Straddling atrioventricular valve with absent atrioventricular connection

Report of 10 cases

SIEW YEN HO, SIMCHA MILO,* ROBERT H ANDERSON,† FERGUS J MACARTNEY,* ANTHONY GOODWIN, ANTON E BECKER, ARNOLD C G WENINK, LEON M GERLIS, JAMES L WILKINSON

From the Department of Paediatrics and Surgery, Cardiothoracic Institute, Brompton Hospital, Fulham Road, London; Thoracic Unit, The Hospital for Sick Children, London; Freeman Road Hospital, Newcastle-upon-Tyne; Department of Pathology, University of Amsterdam, Holland; Department of Anatomy, State University of Leiden, Holland; Department of Pathology, Grimsby General Hospital, South Humberside; and Institute of Child Health, University of Liverpool

SUMMARY Six hearts with absent right connection and four hearts with absent left connection in association with straddling of an atrioventricular valve are described. Their categorisation is made easier by adopting a segmental approach. The conduction tissue was studied in two hearts and the arrangements were unusual.

Straddling of an atrioventricular valve may be defined as the arrangement in which its tension apparatus is attached to both sides of the ventricular septum.1 2 When defined as such, the most frequent example of a straddling valve is an atrioventricular septal defect with a common atrioventricular orifice ("complete endocardial cushion defect" or "complete atrioventricular canal malformation"), though some authorities question whether "straddling" should be used in this context.3 4 Less frequent are the hearts in which there are two atrioventricular valves and either the right valve or the left valve, or, exceptionally, both valves, have their tension apparatus attached to both sides of the septum.2-4 These hearts pose a major problem in surgical treatment.1 Hearts have also been reported, however, in which either the right or left valve straddles when the other valve is atretic. These latter hearts are exceedingly rare,3 5-8 but they pose particular problems in classification, particularly concerning their relation to common atrioventricular orifice.9 We have now examined 10 such hearts from various centres and studied their morphology and, in two cases, the distribution of their conduction tissues. The unusual findings encountered form the basis of this report.

Definitions and terms

A straddling atrioventricular valve is one in which the tension apparatus is attached to both sides of a ventricular septum. Usually when an atrioventricular valve straddles, its annulus is committed to both chambers in the ventricular mass. The degree of commitment is termed the override, and is described according to the percentage of the valve annulus attached to each ventricular chamber.

It is our convention in hearts with overriding valves to distinguish the atrioventricular connection present according to the precise connection of the abnormal valve. Thus, the atrioventricular connection must be either double inlet, or else concordant, discordant, or ambiguous according to the morphology of the chambers connected together. Irrespective of the connection, of necessity one of the ventricular chambers must be more or less rudimentary while the other ventricle will unequivocally be the main or primary chamber. When one atrioventricular orifice is atretic and the other orifice overrides, this condition does not apply. There is always absence of one atrioventricular connection. The sole remaining ventricular inlet por-

*RHA and FJM are supported by the British Heart Foundation together with the Joseph Levy and Vanderwell Foundations, respectively; and SM was a Visiting Fellow from Chaim Sheba Medical Centre, Tel Aviv, Israel.

†RHA is also Honorary Senior Lecturer to The Hospital for Sick Children and Honorary Lecturer to the University of Liverpool.

Accepted for publication 2 December 1981
Straddling AV valve with absent connection

...tion, together with the straddling valve, is then shared between the two ventricular trabecular components, meaning that both ventricular chambers are more or less rudimentary. None the less, except when the override is exactly equal, one of the chambers can be nominated as the main ventricular chamber and the other as the secondary chamber. It is also possible to describe the pattern of ventricular architecture present. For the sake of clarity, the overriding valve will be described simply as right or left, depending on whether it drains the right-sided or left-sided atrium. But, for those who wish to know whether the valve is basically tricuspid (morphologically right) or mitral (morphologically left), this can be deduced knowing the atrial situs and ventricular architecture of a given heart. Where the valve was recognisably mitral or tricuspid, this was stated.

Material and methods

Ten hearts were included in the study (Table). They came from cardiological collections of the Brompton Hospital, London; The Hospital for Sick Children, London; Royal Liverpool Children's Hospital; Killingbeck Hospital, Leeds; Grimsby General Hospital; Newcastle General Hospital, Newcastle; Wilhelmina Gasthuis, Amsterdam; and the State University Hospital of Leiden.

Careful studies of each heart were undertaken to determine the segmental arrangement and the degree of override of the straddling valve. This was done by viewing the annulus of the valve from its atrial side and thus determining the degree of commitment of the annulus to each of the ventricular chambers. Two hearts were studied so as to determine the disposition of their conduction system, using the technique described by Smith et al. The cardiac apex and top of the atria were removed, leaving the atrioventricular junction. The heart with left atrioventricular valve atresia was then divided into three blocks—a left parietal, a right parietal, and a septal block. The heart with right atrioventricular valve atresia was processed as one piece after the removal of the left parietal wall. Serial sections were cut from each block at 10 μm thickness. All the sections were kept and every 25th section mounted and stained with the Masson's trichrome technique. Further sections were mounted and stained wherever deemed necessary.

Results

The hearts studied could be divided into two basic groups, those with absent right atrioventricular connection (atresia of the right atrioventricular valve) and those with absent left connection (atresia of the left atrioventricular valve). The hearts were further subdivided into two groups according to the trabecular morphology of the main ventricular chamber, that is of right or left ventricular type. The position and the morphology of the atrioventricular valves were also noted. In each of these groups, we identified hearts with rudimentary secondary ventricular chambers to the left or to the right of the main chamber, the morphological pattern always being complementary to that of the main chamber. Among the hearts studied, six had absent right and four had absent left atrioventricular connection.

Hearts with absent right atrioventricular connection

In this group, four hearts had a main chamber with right ventricular trabecular pattern and two had main chambers of left ventricular type. The rudimentary chamber of left ventricular morphology was positioned to the right and posterior to the main chamber in cases 1-3 (left-hand pattern ventricular architecture—Fig. 1) and to the left and posterior in case 4 (right-hand pattern ventricular architecture). In the two specimens with rudimentary secondary chamber of right ventricular type, the rudimentary chamber was right anterior in one case (right-hand pattern) and left anterior in the other (left-hand pattern). In this respect the terms “right-hand pattern” and “left-hand pattern” are used to describe the ventricular architecture as suggested by Van Praagh and his colleagues and are independent of either the atrioventricular connection or the ventricular relation.

The straddling and overriding valve in this group was the left atrioventricular valve. Its morphology was as anticipated from the presumed ventricular architecture in all cases. In the three cases with left-hand pattern architecture, the left-sided atrioventricular valve was a morphologically tricuspid valve. In the case with right-hand architecture in which the main chamber was of right ventricular morphology the atrioventricular valve was bicuspid in appearance. Both the left valves in the hearts with left ventricular main chambers were of mitral morphology. The least commitment of the atrioventricular valve to the main chamber in this group was 65% and the maximum was 80%. The trabecular septum in the hearts with main chambers of right ventricular type was without exception found to extend to the crux cordis. In the hearts with main chambers of left ventricular type, the septum extended to the crux in one but was an anterior structure in the other heart.

The ventriculoarterial connections varied. Three of the four hearts with right ventricular main chambers had double outlet from the main chamber and the other had pulmonary atresia, the aorta arising from the main chamber. The two specimens with main chamber of left ventricular morphology had discor-
Table 1  

<table>
<thead>
<tr>
<th>Mode of connection chamber</th>
<th>Main chamber morphology</th>
<th>Position and morphology of rudimentary chamber</th>
<th>Straddling valve</th>
<th>Septum to crux</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absent right connection</td>
<td>Right ventricular type</td>
<td>Left ventricular type right posterior</td>
<td>LAVV(TV)</td>
<td>Yes</td>
</tr>
<tr>
<td></td>
<td>Left ventricular type</td>
<td>Left ventricular type left posterior</td>
<td>LAVV(TV)</td>
<td>Yes</td>
</tr>
<tr>
<td>Absent left connection</td>
<td>Right ventricular type</td>
<td>Right ventricular type right anterior</td>
<td>LAVV(MV)</td>
<td>Yes</td>
</tr>
<tr>
<td></td>
<td>Right ventricular type</td>
<td>Right ventricular type left anterior</td>
<td>RAVV(TV)</td>
<td>No</td>
</tr>
<tr>
<td></td>
<td>Left ventricular type</td>
<td>Left ventricular type left anterior</td>
<td>RAVV(TV)</td>
<td>No</td>
</tr>
</tbody>
</table>

LAVV, left atrioventricular valve; RAVV, right atrioventricular valve; DOMC, double outlet main chamber; MV, morphologically mitral valve; TV, morphologically tricuspid valve; PFO, patent foramen ovale; DORC, double outlet rudimentary chamber; PA, pulmonary atresia; ASD, atrial septal defect; L SVC, left superior vena cava; PDA, persistent ductus arteriosus.

Rudimentary ventriculoarterial connections. The aorta was anterior to the pulmonary trunk or its atretic remnant in two cases, right anterior in three cases, and left anterior in the other. A common finding in all specimens of this group was an interatrial communication, either a patent foramen ovale or a secundum atrial septal defect. Interruption of the aortic arch was found in case 1.

HEARTS WITH ABSENT LEFT ATRIOVENTRICULAR CONNECTION

Four hearts were found in this group, two (cases 7, 8)

Fig. 1  The heart from case 1.  
(a) The heart viewed from the front. The aorta arises anterior to the left of the pulmonary trunk.  
(b) The blind-ending right atrium opened and viewed from above and behind.  
(c) The right-sided rudimentary chamber with left ventricular pattern. The left atrioventricular valve straddles through the septal defect.  
(d) The left-sided main chamber with right ventricular trabeculations.
Straddling AV valve with absent connection

<table>
<thead>
<tr>
<th>% to main chamber</th>
<th>Ventriculoarterial connection</th>
<th>Aortic position</th>
<th>Cardiac position</th>
<th>Associated anomalies</th>
<th>Case No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>70%</td>
<td>DOMC</td>
<td>Left anterior</td>
<td>Left</td>
<td>ASD, interrupted arch</td>
<td>1</td>
</tr>
<tr>
<td>80%</td>
<td>DOMC</td>
<td>Anterior</td>
<td>Mid-line</td>
<td>PFO</td>
<td>2</td>
</tr>
<tr>
<td>80%</td>
<td>PA, aorta from main chamber</td>
<td>Anterior to atretic pulm trunk</td>
<td>Right</td>
<td>PFO, PDA</td>
<td>3</td>
</tr>
<tr>
<td>75%</td>
<td>DOMC</td>
<td>Right anterior</td>
<td>Left</td>
<td>Secundum ASD</td>
<td>4</td>
</tr>
<tr>
<td>80%</td>
<td>Discordant</td>
<td>Right anterior</td>
<td>Left</td>
<td>PFO</td>
<td>5</td>
</tr>
<tr>
<td>65%</td>
<td>Discordant</td>
<td>Right anterior</td>
<td>Not known</td>
<td>PFO</td>
<td>6</td>
</tr>
<tr>
<td>70%</td>
<td>DOMC</td>
<td>Right anterior</td>
<td>Left</td>
<td>PFO, persistent LSVC</td>
<td>7</td>
</tr>
<tr>
<td>95%</td>
<td>PA, aorta from main chamber</td>
<td>Anterior to atretic pulm trunk</td>
<td>Left</td>
<td>PFO, PDA</td>
<td>8</td>
</tr>
<tr>
<td>65%</td>
<td>Concordant</td>
<td>Right posterior</td>
<td>Left</td>
<td>Secundum ASD</td>
<td>9</td>
</tr>
<tr>
<td>75%</td>
<td>DORC</td>
<td>Right</td>
<td>Left</td>
<td>PFO</td>
<td>10</td>
</tr>
</tbody>
</table>

with a main chamber of right ventricular morphology (Fig. 2) and two with main chambers of left ventricular morphology (Fig. 3). In hearts with right ventricular main chambers, the rudimentary chamber was found to the left and posterior to the main chamber (right-hand pattern ventricular architecture). The sole atrioventricular valve was a morphologically tricuspid valve, being committed by 70% and 95% to the main chamber. Though the main chamber in both was of right ventricular morphology, the septum extended to the crux cordis only in one. This particular heart was unusual in that though it was in the left chest with its apex directed to the left, the ventricular mass was rotated so that the crux was in relatively anterior position. None the less, the septum between the ventricular chambers still extended to the point where the atrial septum joined the atrioventricular junction. The ventriculoarterial connection was double outlet main chamber, with the aorta right and anterior to the pulmonary trunk in one heart while in the other the aorta arose from the main chamber anterior to the atretic pulmonary trunk.

The two hearts with main chambers of left ventricular morphology had anterior right ventricular rudimentary chambers located to the right in one (right-hand pattern architecture) and to the left in the other (left-hand pattern). Though it drained predominantly (65%) to a main chamber of left ventricular type situated on the left, the atrioventricular valve had a tricuspid morphology in one of these hearts. In the other the atrioventricular valve had four cusps and was neither tricuspid nor mitral in appearance. It was committed by 75% to the main chamber. The septum did not reach the crux in either heart. The ventriculoarterial connection was concordant in one heart, the aorta arising to the right and posterior of the pulmonary trunk. Both great arteries originated from the rudimentary chamber in the other with the aorta to the right of the pulmonary trunk.

CONDUCTION TISSUE DISPOSITION

(a) With absent right connection, straddling left valve, main chamber with right ventricular morphology, and rudimentary left ventricular chamber in right posterior position (left-hand pattern ventricular architecture)

The atrioventricular conduction system was in the form of a sling between two atrioventricular nodes (Fig. 4, 5). The posterior node was located in the posterior interatrial septum above the fibrous annulus of the left valve. The penetrating bundle perforated the fibrous annulus and the long non-branching bundle then descended steeply along the posterior septum. The bundle then ran along the crest of the defect beneath some chordae tethering the straddling valve. Towards the anterior rim, the bundle dropped off the edge to the left side and gave off a large right bundle-branch to the left sided morphologically right ventricle. The anterior limb of the bundle ascended sharply as it looped backwards to contact an anterior node. This node was well formed and situated in the right atrial aspect of the anterior part of the atrial septum.

(b) With absent left connection, straddling right valve, main chamber with left ventricular morphology, and rudimentary left ventricular chamber in left posterior position (right-hand pattern ventricular architecture)

There was gross malalignment between the atrial
septum and the ventricular septum. The atrial septum lined up with the left lateral wall of the heart such that the left posterior quadrant of the right atrioventricular valve straddled the ventricular septum (Fig. 6). Atrioventricular ring conduction tissue was found in clusters at various points around the right atrioventricular orifice on the atrial aspect. The rudimentary left ventricular chamber was very small and a left bundle-branch could be seen on the left side of the septum. This led to a non-branching bundle which ran up a free-running trabecula in the back of the rudimentary chamber. The trabecula was adherent to the septum at its base. The bundle perforated the valve annulus in its posterior border and made contact with an atrioventricular node formed from a cluster of ring tissue in the back of the right atrium. Further to the left but more anteriorly along the orifice there was a cluster of specialised tissue which resembled an atrioventricular node. This node had an arterial supply but did not give rise to a penetrating atrioventricular bundle.

Discussion

Hearts with straddling of one atrioventricular valve and atresia of the other are exceedingly rare. Though difficult to categorise, we have shown that adoption of a segmental approach makes their description much easier. The necessary information for full description is whether the atrium lacking an atrioventricular connection is right- or left-sided. As a corollary the straddling valve must be on the other side. The ventricular architecture together with the relation of the ventricular chambers must then be described and the commitment of the straddling valve to the ventricular chambers must be specified. The ventriculoarterial connection and relations, together with a catalogue of associated malformations, complete the description.

Fig. 2. (a) The heart from case 7 viewed from the front. The aorta is anterior and to the right of the pulmonary trunk. The asterisks mark the line of dissection shown in (d). (b) A view of the left atrium from the back. The open arrows indicate the orifices of the pulmonary veins. The “dimple” represents the floor of the left atrium but this has no connection with the ventricular mass. (c) The main chamber which receives the main part of the single atrioventricular valve. There is a bilateral infundibulum supporting the outlets. (d) An apical view of the heart displayed by opening the ventricular mass along the line of dissection (asterisks marked in (a)). The right atrioventricular valve straddles into the rudimentary chamber of left ventricular type.
This can then be augmented by describing the morphology of the straddling valve and determining whether the septum runs to the crux of the heart.

Although use of such an approach gives no difficulties in description, problems are encountered when the hearts are categorised in the hierarchy of congenital anomalies, specifically as to whether they should be considered as having biventricular or univentricular connections. Associated with this are problems in interpretation of the nature of the sole atrioventricular valve, specifically as to whether it is a mitral valve, a tricuspid valve, or a common valve. We now use the terms "biventricular" and "univentricular" simply as a means of grouping together hearts with comparable atrioventricular connections. In hearts with concordant, discordant, or ambiguous connections, each atrium is connected to its own ventricle. These are the hearts we term biventricular. In most hearts with double inlet or with absence of one atrioventricular connection, only one ventricular chamber has an atrioventricular connection and these are the hearts grouped together as having univentricular connections. In hearts with overriding of one of two or a common valve, the connection is determined according to the commitment of the overriding valve and the hearts categorised accordingly as having biventricular or univentricular connections. In the hearts presently described there is clearly a problem. One atrioventricular connection is absent, but both ventricular chambers are connected to the remaining atrial chamber.

Since there is only one ventricular inlet portion shared between the ventricular trabecular components, each ventricular chamber is more or less rudimentary. Arguments about whether one chamber is a ventricle and the other not in this context are non-productive, as indeed they have proved to be in all hearts with two ventricular chambers. Both are ventricular chambers. Each is recognisable according to its trabecular pattern as right or left ventricular in type. In each heart we have examined it was possible to determine the pattern of ventricular architecture present, and which ventricular chamber was connected to most of the overriding valve. In the necropsy material studied here we had no problem in making this distinction, the most nearly equal sharing being 65% to one chamber and 35% to the other.
Thus, we had no difficulty in distinguishing a main ventricular chamber from a secondary ventricular chamber. All of this information is required for a full description of these complex cases, together with the precise relations of the ventricular chambers. When given in this fashion, the description provided transcends possible arguments as to whether the hearts have univentricular or biventricular connections, or as to whether they are "single" ventricles. With regard to the connection, it does not really matter. With regard to "singleness", they clearly have two ventricular chambers. Some have categorised the hearts as "double outlet atrium." Thus, in hearts with right ventricular main chambers, the left-handed valve was a tricuspid valve with left-hand pattern ventricular architecture and a mitral valve with left-hand pattern architecture. The association was less perfect when the main chamber was of left ventricular morphology. But this is to be expected, since, when there is double inlet to a left ventricular chamber, both the valves resemble mitral valves. Some have suggested that the sole valve is a common valve. Though on occasions the valve had morphological features of a common valve, in all cases it drained only one atrial chamber. We believe the term "common valve" should be reserved for those valves draining both atrial chambers. The valves in these hearts are best described as simply right valves or left valves depending on whether they drain the right-sided or left-sided atrial chambers, respectively. This convention is unambiguous and cannot lead to confusion. Morphology of the valve can always be specified if required, and can usually be inferred from the ventricular architecture present. It should also be appreciated that the nature of the atretic valve can also be inferred from the ventricular architecture for those

![Diagram of heart anatomy](image-url)
who are concerned with morphogenesis. Thus, in our grouping of hearts in terms of absent right or absent left connection, we have included cases together which can be interpreted in terms of mitral or tricuspid atresia. For those requiring knowledge of the presumed morphology of the atretic valve, this can be inferred knowing the situs, the ventricular architecture, and which connection is absent.

The conduction tissue was unusual in both the hearts we examined morphologically. Both hearts had the straddling valve, of tricuspid morphology, committed in its greater part to a right ventricular chamber. In one of the hearts, it was the left connection which was absent and the rudimentary left ventricular chamber was left-sided (right-hand pattern architecture). The trabecular septum did not reach the crux and was malaligned relative to the atrial septum. As in hearts with atrioventricular concordance and straddling tricuspid valve, where the same septal orientation is found, the connecting node was formed posterolaterally in the right atrioventricular orifice, at the point where the malaligned trabecular septum reached the atrioventricular junction. A regular node at the apex of the triangle of Koch did not give rise to a penetrating atrioventricular bundle.

The second heart studied had absence of the right atrioventricular connection, and in this heart the straddling left valve was committed mostly to the right ventricular chamber, but the rudimentary left ventricular chamber was right-sided and posterior (left-hand pattern ventricular architecture). It is well established that in either situs solitus or with atrial isomerism13-14 this pattern is accompanied by an abnormal conduction tissue disposition, frequently with a “sling” between two connecting nodes.15 This was indeed the arrangement found in this particular case, but surprisingly the posterior connecting node was not the regular node in the triangle of Koch.

Instead the posterior node was formed in the left atrioventricular orifice, but was again formed at the point where the malaligned trabecular septum made contact with the atrioventricular junction. Thus, it seems that these cases conform with our basic rules for conduction tissue disposition,16-17 namely that a trabecular septum joining the atrioventricular junction at a point other than the crux and a left-hand pattern of ventricular architecture with situs solitus both presage an anomalous location for the connecting atrioventricular node. The present findings show that this anomalous connecting node cannot only be formed at any point within the right atrioventricular orifice, but also, in patients with situs solitus atrial chambers, can be formed within the left atrioventricular orifice. As far as we are aware, only Lev et al.18 have previously described a connecting atrioventricular node in the posterior wall of the left atrium. As yet we have not had the opportunity to study histologically hearts with absent connection and straddling valve in which the valve is committed mostly to the left ventricular chamber. In that in all such hearts except one studied grossly in this series the trabecular septum never reached the crux, we anticipate anomalous conduction systems in the majority. Only further histological examination will test the veracity of this prediction, and show what happens when a trabecular septum does extend to the crux.

References


Requests for reprints to Professor RH Anderson, Cardiothoracic Institute, Brompton Hospital, Fulham Road, London SW3 6HP.