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Case Report

Novel Method for Delivering the Amplatzer® Muscular VSD Occluder in a Patient With Double Outlet Right Ventricle After Bidirectional Glenn Procedure and Pulmonary Artery Band

Martin L. Bocks, MD and Aimee K. Armstrong, MD

We report the first use of bilateral femoral venovenous rail creation for the delivery of an Amplatzer® Muscular Ventricular Septal Defect Occluder in a patient with a large mid-to-apical muscular ventricular septal defect before Rastelli operation. The presence of a right-sided bidirectional Glenn shunt, a banded main pulmonary artery, and double outlet right ventricle anatomy precluded the use of standard delivery techniques. The patient underwent successful transcatheter device placement followed by Rastelli operation on the following day. © 2009 Wiley-Liss, Inc.

Key words: congenital heart defects; heart catheterization; heart septal defects; ventricular; infants; echocardiography; transesophageal

INTRODUCTION

The standard percutaneous technique for closing mid-to-apical muscular ventricular septal defects (mVSDs) with the Amplatzer® Muscular Ventricular Septal Defect Occluder (AMVSDO) has been well described [1–3]. This procedure involves either the formation of an arteriovenous loop from a femoral artery to the right internal jugular (RIJ) vein (Fig. 1a) or a venovenous loop from a femoral vein to the RIJ vein, using an existing interatrial communication or via transseptal atrial puncture. The device is typically delivered antegrade into the left ventricle (LV) from a right ventricular (RV) approach via the RIJ venous sheath. Retrograde delivery of the AMVSDO from the aorta can also be used, if kinking of the sheath is encountered when approaching from the RIJ vein [1], but this technique can lead to femoral arterial injury and aortic insufficiency in smaller children [1,4,5]. There are, however, particular congenital and post-surgical cardiac configurations that preclude the use of these standard delivery techniques. First, the presence of a cavopulmonary anastomosis, such as a bidirectional Glenn shunt (BDG), or of superior vena cava occlusion prohibits standard rail or loop formation, due to the lack of accessibility to the RIJ vein. Second, patients with unrepaired double outlet right ventricle (DORV) with D-malposed great arteries are not candidates for standard arteriovenous rail creation, because of the presence of the aorta on the RV side of the defect. Any rail involving the aorta requires crossing through both VSDs (RV→LV→RV) and could increase the risk of kinking the delivery sheath and of developing heart block. Lastly, creating the through-and-through loop is difficult in the presence of a main pulmonary artery band (PAB), which makes the standard location for snaring the wire inaccessible. We report a novel technique using a bifemoral venovenous loop, to deliver an AMVSDO in a patient with DORV, D-malposed great arteries, large subpulmonary VSD, and multiple apical and mid mVSDs, who had previously undergone right BDG and PAB in preparation for single ventricle palliation at an outside institution.

Division of Pediatric Cardiology, University of Michigan Health System, Ann Arbor, Michigan

Conflict of interest: Nothing to report.

*Correspondence to: Martin L. Bocks, MD, Division of Pediatric Cardiology, University of Michigan Health System, L1242 Women's, SPC 5204, 1500 E. Medical Center Drive, Ann Arbor, MI 48109-5204. E-mail: mbocks@umich.edu

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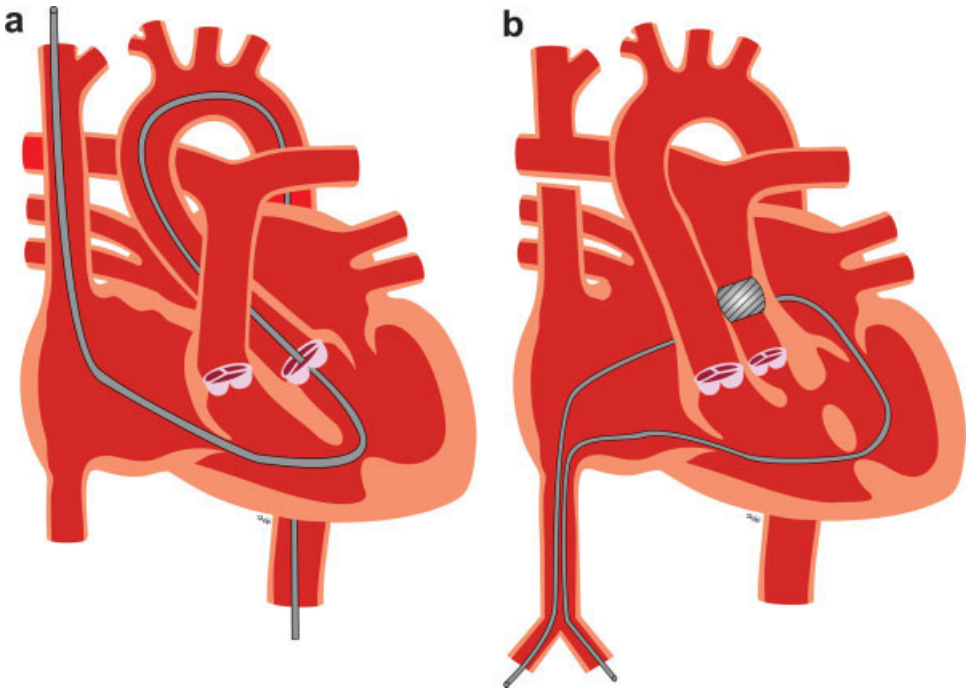

Fig. 1. (a) Diagram of standard arteriovenous loop formation for closure of apical and mid-muscular VSDs using the AMVSDO. (b) Diagram of bifemoral venovenous loop used in delivering the AMVSDO in this patient with DORV status-post PAB and BDG. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

CASE REPORT

A 23 month-old, 7.5 kg female was referred for transcatheter closure of a large mid-to-apical mVSD before undergoing Rastelli procedure. The patient was born prematurely at 30 weeks of gestation, weighing approximately 900 grams. Her cardiac diagnoses included DORV, D-malposed great arteries, a large sub-pulmonary VSD, multiple mVSDs, a large secundum atrial septal defect, an aberrant right subclavian artery, a bicuspid pulmonary valve, and mild subpulmonary stenosis. Her small size and the presence of abnormal tricuspid valve attachments to the crest of the outlet septum initially precluded her from a complex biventricular repair. She underwent PAB and atrial septectomy at 2 months of age at an outside institution. At 6 months of age, as part of a single ventricle palliation pathway, a right BDG was performed, and her banded pulmonary artery was left intact. The patient was subsequently referred to our institution for consideration of a biventricular repair. Upon evaluation of the patient's echocardiogram, it was determined that a Rastelli operation was possible, despite the presence of abnormal septal attachments of the tricuspid valve. The largest of the multiple mVSDs, a large mid-to-apical mVSD, was in a location that would make it difficult to close surgically. Because of the extensive nature of the proposed surgery, perventricular device placement was not considered, as not to prolong the length of the already complex operation. Therefore, she was referred for transcatheter closure of the largest of the mVSDs on the day before surgical intervention.

Preoperative 2D echocardiography confirmed the above described anatomy, with the largest mVSD measuring approximately 10 mm on the LV side (Fig. 2). Three dimensional transthoracic echocardiography further demonstrated that this defect was divided on the RV side by a prominent muscle bundle/moderator band. The muscle bundle divided the defect on the RV side into a 5–7-mm defect superiorly and a smaller 2–3-mm defect inferiorly.

The patient's aspirin was discontinued 2 days before the cardiac catheterization, as the surgery was planned for the following day. The cardiac catheterization was performed under general anesthesia with transesophageal echocardiographic (TEE) guidance. Access was obtained in the right femoral artery, bilateral femoral veins, and right subclavian vein. Heparin (100 units/kg) was administered, and activated clotting times (ACT) were maintained above 200 sec. A right and left heart catheterization was performed. Pulmonary artery systolic pressure was 12 mm Hg with a right ventricular pressure of 77/7 mm Hg, suggesting a tight PAB. A retrograde left ventriculogram was performed

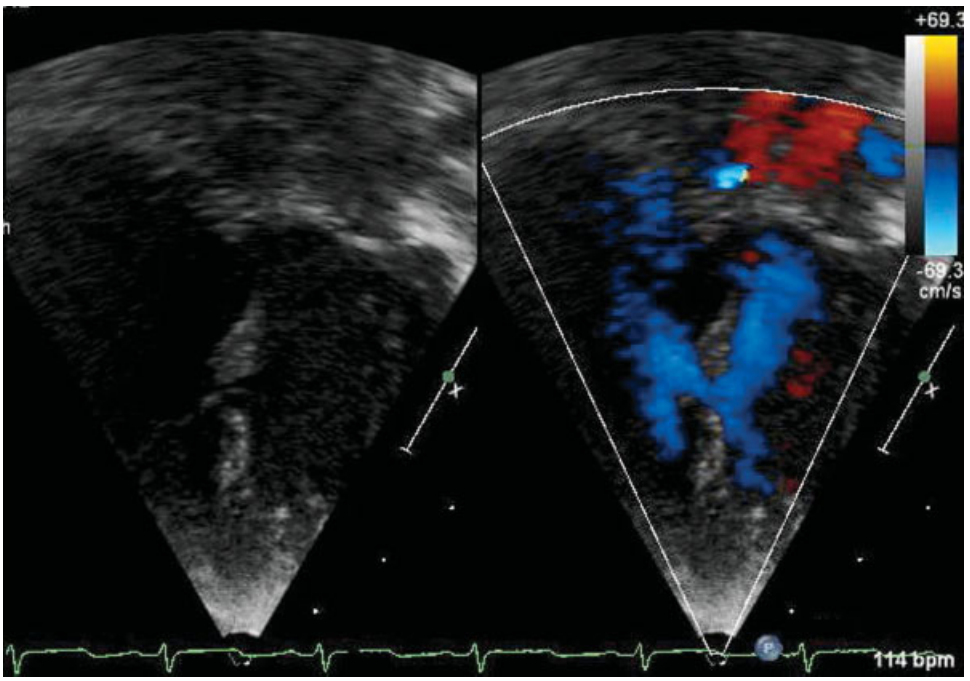

Fig. 2. Apical four-chamber view demonstrating muscular VSD in 2D and with color flow mapping. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

detailing the location and size of the mVSDs (Fig. 3). A 6-Fr balloon wedge catheter was placed into the left femoral venous (LFV) sheath and was advanced up through the atrial septal defect and into the apex of the left ventricle. A 0.035-inch stiff-angled Glidewire® (Terumo Medical, Somerset, NJ) was used to access the largest mVSD. Following over the wire the catheter was advanced into the RV and out the aortic valve to the ascending aorta. A second balloon wedge catheter was placed in the right femoral venous (RFV) sheath and advanced through the right atrium, into the RV, and out the ascending aorta. Care was taken to ensure that both balloons were inflated at all times while passing through the right ventricle and into the ascending aorta. A 10-mm Amplatz Goose Neck® Snare retrieval device (EV3, Plymouth, MN) placed in the RFV balloon catheter was then used to snare a 0.035-inch J-exchange guidewire from the LFV catheter. The guidewire was pulled across the aortic and tricuspid valves and exteriorized out of the RFV sheath, creating a bifemoral venovenous rail across the large mVSD (Fig. 1b).

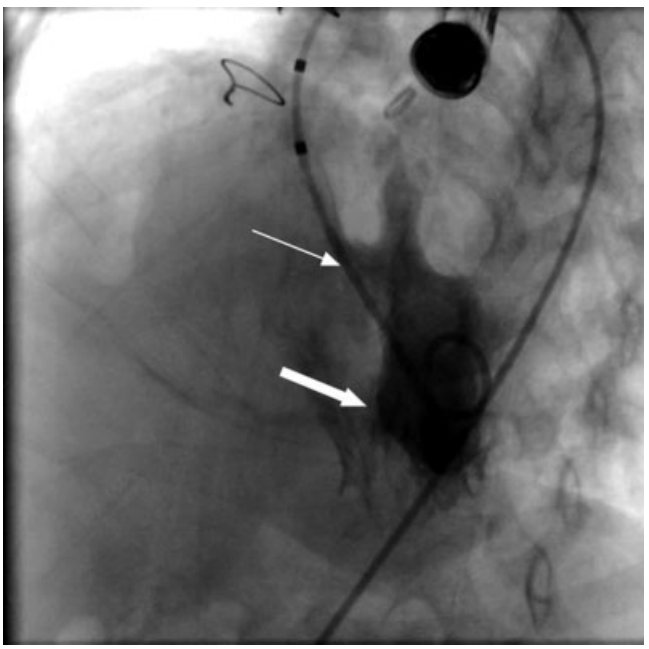

Fig. 3. Left ventriculogram in the long axial oblique projection revealing location of muscular VSD (thick arrow) and outlet VSD (thin arrow).

An 8-mm AMVSDO was selected for transcatheter closure based on angiographic and TEE measurements and taking into consideration the prominent right ventricular muscle bundle. A 7-French, 45 degree Amplatzer delivery sheath (AGA Medical, Golden Valley, MN) was prepared, and an 0.035-inch Amplatz Super Stiff™ exchange wire (Boston Scientific, Natick, MA) was advanced through the rail catheter antegrade across the tricuspid valve for a right-sided approach. Passage of the Super Stiff™ wire resulted in severe, reversible

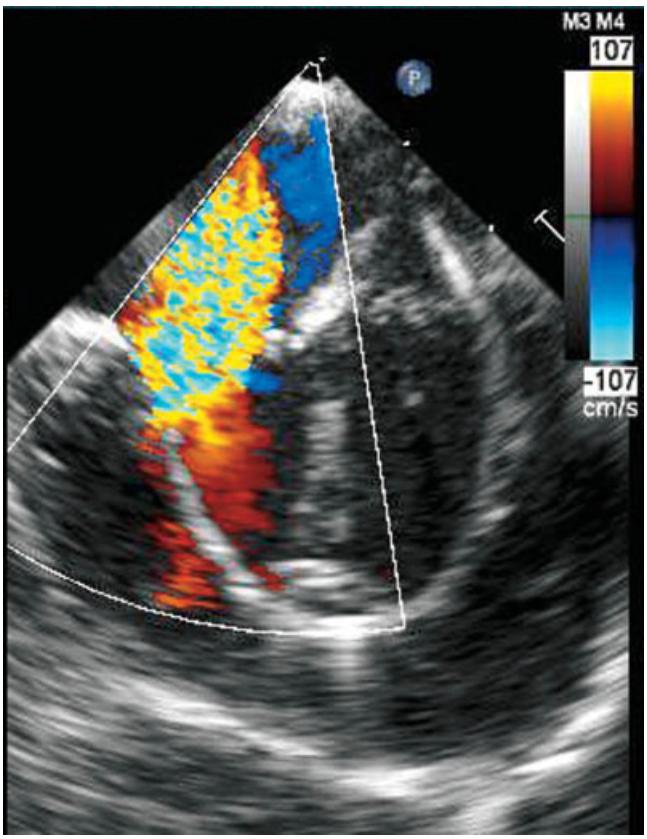

Fig. 4. Transesophageal echocardiogram with color flow mapping demonstrating severe TVR during use of Super-stiff exchange wire through the loop. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

tricuspid valve regurgitation (TVR) and subsequent systemic hypotension (Fig. 4). This wire was replaced with a less stiff 0.035-inch J-exchange guidewire (Argon Medical, Athens, TX), which was used to advance the delivery sheath across the mVSD into the LV with minimal TVR. Advancement of the AMVSDO toward the LV resulted in significant kinking of the sheath at the septum, despite keeping the LFV catheter and guidewire within the delivery sheath, using the "kissing" technique. The device and delivery cable were removed. A 0.018-inch Platinum Plus™ stiff wire (Boston Scientific, Natick, MA) was then loaded alongside the device and delivery cable and was advanced out the end of the delivery sheath and into the LFV catheter. This provided the needed stiffness with which to advance the AMVSDO across the ventricular septum without kinking of the delivery sheath. The device was then delivered in the mVSD under fluoroscopic and TEE guidance using techniques previously described [1–3]. Before releasing the device, the arterial angiographic catheter was advanced retrograde into the left ventricle, and an angiocardiogram was performed. The presence of the

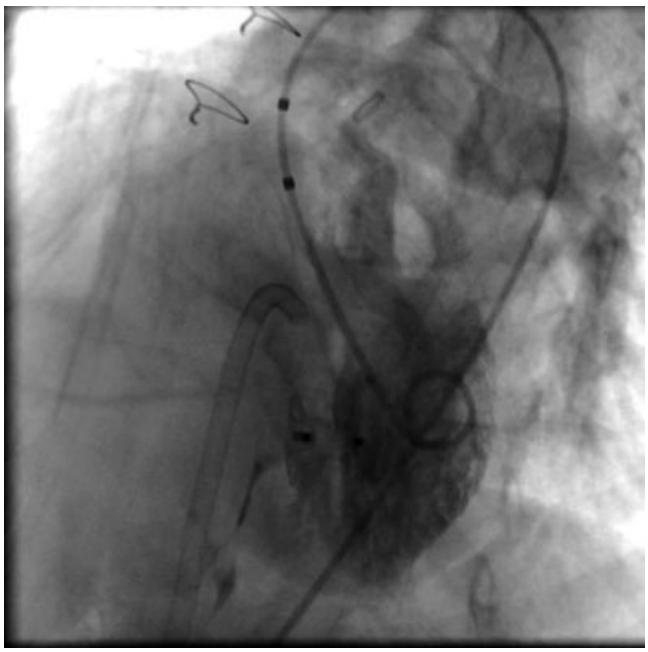

Fig. 5. Repeat left ventriculogram after AMVSDO delivery showing proper placement of the device across the large muscular VSD.

large subpulmonary VSD made it difficult to discern the degree of residual shunt through the mid-to-apical mVSD, but the TEE suggested that there was significantly less mVSD shunt with the AMVSDO in place. The device was subsequently released from the delivery cable. After waiting a period of 10 min, a left ventriculogram revealed good placement of the device and less residual shunting across the apical portion of the ventricular septum (Fig. 5). On the following day, the patient underwent Rastelli procedure with surgical enlargement of the outlet VSD, atrial septal defect closure, and placement of a 14-mm Contegra® heterograft (Medtronic, Minneapolis, MN) from the right ventricle to the distal end of the divided main pulmonary artery. Direct pressure measurements at the end of the surgical repair revealed a RV systolic pressure of 35 mm Hg and a LV systolic pressure of 90 mm Hg. Intraoperative TEE revealed multiple small residual leaks at the VSD patch margin, with estimated cumulative shunt being small-to-moderate. The remainder of the hospital course was unremarkable, and the patient was discharged from the hospital on the eighth postoperative day.

DISCUSSION

Surgical closure of multiple mVSDs, especially in patients with other associated complex congenital heart defects, is often associated with significant morbidity and mortality. Because of the difficulty in accessing and completely closing these defects, multiple mVSDs continue

to pose a major challenge to the surgeon and cardiologist despite improvements in surgical technique and pre- and intraoperative echocardiographic imaging [6–9]. The development of devices specifically designed to close mVSDs via the transcatheter route has provided an alternative approach to surgical closure and a means for patients potentially to avoid surgery altogether. With the transcatheter approach, it has been well established that mid and apical mVSDs are best closed from a rail system that includes RIJ venous access. This provides the smoothest sheath course with the fewest number of bends and usually results in less delivery sheath kinking [1,10,11]. As the hybrid approach for repairing both simple and complex congenital heart defects becomes more common, novel methods of intervention and device delivery need to evolve. Although the use of the bifemoral venovenous approach has been reported in a patient with a membranous VSD and prosthetic aortic valve to deliver the AMVSDO, we report the first use of the bifemoral venovenous rail to deliver this device into a mid-to-apical mVSD [12]. Although technical success was achieved in this case, several observations are worth noting.

First, atrioventricular valve regurgitation, specifically TVR, is a known complication of AMVSDO delivery [13–15]. It is usually secondary to the wire loop or sheath crossing through the chordae tendineae of the tricuspid valve and is rarely permanent or severe. Irreversible TVR, however, has been reported when device delivery results in entrapment of the chordal apparatus or when the device directly impinges on the tricuspid valve leaflets [3,14–16]. Despite making efforts to establish a wire loop that did not involve the chordae of the tricuspid valve in this case, the bifemoral venovenous loop still impinged on the tricuspid valve and caused reversible, severe TVR. The acute wire angle from the inferior vena cava across the tricuspid valve and down into the mid-to-apical portion of the RV put significant stress on the posterior and lateral leaflets of the tricuspid valve. This stress was increased with the use of the very stiff Amplatz Super Stiff guidewire, and in our patient with unrepaired DORV, severe and acute TVR resulted in considerable systemic hypotension. A less stiff wire, such as a standard J-exchange wire or the Amplatz Noodle™ wire, results in less stenting-open of the tricuspid valve leaflets.

Second, delivery sheath kinking, which is a well known problem with AMVSDO delivery, was encountered due to the acute angle formed when crossing the VSD [1,3,10,14,17]. Although it was not available at the time of this case, the more recently developed Amplatzer TorqVue Delivery and Exchange System (AGA Medical, Golden Valley, MN) offers greater flexibility and resistance to kinking and is now the recommended sheath for delivering the AMVSDO. In addition, the use of a 180 degree sheath may have caused less kinking than the 45 degree sheath. As has been previously reported, using a 0.018-inch stiff wire alongside the delivery cable provided the necessary stiffness to successfully position the device in the left ventricle [1,10].

Lastly, perventricular delivery of the AMVSDO should be considered for patients with complex anatomy when the ideal route of device delivery is not an option. The safety and efficacy of perventricular delivery of the AMVSDO when additional cardiac surgeries are required has been well established [15,17,18]. Bacha, et al. reported the largest series of patients ($n = 10$) in whom simultaneous perventricular AMVSDO delivery and cardiac surgical interventions were performed [15]. Most of the additional procedures performed included takedown of PAB, pulmonary artery augmentation, and coarctation repair. One patient in the series had congenital heart defects similar to the patient in this case, including previous surgical intervention with BDG and PAB. This patient underwent perventricular AMVSDO placement after initial VSD enlargement and interventricular baffling of the left ventricle to the aorta. Preoperatively, the defect was thought to be insignificant but, in the operating room, was found to be causing a large residual left-to-right shunt that prevented weaning from cardiopulmonary bypass. The device was placed in the operating room, using the perventricular approach. To our knowledge, there has been no patient described in the literature in whom the preoperative plan included perventricular device placement simultaneously with such a complex surgery. Although a perventricular approach may have made device delivery technically easier, preoperative closure of this remote mVSD allowed the surgeons to spend valuable time focusing on the more complex part of the operation.

CONCLUSIONS

A transcatheter bifemoral venovenous rail can be used for successful delivery of the AMVSDO into mid-to-apical mVSDs in patients with complex congenital heart lesions, including DORV and particularly when the RIJ vein is not accessible. Severe TVR caused during delivery sheath placement can be overcome by using a less stiff guidewire, and sheath kinking can be prevented with the use of a kink resistant delivery sheath. Perventricular device delivery should be considered in patients with complex congenital heart disease, along with novel methods of transcatheter delivery.

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