“Intramural” Residual Interventricular Defects After Repair of Conotruncal Malformations

Tamar J. Preminger, MD; Stephen P. Sanders, MD; Mary E. van der Velde, MD; Aldo R. Castañeda, MD, PhD; James E. Lock, MD

Background We report an unusual type of residual interventricular communication in patients with conotruncal malformations in which the aorta is completely or partly aligned with the right ventricle (RV). Interventricular communications after surgical repair usually result from additional defects, patch dehiscence, or incomplete close and lie in the septal plane. However, after a right ventricular aorta is baffled to the left ventricle, the ventricular septal defect (VSD) patch and RV free wall form part of the systemic outflow tract. This “neo-left ventricular” outflow tract may provide a location for residual interventricular communications out of the septal plane.

Methods and Results We reviewed echocardiographic, angiographic, and clinical records of patients who had one or more residual interventricular communications out of the plane of the ventricular septum after repair of a conotruncal anomaly. Between June 1990 and October 1992, we observed such defects in eight patients, 5 to 26 years old, after repair of double-outlet right ventricle (n=6), tetralogy of Fallot (n=1), or truncus arteriosus (n=1). In each, the VSD patch was anchored to the RV free wall near the aortic root. None-the-less, channels were observed around the edge of the patch, between the neo-systemic outflow tract and the right ventricle. All patients had right ventricular hypertension; in seven, the pulmonary-to-systemic flow ratio (Qp:Qs) was ≥2. At multiple unsuccessful reoperations (two to four per patient), the patch edges appeared securely attached to myocardium. Angiographic views profiling the septum failed to localize these defects, since they are not in the native septum. Echocardiographic detection of such anterior defects can be difficult. Transcatheter umbrella closure was attempted in the seven patients with large shunts; success was limited by the multiplicity of flow channels. Umbrella closure eliminated the need for further reoperation in four of seven patients, one patient died suddenly awaiting reoperation, and two deaths followed reoperation.

Conclusions “Intramural” residual interventricular defects are difficult to diagnose by all modalities. Umbrella placement may reduce the left-to-right shunt. Successful surgical closure may require removal and reattachment of the anterior portion of the patch. (Circulation. 1994;89:236-242.)

Key Words • ventricles • defects • surgery

Residual interventricular communications after surgical closure of conoventricular (usually malalignment) ventricular septal defects (VSDs) are most commonly due to patch dehiscence, additional unrecognized VSDs, or incomplete closure of the main defect.1,3 These residual defects generally lie in the plane of the ventricular septum.

In most conotruncal anomalies (in particular, transposition of the great arteries with VSD, double-outlet right ventricle, and tetralogy of Fallot), the aorta is completely or partly aligned with the right ventricle. Surgical correction involves directing the left ventricular outflow through the VSD to the aorta by means of a baffle.4,5 After these operations, the systemic outflow tract is formed by the baffle and the right ventricular (RV) free wall, which is often hypertrophied, with coarse trabeculations.

We have observed an unusual type of residual interventricular communication after repair of conotruncal anomalies that is both difficult to localize and a major cause of postoperative morbidity and mortality. These multiple communications course through intertrabecular spaces in the RV free wall where the VSD patch is anchored to trabeculae rather than the infundibuloven-tricular fold, close to the aortic valve annulus (Fig 1). We reviewed the angiographic and echocardiographic recognition of these residual defects and strategies for their management.

Methods

Patient Selection

All patients who had undergone repair of conotruncal anomalies and who had residual interventricular communications out of the plane of the ventricular septum recognized by echocardiography or angiography at Children’s Hospital, Boston, since 1985 were identified. The echocardiograms, cineangiograms, and clinical records of these patients were reviewed.

Echocardiographic Evaluation

A two-dimensional transthoracic echocardiogram (TTE) was performed on each patient before and after cardiac catheterization and surgery. Images were obtained in subxyphoid, apical, and parasternal views. Pulsed and color Doppler interrogations were performed when technically possible. In patients with suboptimal transthoracic windows or who were undergoing umbrella closure of “intramural” defects, transesophageal echocardiograms (TEE) were performed.

Hemodynamic and Angiographic Evaluation

Each patient underwent right and left heart catheterization. The location, size, and configuration of suspected or previously

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From the Departments of Cardiology and Cardiothoracic Surgery (A.R.C.), Children’s Hospital, and the Departments of Pediatrics and Surgery (A.R.C.), Harvard Medical School, Boston, Mass.
Correspondence to Tamar J. Preminger, MD, Department of Cardiology, Children’s Hospital, 300 Longwood Ave, Boston, MA 02115.
identified defects were defined angiographically; multiple angled views profiled the defects, displaying their geometry and relation to surrounding structures.6

Management

Patients were considered candidates for transcatheter umbrella VSD closure if the residual shunt caused significant morbidity and the patient was deemed a high-risk candidate for reoperation. Informed consent was obtained in all cases, and device placement was performed under a protocol approved by the US Food and Drug Administration and the Committee on Clinical Investigation at Children's Hospital. The technique for transcatheter VSD closure has been described previously.7-10

Terminology

We have used the segmental approach of Van Praagh11,12 to describe cardiac anatomy.

Permission for this retrospective study of residual interventricular communications after repair of conotruncal anomalies was granted by the Committee on Clinical Investigation.

Results

Patient Population

Between June 1990 and October 1992, eight patients 5 to 26 years old (median, 8 years) were identified who had a residual left-to-right shunt secondary to intertrabecular communications between the neo–left ventricular outflow tract and the RV sinus after surgical repair of a conotruncal malformation. Six of the eight patients (patients 2, 3, 4, 5, 6, and 8) had undergone surgery at six different referring institutions. Of note, seven of the eight patients had been referred for catheter closure of important VSDs that persisted despite two or three attempts at surgical closure. Six of the eight patients underwent a Rastelli-type repair for double-outlet right ventricle or D-transposition of the great arteries, one patient underwent repair of tetralogy of Fallot, and one patient underwent repair of truncus arteriosus. The patient profiles are outlined in Table 1.

All patients had previously undergone catheterization on two to four occasions (median, 3) to assess hemodynamics and identify residual defects. These eight patients underwent a total of 16 reoperations directed at closing residual VSDs. Nonetheless, seven of eight patients were left with major (Qp:Qs ≥2) residual shunts (Table 2). Five of the patients (patients 1, 2, 4, 5, and 6) were severely disabled by chronic congestive heart failure, cardiac dysrhythmias, and/or endocarditis.

Echocardiographic Evaluation

In five of the eight patients (patients 1, 2, 4, 5, and 6), multiple residual interventricular communications between the lateral margin of the patch and the RV free wall were identified by TTE and color Doppler flow mapping (Fig 2). TTE also revealed one or more coexisting defects in seven of the eight patients. In most patients, the “intramural” defect appeared by color Doppler to constitute the dominant location for left-to-right shunting. “Intramural” defect size ranged from 5 to 13 mm. TTE failed to reveal “intramural” defects in three patients because of the anterior location of the defects in two and poor echo windows in one.

Four patients (patients 3, 5, 6, and 8) had TEEs performed at the time of cardiac catheterization to guide umbrella placement. In patients 3 and 8, TEE identified “intramural” defects that had not been detected by surface echocardiography.

Hemodynamic and Angiographic Evaluation

RV pressure was systemic in five patients (patients 3, 4, 5, 6, and 7). Patients 1, 2, and 8 had restrictive defects with subsystemic RV pressure. The Qp:Qs ratio was ≥2 in all but one patient. All patients had associated obstruction of the RV outflow tract that contributed to elevation in RV pressure and may have caused hemodynamic underestimation of the size of residual communications.

At angiography, contrast crossed the true ventricular septum in all seven patients with residual peripatch defects, but other ill-defined contrast streams also opacified the right heart. These were not profiled with the native septum (as assessed from a long-axial oblique projection) but were oriented more anteriorly. Angiographically, the “defects” measured 4 to 13 mm (median, 9 mm), correlating well with the echocardiographic assessments (Fig 3). Test balloon occlusion confirmed
TABLE 1. Patient Characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, y</th>
<th>Cardiac Defects</th>
<th>Other Diagnoses</th>
<th>Previous Surgeries</th>
<th>Number of Postoperative Catheterizations</th>
<th>Number of Surgeries</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8</td>
<td>(S,D,D) TGA, VSD, PS</td>
<td>Bacterial endocarditis: severe, chronic CHF</td>
<td>RMBTS and takedown; Rastelli and subsequent revision</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>(S,D,S) TOF</td>
<td>Severe, chronic CHF</td>
<td>Complete repair; attempted closure residual VSD×2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>26</td>
<td>(S,D,D) TGA, VSD, PS</td>
<td>Bacterial endocarditis×2; postpericardiotomy syndrome; chronic CHF</td>
<td>RMBTS and takedown; Rastelli-type repair; conduit revision</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>19</td>
<td>(S,D,D) DORV, PA</td>
<td>Chronic CHF</td>
<td>Waterston shunt and takedown; Rastelli-type repair; VSD patch replacement</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>5</td>
<td>17</td>
<td>(S,D,D) DORV, PA; cor triatratium</td>
<td>Chronic CHF</td>
<td>Rastelli-type repair; resection of cor triatratium; RV-PA conduit replacements and VSD closures×2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>6</td>
<td>8</td>
<td>(S,D,D) DORV, multiple VSDs, subAS</td>
<td>Chronic CHF</td>
<td>Rastelli-type repair with intraoperative umbrella placement in muscular septum; Stansel anastomosis</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>7</td>
<td>5</td>
<td>(S,D,S) truncus arteriosus</td>
<td>left hemidiaphragm paralysis</td>
<td>Complete repair (VSD closure, RV-PA valved conduit)</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>8</td>
<td>10</td>
<td>(S,D,D) DORV, PA</td>
<td>Rastelli-type repair</td>
<td></td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

TGA indicates transposition of the great arteries; VSD, ventricular septal defect; PS, pulmonary stenosis; TOF, tetralogy of Fallot; DORV, double-outlet right ventricle; PA, pulmonary atresia; subAS, subaortic stenosis; CHF, congestive heart failure; RMBTS, right modified Blalock-Taussig shunt; and RV-PA, right ventricle to pulmonary artery.

Letters in parentheses refer to the classification system for segmental anatomy.11,12

The results of the catheterization and attempted umbrella closure for these patients are listed in Table 2.

Evaluation After Closure

At least one device was successfully implanted in all seven patients. This resulted in a diminution in Qr:Qs in all patients (Table 2). Angiography was performed after umbrella placement in six of seven patients and demonstrated significantly reduced flow in all (Fig 3). In patients with coexisting VSDs, the channel with the umbrella appeared to be effectively closed, although RV opacification by other channels persisted.

Follow-up echocardiography was performed in these seven patients within 24 to 72 hours after catheterization; transesophageal studies were performed in three. All the umbrellas appeared stable in position. Color flow mapping confirmed the angiographic findings; the degree of residual shunting through the intertrabecular channels was quantified as mild in five patients (patients 1, 2, 4, 6, and 8), moderate in one (patient 2), and large in one (patient 3). Patient 4 had an additional significant left ventricular-to-right atrial shunt.

Complications. No significant early complications occurred. There were no reported episodes of device embolization, hemolysis, stroke or other embolic phenomena, dysrhythmia, or infection.

Follow-up. Clinical follow-up on these eight patients ranges from 6 to 26 months (mean, 13 months) after cardiac catheterization. Of the five patients with severe clinical disability (patients 1, 2, 4, 5, and 6), two (patients 2 and 6) remain clinically asymptomatic 14 and 10 months after device placement, respectively. The other three patients (patients 1, 4, and 5) had no improvement in clinical status. Patient 1 had symptomatic improvement but remained in congestive failure and underwent reoperation 5 months later, with removal of the umbrella devices and patch closure of the multiple residual defects. At surgery, the umbrella devices were found to be partially occlusive. After surgery, he demonstrated marked clinical improvement, with evidence of a small residual shunt. Approximately 8 months later he developed recurrent congestive failure and died at an outside institution while awaiting reoperation. Patient 4 demonstrated only modest clinical improvement and was awaiting reoperation. Fol-
TABLE 2. Hemodynamic and Angiographic Assessments

<table>
<thead>
<tr>
<th>Patient</th>
<th>Total No. of Defects</th>
<th>&quot;Intramural&quot; Defect Size</th>
<th>Transcatheter Intervention</th>
<th>Qp:Qs Preumbrella</th>
<th>Qp:Qs Postumbrella</th>
<th>Results and Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2+</td>
<td>9 mm</td>
<td>Intramural defect closed with 23-mm and 28-mm umbrellas; peripatch defect and LV-RA defect closed with 28-mm umbrellas</td>
<td>&gt;4:1</td>
<td>3:1</td>
<td>Persistent CHF; underwent surgical repair of residual defects 5 mo after umbrella placements. Recurrent CHF; died 8 mo postop awaiting reoperation</td>
</tr>
<tr>
<td>2</td>
<td>2 or 3</td>
<td>9 mm</td>
<td>Intramural defect closed with 23-mm umbrella</td>
<td>&gt;4:1</td>
<td>1.2:1</td>
<td>Clinically well; 2 or 3 smaller residual defects</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
<td>10 mm</td>
<td>Intramural defect closed with 28-mm umbrella</td>
<td>2.5:1</td>
<td>1.4:1</td>
<td>Clinically well; residual flow through other intertrabecular channels</td>
</tr>
<tr>
<td>4</td>
<td>3+</td>
<td>6 to 7 mm</td>
<td>Intramural defect closed with 23-mm umbrella; peripatch defect closed with 23-mm umbrella; LV-RA defect closed with 28-mm umbrella</td>
<td>3:1</td>
<td>2:1</td>
<td>Clinically improved, but with persistent LV-RA shunt; sudden death 3 wk after umbrella placement while awaiting reoperation</td>
</tr>
<tr>
<td>5</td>
<td>3</td>
<td>10 mm</td>
<td>Intramural defect closed with 28-mm umbrella; anterior muscular VSD closed with 40-mm umbrella; peripatch VSD closed with 33-mm umbrella</td>
<td>2.2:1</td>
<td>1.6:1</td>
<td>Bacterial endocarditis x2; 3 AVB requiring pacemaker implantation; SVT; surgical closure of VSDs and conduit revision with postoperative death</td>
</tr>
<tr>
<td>6</td>
<td>2</td>
<td>13 mm</td>
<td>Intramural defect closed with 33-mm umbrella</td>
<td>2:1</td>
<td>1.5:1</td>
<td>Clinically improved; residual small anterior and superior defects</td>
</tr>
<tr>
<td>7</td>
<td>1</td>
<td>3.5 mm</td>
<td>None</td>
<td>1.4:1</td>
<td>Not performed</td>
<td>Clinically well</td>
</tr>
<tr>
<td>8</td>
<td>2</td>
<td>9 mm</td>
<td>Intramural defect closed with 23-mm umbrella; peripatch VSD closed with 33-mm umbrella</td>
<td>3.5:1</td>
<td>1.3:1</td>
<td>Clinically well</td>
</tr>
</tbody>
</table>

Qp:Qs indicates pulmonary-to-systemic flow ratio; LV-RA, left ventricular to right atrial; VSD, ventricular septal defect; CHF, congestive heart failure; AVB, atrioventricular block; and SVT, supraventricular tachycardia.

low-up echocardiograms confirmed decreased flow through the "intramural" defects; however, a significant left ventricular-to-right atrial shunt persisted secondary to flail septal leaflet of the tricuspid valve. He died suddenly 3 weeks after catheterization. He had a prior history of ventricular ectopy as well as supraventricular tachycardia. Gross pathological cardiac examination demonstrated partial occlusion of two of the defects and complete occlusion of the third. The arms of the umbrella placed in the left ventricular-to-right atrial shunt penetrated the anterior leaflet of the mitral valve. Patient 5 developed bacterial endocarditis approximately 3 months after umbrella placement. Cardiac dysrhythmias, including supraventricular tachycardia and complete heart block, occurred secondary to the endocarditis, the latter prompting pacemaker implantation. Approximately 18 months after device placement, he underwent reoperation at another institution, with removal of the umbrellas, patch closure of residual "intramural" and peripatch defects, and revision of the right ventricular-to-pulmonary artery conduit. He died early after surgery. The other patient (patient 8) who underwent umbrella closure of an "intramural" defect remains well 6 months after catheterization. Patient 7 was clinically asymptomatic despite an "intramural" defect.

Resource utilization. The common failure to make an accurate diagnosis in these patients resulted in considerable consumption of medical care. Before referral, patients had undergone an average of two unsuccessful operations to close residual VSDs; one of two patients managed at our center underwent one unsuccessful reoperation.

Discussion

As with any postoperative VSD, residual shunting through intertrabecular communications in the right ventricular free wall after repair of conotruncal malformations can cause postoperative morbidity and mortality. These defects appear to result from suturing of the VSD patch to trabeculations within the ventriculo-"intramural" fold rather than to the ventriculo-infundibular fold itself. Consequently, blood can flow from the neo-left ventricular outflow tract, between the trabeculae, into the right ventricle. The entrance site into the RV sinus is often some distance from the margin of the VSD patch (Fig 1). In each of the cases, the trabeculae on the RV free wall extend close to the aortic valve...
annulus. At direct operative inspection, the defects appear to be closed at that margin; the “septum” nonetheless permits left-to-right shunting via tunnels within the RV wall that open into the RV sinus remote from the edge of the VSD patch. The presence of additional residual defects in the ventricular septum (dehiscence of the patch or an additional VSD) often complicates the detection of these RV anterior free wall communications.

To prevent these residual defects, the surgeon must be certain that the sutures along the posterosuperior aspect of the VSD, in the presence of trabeculations, be placed within the infundibuloventricular fold itself, usually very close to the annulus of the aortic valve (Fig 4). These “intramural” defects not only cause major morbidity but also are difficult to diagnose and manage. Conventional echocardiography and standard angiographic views may fail to localize and detect them because they are not located in the native ventricular septum. We detected five of eight of these defects by TTE; the other three defects were identified angiographically. Two of these three defects were seen well by TEE during cardiac catheterization. These “intramural” channels may not be seen well by TTE from the right side of the patch because of shadowing. Because of the frequent coexistence of RV outflow tract obstruction, hemodynamic assessments, including RV pressure and pulmonary-to-systemic flow ratios, can be misleading. Surgical relief of RV outflow obstruction in the presence of unrecognized residual “intramural” defects, is likely to increase the pulmonary-to-systemic flow ratio and result in left heart failure. To further complicate their identification, we suspect that these channels are small early after initial surgical repair of the conotruncal defect because of RV hypertrophy and enlarge to become hemodynamically significant with regression of hypertrophy after RV decompression.

In symptomatic patients, our management involved attempted transcatheter umbrella closure, which diminished the shunt in all cases and eliminated the need for further reoperation in four of seven patients. Surgical detachment of the previously positioned VSD patch and reattachment or replacement proximal to the trabeculae seems to be the most effective technique of eliminating these “intramural” communications. However, given the poor clinical status of these patients, operation was associated with considerable morbidity and mortality.

Technical Considerations

Echocardiography. TEE appears to be a more sensitive method for detecting these residual “intramural” defects than TTE. TEE usually provides superior images in postoperative patients, especially older patients, in whom TTE image quality is compromised by scarring and implantation of prosthetic material. The sites of entry of the “intramural” flow channels into the RV.
sinus tend to be more lateral and posterior than the edge of the VSD patch and are more likely to be seen by TEE than by TTE. Acoustic interference by the patch material may produce shadowing of the RV sinus with the transducer on the anterior chest wall, obscuring the site of entry of the defect into the RV sinus. Apical and subxyphoid views are often useful for detecting these flow channels in smaller children.

**Catheterization and device closure.** These “intramural” defects are not profiled angiographically by the long-axial oblique views typically used to delineate defects in the native ventricular septum but rather are best seen from a straight lateral view or occasionally a steep right anterior oblique view, sometimes with cranial angulation. The multiplicity of flow jets and their serpiginous courses through the RV free wall make balloon sizing and effective umbrella closure of such defects difficult. Furthermore, delivery of the device within the channel via the RV trabeculations may limit full expansion of the arms, complicating fluoroscopic assessment of position. Simultaneous TEE facilitates correct positioning of the umbrella device.

**Summary**

During a 28-month period, eight patients were identified as having a previously undescribed type of residual interventricular communication after surgical repair of a VSD with a conotruncal malformation. These defects consisted of multiple intertrabecular communications within the RV free wall between the neo–left ventricular outflow tract and the RV sinus, at the anterior and lateral attachment of the patch to the RV free wall. These defects were difficult to detect and to localize by conventional echocardiographic and angiographic means. We attempted umbrella closure in seven patients and were able to reduce the shunt in all. Although most of these “intramural” communications were not completely eliminated by transcatheter umbrella closure, the need for further reoperation was circumvented in four of seven patients. These four patients remain clinically improved at 6 to 26 months of follow-up. Among the patients in whom transcatheter umbrella closure was unsuccessful, one patient died suddenly awaiting reoperation and two died after reoperation. Definitive closure of such residual “defects” may require surgical removal and reattachment of the VSD patch to a non trabeculated area.

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T J Preminger, S P Sanders, M E van der Velde, A R Castañeda and J E Lock

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