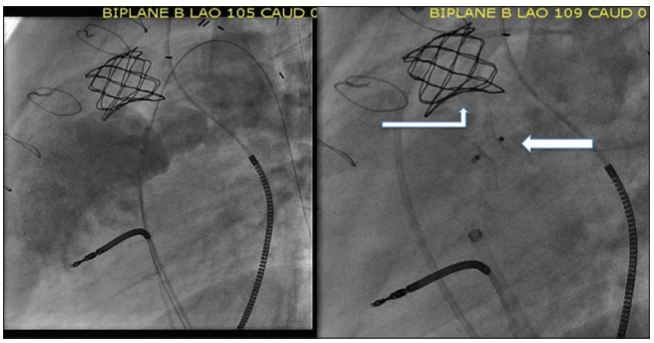

Figure 2: Fluoroscopy image from the cardiac catheterization showing the regurgitation of the native pulmonary tract (left) and the placement of the AMPLATZER™ muscular ventricular septal defect occluder (right). The Melody® valve (elbowed arrow) is shown in the anterior right ventricle to pulmonary artery homograft and the AMPLATZER™ muscular ventricular septal defect occluder (horizontal arrow) is seen below it parallel to the Melody® valve in the native right ventricular outflow tract

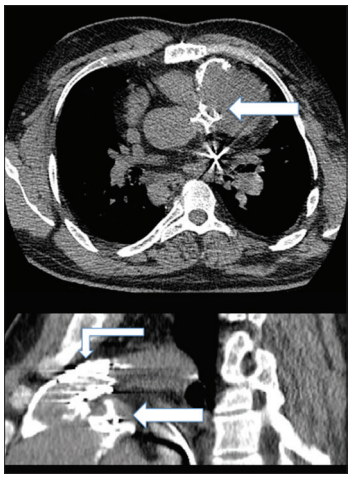

Figure 3: Computed tomography scan of the patient showing the AMPLATZER™ muscular ventricular septal defect occluder (arrow) in the native right ventricular outflow tract in axial (top) and sagittal (bottom) planes. The Melody® valve (elbowed arrow) is seen in the conduit anterior to the device

stenosis.[12] The major drawback of leaving the native RVOT is development of progressive regurgitation leading to RV dilatation and associated symptoms.[13] When a patient develops symptoms or significant RV dilatation due to native RVOT insufficiency, the native tract can be closed surgically or by transcatheter device placement.

The AMPLATZER™ muscular VSD occluder has been described twice in the past to close the native RVOT. The first case was in a 12-month-old child with bidirectional cavopulmonary anastomosis to prevent excessive antegrade pulmonary blood flow.[10] The second case report was in a 13-year-old patient, similar to our case, with TOF and a conduit repair. The patient had a limited follow-up of only 18 months after closure of the native RVOT with no complications.[11]

In our patient, surgical closure of the native RVOT would have entailed dissection near the aberrant coronary artery as well as the removal of the competent Melody® valve, which was not clinically necessary. Percutaneous closure of the native RVOT allowed a less invasive, nonsurgical procedure that spared the Melody® valve and alleviated his symptoms of pulmonary insufficiency. The patient has since undergone homograft replacement 5 years after RVOT device closure, giving him those 5 additional years without a need for surgical intervention and a persistently occluded native RVOT for the future. At the recent conduit replacement surgery, the native RVOT device was found to be in satisfactory position and working well; thus, it was left untouched [Figure 4].

CONCLUSION

We believe that the cause of his symptoms was a combination of the regurgitation from the native RVOT as well as the conduit. He had improvement in his symptoms post-RVOT occlusion and a conduit change was deferred for 5 years.

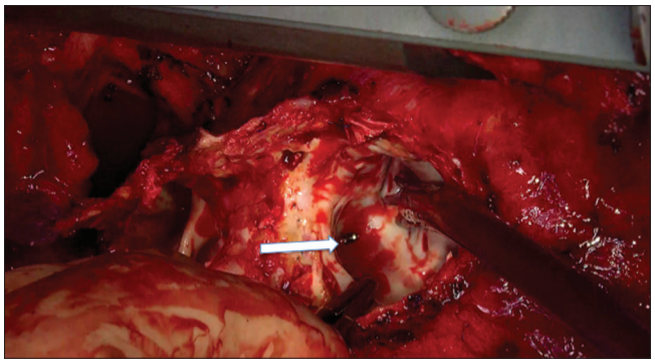

Figure 4: Intraoperative picture of the AMPLATZER™ muscular ventricular septal defect occluder (arrow) 5 years later. Only the screw is visible and the rest of the device is completely endothelialized in good position. This was left untouched during the surgical homograft replacement

Our case is unique due to the patient's adult age, longer follow-up, and direct visualization of the device by the surgeon at conduit revision. The anatomic similarity of the stenosed native RVOT to the congenital muscular VSD (for which this device was initially intended) makes the AMPLATZER™ muscular VSD occluder an attractive choice for these kinds of procedures. Longer follow-up is necessary in all patients in whom devices are used in nonconventional ways.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Berger F, Ewert P, Björnstad PG, Dähnert I, Krings G, Brilla-Austenat I, *et al.* Transcatheter closure as standard treatment for most interatrial defects: Experience in 200 patients treated with the amplatzer septal occluder. Cardiol Young 1999;9:468-73.
2. Awasthy N, Tomar M, Radhakrishnan S, Shrivastava S. Unconventional uses of septal occluder devices: Our experience reviewed. Indian Heart J 2015;67:128-35.
3. Baspinar O, Irdem A, Kilinc M. Off-label use of amplatzer

devices in congenital heart disorders during childhood. Acta Cardiol 2013;68:31-5.

4. Tofeig M, Patel RG, Walsh KP. Transcatheter closure of a mid-muscular ventricular septal defect with an amplatzer VSD occluder device. Heart 1999;81:438-40.
5. Bilkis AA, Alwi M, Hasri S, Haifa AL, Geetha K, Rehman MA, *et al*. The amplatzer duct occluder: Experience in 209 patients. J Am Coll Cardiol 2001;37:258-61.
6. Fernando R, Koranne K, Loyalka P, Kar B, Gregoric I. Patent ductus arteriosus closure using an amplatzer(™) ventricular septal defect closure device. Exp Clin Cardiol 2013;18:e50-4.
7. Gioia G, Zheng J, Ray A, Gioia M. Perforated sinus of valsalva (PSOV) aneurysm closure with a muscular VSD occluder. Cardiovasc Revasc Med 2014;15:165-70.
8. Ece I, Uner A, Cuce F, Balli S. Transcathether closure of a right pulmonary artery-to-left atrial fistula using amplatzer muscular ventricular septal defect occluder. Cardiovasc Interv Ther 2014;29:359-62.
9. Rad EM, Arabi MY. Transcatheter closure of direct communication of main pulmonary artery to left atrium using muscular VSD occluder in an infant. Catheter Cardiovasc Interv 2011;78:276-9.
10. Thanopoulos BV, Georgakopoulos DJ, Tsaousis GS, Eleftherakis NG. A novel use of the amplatzer muscular ventricular septal defect occluder. Catheter Cardiovasc Interv 2002;56:234-7.
11. Momenah TS, El Houry MT, Khan MA, Galal MO. Post melody valve implant in a conduit with regurgitant native outflow tract successful closure by amplatzer muscular device. Cardiol Young 2015;25:324-7.
12. Dandolu BR, Baldwin HS, Norwood WI Jr., Jacobs ML. Tetralogy of Fallot with anomalous coronary artery: Double outflow technique. Ann Thorac Surg 1999;67:1178-80.
13. Tchervenkov CI, Pelletier MP, Shum-Tim D, Béland MJ, Rohlicek C. Primary repair minimizing the use of conduits in neonates and infants with tetralogy or double-outlet right ventricle and anomalous coronary arteries. J Thorac Cardiovasc Surg 2000;119:314-23.