Straddling and Displaced Atrioventricular Orifices and Valves with Primitive Ventricles

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SUMMARY
Fourteen cases of straddling or displaced atrioventricular (A-V) orifices and valves are presented. There are three types: (1) straddling tricuspid orifice entering both primitive ventricles with the mitral valve entering the left ventricle (nine cases), (2) displaced tricuspid orifice into the primitive left ventricle, with no A-V orifice in the primitive right ventricle (three cases), and (3) displaced tricuspid orifice with straddling mitral valve, in which the mitral valve has connections in both primitive ventricles, with the tricuspid entering the primitive left ventricle (two cases). Pathologically, these hearts are not single ventricle since there is a primitive right ventricle containing a portion of sinus as well as conus. Embryologically, however, these hearts are related to single ventricle, all being due to insufficient or no passage of the atrial canal region to the right during the process of absorption of the bulbus. Selective angiocardiography may differentiate straddling A-V orifices from single ventricle by showing immediate filling of both ventricles by injection into the right or left atrium.

Additional Indexing Words:
Angiocardiography  Congenital heart disease  Displaced orifices
Embryology  Single ventricle  Straddling A-V orifices

A STRADDLING atrioventricular (A-V) orifice and valve is defined in this work as that condition in which either A-V orifice enters both ventricular chambers, and has papillary or chordal attachments in both. Only those hearts with two distinct A-V orifices, or those in which one is atretic are included in this study. Hearts with common A-V orifices are excluded for semantic reasons, for they normally straddle, although conceptually some of them belong to this group. All hearts studied have morphologic right and left ventricles, containing both sinus and conus portions, thereby excluding a single ventricle. The ventricles, however, are primitive, in the sense that only a small sinus is present in the right ventricle, whereas a portion of right ventricular sinus in addition to the entire left

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ventricular sinus is present in the left ventricle.

The term displaced A-V orifice is reserved for that condition in which one ventricle receives both A-V orifices entirely, while the other receives none. Here again, these hearts have two distinct A-V orifices, or one orifice may be atretic, and two grossly identifiable but primitive ventricles with both sinus and conus elements in each.

The concept of straddling and displaced A-V orifices has received little attention in the literature. Straddling right or left A-V ("tricuspid") valve has been named and described by Rastelli et al.,\(^2\) Keith et al.,\(^3\) and Lev et al.\(^1\) Hearts of this description have been called double-inlet left ventricle by Mehrizi et al.\(^4\) and by de la Cruz and Miller.\(^5\) They have been described by Lambert\(^6\) and subsequently called Lambert's heart,\(^7\) and have been included by Van Praagh et al.\(^8\) as a type of single ventricle. Displaced right A-V ("tricuspid") orifice, and straddling left A-V ("mitral") orifice associated with displaced tricuspid orifice have not, to our knowledge, been described in the literature.

Of the 3,110 hearts seen at the Congenital Heart Disease Research and Training Center from 1957 to the present, eight have had straddling right A-V ("tricuspid") orifices, one has had a straddling left A-V ("tricuspid") orifice with ventricular inversion, three have had displaced right A-V ("tricuspid") orifices, and two have had a straddling left A-V ("mitral") orifice with a displaced right A-V ("tricuspid") orifice.

**Definition of Terms**

In this work, the diagnosis of atria and ventricles is made on the basis of septal morphology.\(^9\) To qualify as a ventricle, a chamber must have both a sinus and a conus directly related to it.* The sinus is that portion of the ventricle which normally houses the tensor apparatus of an A-V valve, i.e., the papillary muscles and chordae; but the sinus of the right ventricle also includes the apical recess. When this tensor apparatus is absent, the presence of the apical recess portion of the sinus may be inferred by the presence of a cavity demarcated by a well-recognized posterior portion of a ventricular septum. The conus is that area of the ventricle which serves as an outflow tract. In this study, we qualify the definition of mitral and tricuspid valves by placing them in quotations, because in the strict sense an A-V valve is identified by its attachments, which in our study in some instances are markedly deranged. In addition, the identity of the receiving ventricle is used to help identify the A-V valves. The terms primitive right and primitive left ventricle are used because, in the former case, only part of the sinus of the right ventricle is in that chamber, and, in the latter case, there are portions of right ventricular sinus present in addition to the normal left ventricular sinus. Transposition of the arterial trunks is that condition in which the arterial trunks emerge from the wrong chambers, or are abnormally related to each other and to the atrioventricular orifices. The latter part of the definition is necessary in order to include transposition in a single ventricle, in which emergence from the wrong chambers does not pertain. Regular transposition refers to a disturbance in anteroposterior position of the bulbo-truncal area without any gross disturbances in laterality, while inverted transposition refers to a disturbance in laterality as well. A diagnosis of regular or inverted transposition must take into account the position of the chambers. Inversion of chambers refers to a disturbance of laterality of chambers, whereby the morphologic right occupies the position of the morphologic left and vice versa. Regular noninverted transposition usually (but not always) is associated with normal position of ventricles. Inverted transposition is usually associated with inverted ventricles.

**Characteristics of Types of Straddling and Displaced A-V Orifices**

The age, sex, and diagnoses of cases of straddling and displaced A-V orifices are summarized in table 1. We divided these hearts into three groups.

**Group 1: Straddling Right or Left A-V ("Tricuspid") Orifice (Figs. 1-3)**

The "tricuspid" orifice was small, normal, or large. The anterolateral "tricuspid" leaflet in its lateral aspect was connected to papillary muscles in the right ventricle (figs. 1L and 2L.), medially to the rim of the ventricular septal defect (VSD) and the muscle of Lancisi, and in some cases it was connected to the

\(^*\)This definition does not hold for double-outlet right ventricle, in which both coni may be related to the right ventricle, and no conus directly related to the left ventricle.
### Table 1

**Details of Hearts Examined**

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Sex</th>
<th>Associated abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Straddling tricuspid valve with normally related arteries</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>6 wk</td>
<td>F</td>
<td>PDA, partial common brachiocephalic trunk, PFO, thebesian attached to eustachian valve.</td>
</tr>
<tr>
<td>Straddling tricuspid valve with regular transposition</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>6 mo</td>
<td>F</td>
<td>Complete transposition with mitral stenosis. Fetal coarctation, dilatation of pulmonary trunk, right aortic arch, double ductus arteriosus, left obliterated, absent eustachian and thebesian valves, abnormal bands at mouth of SVC, PFO.</td>
</tr>
<tr>
<td>3</td>
<td>29 days</td>
<td>F</td>
<td>Complete transposition. PFO, PDA, common eustachian and thebesian valve.</td>
</tr>
<tr>
<td>4</td>
<td>20 days</td>
<td>M</td>
<td>Complete transposition with pulmonic stenosis, bicuspid pulmonic valve, abnormal fossa ovalis with ASD, combined eustachian and thebesian valves, accessory bands at mouth of SVC.</td>
</tr>
<tr>
<td>5</td>
<td>2 mo</td>
<td>F</td>
<td>Complete transposition. Absent coronary sinus.</td>
</tr>
<tr>
<td>6</td>
<td>6 wk</td>
<td>M</td>
<td>Double-outlet RV with mitral atresia. ASD, fossa ovalis type, PDA, eustachian valve enlarged. Thebesian valve absent.</td>
</tr>
<tr>
<td>7</td>
<td>3½ yr</td>
<td>F</td>
<td>Complete transposition. PFO, combined eustachian and thebesian valves.</td>
</tr>
<tr>
<td>8</td>
<td>4 mo</td>
<td>M</td>
<td>Complete transposition. Fetal coarctation, left superior vena cava entering the coronary sinus, PFO, right infundibular stenosis.</td>
</tr>
<tr>
<td>Straddling tricuspid valve with inverted transposition</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>7 mo</td>
<td>F</td>
<td>Mixed levocardia with ventricular inversion (corrected transposition). Combined eustachian and thebesian valves.</td>
</tr>
<tr>
<td>Displaced tricuspid orifice and valve</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>2½ mo</td>
<td>F</td>
<td>Atypical complete transposition with straddling aorta. Fetal coarctation, PDA, PFO, subaortic stenosis, absent thebesian valve.</td>
</tr>
<tr>
<td>11</td>
<td>12 yr</td>
<td>F</td>
<td>Pulmonary infundibular and subaortic stenosis. PFO, aneurysmal dilatation of pulmonary trunk (Potts anastomosis), absent coronary sinus.</td>
</tr>
<tr>
<td>12</td>
<td>6 days</td>
<td>M</td>
<td>Atypical complete transposition with straddling aorta and pulmonary stenosis. PFO, PDA, cleft aortic leaflet of the mitral valve, absent eustachian and thebesian valves.</td>
</tr>
<tr>
<td>Straddling mitral with displaced tricuspid valves</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>3 wk</td>
<td>F</td>
<td>Mitral and pulmonic stenosis, PDA, prominent eustachian valve. Thebesian valve anchored on eustachian valve.</td>
</tr>
<tr>
<td>14</td>
<td>22 yr</td>
<td>M</td>
<td>Complete transposition with pulmonic stenosis. ASD, fossa ovalis type, absent thebesian valve.</td>
</tr>
</tbody>
</table>

**Group 1**

**Group 2**

**Group 3**

Abbreviations: ASD = atrial septal defect; PDA = patent ductus arteriosus; PFO = patent foramen ovale; RV = right ventricle; SVC = superior vena cava.

Accessory inferior papillary muscle of the left ventricle. In case 9, with inverted transposition, its medial portion traversed the VSD to attach to a posterior papillary muscle in the morphologic left ventricle (fig. 3R). The medial leaflet was usually connected to the muscle of Lancisi, to the rim of the VSD, and to the accessory inferior papillary muscle in...
the left ventricle (figs. 1R and 2R). The inferior leaflet was attached in most cases to the accessory papillary muscle in the left ventricle. In some cases, it was attached to the rim of the VSD, and in some to the posterior wall of the right ventricle and to the anterolateral papillary muscle in the right ventricle. In cases with no inversion of chambers, the primitive right ventricle lay anterior, superior, and to the right of the primitive left ventricle. The former was small and thick-walled, and consisted mostly of conus, though it had a small sinus. The left ventricle was enlarged, usually hypertrophied and had, in addition to the normal posterior papillary muscle group, an accessory group of posterior papillary muscles which held the aberrant portions of the straddling "tricuspid" valve. A median ridge either independent of or fused with the accessory posterior papillary muscle of the left ventricle often extended from the central fibrous body to the apex, on the posterior wall (fig. 2R). The conal portion of this chamber was abnormal. Muscle ridges extended from the efferent vessel downward on the septum (fig. 1R), and in some instances muscle lay beneath the efferent vessel separating it from the mitral valve. The communication between the ventricles was of the posterior type. In all but two cases, the right atrium lay lateral, slightly to the right, and inferior to the left atrium. Both atria showed hypertrophy and
Case 7. Straddling tricuspid orifice with regular transposition. (Left) Right atrial and right ventricular view. (Right) Left ventricular view. Abbreviations: PR = posterior ridge; RA = right atrium; RV = right ventricle; others = same as in figure 1.

Figure 2

Case 9. Mixed levocardia with ventricular inversion with straddling left A-V ("tricuspid") valve. (Left) Morphologic right ventricular (left-sided) view. (Right) Morphologic left ventricular (right-sided) view. Abbreviations: AP = accessory posterior papillary muscle to which tricuspid valve is attached; others = same as in figure 1.

Figure 3
enlargement, and endocardial hypertrophy. One heart had normally related great vessels (figs. 1L and R). Six had regular complete transposition (figs. 2L and R), one was a case of double-outlet right ventricle, and one a case of mixed levocardia with ventricular inversion and inverted transposition (corrected transposition) (figs. 3L and R).

Among those with regular complete transposition, there were two cases in which the aorta was slightly to the left of the pulmonary trunk. The coronary distribution with regular or inverted transposition was typical of that found in those anomalies. In addition, most cases with or without regular transposition showed a tendency for the right-sided coronary artery to dominate the circulation, while the left-sided one was limited in most cases to the anterior descending branch.

In this group there were two patients with mitral stenosis, one with mitral atresia and one with enlargement of the mitral orifice. Fetal (preductal) coarctation occurred in two cases. Patent foramen ovale or atrial septal defect of the fossa ovalis (secundum) type occurred in seven cases. Patent ductus arteriosus occurred in four cases, pulmonary stenosis in one, bicuspid pulmonic valve in two, bicuspid aortic valve in one, and subaortic stenosis in one.

**Group 2: Displaced "Tricuspid" Orifice (Fig. 4)**

The "tricuspid" orifice was completely displaced into the primitive left ventricle. The orifice was minute (fig. 4), normal, or large. In one case it was not at all related to the VSD, and in two it was related to the right side of its rim. The "tricuspid" valve was in one case connected to anterior and posterior papillary muscles in the primitive left ventricle as in single ventricle. In another, the leaflet structures were not distinct; parts consisted of fragments of tissue; parts were very short and attached by chordae to the septum; and parts were attached to a thin posterior papillary muscle in the primitive left ventricle. In the third case there were two attenuated leaflets, one leaflet attached by chordae to the summit of the septum on the left side, and one leaflet attached to a small posterior papillary muscle in the left ventricle and to the summit of the ventricular septum. The mitral orifice varied in size and its valve was attached to anterior and posterior papillary muscles in the primitive left ventricle. The primitive right ventricle was again a small or flattened chamber with a normal or hypertrophied wall. It consisted partly of conus and partly of sinus, as defined by a well-recognized posterior portion of the ventricular septum. The sinus, however, had no tensor apparatus (fig. 4). It was rather elongated, extending to the apex (apical recess). The primitive right ventricle lay anterior and to the right of the primitive left ventricle. The primitive left ventricle was hypertrophied and enlarged. Two cases had atypical regular complete transposition with straddling coni (fig. 4). In one of these the aorta was anterior to and slightly to the left of the pulmonary trunk. The communication between the ventricles was in the anterior part of the septum (fig. 4). Where there was transposition, the coronary artery distribution was typical of that anomaly. In the case...
Case 13. Displaced tricuspid orifice with straddling mitral orifice. Mitral and pulmonic stenosis without transposition. (Left) Biatrial and left ventricular view. (Right) Right ventricular view. (Lower) Right atrial and left ventricular view. Abbreviations: CP = combined posterior papillary muscle and ridge; LA = left atrium; MV = mitral valve passing through VSD; PT = pulmonary trunk; TV = tricuspid valve; others = same as in preceding figures.
without transposition there was a dominant right circulation.

**Group 3: Straddling Left A-V (Mitral Orifice) with Displaced "Tricuspid" Orifice (Fig. 5)**

In one case the "mitral" orifice was stenotic, with the valve ring displaced downward to the roof of the VSD at the anterior comissure. The anterior leaflet of the left A-V ("mitral") valve was attached to the anterolateral papillary muscle in the right ventricle, to the wall of the sinus of the right ventricle, and to the posterior papillary muscle in the right ventricle (figs. 5L and R). The posterior leaflet was connected to a posterior papillary muscle in the left ventricle, to the rim of the VSD, and to the wall of the sinus of the right ventricle (figs. 5L and R). In the other case, the mitral orifice was normal in size, and the "mitral" valve consisted of three leaflets. The medial leaflet was attached to a small papillary muscle on the right side of the rim of the VSD, to a papillary muscle in the accessory sinus of the right ventricle, and to the posterior papillary muscle in the left ventricle. The lateral leaflet was connected to the posterior papillary muscle in the left ventricle and to the rim of the VSD. The anterior leaflet was attached to the papillary muscle (mentioned above) in the accessory sinus of the right ventricle. The "tricuspid" orifice was normal in size in the first case (fig. 5, Lower), and increased in size in the second case. In both cases, the orifices entered the left ventricle. Their valves consisted of two leaflets which attached to anterior and posterior papillary muscles, as in single ventricle (fig. 5, Lower). The primitive right ventricle was again a small, hypertrophied chamber that lay superior, anterior, and to the right of the left ventricle, consisted mostly of conus, but also had a small sinus (fig. 5R). The left ventricle was hypertrophied and enlarged and had no anterior papillary muscle for the mitral valve (fig. 5L). The right and left atria lay side by side. The right atrium was hypertrophied, enlarged, and had a thickened endocardium. The left atrium was normal in size in one case, enlarged in the other, and hypertrophied in both. The communication between the ventricles was in the anterior position (fig. 5L). The arterial trunks were normally related in the first case, and showed regular (noninverted) transposition in the second. The coronary artery distribution was normal in the first, and typical of complete transposition in the second. Of our two cases, one had pulmonary stenosis, one had a patent ductus arteriosus, and both had bicuspid pulmonic valves, and atrial septal defects of the fossa ovalis type.

**Clinical Data**

The clinical picture of the three malformations was similar, and will be dealt with together. It was characterized generally by cyanosis, respiratory difficulties, poor growth, and heart failure very early in infancy. The auscultatory findings were nonspecific, consisting chiefly of a loud single or closely split second sound, and an ejection or pansystolic murmur along the left sternal border. Chest roentgenograms revealed cardiomegaly and increased pulmonary vascular markings in cases without pulmonary stenosis. In some cases, a bulge at the left upper cardiac border representing the small right ventricle was noted in the frontal projection.

Used in 11 cases, the electrocardiogram revealed right atrial enlargement in eight, and left ventricular hypertrophy in eight, usually manifested by prominent postero-leftward QRS forces. RS complexes in the entire precordial tracing such as one often observes in ventricular inversion were noted in five cases. Right ventricular hypertrophy was observed in three cases. The frontal QRS axis ranged from +60 to +100° in eight cases, and from +10 to −100° in three cases.

The cardiac catheterization findings were generally inconclusive. In nine cases in which this was performed, systemic arterial oxygen unsaturation of various degrees of severity was observed, and was marked in those cases with pulmonary stenosis. Evidence for left-to-right shunting at the ventricular level was invariably present. In the cases with regular transposition of the great vessels, the oxygen content of the right ventricle was sometimes higher than that of the aorta, presumably due
Figure 6

Case 5. Cineangiocardiograms. Straddling tricuspid valve and regular transposition of the great vessels. Frontal projection (Upper Left) Right atrial injection, demonstrating gross enlargement of right atrium. (Upper Right) With atrial contraction, simultaneous opacification of both ventricles is observed through separate atrial flow streams. Note position of ventricular septum and of right ventricle, and hemoilution of contrast substance in left ventricle. (Lower Left) During ventricular systole, there is striking reduction in size of right ventricle. (Lower Right) Left ventricular injection, ventricular diastole, demonstrating relative size and position of ventricular chambers and great vessels. Abbreviations: AO = aorta; IVS = interventricular septum; PA = pulmonary trunk; others = same as in preceding figures.
to streamlining of the right atrial blood into the latter. The pulmonary arterial samples also revealed higher oxygen saturation than that of the aorta.

Angiocardiography obtained with injection of the contrast material into either ventricle revealed almost simultaneous opacification of both ventricles and great vessels. In some cases, a small ventricular chamber thought to represent an accessory outlet chamber in single ventricle was observed. In one case with ventricular inversion, VSD, and inverted transposition (case 9), both ventricles appeared large. In some cases, A-V valve incompetence was not demonstrated, whereas in others this was clearly notable. In some cases of regular (noninverted) transposition of the great vessels, the ascending aorta lay to the left of the pulmonary trunk (figs. 6 and 7).

Right atrial injection in a case of straddling tricuspid valve and noninverted transposition of the great vessels (case 5) was diagnostic. The right atrium was large (fig. 6, Upper L). In addition, two streams of contrast material could be clearly delineated across the tricuspid orifice—one leading into a small, antero-superiorly situated right ventricle from which the aorta originated, and the other into a large inferiorly located left ventricle which gave rise to the pulmonary trunk (fig. 6, Upper R). This distinct separation of flow streams was obviously related to the abnormal attachments of the "tricuspid" valve into both ventricles. The small right ventricle which appeared larger than the usual rudimentary outlet chambers of single ventricle, changed significantly in size from systole to diastole (figs. 6, Upper R and Lower L). In addition, the position of the interventricular septum was most unusual, running to the left almost horizontally (fig. 6, Lower R). In another case with straddling tricuspid valve and noninverted transposition of the great vessels (case 7), left ventricular injection demonstrated striking tricuspid regurgitation (fig. 7L). Repeat injection into this chamber in the right posterior oblique projection outlined the unopacified mitral and tricuspid orifices, the latter apparently overriding a large left ventricle and a small right ventricle (fig. 7R).

In one case of displaced tricuspid orifice and straddling mitral valve (case 13), injection of contrast material into the inferior vena cava demonstrated selective filling of the left ventricle.

**Figure 7**

Case 7. Cineangiograms. Straddling tricuspid valve and complete transposition of great vessels. (Left) Left ventricular injection, frontal projection, with catheter advanced from right atrium across tricuspid valve into left ventricle. Note tricuspid regurgitation and unusual position of great vessels. (Right) Left ventricular injection, 70° right posterior oblique projection, with catheter advanced through right atrium, left atrium, and mitral valve. Note unopacified mitral and tricuspid orifices of approximately similar sizes, with the tricuspid valve apparently overriding a small right ventricle and large left ventricle. Abbreviations: MV = mitral valve orifice; TV = tricuspid valve orifice; others = see preceding figures.
Case 13. Cineangiograms. Displaced tricuspid valve, straddling mitral valve, and normal position of great vessels. (Left) Inferior vena caval injection, frontal projection, demonstrating immediate filling of left ventricle from right atrium. (Right) During ventricular systole, simultaneous opacification of both vessels in normal right-left positional arrangement is observed. (Lower Right) Left ventricular injection, 70° right posterior oblique projection, with catheter advanced from right atrium into left ventricle. Note small right ventricle and pulmonary artery, and normal position of great vessels. Abbreviations as in figure 6.

A small right ventricle and pulmonary trunk were opacified shortly after by way of a ventricular septal defect, thus simulating somewhat the findings in tricuspid atresia with ventricular septal defect. This was also demonstrated by left ventricular injection in the frontal and right posterior oblique projections (figs. 8R and Lower R). There was, however, no demonstrable separation of the left atrial flow stream entering the left ventricle. This could have been due to hemodilution of contrast.
Successive stages in the development of the bulbus and atrial canal during the second phase of the development of the heart (after Asami). The view is from the posterior aspect with the atria removed. Upper embryo—horizon XVI. Lower left—horizon XVII. Lower middle—horizon XVIII. Lower right—horizon XIX. Abbreviations: P = pulmonary artery; Ao = aorta; O = anterior endocardial cushion; U = posterior endocardial cushion; B = bulbus; Pr = proampulla; Mt = metaampulla; MV = mitral valve; TV = tricuspid valve; Ba-Sp. = bulboauricular spur. Arrows point to the embryos representing the time of malformation in straddling tricuspid orifice.

Discussion

The anomalies that form the basis of this report are to be differentiated from single ventricle, as pointed out by Mehrizi et al. In single ventricle, both A-V orifices enter a common ventricular chamber which gives rise to a conus-like chamber. The latter cannot be called a ventricle since it has no sinus. In the hearts discussed in this report, although two A-V orifices enter at least in part into a common chamber, there is a second chamber which can be called a primitive right ventricle since it has a distinct conus and part of a sinus of the right ventricle. This is obviously true of straddling tricuspid orifice. It is also true of displaced tricuspid orifice, where, although there is no tensor apparatus in the primitive right ventricle, there is an apical recess associated with the distinct conus of the right ventricle. It is likewise true of displaced tricuspid with straddling mitral orifice, where the primitive right ventricle possesses the distinct conus of a right ventricle, and, in addition, a sinus with tensor apparatus, connected surprisingly enough, to the mitral valve. It is obvious that all of these malforma-
ATRIOVENTRICULAR ORIFICES AND VALVES

Our cases of straddling tricuspid orifice have a posterior type or common A-V canal type of communication between the ventricles, thereby resembling those of Rastelli et al.,2 although the connections of the tricuspid valve in our cases are not exactly identical with theirs. The cases of displaced tricuspid orifice and those with straddling mitral orifice are quite different from those of Rastelli et al., for the defects in the former are more anterior and are not of the common A-V canal type.

The position of the aorta slightly to the left in some cases of regular transposition in this series has been previously noted.1 10 11

From the embryologic standpoint, the anomalies under discussion are similar to single ventricle. Our understanding of them is related to a knowledge of the second phase of the development of the heart. This has been described by Pernkopf and Wirtinger12 and confirmed by Asami13 (fig. 9). In this phase, the atrial canal moves to the right and the bulbus to the left. In these movements portions of the descending and ascending limbs of the ventriculo-bulbar loop are distributed to both sides of the heart. The descending limb consists of the proampulla, and the ascending limb of metaampulla and bulbus. The definitive left ventricle, therefore, consists mostly of proampulla (sinus) and, in addition, a small part of metaampulla and bulbus (left ventricular outflow), which are largely absorbed into the proampullar mass. The definitive right ventricle consists in part of proampulla (sinus), in part of metaampulla (apical recess and lower outflow tract), and in part of a well-developed bulbus (upper outflow tract).

The anomalies under discussion, as well as single ventricle, may be conceived of as caused by a lack of passage towards the right of the atrial canal during the time the bulbus is being absorbed into the ventricles (fig. 9). Thus, although portions of the bulbus are distributed to each side of the heart, the atrial canal remains partly or completely on the left in the descending limb of the ventriculo-bulbar loop. Asami13 has pointed out that there is an intermediate stage in the passage of the atrial canal to the right, in which the developing tricuspid orifice straddles (fig. 9—arrows). If the passage stops at this stage, then the anomaly straddling tricuspid orifice occurs. If the passage does not occur at all, then single ventricle, or displaced tricuspid orifice occurs. In single ventricle, the metaampulla fails to develop at all, while in straddling and displaced tricuspid orifice it remains in part.

Straddling mitral orifice with displaced tricuspid orifice is more difficult to explain, for no such stage exists in embryologic development. One might postulate that in rare cases, where the atrial canal fails to move towards the right, its left side may become tilted upward and to the right, so that the developing mitral orifice obtains connections with the metaampulla, while the anterior papillary muscle of the left ventricle fails to form.

The clinical and hemodynamic findings in the three malformations under discussion closely resemble those observed in common ventricle or in single ventricle with rudimentary outlet chamber. Bidirectional shunting at the ventricular level is present, and, in the absence of pulmonary stenosis, pulmonary hypertension at systemic levels occurs. Systemic arterial oxygen saturation may be significantly lower than that of the ventricles or pulmonary artery.

Of some interest in the present series was the greater incidence of straddling or displaced right A-V valve in females (nine cases) than in males (five cases).

The electrocardiographic findings of right atrial hypertrophy and left ventricular hypertrophy in a cyanotic infant may strongly suggest tricuspid atresia. However, the left axis deviation usually observed in the latter condition is not common in the anomalies under discussion. In addition, right ventricular hypertrophy may sometimes be observed.

It appears that straddling A-V valves can be
diagnosed during life by selective angiography. A high index of suspicion, however, is necessary to alert one to this possibility and thus perform selective angiography at proper injection sites. It has been suggested that the angiographic demonstration of an anterior chamber that is bigger than the usual rudimentary outlet chamber in single ventricle, shows a coarser surface, and is situated to the right of the left ventricle, allows differentiation of the small right ventricle in straddling tricuspid valve from the rudimentary outlet chamber in single ventricle.4

We believe that a definitive diagnosis can be established in straddling tricuspid orifice, by right atrial angiography which demonstrates two separate and distinct streams coming from the right atrium—one into the bigger and posteriorly situated left ventricle and the other into the smaller and anterosuperior right ventricle. The levoangiogram shows emptying of the left atrium only into the large posterior left ventricle. In single ventricle with a rudimentary outlet chamber, filling of the small chamber is solely from the single ventricle and never from an atrium, thus further attesting to the absence of a functional sinus in the rudimentary chamber.

The angiocardiographic findings in displaced tricuspid orifice resemble very closely those of tricuspid atresia. However, right atrial or inferior vena caval injection demonstrates immediate filling of the left ventricle directly from the right atrium in the former, whereas in tricuspid atresia left ventricular opacification occurs only following that of the left atrium by way of a right-to-left atrial shunt.

In the case of straddling left A-V ("mitral") valve, the diagnosis may likewise be angiographically demonstrable in the form of two separate left atrial streams filling each of the two ventricles. Although this was not noted in one of our cases submitted to this procedure, we believe that this negative finding could have been due to hemodilution of the contrast substance at the time of left atrial opacification. Direct left atrial injection could have been diagnostic.

References
Straddling and Displaced Atrioventricular Orifices and Valves with Primitive Ventricles

RICHARD R. LIBERTHSON, MILTON H. PAUL, ALEXANDER J. MUSTER, RENÉ A. ARCILLA, FRIEDRICH A. O. ECKNER and MAURICE LEV

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