Doubly Committed and Juxtaarterial Ventricular Septal Defect: Outcomes of the Aortic and Pulmonary Valves


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Background. The morphology of ventricular septal defects (VSDs) that are doubly committed and juxtaarterial places the patient at risk for aortic valvar prolapse and aortic valvar insufficiency (AI). Surgical repair of this type of defect often involves placing sutures through the base of one or more of the leaflets of the pulmonary valve, raising concern for late pulmonary valvar insufficiency (PI). The purpose of this review was to analyze the postoperative follow-up relating to potential late complications with the aortic and pulmonary valves.

Methods. Between 1980 and 2012, 106 patients with doubly committed juxtaarterial VSD underwent intracardiac repair. Median age at repair was 1.1 years. Preoperative evaluation showed 69 patients (65%) had aortic valvar prolapse and 51 (48%) had AI. Operative approach was through the pulmonary trunk in 88 (83%) of the patients. In 81 patients (76%), sutures securing the VSD patch had been placed through the base of the pulmonary valvular leaflets.

Results. Operative survival was 100%. Follow-up ranges from 6 months to 17 years, with a mean of 4.9 years. No patient had heart block or residual shunting. Of the 70 patients with long-term contemporary echocardiographic follow-up, 66 (94%) had trivial or no AI and 4 (6%) had mild AI. Of these patients, 49 (70%) had trivial or no PI, and 21 (30%) had mild PI. In 1 patient having aortic valvoplasty at the time of VSD closure, the aortic valve was replaced 7 months later. No other patient had worrisome progression of their AI or PI.

Conclusions. The incidence of aortic valvar prolapse and AI in the setting of doubly committed juxtaarterial VSD is quite high. The optimal surgical approach is through the pulmonary trunk. Sutures placed through the base of the pulmonary valvular leaflets do not predispose to clinically significant late pulmonary valvar insufficiency. Timely surgical closure of this type of defect prevents progression of AI.


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valvar prolapse, the prevalence and degree of aortic and pulmonary valvar insufficiency before and after repair, the operative approach and techniques, and late postoperative outcomes.

Patients and Methods

Between January 1990 and February 2012, 106 patients with doubly committed and juxtaarterial VSDs underwent intracardiac repair at Ann and Robert H. Lurie Children’s Hospital of Chicago. We reviewed the clinical records, echocardiograms, operative findings, and surgical outcomes of all these patients. Lurie Children’s Institutional Review Board approved this study and granted a waiver of informed consent on June 13, 2012.

Preoperative echocardiograms were performed to evaluate the intracardiac anatomy, with emphasis on identifying the lack of a muscular outlet septum, and the relationships between the leaflets of the aortic and pulmonary valves. During slow rotation from a parasternal long to a modified short-axis view, we interrogated the anticipated location of the outlet septum. If a structure was found separating the outflow tracts, it proved to be fibrous rather than muscular. With further rotation to a standard short-axis view, the anatomic site of the VSD was demonstrated. The leaflets of the aortic valve, in particular the right coronary leaflet, were carefully evaluated for prolapse. Aortic and pulmonary valvar insufficiencies were assessed by pulsed and color flow Doppler examination, and were graded as trivial, mild, moderate, or severe.

The patients underwent surgery using bicaval venous cannulation, moderate hypothermia at 28°C to 32°C, and with the left ventricle vented through the right superior pulmonary vein. In Figure 3, we show the typical operative exposure as achieved through the opened pulmonary trunk. After cross-clamping the aorta, and delivering cold blood cardioplegic solution, we placed interrupted pledgeted supported sutures circumferentially around the defect. A critical part of the process of closure involves placing sutures directly in the base of the pulmonary valvar leaflets as an anchoring point where there is no muscular septum separating the aortic and pulmonary valves, as shown in Figure 4. All defects were closed with a round polytetrafluoroethylene patch, which supports the leaflets of the aortic valve (Fig 4). The pulmonary trunk was closed primarily.

Fig 1. (A) In the normal heart, the free-standing muscular subpulmonary infundibulum lifts the pulmonary trunk away from the base of the heart. The leaflets of the aortic and pulmonary valves are separated, and are at different levels. (B) This subpulmonary infundibular sleeve is not present in the heart with the doubly committed juxtaarterial ventricular septal defect, pictured at right, a feature that brings the aortic and pulmonary valves into alignment, and allows for the fibrous continuity between the leaflets of the two valves.

Fig 2. Two images of doubly committed juxtaarterial ventricular septal defects (VSDs). (A) A doubly committed juxtaarterial VSD viewed from the right side of the heart. This shows the VSD located superior to the limbs of the septal band, and inferior to the conjoined leaflets of the pulmonary and aortic valves (yellow dotted line). Note the continuity between the caudal limb of the septal band and the ventriculoinfundibular fold, which produces a muscular posteroinferior rim to the defect. The prolapsed aortic valve is visualized through the defect. (B) A doubly committed juxtaarterial VSD viewed from the left side of the heart. Here one can appreciate its positioning just inferior to the aortic valve, and its proximity to the right coronary leaflet of the aortic valve.
Results

Age at the time of operation ranged from 13 days to 26 years, with a median age of 1.1 years; mean age was 3.9 years. Of the patients, 60 (57%) were male and 46 (43%) were female. We provide the complete demographic data in Table 1. All patients had preoperative and postoperative echocardiograms. All patients survived the initial operation, and all remain alive apart from 1 patient who died 2 years postoperatively owing to complications from a pre-existing seizure disorder. Operative exposure was through the longitudinally opened pulmonary trunk in 88 (83%) of the patients. Exposure was at the operating surgeon’s discretion. Early in the series, operative exposure was through the right atrium in 7 patients, the aorta in 6, and the right ventricle in the remaining 5. All recent patients have been approached through the pulmonary trunk. Sutures were placed in the base of the pulmonary valvar leaflets in 81 patients (76%). Sixty of those patients have long-term follow-up echocardiogram data available for review. In the remaining 25 patients (24%), sutures were placed through the fibrous outlet septum. All patients are in normal sinus rhythm. No patient has required reoperation for residual VSD. Preoperative evaluation showed that 69 patients (65%) had aortic valvar prolapse, and 51 (48%) had AI. The size of the defect ranged from 2.5 to 18 mm, with a mean of 7.0 mm. In some patients, the indication for surgery was failure to thrive, congestive heart failure, elevated pulmonary artery pressures, or other standard indications for VSD closure.

Mean cross-clamp time was 59.3 ± 22.5 minutes. Mean CPB time was 85.5 minutes.

Simultaneous repair of the aortic valve because of preoperative severe valvar insufficiency was performed in 2 patients. One patient presented with a 5-mm residual VSD after previous repair of a doubly committed juxtaarterial VSD 1.5 years earlier at another hospital. His AI had progressed to become severe, with prolapse of the right coronary leaflet of the aortic valve.

Table 1. Demographics of 106 Patients With Doubly Committed and Juxtaarterial Ventricular Septal Defects at Ann and Robert H. Lurie Children’s Hospital of Chicago (1990 to 2012)

<table>
<thead>
<tr>
<th>Demographics</th>
<th>Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Race</td>
<td></td>
</tr>
<tr>
<td>Asian</td>
<td>15 (14)</td>
</tr>
<tr>
<td>African-American</td>
<td>10 (9)</td>
</tr>
<tr>
<td>Caucasian</td>
<td>40 (38)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>40 (38)</td>
</tr>
<tr>
<td>Other</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Male</td>
<td>60 (57)</td>
</tr>
<tr>
<td>Female</td>
<td>46 (43)</td>
</tr>
<tr>
<td>Age, years</td>
<td>3.9 ± 5.2</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>13.7 ± 14.3</td>
</tr>
<tr>
<td>Follow-up period, years</td>
<td>4.9 ± 3.6</td>
</tr>
</tbody>
</table>

Data are n (%) or mean (SD).
redundancy was treated by placing a stay suture in the nodules of Arantius of the normal leaflets, and the center of the prolapsed leaflet. Excessive valvar tissue was plicated and sutured with pledgets to the margin of the leaflet close to the sinotubular junction (Trusler repair) [9]. Within 7 months, the repair had failed, and it proved necessary to replace the valve through an aortotomy using a no. 23 St. Jude prosthesis (St. Jude Medical, St. Paul, MN). Simultaneous replacement of the aortic valve was performed in another patient found to have severe AI preoperatively. This patient had initially been diagnosed with a doubly committed juxtaarterial VSD at our hospital as an infant, but without aortic valvar prolapse or AI. The patient was then lost to follow-up, not having undergone operative closure of the defect. He returned at the age of 25 years, presenting with prolapse of the right coronary leaflet of his aortic valve, and severe AI. The Yacoub repair was attempted but failed owing to AI [10]. The VSD was closed with a patch, and the aortic valve was replaced.

Long-term follow-up data are available for 70 patients. These are the patients operated on after 1990 when our cardiac surgery electronic database was established. The 36 patients for whom long-term follow-up is not available were operated on before 1990. In the recent cohort of 70 patients, follow-up is as long as 16.9 years (mean 4.9). In this subset, preoperative AI had been present in 35 patients (50%). Of these patients, 18 had AI that was graded trivial, 13 patients had mild AI, 2 patients had moderate AI, and 2 patients had severe AI. On postoperative follow-up, 51 (73%) have no evidence of AI. In the remaining 19 patients with AI, the regurgitation has been assessed as trivial in 15 and mild in 4 (Fig 5). As described previously, 1 patient underwent concurrent replacement of the aortic valve, and 1 patient had failure of valvar repair and required early replacement of the aortic valve. No other patient had worsening of AI over the period of follow-up (Fig 6).

In 13 of these 70 patients with long-term follow-up, pulmonary valvar insufficiency was noted preoperatively, being graded trivial in 10 and assessed as mild in 3. Over the long-term follow-up of the 70 patients with echocardiogram reports, 49 (70%) have trivial or no pulmonary valvar insufficiency. Mild pulmonary insufficiency is present in 21 (30%) of the patients (Fig 7). No patient required any therapeutic intervention for pulmonary valvar insufficiency during the period of follow-up, and no patients have more than mild pulmonary insufficiency. An additional point of information is that even though all longitudinal pulmonary arteriotomies were closed primarily, no patients had supravalvar pulmonary stenosis.

Comment

The natural history, and anatomic location, of doubly committed and juxtaarterial VSDs critically affects their surgical management. The magnitude of the left-to-right shunt may vary over time because of anatomic restriction of the defect caused by prolapse of the leaflets of the aortic valve. That leads to decreased left-to-right shunting on one hand, but potentially dangerous injury to the aortic valve on the other. The primary indications for closure of these defects, therefore, are aortic valvar prolapse and AI [5, 11]. Secondary indications include the degree of left-to-right shunting, and the known potential for aortic valvar prolapse.

There are at least two reasons why the right coronary leaflet of the aortic valve is prone to prolapse into the doubly committed juxtaarterial VSD. The loss of anatomic support directly beneath the aortic valve leads to herniation of the leaflet through the defect [1]. Additionally, the Venturi effect created by the shunting of blood across the defect during systole pulls the sagging leaflet into the low-pressure, high-velocity shunt between the ventricles [12]. This prolapse is progressive, leading to AI when
the elongated leaflet can no longer close during diastole (Fig 8) [13]. Eventually, an aneurysm of the supporting aortic sinus of Valsalva can develop. A distinct relationship has been shown between the age of the patients and the development of aortic valve prolapse. In a series of 209 patients with doubly committed and juxtaarterial VSD, 100% of patients had aortic valve prolapse by age 15 years, the mean age of onset of aortic valve prolapse was 4.9 years [4]. In a series of 395 patients with doubly committed juxtaarterial VSD, half had AI by the age of 8 and almost nine tenths by the age of 20 [7]. Aneurysm of the sinus of Valsalva was not found before the age of 10 years, but began to develop during the second decade of life, and was diagnosed most frequently in the third decade of life. These investigators [7] also confirmed that defects of this type rarely close spontaneously, but when they do, it is usually secondary to unwanted aortic valvar prolapse.

Early surgical intervention prevents the progression of AI [5, 14]. Closure of the defect in itself has been shown to be sufficient to arrest progression of AI in patients with mild insufficiency [5, 15]. In our series, we have 2 patients who presented with severe AI. Both underwent significant delays before repair, not being under vigilant supervision for the onset of AI or aortic valvar prolapse. The patient in whom the defect was diagnosed in infancy, but not repaired, had neither AI nor aortic valvar prolapse before he was lost to follow-up. When he returned 25 years later, he had severe AI, coupled with severe prolapse of the right coronary leaflet of the aortic valve. The other patient underwent attempted repair of his defect at the age of 2.5 years at another institution. When presenting at our hospital at the age of 4 years, he had a residual defect of moderate size accompanied by severe AI and prolapse of the right coronary leaflet of the aortic valve. There are essentially four techniques reported for repairing the AI with doubly committed and juxtaarterial VSD. These are the Trusler repair [9], Yacoub repair [10], Carpentier repair [16], and Lecompte repair [17]. We have only used these in 2 patients because in all other patients we have operated before the development of moderate or severe AI.

One potential problem that has been posed regarding the surgical closure of defects that are doubly committed and juxtaarterial pertains to the placement of sutures in the base of the pulmonary valve, the fear that the technique might possibly cause subsequent pulmonary valvar insufficiency [1]. We addressed this question by reviewing the immediate postoperative and late follow-up echocardiograms of our patients. We have 70 patients undergoing long-term follow-up with echocardiographic evaluation. Of these, 60 had sutures placed through the base of the pulmonary valve, and 29 had no pulmonary insufficiency. In the remaining 41 patients, pulmonary insufficiency was mild or trivial. These data indicate that the technique of securing the VSD patch with sutures placed through the base of the pulmonary valvar leaflets does not put the pulmonary valve in danger of becoming seriously insufficient. Also of interest, all of the longitudinal pulmonary artery incisions were closed primarily, and no patient had late supravalvar pulmonary stenosis.

The anatomic location of the doubly committed defect makes surgical closure difficult if approached from the right atrium [18]. Some researchers have recommended a right ventriculotomy for repair, but more recently the transpulmonary approach has become increasingly favored [1, 13, 19]. Our results endorse the approach through the pulmonary trunk as the method of choice. Such transpulmonary closure was first reported by Kawashima and coworkers [20]. They noted the advantages of avoiding a
right ventriculotomy, with the attendant risk of a focus for late ventricular ectopy, and possible decreased right ventricular contractility. Successful transpulmonary repair was subsequently reported in infancy [21], with others [14, 22] endorsing the transpulmonary route as the approach of choice for defects that are doubly committed and juxtaarterial.

We, and others, have used a patch to close the defects in all of our patients. We agree that it is very important to repair the defect with a patch, because if direct closure is attempted, there is danger of pulling the valve down to the septum, making worse any AI. The patch helps support the leaflets of the aortic valve, preventing prolapse and progressive AI.

A limitation of this study is that 36 patients operated on before 1990 do not have long-term follow-up. However, we do have as many as 23 years of follow-up in a cohort of 70 patients, which is a substantial number for this study.

In conclusion, we suggest that surgical management of the doubly committed and juxtaarterial defect should be based on knowledge of its anatomic features and natural history. The incidence of aortic valvar prolapse and AI is quite high in these patients. Surgical closure with a patch placed through the pulmonary trunk is our operative technique of choice. That allows the best exposure of any fibrous remnant of the outlet septum, confirms the continuity between the leaflets of the pulmonary and aortic valves, and facilitates safe suture placement through the base of the pulmonary valve leaflets. Sutures placed through the base of the pulmonary valvar leaflets does not predispose to clinically significant pulmonary valvar insufficiency. Timely surgical closure of this type of defect prevents progression of AI. We strongly recommend early elective closure of such defects regardless of shunt volume before progressive aortic valvar prolapse can lead to AI.

Fig 8. Pathophysiology of aortic valvar insufficiency in juxtaarterial and doubly committed ventricular septal defect (VSD). (A, C, E) In early systole, blood ejected from the left ventricle is shunted through the VSD, forming a high velocity, low pressure shunt. As a result, the anatomically unsupported right coronary leaflet, due to the loss of septal support, is pulled into the right ventricle by the Venturi effect (essentially being sucked into the low pressure zone within the defect). (B, D, F) In diastole, the intraventricular pressure forces the aortic valvar leaflet to close, but the unsupported prolapsed leaflet is pushed down into the VSD, away from the opposed coronary leaflet, resulting eventually in aortic insufficiency. (Reprinted from Tweddell et al [13], with permission from Elsevier.)
CONGENITAL HEART

DISCUSSION

DR E. DEAN MCKENZIE (Houston, TX): Paul, beautifully presented. I suspect we’ll be seeing more of you here. Another great presentation from Lurie Children’s Hospital. Dr Backer runs a tight ship. I received your manuscript a couple of months ago. I noticed that Professor Anderson is a coauthor on your paper, and I agree with the nomenclature used to describe the defect. Doubly committed VSD, however, has a very different connotation in the context of double outlet right ventricle, which is I think where we are more accustomed to assigning that term. I have actually heard Bob Anderson say that we should dispense with the term VSD when we talk about double outlet right ventricle and use the term interventricular communication instead. This VSD has been called supravalvular and subarterial in the past. What is your understanding of the nomenclature and how we arrived at assigning this defect this term?

MR DEVLIN: As you can see on our anatomy slide, the VSD is immediately adjacent to the pulmonary artery and the aorta. Hence, the term juxtaarterial. From the right side, the defect is committed to the pulmonary trunk and its valve; from the left side, the defect is committed to the aorta and aortic valve. Hence, the term doubly committed. Indeed there is truly fibrous continuity between the aortic and pulmonary valves, quite different than the usual relationship between these two valves where they are offset. The combination of the phrases doubly committed and juxtaarterial make the unique location of this defect very clear. Professor Anderson was actually a visiting professor at Lurie Children’s when we were researching this, and I made the mistake of calling it a conal VSD, and he corrected me quite quickly. I don’t know if Dr Backer has anything more he wants to say about that.

DR BACKER: Paul, that was a great answer. I personally never really understood, even back when I wrote that first paper, as Dr Tweddell pointed out, 20 years ago—thanks, Jim—that the infundibular sleeve prevents you from having a doubly committed juxtaarterial VSD in a normal heart. To have this defect, you have to have the aortic and pulmonary valves at the same level with the VSD below both valves; hence the phrase, doubly committed and juxtaarterial. I initially resisted using that phrase myself, but Professor Anderson convinced me of the clarity of this terminology, as he has now convinced most of us. I believe it is the most accurate nomenclature to use.

DR MCKENZIE: And one other question. Our experience is similar to yours in that usually these VSDs, once you get the

References
heart open and the valves out of the way, look much larger than they had on echo. So our preference has been to close it when initially diagnosed, and I think we have most of our cardiologists educated to that way of thinking.

I noticed that in this series the average age of closure was almost 5 years. Is that a result of practice in the past era, this is a very long series, or is that just a continued lack of education and delayed referral?

MR DEVLIN: There are two factors at work here. The median age is a more accurate representation of the timing for surgical repair in this population, and in our series was actually 1.1 years. There was a 26-year-old in the series, and that skewed the mean age. There also has been a shift over time; we are now repairing these defects much earlier in life. Dr Backer and I were discussing this, and now the usual age for repair is 6 months to a year.

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Mark your calendar for the 51st Annual Meeting of The Society of Thoracic Surgeons (STS) to be held at the San Diego Convention Center in San Diego, California, January 24-28, 2015. The STS Annual Meeting offers you a chance to meet the experts, network with colleagues from around the world, and participate in a dynamic learning experience.

This preeminent educational event is open to all physicians, residents, fellows, research scientists, perfusionists, physician assistants, nurses, and others interested in cardiothoracic surgery.

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An advance program with information about housing and registration will be mailed to STS members this fall. Nonmembers may contact the Society to receive a copy of the printed advance program; however, detailed up-to-date meeting information will be available on the STS website at www.sts.org/annualmeeting.

I hope to see you in San Diego.

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