

## Multicenter Experience with Periventricular Device Closure of Muscular Ventricular Septal Defects

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**Abstract.** Hybrid procedures are becoming increasingly important, especially in the management of congenital heart lesions for which there are no ideal surgical or interventional options. This report describes a multicenter experience with periventricular muscular ventricular septal defect (VSD) device closure. Three groups of patients ( $n = 12$ ) were identified: infants with isolated muscular VSDs ( $n = 2$ ), neonates with aortic coarctation and muscular VSDs ( $n = 3$ ) or patients with muscular VSDs and other complex cardiac lesions ( $n = 2$ ), and patients with muscular VSDs and pulmonary artery bands ( $n = 5$ ). Via a sternotomy or a subxyphoid approach, the right ventricle (RV) free wall was punctured under transesophageal echocardiography guidance. A guidewire was introduced across the largest defect. A short delivery sheath was positioned in the left ventricle cavity. An Amplatzer muscular VSD occluding device was deployed across the VSD. Cardiopulmonary bypass was needed only for repair of concomitant lesions, such as double-outlet right ventricle, aortic coarctation, or pulmonary artery band removal. No complications were encountered using this technique. Discharge echocardiograms showed either mild or no significant shunting across the ventricular septum. At a median follow-up of 12 months, all patients were asymptomatic and 2 patients had mild residual ventricular level shunts. Periventricular closure of muscular VSDs is safe and effective for a variety of patients with muscular VSDs.

**Key words:** Congenital heart defects — Heart septal defects

Management of muscular ventricular septal defects (VSDs) remains suboptimal [11, 15, 16]. Isolated muscular VSDs are increasingly being successfully closed in the catheterization laboratory [9, 17]. However, when encountered in small infants or patients with poor vascular access, catheter closure remains challenging. Managing multiple muscular VSDs remains a challenge for both surgeons and interventionalists, sometimes culminating in the need for heart transplantation [11, 14, 15]. Left ventriculotomy or the division of multiple important right-sided muscle bands is associated with long-term morbidity [11, 15, 16]. On the other hand, placing several devices percutaneously across the ventricular septum during several sessions in the catheterization laboratory can also result in significant arrhythmias, hemodynamic compromise, tricuspid valve injury, or incomplete closures [17]. Furthermore, the presence of multiple muscular VSDs has been found to be an independent risk factor for early mortality after repair of complex congenital heart lesions such as double-outlet right ventricle (DORV) or transposition of the great arteries (TGA) [5, 12]. Therefore, eliminating this risk factor prior to complex repair is beneficial, especially if it can be done in one setting in the operating room.

Hybrid therapies aim to combine the advantages of surgical and interventional techniques in an effort to reduce the “invasiveness” (cardiopulmonary bypass, cardioplegic arrest, incisional trauma, groin vessel injury, risk to the tricuspid valve, etc.) of a given procedure [7]. The combination of interventional and surgical techniques has been tried before

but with limited success. Cardiopulmonary bypass and cardioplegic arrest were needed in all cases and VSD devices were deployed under direct vision on the arrested heart [8, 10, 13, 14]. Results were not satisfactory, mainly due to residual shunting, with mortality rates ranging from 14 to 25%.

This report summarizes the combined experience with periventricular muscular VSD closure acquired by three separate groups actively engaged in hybrid techniques. The device used was the Amplatzer muscular VSD occluder (AGA Medical Corporation, Golden Valley, MN, USA [9]), a self-expandable double-disc device made from nitinol wire mesh. The device size corresponds to the waist diameter of the device, with the length being 7 mm, corresponding to the thickness of the muscular septum in most infants and children.

## Methods

This study started in September 2002. Informed consent was obtained from the patients' guardians. The study was approved by each hospital investigation review board. Excluding one patient from Chile, this study was part of a Food and Drug Administration Investigation Device Exemption clinical trial.

Selection criteria were as follows:

Group 1: Small infants with muscular VSD(s) or patients with poor vascular access

Group 2: In addition to one or multiple muscular VSDs, the presence of cardiac lesions necessitating operative repair

Group 3: Patients with muscular VSDs and pulmonary artery (AA) bands

Standard pediatric cardiac surgical intra- and postoperative monitoring, protocols, and equipments were used.

### *Description of the Technique*

The heart was approached via a median sternotomy or a subxyphoid minimally invasive incision without sternotomy. Under continuous transesophageal echocardiography (TEE) guidance, the best location for right ventricular (RV) puncture was chosen, paying being attention away from any papillary muscles but far enough from the septum so as to approach it from a perpendicular angle with the needle and wire (Fig. 1). A 5-0 polypropylene purse-string was placed at the chosen location. An 18-gauge needle (Cook, Bloomington, IN, USA) was introduced into the RV cavity and a 0.035-in. angled glide wire (Boston Scientific, Medi-Tech, Natick, MA, USA) was passed through the needle and manipulated into the left ventricular (LV) cavity through the defect. A 7- to 10-Fr short (8-13 cm) introducer sheath with a dilator was fed over the wire and carefully advanced into the LV cavity. Care must be taken to keep the tip of the dilator in the middle of the LV cavity (monitored by TEE) because it could perforate the LV free wall. The dilator was removed and the sheath tip positioned in the LV cavity. The appropriate device size was chosen to be 1 or 2 mm larger than the VSD size as assessed by color doppler TEE. The device was presoaked in nonheparinized patient's blood for 10

minutes to allow for the tiny fenestrations of the nitinol mesh to thrombose. The device was then screwed to the cable and pulled inside a 6- to 9-Fr loader under blood seal to prevent any air bubbles. The device was advanced inside the short delivery sheath until it was seen by TEE to be close to the tip of the delivery sheath. The LV disc was deployed in the mid-LV cavity by gentle retraction of the sheath over the cable. The entire assembly (cable/sheath) was withdrawn gently until the LV disc was against the septum. Further retraction of the sheath over the cable would deploy the waist inside the septum. Continuous TEE to confirm the device position is of extreme importance. Once the position was confirmed, further retraction of the sheath to expand the RV disc was performed. If the device position was satisfactory, the device was released by counterclockwise rotation of the cable using the pin vise. A complete TEE study in multiple planes was done to confirm device placement and assess for residual shunting and any obstruction or regurgitation induced by the device.

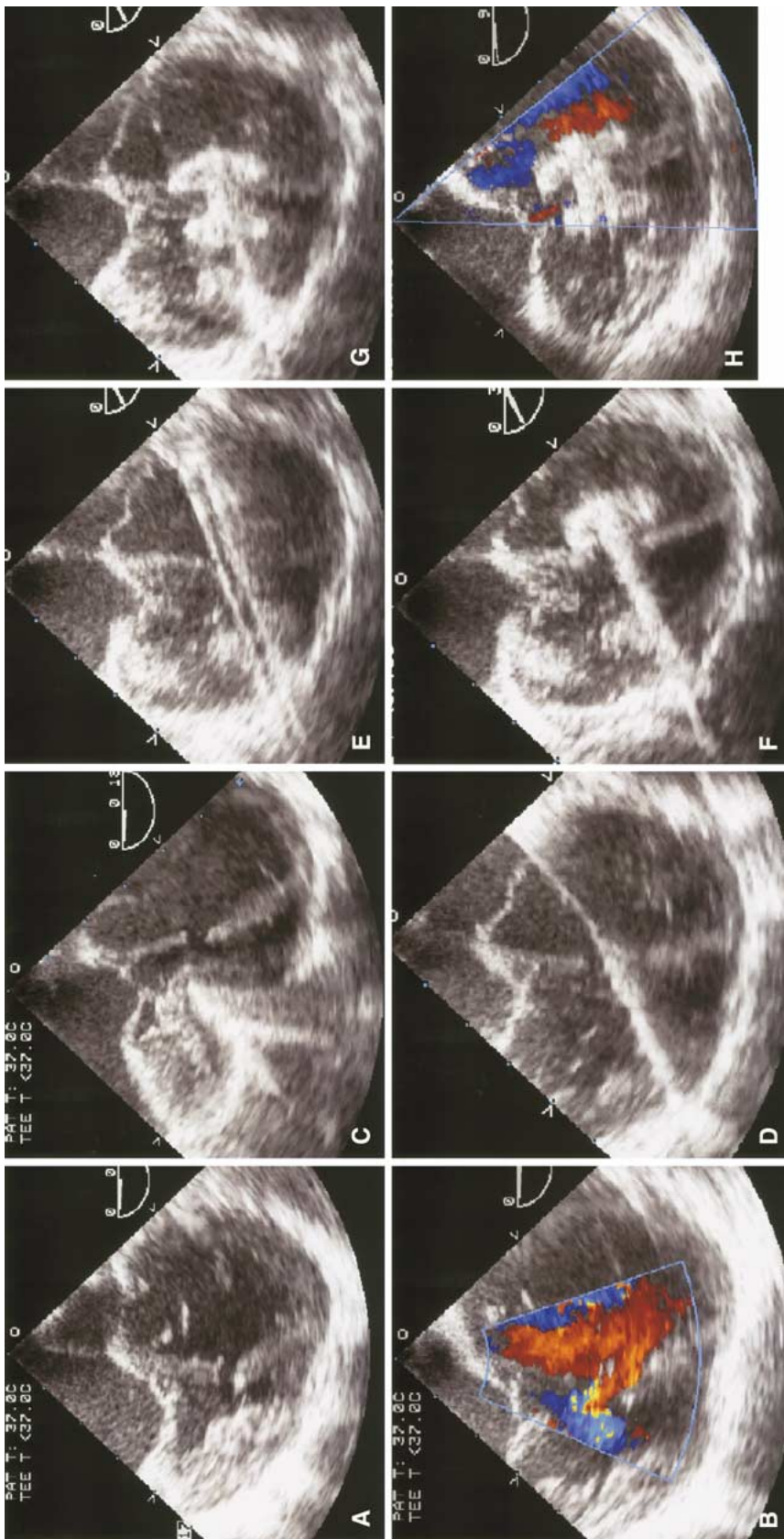
## Results

Table 1 lists the clinical details. Six patients have been presented previously [6]. The procedure was attempted in 13 patients and was successful in 12. One 3-kg neonate with a large high posterior muscular VSD was converted from a periventricular approach to an open-heart approach due to the inability to open the RV disc without impingement of the septal leaflet of the tricuspid valve. The repair was successfully completed surgically. One patient with a posterior muscular VSD had a peratrial approach with complete closure after attempts to cross the VSD via a periventricular approach were unsuccessful. There were no intraoperative complications. One patient needed reintubation for pulmonary reperfusion injury secondary to a PA band takedown. One neonate with CoA/VSD had postoperative mediastinitis that required sternal plating and a prolonged intensive care unit stay. At a median follow-up of 12 months (range, 3-23), all patients were asymptomatic. One patient in group 3 has mild to moderate shunting across separate small apical defects, and one mild patient has mild shunting across the edge of the device. Neither patient has echocardiographic or clinical evidence of significant congestive heart failure, volume overload, or pulmonary hypertension.

## Discussion

The first successful case of intraoperative periventricular device closure on the beating heart in an infant was reported in 1998 [2]. Subsequently, this technique's safety has been validated in animal experiments [3, 4], and it has even been successfully applied for periventricular membranous VSD closure in animals [1].

Using the technique described here, we have identified four groups of congenital heart disease patients that can be better managed with a hybrid approach. This is the largest reported series of per-



**Fig. 1.** (A) Four-chamber view of a mid- to posterior muscular ventricular septal defect (VSD) via intraoperative transesophageal echocardiography (TEE). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. (B) Color Doppler flow across the VSD. (C) Using a forceps tip, the right ventricular free wall is probed to determine the shortest distance from the free wall to the VSD, staying away from the anterior papillary muscle of the tricuspid valve. (D) A needle is introduced into the right ventricular cavity and a guidewire manipulated into the left ventricle. (E) An introducer sheath is passed over a dilator and positioned into the left ventricular cavity. (F) The device is pushed into the sheath and the LV disc deployed by gently retracting the sheath. (G) The RV disc is similarly deployed, but the device is still attached to the delivery cable. This allows for TEE confirmation of valve function and VSD closures, as well as recapture of the device into the sheath (by advancing the sheath), necessary. (H) Final confirmation of lack of shunting.

**Table 1.** Clinical data (11 patients)

| Age/weight   | Diagnosis  | Pre-op. Symptoms                      | Exposure         | Device size (mm)     | Additional procedures                         | CPB <sup>a</sup> | Cardioplegic arrest | Intraop. complications                             | $Q_p/Q_s$ | Post-op. complications             | D/C day | Echo and status at followed (months)   | Meds at follow-up           |
|--|--|---------------------------------------|------------------|----------------------|---|------------------|---------------------|--|-----------|------------------------------------|---------|--|-----------------------------|
| <b>Group 1: Isolated muscular VSD in infants</b>         |  |                                       |                  |                      |   |                  |                     |  |           |                                    |         |  |                             |
| 17 days/3 kg   | Large anterior musc. VSD   | CHF                                   | Xyphoid incision | 12                   | No  | No               | No                  | No   | 1.2       | No                                 | 4       | No shunt, asympt. (16)   | None                        |
| 4 months/4 kg  | Anterior musc. VSD, ASD  | FIT, CHF                              | Xyphoid incision | 10 (VSD)<br>11 (ASD) | No  | No               | No                  | No   | 1.2       | No                                 | 3       | No shunt, asympt. (17)   | None                        |
| <b>Group 2: Muscular VSD with other surgical lesions</b> |  |                                       |                  |                      |   |                  |                     |  |           |                                    |         |  |                             |
| <b>Group 2a: CoA/musc VSD in neonates</b>                |  |                                       |                  |                      |   |                  |                     |  |           |                                    |         |  |                             |
| 14 days/3 kg   | Hypoplastic aortic arch + coarctation, mid-musc. VSD                   | Intubated, on PGE                     | Median sternot.  | 8                    | Coarctation repair and arch augmentation      | Yes              | No                  | No   | 1.2       | No                                 | 18      | No shunt, asympt. (12)   | None                        |
| 20 days/3 kg   | Aortic CoA + arch hypopl., large ant. musc. VSD, LV hypoplasia         | Metabolic acidosis, cardiogenic shock | Median sternot.  | 6                    | Coarctation repair and arch augmentation      | Yes              | No                  | No   | 1.1       | No                                 | 20      | tiny apical VSD, asympt. (19)  | None                        |
| 14 days/3kg  | Hypoplastic aortic arch + coarctation, mid-musc. VSD                   | Metabolic acidosis, cardiogenic shock | Median sternot.  | 8                    | Coarctation repair and arch augmentation      | Yes              | No                  | No   | 1.1       | Yes (mediastinitis)                | 90      | No shunt, asympt. (4)  | None                        |
| <b>Group 2b: Other infants and children</b>              |  |                                       |                  |                      |   |                  |                     |  |           |                                    |         |  |                             |
| 3 years/20 kg  | DORV, TGA, inlet VSD, apic. musc. VSD, hypopl. LV, s/p PA band and BDG | No                                    | Median sternot.  | 14                   | VSD enlargement and LV aortic baffle, t/d BDG | Yes <sup>b</sup> | Yes                 | LV failure, ECMO; t/d of repair, removal of device | 1.1       | Pleural effusions                  | 20      | Widely patent ASD and VSD, asympt. (15)  | Captopri<br>Lasix           |
| 5 months/7 kg  | DORV, subaortic VSD, subPS/PS, mult. apical VSDs                       | No                                    | Median sternot.  | 8                    | Subaortic VSD patch, RVOT reconstruction      | Yes              | Yes                 | No   | 1.2       | No                                 | 6       | No shunt, asympt. (22)   | None                        |
| <b>Group 3: Muscular VSDs s/p PA band</b>                |  |                                       |                  |                      |   |                  |                     |  |           |                                    |         |  |                             |
| 3 years/15 kg  | S/p PA band for post musc. VSD   | No                                    | Median sternot.  | 14                   | Band removal and PA plasty                    | Yes              | No                  | No   | 1.1       | No                                 | 4       | No residual VSD, asympt. (11)  | None                        |
| 1 year/8 kg  | S/p PA band for post. musc. VSD + mult. apical VSDs                    | No                                    | Median sternot.  | 8                    | Patch closure post. Musc. VSD and PA plasty   | Yes              | Yes                 | No   | 1.1       | No                                 | 5       | Min. residual apical shunt, asympt. (15)   | None                        |
| 4.3 years/6.9 kg   | s/p PA band; s/p BTS, mult. apical musc. VSDs                          | FIT                                   | Mid sternot.     | 12                   | PA band and BTS take down                     | Yes              | No                  | No   |           |                                    | 8       | Multi. separate anterior apical VSDs with small-moderate shunt (not through devices), asympt. (16) | Digoxin<br>Lasix<br>Aspirin |
| 2.5 years/12 kg  | s/p PA band for mult. apical VSDs                                      | No                                    | Median sternot.  | 18                   | Band removal and PA plasty                    | Yes              | No                  | Difficult deployment of RV disk <sup>c</sup>       | 1.5       | POD 1: reperfusion pulmonary edema | 7       | No residual VSD, asympt. (17)  | Lasix                       |

| 2 years/13 kg | s/p PA band for large posterior musc. VSD | No | Median sternot. | 14 | Band removal and PA plasty | Yes | No | Needed peratrial approach after failed perventricular approach | 1.1 | No | 7 | No residual VSD, asympt. (6) | None |
|---------------|---|----|-----------------|----|----------------------------|-----|----|--|-----|----|---|------------------------------|------|
|---------------|---|----|-----------------|----|----------------------------|-----|----|--|-----|----|---|------------------------------|------|

ASD, atrial septial defect; BDG, bidirectional Glenn shunt; BTS, Blalock-Taussig shunt; CHF, congestive heart failure; coarct, coarctation; CPB, cardiopulmonary bypass; D/C, discharge; FTT, failure to thrive; hypopl., hypoplasia; LV, left ventricle; PA, pulmonary artery; PS, pulmonary stenosis; POD, postoperative day; reperf., reperfusion; RV, right ventricle; RVOT, right ventricular outflow tracts; s/p, status post; t/d, takedown; VSD, ventricular septal defect.

<sup>a</sup>Cardiopulmonary bypass was needed only for additional procedures, not for placement of the device.

<sup>b</sup>Failure of biventricular repair due to LV hypoplasia led to repair takedown to an extracardiac Fontan and removal of device.

<sup>c</sup>The LV disc obliterated the entire left apical septum, resulting in no residual shunting. However, the RV disc was difficult to deploy due to a severely hypertrophied moderator band and apical RV muscle bundles and prevented the expansion of the RV disc. The RV disc was eventually positioned within the apical muscle bundles with the device microscrew protruding through the RV free wall puncture site. The screw was secured to the epicardium with a pledgetted suture.

ventricular muscular VSD device closure. The technique does not require cardiopulmonary bypass (CPB) support or full sternotomy. The heart can be approached via small incisions, thus providing the possibility of performing a large variety of peratrial or perventricular procedures. The technique is simple and requires mainly excellent collaboration between echocardiographer, surgeon, and interventionalist. Although not formally measured, the time needed to cross the VSD and position the device was less than 30 minutes in all cases. This compares very favorably with the much longer procedure times when the equivalent maneuver is attempted in the catheterization laboratory. The advantages of this technique over standard surgical techniques include avoidance or reduction of CPB time. Other advantages include avoidance of transection of the moderator band or other RV muscle bundles, immediate confirmation of adequate closure, and avoidance of any ventricular incisions. The technique is not limited by low weight or vascular access problems. Particularly in malformations with unusual septal planes, such as DORV or TGA, the percutaneous retrograde approach to cross muscular VSDs can be very challenging if not impossible. This technique eliminates that issue because the septum is approached from a perpendicular and not a parallel (i.e., tricuspid valve) plane. As opposed to a percutaneous approach, the perventricular approach offers a "straight shot" at the lesion to be addressed, with no wire kinks or loops to be negotiated by the sheath or the device. Delivery of self-expandable valves may also be simplified by this technique [18].

Based on our data, we believe that palliating symptomatic infants with large muscular VSDs with PA banding is not indicated anymore. Irrespective of weight, these infants can undergo perventricular VSD closure if they are not candidates for a percutaneous approach. This approach is also indicated for neonates with aortic coarctation and large muscular VSDs, where a "one-stage" repair via sternotomy can be applied. The technique also offers the possibility to do a one-stage PA band takedown, identify all VSDs under nonbanded conditions, and close them using the perventricular approach. However, in this setting, severely hypertrophied RV muscle bundles can make the recognition of the true VSD margins difficult and render the correct deployment of the RV disc more challenging. Muscular VSDs that are in close proximity to atrioventricular valves, such as high posterior muscular VSDs present a unique challenge. As seen in one of the patients, they may be better approached via a peratrial puncture, allowing the wire and the device to glide under the tricuspid subvalvar apparatus. The muscular septal thickness is typically only 4 or 5 mm. The waist of the current Amplatzer

muscular VSD occluder is 7 mm long, making it too long for this location, with likely interference with tricuspid or mitral valve function, seen in the one patient in whom the technique was unsuccessful. A custom-made device may alleviate this problem in the future.

The limitations of this study include a small patient sample, short follow-up, and the lack of randomization.

In conclusion, this multicenter study demonstrates that perventricular muscular VSD closure is safe and effective for a variety of patients with muscular VSDs. With advances in introducer and robotic technologies, peratrial or perventricular procedures may be performed via port approaches.

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