Partial Zone of Apposition Closure in Atrioventricular Septal Defect: Are Papillary Muscles the Clue

Timothy M. Colen, MBBS, Nee S. Khoo, MBChB, David B. Ross, MD, and Jeffrey F. Smallhorn, MBBS

Departments of Pediatrics and Surgery, University of Alberta and Stollery Children’s Hospital, Walter C. Mackenzie Health Sciences Centre, Edmonton, Alberta, Canada

Background. Long-term survival after atrioventricular septal defect repair is excellent; however, postoperative left atrioventricular valve regurgitation affects morbidity and quality of life. Left atrioventricular valve regurgitation is the most common reason for reoperation after repair and it is critical that clinicians recognize pathologic mechanisms pre-repair.

Methods. In this single-center experience, we identified a pattern of left atrioventricular valve abnormality in 5 cases presenting for routine surgical repair between 1 month and 24 years of age. We reviewed two-dimensional and real-time three-dimensional echocardiographic and surgical findings to assess for specific valvar or sub-valve abnormalities, including short chordae, commissural deformities, and an eccentric zone of apposition. Two-dimensional echocardiography was used to assess the degree of preoperative and postoperative left atrioventricular valve regurgitation.

Atrioventricular septal defect (AVSD) is one of the more common congenital defects encountered by both pediatric and adult congenital surgeons. Mortality at the time of initial repair has declined; however, in some, ongoing surgical intervention is necessary to address progressive left atrioventricular valve regurgitation (LAVVR) [1, 2]. Although two-dimensional (2D) echocardiography has been the main mode of investigation prior to surgical intervention [3], the technique has been less sensitive to determine the mechanisms of LAVVR, both preoperatively and postoperatively. More recently, real-time three-dimensional echocardiography (RT3D) has been employed to overcome some of the limitations of 2D techniques [4–6] and permits detailed evaluation of commissures and more subtle morphologic variations that might impact on left atrioventricular valve (LAVV) function [7].

Results. Abnormal features identified included short, thickened chordae, poorly formed superior-mural commissure, and an eccentric zone of apposition. At surgical repair, 2 patients had limited closure of the zone of apposition, as part of a complete repair, and developed only mild left atrioventricular valve regurgitation in short-term follow-up. Two further patients had attempted complete closure of the zone of apposition with moderate postoperative regurgitation ultimately necessitating left atrioventricular valve replacement.

Conclusions. This uncommon form of atrioventricular septal defect is identifiable with echocardiography and may be associated with significant postoperative regurgitation if the zone of apposition is completely sutured at time of repair. Limited closure of the zone of apposition may improve postoperative regurgitation.


Surgical philosophy has been to close the “cleft,” which is now being referred to by morphologists as the zone of apposition, in the LAVV if there are 2 supporting papillary muscles, unless there is a double orifice where intervention may cause significant stenosis [8, 9]. Despite this, there are some instances where minimal preoperative LAVVR is followed by moderate regurgitation, in the early or late postoperative period, despite a relatively straightforward surgical procedure.

Our index case, seen in 2010 (patient number 1, Table 1), was put forward for routine closure of a primum atrial defect. There was moderate LAVVR on the preoperative 2D echocardiogram. A RT3D echocardiogram was performed as part of our ongoing research; however, this was not used in the surgical decision process. The 2D echocardiogram report indicated 2 well-spaced papillary muscles and a zone of apposition between the bridging leaflets. The surgeon closed the primum defect and sutured the zone of apposition as per our usual policy at that time. The patient developed significant LAVVR that required early reintervention to repair the valve and subsequent early mechanical valve replacement.

On review of the 2D and RT3D echocardiograms we identified an uncommon, but significant abnormality...
Table 1. Echocardiographic and Surgical Characteristics of Patient Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis</td>
<td>Primum AVSD</td>
<td>Primum AVSD</td>
<td>Primum AVSD</td>
<td>Complete AVSD</td>
<td>Complete AVSD</td>
</tr>
<tr>
<td>Age at primary surgery</td>
<td>18 months (2 years)</td>
<td>4 years (24 years)</td>
<td>19 years (24 years)</td>
<td>2 months</td>
<td>2 months</td>
</tr>
<tr>
<td>Preoperative RT3D echocardiographic findings:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zone of apposition</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chordae and PM</td>
<td>Short and muscularized chordae, thickened ALPM</td>
<td>Large ALPM and small PMPM</td>
<td>Eccentric</td>
<td>Dominant ALPM; short and muscularized chordae</td>
<td>Eccentric</td>
</tr>
<tr>
<td>Commissure</td>
<td>Blunted</td>
<td>Blunted</td>
<td>Deformed and blunted</td>
<td>Blunted &amp; abnormally formed</td>
<td>Blunted</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
<td>Large</td>
<td>Small</td>
</tr>
<tr>
<td>LAVVR</td>
<td>Moderate</td>
<td>Moderate</td>
<td>Mild</td>
<td>Moderate</td>
<td>Mild</td>
</tr>
<tr>
<td>Surgical findings at primary repair</td>
<td>Oblique zone of apposition</td>
<td>Oblique zone of apposition</td>
<td>Oblique zone of apposition</td>
<td>Oblique zone of apposition</td>
<td>Oblique zone of apposition</td>
</tr>
<tr>
<td>Surgical findings at subsequent repair</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chordae to ALPM thickened and shortened. Mural leaflet tethered and restricted. Prolapse of SBL.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subsequent surgical repair</td>
<td>Leaflet extension to anterior leaflet (combined SBL and IBL) using autologous pericardium</td>
<td>Closure of the zone of apposition and ring annuloplasty. Subsequent valve replacement (33 mm St. Jude valve) due to significant post-operative regurgitation.</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>

(Continued)
where “complete” closure of the zone of apposition exacerbated postoperative LAVVR. This was characterized by a specific abnormality of the anterolateral papillary muscle, a malformed superior-mural commissure, and eccentricity of the zone of apposition. This report describes the echocardiographic and pathologic features, the surgical findings, and clinical outcome of this patient, as well as four others that we have subsequently identified.

Patients and Methods

Since the index case in 2010 we have identified 4 other patients from our institution with AVSD and this form of LAVV abnormality. These patients are being reported with the approval of the Institutional Research Ethics Board.

Patient number 2 (Table 1) was identified from our recently published series on quantitative analysis of LAVV function post repair of AVSD [10]. While reviewing all the surgical reports and studies it was clear this patient had the same abnormality as case number 1. Since the index case another 3 patients were identified preoperatively, and the findings transmitted to the surgeons who modified their surgical approach based on this information.

All patients had transthoracic 2D and RT3D echocardiographic studies performed preoperatively. Echocardiograms were performed using IE33 (Phillips Healthcare, Andover, MA) with appropriate-sized probes for the patient. For retrospective review, both transthoracic and transesophageal images, when available, were examined. Echocardiographic images were available for viewing through our electronic image database (Xcelera; Phillips Healthcare, Andover, MA). The 3D echocardiography data sets were manipulated and reconstructed using Q-Lab (Phillips Healthcare). The RT3D echocardiographic datasets were reconstructed to examine the LAVV and sub-valve structures. Data sets acquired from the apical 4-chamber view were cropped from above to view the LAVV en face and to assess the zone of apposition position. The same data sets were then cropped from below to assess the commissural and papillary muscle morphology, as well as the zone of apposition position. Images acquired from a parasternal long-axis view made it possible to view the papillary muscles and chordae in their entire length to assess for abnormalities in structure and size.

Left atrioventricular valve regurgitation was assessed using color Doppler echocardiography and characterized qualitatively as nil, mild, or moderate. Surgical reports were obtained from our institutional surgical database and reviewed for surgical findings.

Valve anatomy and morphology was assessed qualitatively using 2D and RT3D echocardiography, and compared with surgical findings. Specifically, papillary muscle number, size, and structure, commissural size, leaflet size, and zone of apposition position were assessed. Associated cardiac lesions were noted.
Results
Clinical data are summarized in Table 1. One patient had Down syndrome. There was no associated left ventricular outflow tract obstruction, either preoperatively or postoperatively, in any of the cases. Three patients had a primum defect and 2 had complete AVSD with a common atrioventricular valve orifice. In the latter 2, 1 had a large and the other a small ventricular component. One patient had a coarctation of the aorta that was repaired in the neonatal period (patient number 5), with associated pulmonary valve stenosis that was relieved at the time of repair of the AVSD.

Two-Dimensional Echocardiography Findings
The common feature on 2D echocardiography was a particularly dominant anterolateral papillary muscle that could be identified from its normal attachment at the base, continuing to be observed as the probe was swept toward the left ventricular outflow tract (Figs 1; 2). This appearance reflected the fused and shortened chords, and in these patients indirectly represented the abnormal superior-mural commissure that was identified by RT3D echocardiography. Of note, direct imaging of the abnormal superior-mural commissure was more challenging by 2D echocardiography.

RT3D Echocardiography Findings
By RT3D echocardiography a shortened or deformed superior-mural commissure was the most common abnormality described in this series, present in all patients (Figs 1, 2). Short and thickened, or absent chordae were present in all and the anterolateral papillary muscle was dominant in each case. In each patient the commissure, chord, and papillary muscle abnormalities affected the superior-mural commissure rather than its inferior counterpart. In 2 patients an eccentric zone of apposition pointing more toward the left ventricular outflow tract was identified by RT3D echocardiography and represented a somewhat deficient superior bridging leaflet.

The degree of preoperative LAVVR was qualitatively graded as mild in 2, and moderate in 3. In patient number 1, where the zone of apposition was completely closed, the postoperative RT3D echocardiogram demonstrated the mechanism of regurgitation was poor leaflet coaptation due to the tethered superior bridging leaflet (Fig 3).

Surgical Findings
The description in Table 1 represents the actual surgeon’s “words” in their reports. Of importance, in each case the surgeons recognized that the LAVV was abnormal. The most common abnormality described was of the papillary muscle, with either direct attachment to the leaflet or significantly short and thickened chordae. This was identified in all patients at either initial or subsequent procedure. Other observations included an oblique zone of apposition in the LAVV, a dominant anterolateral papillary muscle, and an abnormal superior-mural commissure. There was reasonable correlation between the surgical and RT3D echocardiographic description, with the appreciation that the latter technique provides real-time
data and enhanced imaging of the LAVV from below (the ventricular aspect).

The surgical technique was through a median sternotomy and right atrial incision in all patients. Specific approach to the LAVV was not influenced by the echocardiographic findings in cases 1 and 2, and in both when complete closure of the zone of apposition was attempted the result was significant LAVVR. The surgical approach in the remainder of the patients was modified by the prior knowledge of the echocardiographic findings such that the zone of apposition was only closed at the base in numbers 3 and 5. In case 4, where the ventricular component was large, the surgeon was able to recruit some of the right-sided component of the superior bridging leaflet.

Fig 2. These images are from case number 1, as seen preoperatively. (B) shows a two-dimensional short axis view of the left atrioventricular valve, and demonstrates a similar appearance to that seen in case number 3, Figure 1. Note the black star, which represents the thickened anterolateral papillary muscle (ALPM), which appears to be stuck to the superior bridging leaflet (SBL). (A) is a real-time three-dimensional echocardiography image (RT3D) as seen from the left ventricular (LV) aspect. The black star is on the ALPM, which can be seen to fuse with the superior bridging leaflet (SBL) having very short chords and a blunt superior-mural commissure, unlike the well-formed inferior-mural commissure. (* = inferior; IBL = inferior bridging leaflet; LVOT = left ventricular outflow tract; ML = mural leaflet; PMPM = posteromedial papillary muscle; RV = right ventricle; S = superior; SBL = superior bridging leaflet.)

Fig 3. These images were taken during the early postoperative period in case number 1. They are seen in the multiplanar reconstruction, or multiplane reformatting image, which represents simultaneous images of the left atrioventricular valve seen in different planes, as well as a real-time three-dimensional echocardiography (RT3D) reconstructed image. (D) is the RT3D image, seen from the left atrial view, and shows the fully sutured zone of apposition, outlined by the white stars. Note that in systole there is a significant area of non-coaptation indicated by the black arrow. The main site of malcoaptation is to the left of the sutured zone of apposition. (C) is a RT3D color Doppler image demonstrating the significant regurgitant jet, which arose from the site of malcoaptation. Note there is also a smaller jet, which arose from the area of the sutured zone of apposition. (A) and (B) are two-dimensional images at right angles to each other. The white ellipse indicates the anterolateral papillary muscle which courses toward the left ventricular outflow tract (LVOT), thus tethering the superior bridging leaflet. (ALPM = anterolateral papillary muscle; AO = aorta; IBL = inferior bridging leaflet; LA = left atrium; LV = left ventricle; ML = mural leaflet; SBL = superior bridging leaflet; SEPT = interventricular septum.)
bridging leaflet to the left side to allow more complete closure of the zone of apposition. Patient number 5 died from multiorgan failure and an autopsy confirmed the RT3D echocardiographic findings (Fig 4). This patient also had left ventricular hypertrophy, despite no evidence of left ventricular outflow tract obstruction.

Comment

This report identifies a subgroup of AVSD patients with abnormalities of the superior-mural commissure region, where closure of the zone of apposition may potentially impact outcome. These abnormalities include dominance of the anterolateral papillary muscle, shortened chordae, commissural blunting, and in some cases an oblique zone of apposition between the superior and inferior bridging leaflets, pointing more toward the left ventricular outflow tract. This pathology is similar to that seen in a “mitral valve arcade” as described by Layman and Edwards in 1967 [11]. Their original description included direct attachment of the papillary muscle to the leaflet, or attachment by short and thickened chordae, and loss of commissures in the context of severe mitral regurgitation. Their supposition was that the lack of chordae restricted leaflet movement during systole so that optimal valve position was not obtained to maintain competence.

In the setting of an AVSD, a similar papillary muscle abnormality has been described in pathologic series, with direct attachment of the papillary muscle to the leaflet in a subset of specimens [12]. However, the clinical findings were not known. Surgical series of patients with complete [13] and partial [14] AVSD described poorly formed commissures, chordal, and papillary muscle abnormalities in association with an increased likelihood of postoperative LAVVR.

Identification of this specific pathology by echocardiography should alert the surgeon to perform a more detailed inspection of the sub-valve apparatus at the time of the primary repair. The clue to these abnormalities by 2D echocardiography in our patients was a dominant anterolateral papillary muscle, which could be observed traversing from its basal insertion toward the left ventricular outflow tract. This new 2D echocardiographic finding should warrant a RT3D echocardiographic assessment of the LAVV prior to surgical repair. Although in general there was good agreement between the echocardiographic and surgical findings, RT3D echocardiography has an advantage as it is easier to see beneath the LAVV, this being the optimal location to interrogate commissures and their supporting apparatus. Although the simulated RT3D surgical en face view is “what the surgeon sees,” it does not indicate what lies beneath the LAVV.

When these abnormalities of the anterolateral papillary muscle and commissure are discovered we recommend that the surgeon consider only partially closing the zone of apposition with a single suture, and perform careful intraoperative saline testing before adding any additional sutures. If the defect is a complete AVSD and the superior bridging leaflet is free floating (Rastelli type C), then another useful technique is to displace the line of suturing of the superior bridging leaflet to the VSD patch (or septum) to the right, thus recruiting tissue from the right AV valve to the left. Careful transesophageal echocardiographic evaluation of the LAVV for regurgitation needs to be performed in the operating room prior to decannulation and modifications to the repair considered if there is significant LAVVR.
In our first case the surgeon saw an oblique zone of apposition, but the significance of this was not fully appreciated. In case number 3 the same surgeon was alerted to the similar findings after their significance was appreciated, resulting in modification of the primary repair with a far more desirable immediate outcome. In case number 2 the first surgery was 20 years earlier and at subsequent repair these new findings were not appreciated, with complete closure of the zone of apposition resulting in significant LAVVR. In cases 4 and 5 we understood the significance of this commissural abnormality, with modification to the approach in each case.

It has been previously recognized, particularly in primitive defects, that occasions may arise when closure of the zone of apposition is not warranted, although this in regard to limiting inflow stenosis [15]. Interestingly in our series, complete closure of the zone of apposition, in those with an intact ventricular septum or small ventricular component, appeared to be related to significant early postoperative LAVVR. This is in contrast to surgical series and expert opinion that suggest that surgical closure should be undertaken in all patients with AVSD [1, 16].

Further surgical intervention and valve replacement 20 years after initial repair in 1 patient (case 2) suggests that although these patients may do well for years, this commissural abnormality may ultimately lead to reoperation in this subgroup of patients. However, valve replacement in early adulthood is preferable to a similar procedure in infancy.

Limitations

Limitations of this study are that this was a small number of patients with a variety of AVSD types. There is also a lack of long-term follow-up because the majority of these patients underwent operation since the identification of our index case in 2010.

Conclusions

We have described an uncommon, but significant abnormality of the LAVV in AVSD that challenges the concept that all patients with 2 papillary muscles should undergo closure of the zone of apposition at the time of repair. This commissural abnormality is identifiable by preoperative echocardiography, with a clue from 2D echocardiography and detailed delineation by its RT3D counterpart. If identified it should alert the surgeon to consider modifying the surgical approach to avoid tethering of the leaflets and subsequent LAVVR.

References


We thank Dr Atialno Lacson, Site Chief, Laboratory Medicine and Pathology, University of Alberta for providing us with the autopsy specimen. We also thank Professor Robert Anderson for his advice with regard to the display of the autopsy specimen to optimize the features seen by RT3D echocardiography.