48 Hybrid Approach: Defect Closure

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48.1 Anatomical Considerations

The hybrid approach to ventricular septal defect (VSD) closure is applicable in a wide range of muscular VSDs. While hybrid closure is possible in most muscular VSDs, the anatomical position will influence the approach to closure and may limit the ability to appropriately position a closure device [1–3].

48.2 Indications and Patient Selection

Whenever a hybrid approach to VSD closure is considered, it should always prompt careful review of all the approaches to VSD closure including transcatheter, hybrid and traditional surgical closure.

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48.3 Indications

1. *The patient is too small to consider transcatheter closure of the septal defect.* Transcatheter VSD closure is usually reserved for patients greater that 10 kg. Most children with haemodynamically significant VSDs (causing heart failure or a risk of pulmonary vascular disease) require closure within the first 6 months of life and are too small to allow predictable morbidity-free success with a transcatheter approach.
2. *There is a relative contraindication to cardiopulmonary bypass.* This may be due to an ongoing neurological concern or a thrombotic or thrombophilia tendency. In the vast majority of cases, hybrid VSD closure is performed without cardiopulmonary bypass.
3. *The anatomical location of the defect is such that a surgical or transcatheter approach may be difficult.* Defects whose RV exit points are placed in the more extreme regions of the ventricular septum, such as apical and anterior mid-muscular and those closely associated with the moderator band, may be more amenable to a hybrid approach.

48.4 Typical Clinical Scenario

A 4.8 kg infant following surgical repair of a perimembranous VSD is unable to progress from ITU respiratory support despite maximal anti-failure treatment. The chest radiograph is consistent with a large left-to-right shunt. The echocardiogram shows a dilated left side of the heart, a surgically repaired perimembranous VSD and a significant left-to-right flow across the muscular ventricular septum. A haemodynamically significant additional muscular VSD is found apical to the moderator band and measures 5 mm. The tricuspid valve regurgitant velocity suggests an RV pressure at least 75 % of systemic pressure.

48.5 Treatment Options

This patient should be considered for muscular VSD closure when appropriate aggressive medical management has failed.

Option 1: surgical device closure. The patient is within 2 weeks of their initial cardiopulmonary bypass run. The position of the defect in such a small infant is likely to provide a major challenge to the surgeon.

Option 2: percutaneous transcatheter device closure. Although theoretically feasible, the practicalities at this weight in this clinical setting are unfavourable. Even if the defect can be crossed from the left side with a wire and catheter, manipulating a delivery sheath through the right side of the heart without major haemodynamic instability and accurate device delivery is likely to be impossible.

Option 3: hybrid periventricular VSD device closure. Given the patient's weight, position of the defect and the clinical condition, this is an attractive option.

48.6 Pre-procedural Imaging

Adequate imaging is usually possible with high-quality transthoracic echocardiography alone at this age. Transoesophageal echocardiography (TOE) and 3D echo imaging can add useful information in delineating the shape of the defect and allowing a better understanding of its orientation on the septal surfaces. Angiographic delineation of the ventricular septum may be particularly useful in larger patients with complex multiple defects, but is unlikely to add much at this age. The key features which need to be recognised and discussed are:

1. Size and position of the target lesion.
2. Relationship to structures such as the moderator band (the defect may straddle the moderator band), the tricuspid valve and its septal attachments and the mitral valve apparatus.

3. Proximity to the apex and the cavity size on either side of the defect (i.e. how much space is available to deploy the left and right discs).
4. The presence and significance of any additional defects – do these also require closure? If not, then they need to be recognised to ensure that the correct defect will be crossed.

The imaging data is carefully scrutinised by the interventionist, the surgeon and the echocardiographer to plan the procedure and the equipment inventory.

48.7 Technique (Step-by-Step)

1. The ideal place is in a fully specified hybrid operating facility. A full description of this can be found elsewhere; however, in brief biplane angiographic imaging equipment should be available in case angiography becomes necessary during the case. The room should have full cardiopulmonary bypass and deep hypothermic circulatory arrest capabilities. TOE imaging is the key imaging modality in these cases and angiography is rarely necessary. Epicardial echocardiography can provide additional useful guidance (Figs. 48.1 and 48.2).
2. When the cardiac position and connections are normal, a sternotomy will usually be the correct approach; however, a thoracotomy or subxiphoid approach may be used in cases where the anatomical orientation is favourable. Exposure of the right ventricular surface is usually adequate, allowing a "limited" sternotomy to be used. Cardiopulmonary bypass should not be necessary in uncomplicated cases.
3. After locating and delineating the defect on TOE, the correct position to puncture the right ventricle is identified. A combination of angle towards the septum, cavity space for device deployment, proximity to the moderator band and the space constraints for the operators to manipulate the catheters and sheaths needs to be considered. Practically, this is done by indenting different parts of the RV free wall with a finger while observing the TOE image.
4. Prior to puncturing the RV, the occlusion device is selected, prepared and loaded, ready for insertion into the sheath. The

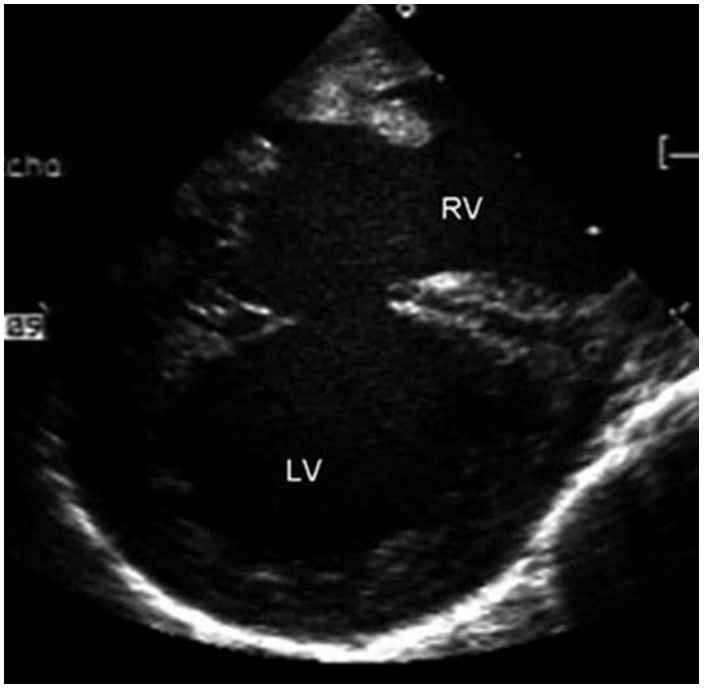

Fig. 48.1 A large mid-muscular VSD delineated with epicardial echocardiography in a 4.8 kg patient. Epicardial echo can be useful as the probe can be used to mimic the desired angle and direction for the wire and sheath passage. *RV* right ventricle, *LV* left ventricle

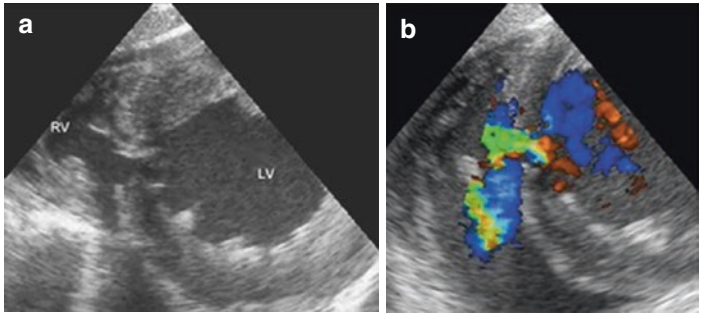

Fig. 48.2 TOE view of a moderate mid-muscular VSD with 2D imaging (**a**) and colour flow Doppler (**b**). Note that the TOE view gives a less 'surgical orientation' of the defect and requires more spatial awareness from the operators compared with the epicardial scan. *RV* right ventricle, *LV* left ventricle

correct device size usually has a waist diameter of 2 mm larger than the maximum measured diameter of the defect. The most frequently used device is the St. Jude AMPLATZER Muscular VSD Occluder; however, VSD occluders by other manufacturers are available. In certain anatomical variants, other device designs such as that used for patent ductus occlusion may be more appropriate although this would be "off-label" use.

5. A purse string is placed on the RV free wall and heparin 100 units/kg is administered. Under TOE guidance the RV is punctured with an 18 g needle and a 0.035″ Terumo J-Tip hydrophilic guidewire guided across the defect into the LV cavity. The guidewire is ideally directed out the left ventricular outflow tract to avoid interference with the mitral papillary muscles and away from the posterior wall of the LV. A short (7.5–15 cm) sheath, large enough (usually 6–10 F) to accommodate the chosen device, is advanced over the wire and across the VSD to the LV cavity (Fig. 48.3). Depending on the anatomy, the VSD may be difficult to cross with the puncture needle and wire. Although attempting to direct the wire with a catheter and wire combination, it is likely that the RV free wall

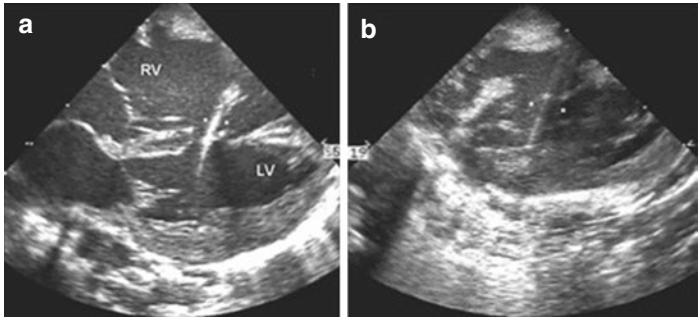

Fig. 48.3 With epicardial imaging the defect shown in Fig. 48.1 has been crossed with the sheath and wire and this has been followed with the dilator and sheath, delineated by the *asterisks* (**a**). After the dilator and wire have been removed, the 'train-track' appearance of the empty sheath is seen (**b**). *RV* right ventricle, *LV* left ventricle

puncture point is suboptimal and needs to be redone. A perpendicular approach from the free wall to the ventricular septum is required so as not to distort the anatomy and enable successful deployment.

6. Using TOE guidance, the LV disc is deployed in the mid-cavity and withdrawn to oppose the disc onto the septum (Fig. 48.4). The waist of the device and subsequently the RV disc are uncovered by withdrawal of the sheath. Several attempts may be needed to conform the RV disc correctly; it is therefore important not to pull the sheath out of the RV during the initial deployment. Indeed the RV disc may not completely conform on the RV septal aspect due to trabeculations, moderator band and limited chamber size near the apex. The operators must then decide whether the RV disc has formed adequately to allow defect occlusion and device stability even if it looks constrained (Fig. 48.5).
7. When the device is in the appropriate position on TOE and not interfering with cardiac function, the device is detached from the delivery cable.
8. The sheath is then withdrawn and the purse string tightened.

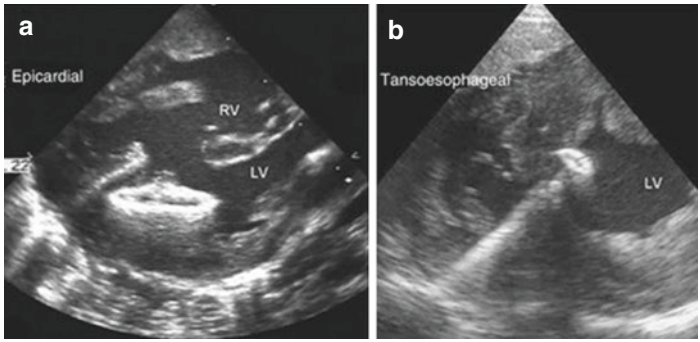

Fig. 48.4 The difference in orientation between epicardial (**a**) and TOE (**b**) guidance is demonstrated here, with deployment of the left ventricular disc. Note that careful planning and good imaging have allowed the disc to be opened free in the LV cavity in both cases. *RV* right ventricle, *LV* left ventricle

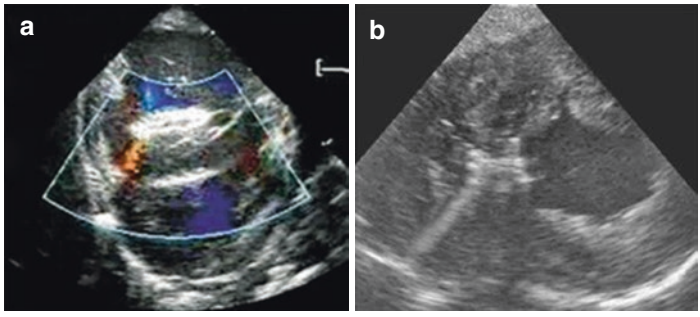

Fig. 48.5 Assessment of the conformation of the waist and RV disc is one of the most important steps. Time should be taken to assess the device and possibly multiple modalities including fluoroscopy can be used. Here we see satisfactory conformation of both discs on the epicardial echo (**a**) and TOE (**b**) – the waist of the device can also be seen to have conformed well (**b**)

48.8 Tips and Tricks

1. Perforation of the posterior wall of the LV with the sheath and dilator is a recognised complication. There are two practical ways of decreasing this risk. Firstly, placing the wire in the aorta should deflect the sheath away from the posterior structures during advancement. Secondly, the dilator should be withdrawn from the sheath until just before the transitional "shoulder" to minimise the length of dilator that needs to be advanced into the LV (Fig. 48.6).
2. Some operators have advocated soaking the device in the patient's blood prior to insertion to decrease the risk of postoperative haemolysis.
3. On occasions where the stability of the RV disc is uncertain, we have sutured the RV disc to the RV trabeculations with a brief period on cardiopulmonary bypass and a limited ventriculotomy.
4. Echocardiography may be supplemented by angiography at any stage although this is rarely necessary with high-quality TOE imaging.

Fig. 48.6 Modification of the sheath to decrease excursion of the stiff dilator towards the LV posterior wall and managing carefully the depth of the sheath into the ventricle can be done by placing two rubber shods onto the ensemble. The first goes onto the dilator and limits its protrusion from the soft sheath to just at the shouldered transition point (*arrow*). The second goes at the point which describes the maximum insertion depth into the heart and can be used as a marker for purse-string placement, avoiding crushing or kinking the sheath at the ventricular puncture site. An AMPLATZER Muscular VSD Occluder is seen on the right of the image

48.9 Expected Results

With careful patient selection, complete occlusion of the muscular VSD or occlusion with only a minimal residual shunt should be achieved. Although no minimum weight has been defined, the size limitation is usually related to the RV and LV cavity being large enough to accommodate the conformed device discs.

48.10 Pitfalls

1. Accepting a suboptimal angle from the RV free wall may result in failure. As mentioned, major difficulty in crossing the VSD should be addressed by relocating the RV access site.
2. Perforation of the LA posterior wall. This has been addressed above and depends on scrupulous communication between the TOE operator and the interventionist.

3. Poor RV disc conformation. This may be unavoidable as described. Usually occlusion is not dependent on apposition of the RV disc; but device stability may be affected and it is important to conform the RV disc with the least tension and distortion possible.
4. Failure to identify additional defects which may be haemodynamically significant may render a difficult and expensive procedure fruitless, and a pragmatic approach must be taken if multiple VSDs, which cannot all be closed, are found at any stage in the assessment or during the procedure.

48.11 Complications

Device embolism, heart block, LV wall rupture, tricuspid valve or mitral valve support apparatus damage, air embolism, thromboembolic stroke and haemolysis are all possible. These complications can be minimised by taking into account the steps and tips above.

48.12 How to Manage Complications

The keys to managing the significant complications are preparation for conversion to cardiopulmonary bypass and the availability of angiographic fluoroscopic imaging. Complications occurring while performing these types of procedures without the necessary personnel and infrastructure back-up may lead to avoidable morbidity and mortality.

48.13 Post-procedural Care and Follow-Up

This should involve an appropriate period of recovery in a cardiac ICU. Careful confirmatory imaging of the implanted device and assessment of any residual shunt should be made over the first 24–48 h. Aspirin at a dose of 3–5 mg/kg should be administered

for 6 months after the procedure to aid non-thrombotic endothelialisation of the device. Intermittent assessment of heart rhythm and RV function should be continued along with monitoring of any concomitant cardiac defects.

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