Straddling Right Atrioventricular Valves in Atrioventricular Discordance

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SUMMARY Four hearts are described in which the right atrioventricular valve, draining a morphologically right atrium, straddled and overrode a septum between a right-sided chamber of left ventricular morphology and a left-sided chamber of right ventricular morphology. The degree of override varied between the straddling valve and was committed by 20–45% to the morphologically right ventricle. The ventriculoarterial connections in the hearts were discordant in one, double outlet from the morphologically right ventricle in two and single outlet via an aorta from the morphologically right ventricle with pulmonary atresia in the other. The straddling valve in one of the cases with double outlet had a dual orifice. Pulmonary stenosis was present in three cases, and pulmonary atresia in the fourth. Study of the conduction system in three of the hearts revealed subtle but important differences from the pattern expected in atrioventricular discordance. Each case had an anterior atrioventricular node and penetrating bundle, but the connection thus formed was more lateral than usual, and in the case with ventriculoarterial discordance, the nonbranching bundle was unrelated to the pulmonary outflow tract. A sling of conduction tissue between the anterior node and the regular node was found in the case with single outlet and pulmonary atresia.

STRADDLING VALVES have usually been described either in hearts in which they connect the right atrium to both the right and left ventricles, with the ventricles normally related or in which they connect the left atrium to both normally related ventricles. More rarely, the left atrioventricular (tricuspid) valve may straddle a septum between a larger right-sided chamber of left ventricular morphology and a left-sided smaller chamber of right ventricular morphology. These hearts are intermediate in morphology between congenitally corrected transposition and “single ventricle with inverted outlet chamber.” Another type of straddling valve, a right-sided atrioventricular (mitral) valve straddling a septum in the setting of atrioventricular discordance, is exceedingly rare; we know of only three cases that have been described.

We have encountered four such hearts with various arterial connections. Presence of the straddling valve markedly influences surgical correction of hearts with atrioventricular discordance, and examination of the conduction systems of three of the hearts studied revealed subtle but important differences from the anticipated pattern usually seen in atrioventricular discordance. The findings in these hearts with straddling mitral valves and atrioventricular discordance are the subject of this report.

Materials and Methods

The hearts were from patients who had been attended at the Binnen Gasthuis, University of Amsterdam; the Hospital for Sick Children, London; the Brompton Hospital, London; and the Royal Infirmary, Edinburgh. The clinical findings were nonspecific and the surgical results not relevant to this paper because our major interest was the cardiac morphology. The patients had died at the ages of 3 days, 3 months, 7 years and 29 years, respectively.

Each heart was carefully studied to determine the segmental arrangement and the degree of override of the straddling valve. To do this the annulus of the valve was viewed from the right atrium and the amount of annulus committed to each ventricular chamber was determined. In three hearts, blocks were then removed for study of the conduction system by the method of Smith et al. In the two smaller hearts the entire right atrioventricular orifice was removed and sectioned as a single block of tissue. In a third, larger heart with double outlet arterial connection only the anterior and posterior junctions of the ventricular septum with the atria were removed as separate blocks of tissue.

Autopsy Findings

In each case the basic malformation was situs solitus and atrioventricular discordance with straddling of the right atrioventricular valve across an anterior ventricular septal defect (table 1, figs. 1–5). The degree of override of the straddling valve was 45% to the morphologically right ventricle in cases 2 and 4 and 20% to the morphologically right ventricle in cases 1 and 3. The overriding part of the valve was tethered by an anomalous anterior papillary muscle that was attached to the trabecula septomarginalis in the morphologically right ventricle in cases 1–3. The posteromedial papillary muscle in these cases was present in the hypoplastic morphologically left ventricle.
Table 1. Autopsy Findings in Four Cases with Situs Solitus, Atrioventricular Discordance and Straddling Right Atrioventricular Valve

<table>
<thead>
<tr>
<th>% of overriding of MV to morph RV</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Size of morph LV compared with morph RV</td>
<td>2:3</td>
<td>1:2</td>
<td>2:3</td>
<td>3:5</td>
</tr>
<tr>
<td>Morphology of straddling atrioventricular valve</td>
<td>Anomalous ant. PM attached to TSM</td>
<td>Anomalous ant. PM attached to TSM</td>
<td>Dual orifice “parachute” ant. orifice to TSM</td>
<td>Keyhole valve attached by chordae to infundibular septum and TSM</td>
</tr>
<tr>
<td>Ventriculoarterial connection</td>
<td>Discordant</td>
<td>Single outlet</td>
<td>DORV</td>
<td>DORV</td>
</tr>
<tr>
<td>Aortic position</td>
<td>Anterior, left-sided</td>
<td>Anterior to pulmonary remnant</td>
<td>Anterior, right-sided</td>
<td>Anterior, left-sided</td>
</tr>
<tr>
<td>Pulmonary outflow tract</td>
<td>Stenotic (“squeeze” + tissue tag)</td>
<td>Atretic</td>
<td>Stenotic (squeeze + valve stenosis)</td>
<td>Stenotic (squeeze + valve stenosis)</td>
</tr>
<tr>
<td>Associated anomalies</td>
<td>ASD</td>
<td>ASD</td>
<td>ASD</td>
<td>ASD</td>
</tr>
<tr>
<td></td>
<td>PDA</td>
<td>PDA</td>
<td>Ebstein’s anomaly LAVV</td>
<td>Acquired incompetence, both AV valves</td>
</tr>
<tr>
<td>Conduction tissues</td>
<td>Anterior connection</td>
<td>“Sling”</td>
<td>Anterior connection</td>
<td>Not studied</td>
</tr>
</tbody>
</table>

In case 3 the straddling valve also had a dual orifice (fig. 4). The orifice presenting to the morphologically right ventricle was tethered in parachute fashion to the anomalous papillary muscle. In case 4 the overriding part of the valve was deformed and keyhole-like (fig. 5). The superior edge of the keyhole was attached to the infundibular septum and the inferior edge was attached by short chordae to the trabecula septomarginalis. The part of the valve remaining in the morphologically left ventricle was attached to the posteromedial papillary muscle. In all four hearts the morphologically left ventricle was smaller than the right, the discrepancy in size being most marked in the cases with 45% override. The ventricular chambers were arranged in superoinferior fashion in each case, with the morphologically left ventricle the inferior chamber. The ventricular septum was therefore horizontally positioned. In all cases it extended posteriorly to the crux, where its position was marked externally by the posterior descending coronary artery. Its anterior component ran toward the right margin of the heart and was marked externally by an anterior descending branch from the right coronary artery (morphologically, a left coronary artery). The

Figure 1. The angiogram obtained from case 1 and the specimen cut in matching fashion. (A) The right atrial injection filling the appendage (append), with splitting of the stream of contrast through the right atrioventricular valve (RAVV). Retrospective comparison with the specimen (B) showed that this splitting was the consequence of the RAVV. The asterisk marks the stenotic outflow tract from the morphologically (morph) left ventricle (LV). RV = right ventricle.
ventricular septal defect in all hearts involved predominantly the anterior part of the septum, but extended posteriorly to the crux.

The ventriculoarterial connection varied. Case 1 had arterial discordance, with the pulmonary outflow tract arising from the morphologically left ventricle and wedged between the overriding part of the right atrioventricular valve and the left atrioventricular valve. It was stenotic, and the narrowing was exacerbated by a subvalvar tissue tag herniated from a par-

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**Figure 2.** Further photographs of case 1. (A) The ventricular septal defect after removal of the overriding part of the straddling right valve (arrows) as viewed from the right atrium. (B) The view from the left-sided morphologically right ventricle (RV). To prepare figure 2B, the infundibular septum has been divided (asterisks) so that the stenotic outflow tract from the morphologically left ventricle (LV) can be visualized. Note the fibrous tissue tag herniating into this outflow tract from the partially formed interventricular membranous septum. RAVV = right atrioventricular valve.

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**Figure 3.** Photographs of the autopsy specimen from case 2. (A) The opened right atrioventricular orifice viewed from behind and slightly from beneath. Note the straddling valve (RAVV). The position of the sling of conduction tissue has been marked. The sections obtained at sites a-a, b-b, c-c and d-d are shown in figure 9. (B) The straddling portion of the right valve as viewed from the morphologically right ventricle. Note the small size of the left valve (AVV), the attachment of the straddling valve to the trabecula septomarginalis (TSM) and the atretic pulmonary artery (pulm art). LV = left ventricle; RV = right ventricle.
**Figure 4.** (A) The straddling right valve from case 3, showing its dual orifice. (B) The attachment of the valve to the annulus in the morphologically left ventricle (LV) has been removed to demonstrate the site of the anterior penetrating bundle (dotted lines). $RAVV =$ right atrioventricular valve.

**Figure 5.** Photographs of the straddling valve in case 4. (A) The valve viewed by opening the right annulus from behind. The valve is committed by 45% to the morphologically right ventricle (RV). (B) View from the morphologically right ventricle (left sided). Note the "keyhole" appearance of the straddling portion of the valve attached superiorly to the infundibular septum. Note also the right diaphragmatic obstruction to the pulmonary outflow tract from the RV. (There is double outlet from this ventricle.) $AV =$ atrioventricular valve; $RAVV =$ right atrioventricular valve.
Straddling Mitral Valve

Right Atrium

Block B

-Morph. Left Ventricle-

Block A

Straddling Mitral Valve (cut away)

Cases 3 and 4 both exhibited double outlet from the morphologically right ventricle. In both, the aorta was anterior, but was to the left in case 4; it was slightly to the right of the pulmonary trunk in case 3. A bilateral infundibulum was present in both hearts and both had subpulmonary infundibular obstruction with the outflow tract wedged between the straddling part of the right-sided mitral valve and the left-sided tricuspid valve. This produced a circular diaphragmatic obstruction (fig. 5). Additional pulmonary valvar stenosis was present in both hearts.

Conduction Tissues

The conduction tissues were studied histologically in cases 1–3. The findings were essentially the same in cases 1 and 3. The regular atrioventricular node was normally positioned at the apex of the “triangle of Koch” formed between the insertion of the com-

FIGURE 6. Photograph illustrating the blocks removed for histologic examination in case 1. Block A contains the regular node, which fails to make contact with the ventricular myocardium. Sections through a-a and b-b are shown in figure 7. Block B contains the anterior node and the penetrating bundle. The sections a-a, b-b and c-c make up figure 8.

FIGURE 7. Photomicrographs of the sections through the regular node in case 1 (see figure 6). The node does not make contact with the ventricular myocardium. AV = atrioventricular. Elastic van Gieson stain; magnification (A) × 12, (B) × 6.5.
misuse of the venous valves to the central fibrous body and the septal attachment of the right atrioventricular valve. The node did not make any contact with the underlying ventricular tissues (figs. 6 and 7).

An anterior node, located at the acute point of the atrioventricular junction (figs. 6 and 8) gave rise to a penetrating bundle. This bundle descended directly onto the anterior limb of the ventricular septum and was unrelated to the pulmonary outflow tract in the case with arterial discordance. Having descended
Discussion

Although the presence of atrioventricular discordance potentially "corrects" the effect of arterial discordance, the segmental arrangement of corrected transposition is usually "uncorrected" by associated malformations.12-14 Of the associated malformations, ventricular septal defects, pulmonary stenosis and Ebstein's malformation of the left atrioventricular valve are well recognized.12 Straddling or overriding atrioventricular valves are less common but important because they make problems in accurate diagnosis and because they present a major challenge to the surgeon, who ideally should be aware of their existence before surgery.6 Straddling of the left (tricuspid) valve in the setting of atrioventricular discordance is becoming recognized with increasing frequency,5,14,16 but as far as we know a straddling right (mitral) valve has been described only three times before.7,8

With increasing sophistication of angiographic17 and echocardiographic18 investigations, diagnosis of straddling and overriding atrioventricular valves may be made with increasing frequency. Thus, the significance of the straddling valve is related to surgical correction. As shown by the experience of Tabry et al.,9 palliative procedures may be necessary in the presence of straddling valves. However, in older patients it may be possible to perform more "corrective" procedures, and several options are then available: first, a modified Fontan procedure, as in our case 3; second, sacrifice of the straddling valve with valve replacement and closure of the ventricular septal defect;8 or finally, a septation procedure in the morphologically right ventricle, giving back to the left ventricle the overriding portion of the right valve. In all these options the demonstrated disposition of the conduction tissues and their subtle but important variation from the anticipated pattern in atrioventricular discordance9,10 are of major significance.

In corrected transposition the penetrating atrioventricular bundle is an anterior structure and the extensive nonbranching atrioventricular bundle is intimately related to the pulmonary outflow tract.8 In all the hearts we studied the penetrating bundle was
anterior, but the connecting anterior node was placed more laterally in the right atrioventricular orifice than anticipated in the “standard” variety of corrected transposition. This was related to the more lateral position of the anterior part of the septum. Because of this, the nonbranching atrioventricular bundle in the case with arterial discordance could descend directly onto the septum without encircling the pulmonary outflow tract. In any case, with double outlet right ventricle the bundle would not be expected to be related to the outflow tract because the left ventricle has no direct outlet. Nonetheless, the connecting node was again more lateral than normal and was the only node present.

Similarly, in the case with pulmonary atresia the conduction tissues took origin more laterally in the right atrioventricular orifice than in other hearts of this type that we have examined.9 However, this case was complicated by the presence of a sling of conduction tissue encircling the septal crest. We have encountered such slings previously in the presence of atrioventricular discordance, and Wenink10 recently reviewed other reported examples of this unusual finding. In the present case, as in our other cases, there was good alignment between the atrial septum and the inlet portion of the ventricular septum, a feature lacking in the other two hearts, which had only an anterior connection. This finding reinforces our concept that the presence of a posterior node in atrioventricular discordance relates to the alignment of the atrial and ventricular septal structures.9, 20

When considering these variations in the light of the surgical options, the more lateral position of the anterior node would be of significance during either valve replacement or closure of the right valve during a modified Fontan procedure. Its lateral position, however, would place the anterior conduction system in relatively safe position should the septation option be attempted through the morphologically right ventricle, the latter perhaps the best option from an anatomic standpoint. However, the presence of a sling of conduction tissue, as in our case 3, could jeopardize a septation procedure because the patch would require insertion between the atrioventricular valves at the crux, the precise site of the posterior penetrating bundle. Intraoperative mapping21 is therefore advisable to exclude this possibility, particularly when good septal alignment at the crux increases the likelihood of a sling.

The fact that surgical correction of these cases could involve a septation procedure, together with the decreasing size of the left ventricular chamber and increasing commitment of the straddling valve to the right ventricular chamber, brings up the whole relationship of hearts with straddling valves to univentricular hearts. Quero-Jimenez et al.3 described hearts with straddling of the mitral valve and double inlet to a chamber of right ventricular morphology. If the hearts we describe are similar, as seems likely, then similar hearts may be found with effectively double inlet ventricle and minimal overriding of the mitral valve above the left ventricular type positioned to the right.22 Of course, the hearts we describe are biventricular with atrioventricular discordance and superoinferior ventricles. But in the setting of a univentricular heart of left ventricular type, it is generally accepted that the criterion for inclusion as a single ventricle is the presence of double inlet.23

Hearts are still classified as single ventricle when there is double inlet despite the presence of minimal straddling of one valve.24, 25 When, in a continuum of transfer of a straddling valve from one chamber to another, does the atrioventricular connection change from discordance (or concordance) to double inlet? We suggest that in terms of the atrioventricular connection, the change in this continuum occurs at its midpoint. Therefore, if hearts similar to those we describe were found in which the straddling valve was committed primarily to the left-sided morphologically right ventricular chamber, we would categorize the atrioventricular connection as double inlet and the hearts as univentricular.

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References