Communication of Mitral Valve with Both Ventricles Associated with Double Outlet Right Ventricle

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SUMMARY
A rare case of an infant with “double inlet right ventricle” (straddling mitral valve) and double outlet right ventricle is described. Coarctation of the aorta was associated.

A left-to-right shunt resulted from part of the mitral valve opening into the right ventricle and also from the presence of a ventricular septal defect. The infant’s clinical features reflected the shunt, although the valvular abnormality had not been suspected clinically. It is anticipated that left atrioventriculography might have identified this rare type of straddling atrioventricular valve.

Additional Indexing Words:
Double inlet right ventricle
Coarctation of aorta
Left-to-right shunt
Straddling mitral valve

The anomaly in which the tricuspid valve straddles the ventricular septum and communicates with both ventricles is rare. It may be associated with either a normal or an atretic mitral valve. This condition is commonly called double inlet left ventricle or straddling tricuspid valve.

We observed a case with the reverse of the above situation. In our patient, the mitral valve straddled the ventricular septum and led to both ventricles, while the tricuspid valve communicated only with the right ventricle. Both great arteries arose from the right ventricle (“double outlet right ventricle”).

To our knowledge, there are only two reported cases of straddling mitral valve while the tricuspid valve opened into the right ventricle.1 In two other reported cases, while the mitral valve straddled the ventricular septum and opened into both ventricles, the tricuspid valve was “displaced” and opened into the left ventricle.2

The rarity of the condition prompts us to place our case on record.

Report of Case

Clinical Features
The patient, a 7-week-old male infant, was born following an uncomplicated pregnancy. Other than unexplained neonatal jaundice, he did well until 3 weeks of age when repeated vomiting occurred and an inguinal hernia and cardiomegaly were discovered. He was transferred to the University of Minnesota Hospitals for further study. There was no history suggesting cardiovascular disease.

On admission, the patient was a well-developed male infant in no acute distress. The blood pressure, obtained by the flush method, was 100 mm Hg in the right arm and 70 mm Hg in the right leg. Tachycardia and tachypnea were present. A grade I/IV short systolic murmur was present along the lower left sternal border. The pulmonic component of the second heart sound was accentuated. Hepatomegaly was present.

Thoracic roentgenograms at the time of admission revealed generalized cardiomegaly. The pulmonary vascular markings were accentuated although it was difficult to determine whether or not these were arterial or venous in nature. The clinical diagnosis was coarctation of the aorta and congestive cardiac failure. A retrograde aortogram demonstrated coarctation of the aorta of severe degree.

Over the next week, a grade III/IV pansystolic murmur became evident along the lower left sternal border and, in addition, a short apical mid-diastolic murmur was present. These findings indicated a ventricular septal defect. Despite intensive treatment for cardiac failure, the patient did poorly so that at 7 weeks of age the coarctation of the aorta was resected. Three hours following the operation, cardiac arrest

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Supported by Public Health Service Research Grants RO1 HL05894 and Research Training Grant 5 T01 HL05570 from the National Heart and Lung Institute.

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Received December 26, 1972; revision accepted for publication April 13, 1973.
occurred and attempts at resuscitation were unsuccessful.

Pathologic Findings

The body configuration was that of situs solitus. The essential pathologic findings were limited to the cardiovascular system. Except for evidence of recent aortic anastomosis for coarctation of the aorta, the principal findings were intracardiac. The heart was grossly enlarged. The apex was directed to the left and formed by the left ventricle. The external appearance of the great arteries showed abnormal relationship. The aorta lay anterior and somewhat to the right of the pulmonary trunk (fig. 1a).

The anatomic right atrium was on the right side and anterior to the left atrium. It received the openings of the coronary sinus and superior and inferior venae cavae normally. The fossa ovalis was present on the right atrial side and associated with a valvular competent patent foramen ovale. The atrial septum was in a normal plane. The right atrium opened through the tricuspid valve into the right ventricle (fig. 1b).

The morphological left atrium was on the left side and posterior to the right atrium. It received the pulmonary veins normally. The mitral valve possessed two leaflets, anterior and posterior. Two main papillary muscles supported the mitral valve, one attached to the lateral wall of the left ventricle, the other to the anterior wall of the right ventricle. With this arrangement, each mitral leaflet straddled a subjacent large ventricular septal defect and communicated about equally with each ventricle (fig. 2). The only outlet for the left ventricle was the aforementioned ventricular septal defect. The plane of the ventricular septum lay at right angles to that of the atrial septum (fig. 2b), the right ventricle lying anteriorly and the left ventricle posteriorly.

In contrast to the small left ventricle, the right ventricle showed a large cavity and its wall was hypertrophied. It received the opening of the tricuspid as well as part of the mitral valve. The posterior wall of the right ventricle showed a vertical ridge starting below the posterior cusp of the pulmonary valve and extending to the apex. The outflow of the right ventricle was in a more posterior position than normal and gave origin both to the pulmonary trunk and to the aorta (fig. 3). The pulmonary valve, which was wider than that of the aorta, lay posteriorly and to the left of the aortic valve. No mass of muscle separated the two tracts leading to the two semilunar valves. Each valve showed three cusps and lay at the same horizontal plane of the body and neither was in continuity with either atrioventricular valve.

The cusps of the aortic valve were oriented left and right and posterior. The coronary arteries arose in relation to left and posterior cusps, the former showing the distribution of a left coronary artery, the latter that of a right coronary artery. The essential abnormalities are summarized in figure 4.

Comment

In the case described, the main abnormalities consisted of (1) the mitral valve straddling the deficient ventricular septum and communicating with both ventricles while the tricuspid valve opened normally into the right ventricle, (2) origin of both great arteries from the right ventricle.

Figure 1

(a) Exterior of heart and great vessels from in front. The aorta (A.) lies anterior and slightly to the right of the pulmonary trunk (P.T.). (b) Right atrium (R.A.) and right ventricle (R.V.). The tricuspid valve (T.V.) opens exclusively into the right ventricle.

Circulation, Volume XLVIII, October 1973
The case reported presents for discussion some of the complexities involving relationships of the atrioventricular valves and the ventricles. This requires certain definitions. A straddling atrioventricular valve is one which, above a ventricular septal defect, joins both ventricles. Straddling valves may be associated either with "displacement" or with normal position of the other valve.

A "displaced" valve, according to use of the term by Libethson, is one which leads into the contralateral ventricle. Thus, a displaced tricuspid valve leads from the right atrium exclusively into the left ventricle.

In classic examples of double inlet ventricle, one valve is normally positioned while the other valve straddles. Thus, in the situation wherein the tricuspid valve straddles, the condition may be termed double inlet left ventricle. Less commonly as in our case, the mitral valve straddles leading to a condition which may be termed double inlet right ventricle.

In two cases described by Libethson et al., the mitral valve opened into both ventricles but the tricuspid valve was "displaced" and opened only into the left ventricle. These cases differ from ours in that in ours the tricuspid valve was properly placed, opening exclusively into the right ventricle, while the mitral valve straddled the ventricular septum.

Seven cases under the title of exaggerated displacement of the atroioventricular canal towards the bulbus cordis (rightward displacement of the mitral valve) were reported by Quero Jimenez et al.

Utilizing our definitions, we would classify the cases of Quero Jimenez et al. as follows: cases 1, 3 and 4 as complete type of persistent common atrioventricular canal; case 2 as single ventricle of the right ventricular type; and cases 6 and 7 as examples of double inlet right ventricle. The remaining case (case 5) had been observed only clinically and complete atrioventricular canal has not been excluded.

As far as we are aware, there are only three reported cases with the type of atrioventricular valvular malformation as in our case. These are cases 6 and 7 of Quero Jimenez and the case of Munoz Castellanos et al. reviewed by Quero Jimenez.

The added feature of double outlet right ventricle seen in our case was present in one case (case 7) of Quero Jimenez and the case of Munoz Castellanos et al.
In our patient, the exact intracardiac abnormalities were not identified clinically. No special intracardiac studies were done but a diagnosis of ventricular septal defect had been made on a clinical basis. Had right ventriculography been performed, it is assumed that the double outlet right ventricle would have been identified. The left-to-right shunt at the ventricular level would, in all probability, have been considered as resulting from flow through the ventricular septal defect, while in fact some of the arterialized blood entering the right ventricle came through that part of the mitral valve which opened into the right ventricle.

Left atrigraphy would have been the ideal method of identifying the straddling mitral valve in a manner comparable to demonstrating the straddling tricuspid valve by right atrigraphy.2,3

Perhaps some insight into the basis for straddling of the mitral valve may be gained from an understanding of the development of the ventricular septum as it relates to straddling of the tricuspid valve.

In the primitive heart, the single atrium is separated from the single ventricle by the atrioventricular canal.4 From the latter, the tricuspid and mitral valves develop. The right ventricle develops as a protrusion from the base of the primitive ventricle, while the left ventricle is derived from the primitive ventricle. This means that, at one stage, the primordia of the mitral and tricuspid valves face the left ventricle. As the right ventricular chamber undergoes further development, there is a shift of

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**Figure 3**
Right ventricle showing origin of aorta (Ao.) and pulmonary valve (P.V.) from the base of the chamber. The tricuspid valve (T.V.) opens into the right ventricle (R.V.) as does part of the mitral valve (M.V.).

**Figure 4**
Diagrammatic portrayal of essential abnormalities observed in case. In the main body of the illustration is shown the deficient ventricular septum beneath a ventricular septal defect. Above this zone, the mitral valve straddles the ventricular septal defect so as to communicate with both ventricles. The tricuspid valve opens only into the right ventricle. In the insert is shown the position of the great vessels, the feature of double outlet right ventricle and the coarctation of the aorta.
the atroventricular canal toward the right so that the tricuspid valve comes to lie over the right ventricular chamber while the mitral valve remains over the primitive or left ventricle. If there is a failure in complete rightward shift of the atroventricular canal, the tricuspid valve may fail to become positioned entirely over the right ventricle and so straddle the underlying ventricular septum, yielding the condition straddling of the tricuspid valve or double inlet left ventricle. As far as we are aware, there is no stage in development of the normal heart at which an arrest in growth could yield the straddling mitral valve (double inlet right ventricle). If, on the other hand, there is an abnormal rightward shift of the atroventricular canal, the mitral valve would come to lie in part over the right ventricle.

A second explanation could be an abnormal direction of the growth of the ventricular cavities as supported by the malposition of the ventricular septum. The papillary muscles and chordae are derived from the walls of the respective ventricular cavities. Hence, if a valve has its attachments to both sides of the ventricular septum, it is probable that the development of the two ventricular cavities was abnormal and at a time subsequent to the appearance of the chordal attachments.

From the viewpoint of double outlet right ventricle, our case has certain points of interest. The anterior position of the aorta with respect to the pulmonary trunk is unusual, though not unique. In the majority of cases of double outlet right ventricle, the ascending aorta lies to the right of the pulmonary trunk rather than anterior to it. Regarding associated anomalies, coarctation of the aorta, which was present, is common. As experience with double outlet right ventricle has increased, it has become apparent that intracardiac anomalies are not unusual. In our case, this took the form of double inlet right ventricle.

References
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_Circulation_. 1973;48:904-908
doi: 10.1161/01.CIR.48.4.904

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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