Intramural ventricular septal defect after repair of conotruncal anomalies: Is there light at the end of the tunnel?

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Patel et al¹ present an interesting article on intraoperative transesophageal echocardiography (TEE) to identify intramural ventricular septal defect (VSD) following the repair of conotruncal anomalies. The article raises a very important issue regarding the sensitivity of a well-established imaging technique to detect a rare, yet potentially life-threatening, complication.

Intramural VSD is defined as a tunnel-like communication that may occur when the VSD patch is not sutured to the edge of the VSD proper, but rather to right ventricular (RV) trabeculations, so that blood can flow from the left ventricle around the trabeculations into the RV cavity (Figure 1). Intramural VSD often has multiple exit points through intertrabecular spaces into the RV free wall. Intramural VSD was originally described in children with conotruncal anomalies undergoing subaortic VSD closure.² It should be emphasized that similar VSDs may occur following closure of any VSD, provided that it is, at least, partially surrounded by trabeculations so that a residual VSD occurs underneath trabeculations with an exit point into the free wall of the RV. VSDs with additional exit points through the free wall of the RV are notoriously difficult to close and can be associated with increased morbidity and mortality.³ Furthermore, although residual intramural VSD may not appear hemodynamically significant immediately after surgery, the interventricular shunting may increase with regression of RV hypertrophy.^{2,4}

Given the clinical importance of intramural VSDs, it is essential that surgeons have a high index of suspicion in all cases of conotruncal defects where VSD margins are in proximity to trabeculae. The best way to resolve the problem would be to prevent it. Thus, preoperative echocardiography in those children with conotruncal anomalies would alert a surgeon of higher risk of postoperative intramural VSD. Once intramural VSD occurs, it would be extremely helpful to visualize its location intraoperatively. This can be achieved by thorough intraoperative TEE assessment. However, intraoperative TEE is not without flaws. Thus,

Intramural ventricular septal defects occur when blood flows underneath the trabeculations.

Central Message

Precise echocardiographic evaluation is essential for successful closure of intramural ventricular septal defect.

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Patel et al¹ excluded 22.7% of patients (105 out of 462) because they did not have adequate imaging. Most importantly, the excluded children were younger and had lower weight. These children also had higher early mortality. It is these patients who would benefit most from intraoperative identification of intramural VSD. Unfortunately, performance of TEE in detecting intramural VSD—and even peripatch VSD—in older children was not perfect either. It was described by the authors as modest, with sensitivities of 56% for intramural VSD, which was not much different from that of 63% for peripatch VSD.¹

Moreover, patients with correct identification of intramural VSD made by TEE were more likely to be older than age 30 days and have higher body weight at operation. Thus, again, smaller children were disadvantaged. In nearly half of the children with missed intramural VSD, a subsequent transthoracic echocardiogram demonstrated residual intramural VSD > 2 mm. Is there a better alternative to TEE intraoperatively? Epicardial echocardiography is sensitive for the detection of residual VSDs.^{5,6}

At the Royal Children's Hospital in Melbourne, surgeons routinely perform epicardial echocardiograms on children with conotruncal anomalies after VSD closure, particularly in those with trabeculations obscuring the edge of the VSD and those with multiple VSDs.⁷ In our experience epicardial echocardiograms give superb imaging quality and are perfect for assessment of residual VSDs. This is of particular importance in small children. Clearly, a randomized controlled trial between TEE and epicardial echocardiography will not be feasible in neonates and smaller children. A surgeon with immediate knowledge of intraoperative anatomy is ideally suited to perform epicardial

FIGURE 1. A, Intramural ventricular septal defect (VSD) may occur when the edge of the VSD is partially covered by trabeculations. It was originally described in subaortic area, but may occur in inlet or anteroapical septum, if the patch is sutured to right ventricular (RV) trabeculations rather than a true septal margin of the VSD. B, The defining feature of intramural VSD is a tunnel underneath the trabeculations that persists with often multiple exit points at the free wall of RV. Ao, aorta; PA, pulmonary artery; LV, left ventricle.

echocardiograms. The importance of proficient acquisition of high-quality epicardial echocardiographic imaging by cardiac surgeons during neonatal surgery cannot be over-emphasized. Once the residual VSD is identified, the shunt is quantified by intraoperative assessment of Qp:Qs. Even if the residual VSD appears small on imaging, it is worth determining Qp:Qs to avoid any postoperative surprises.

Once a significant intramural VSD is identified, it should be closed, because simple pulmonary artery banding may not be a safe option in the presence of large residual VSD and would likely put a child on a rocky postoperative course.⁸ If the large residual VSD is not closed, the echocardiogram, albeit with modest sensitivity, may simply document a very sensitive surgical failure. For successful closure, one must close the entry point into the VSD because the outlets can be multiple.

TRANSATRIAL APPROACH

In the transatrial approach, a VSD patch should be at least partially removed, the edges of the VSD reassessed, and adjacent trabeculations resected if necessary so that the entry point into the VSD can be closed. Any attempt to close the often multiple exit points from this approach would be notoriously imprecise, and would likely fail. Suturing trabeculations together⁸ may open yet another exit point.

TRANSAORTIC APPROACH

To overcome the above problem, a transaortic approach has been proposed for classical intramural VSD,⁹ so that the entry point is closed through the aortic valve. This approach is not foolproof because the entry point into the intramural tunnel may not be easy to visualize.

TRANSAPICAL APPROACH

For defects in the apical region of the septum a small incision in the apex of either ventricle may be performed, allowing the VSD to be closed from the left ventricular side. Whereas this approach provides excellent visualization of

the VSD, an incision in the left ventricular apex is more likely to cause ventricular dysfunction.¹⁰ Thus, we prefer to open the right ventricle to avoid risk of left ventricle dysfunction and aneurysm formation.

Regardless of the approach, precise echocardiographic guidance would dramatically facilitate surgery. *Praemonitus praemunitus!* After all, failure to close intramural VSD occurs when surgeons do not realize how close they were to success when they gave up.

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