Single Ventricle with Transposition

By José Marín-García, M.D., Rajendra Tandon, M.D., James H. Moller, M.D., and Jesse E. Edwards, M.D.

SUMMARY

Fifty-seven cases of single (common) ventricle with transposition of the great vessels are reviewed. The diagnosis had been confirmed by necropsy in 25 and by angiocardiography in 32 cases.

Single (common) ventricle is defined as that condition in which both atrioventricular valves separately enter a single ventricular cavity. According to this definition, cases presenting with common atrioventricular valve, atresia of one atrioventricular valve, or straddling valves have been excluded. Two structural types of common ventricle are identified: the left ventricular type and the primitive type of single ventricle. The former showed anatomic features similar to a left ventricle, whereas in the latter, features characteristic of a left ventricle were not present. The latter was considered a more primitive condition than the former.

A classification based upon the type of single ventricle and its outflow tract and upon the relationship of the great vessels is presented. Of the 25 cases of single ventricle with transposition studied pathologically, 21 were of the left ventricular type and four of the primitive type. Of the four cases with double conus three were among the four examples of primitive ventricle. Among the 21 cases with a single conus, the type of transposition was about equally divided between the d-type (ten cases) and the l-type (11 cases).

Pulmonary stenosis or atresia was observed in seven of the 21 cases with a single conus and in one of the four cases with double conus.

In the clinical cases, only angiocardiography could establish the diagnosis and delineate the different types of great vessel-ventricular relationships.

Additional Indexing Words:

Cor triloculare bia-triatum
Common ventricle

Transposition

Primitive left ventricle

Single or common ventricle is a rare form of congenital cardiac disease which has absorbed attention of many investigators. With the recently developed potential for correction of this condition, a better knowledge of the anatomic details is needed and the clinical features need to be defined so that it can be distinguished from other cardiac anomalies presenting with cyanosis. In reported series of congenital cardiac disease, the incidence of common ventricle has been estimated as being about 3% of all congenital cardiac anomalies.1,2 Although the great vessels may be normally related, most cases of single ventricle exhibit transposition of these structures.

In this paper, we will present the findings in 57 cases of single ventricle with transposition of the great vessels in which both atrioventricular valves were present and patent. The clinical profile of this condition will be presented together with descriptions of the typical angiocardiographic features. The pathologic features will be presented in 25 of the cases and a new classification will be proposed.

Cases of common atrioventricular valve, tricuspid atresia, mitral atresia, double inlet left ventricle, or those with associated splenic anomalies were not included. In no cases was there abdominal heterotaxia. Cases of single ventricle and normally related great vessels observed in our institutions have been reported separately.3

For the purpose of the present work, the terms single and common ventricle have been used synonymously.

We define single ventricle as that condition in which both atrioventricular valves separately enter a single ventricular cavity. There is no evidence of a sinus portion of a second ventricle. Each great vessel arises either directly from the single ventricular chamber or from a conus. According to our definition, the conditions of mitral or tricuspid atresia, double inlet left ventricle and common atrioventricular valve are excluded.

Utilizing this definition of single ventricle, two
morphologic varieties are recognized. In one, the left ventricular chamber has the characteristics of a left ventricle with a smooth wall and finely trabeculated sinus portion. We have defined this as single ventricle of the left ventricular type. The morphologic features of the other variety of single ventricle do not fit specifically the features of a left ventricle. The features suggestive of both ventricles, right ventricle, or neither may be recognizable. Such a chamber has been termed a single ventricle of the primitive type.

Transposition of the great vessels is defined as an external relationship of the great vessels wherein the aorta is abnormally related to the pulmonary trunk. Under this broad designation, the external relationship of the vessels is variable. The aorta may be anterior and to the right of the pulmonary trunk displaying a relationship as in complete transposition of the great vessels (noninverted or d-transposition). The aorta may be anterior and to the left of the pulmonary trunk as seen in corrected (inverted) transposition of the great vessels (l-transposition). Lastly, the aorta may lie to the right of the pulmonary trunk. In the latter situation, both great vessels lie in the same frontal plane or the aorta may be slightly anterior or posterior to the pulmonary trunk. The latter relationship is associated with subaortic and subpulmonary coni (double conus), and the two semilunar valves lie in the same horizontal plane. In contrast, in d-transposition and l-transposition relationship, there is only a subaortic conus while the pulmonary valve shows continuity with the mitral valve.

The conus is defined as a muscular cardiac segment intervening between a semilunar valve and the atriocentral valves.

Materials and Methods
Fifty-seven patients with single (common) ventricle and transposition of the great vessels were reviewed. The diagnosis was confirmed at necropsy in 25 cases (necropsy group) and by angiography in the remaining 32 cases, the latter designated as the "clinical group." In some of the cases in the necropsy group, angiocardiography had also been done.

Clinical data and the radiologic findings were available in all except eight cases of the necropsy group; the electrocardiograms were available in 43 cases.

Schmitt's corrected orthogonal lead vectorcardiogram was available in four cases of the necropsy group and in 16 cases of the clinical group. Selective biplane angiocardiography was performed in 39 cases and includes all of the clinical cases. Cardiac catheterization data were available in 39 cases. Eleven cases of the necropsy group have been included in an earlier report.

Observations
The ages of the patients ranged from one day to 40 years. Among the 32 patients of the clinical group, there were three under one year of age, three between one and five years, 18 between five and 15 years, and eight older than 15 years.

Among the 25 patients of the necropsy group, the age was known in 23 cases. Of these, 14 cases were below one year, three between one and five years, one six years old, another 11 years old, and four over 15 years. The mean age for the necropsy group was five years and six months, and for the clinical group, 12 years and two months. Of the 57 patients, 21 were female and 35 were male. The sex of one case in the necropsy group was not known.

Angiographic Basis for Diagnosis in the Clinical Group
In the 32 patients of the clinical group, the diagnosis was made angiographically. Transposition of the great vessels was demonstrated in each.

A single ventricle morphologically resembling a left ventricle was found in 26 cases and a morphologically primitive ventricle was found in the other six cases. An outflow chamber (located anteriorly) was identified in 21 of the 32 cases. The aorta arose from the outlet chamber, while the pulmonary trunk arose from the main chamber in all cases. The transposition was of the d-type in 11 cases and of the l-type in 17 cases, while in four cases there was evidence suggesting the presence of a double conus. In the last group, a subpulmonary conus, as well as a subaortic conus resulting in discontinuity between the semilunar valves and the atriocentral valves, was identified. Patency of the tricuspid valve and the mitral valve was delineated by the appearance of the valve annuli in ventriculograms, the course of a catheter and/or forward flow of contrast material across the valve.

Pathologic Findings
The 25 specimens of hearts with single ventricle were divided into two types according to the morphology of the ventricular chambers as defined above. There were 21 with the left ventricular type of single ventricle and four with a primitive ventricle. Each variety of single ventricle was further subdivided according to the external relationship of the great vessels and the presence or absence of obstruction to pulmonary blood flow. Table 1 provides a classification of single ventricle and the number of cases of the various subdivisions encountered. The necropsy group of 25 cases herein described, each with transposed great vessels, showed variation in the relationship of the great vessels. In the majority, a subaortic conus was present and pulmonary-mitral valvular continuity was present. In ten the transposition was of the d-type, and in 11 it was of the l-type. In four instances a double conus was present so that
neither semilunar valve showed continuity with the atroventricular valves.

Among the 25 cases obstruction to pulmonary flow was present in eight cases (pulmonary atresia, two cases; subpulmonary with or without pulmonary valvular stenosis, six cases), while in the remaining 17 no obstruction to pulmonary flow was present. The association of pulmonary stenosis appeared to be equally distributed among the cases with d- and l-transposition.

*Single Ventricle of Left Ventricle Type*

Twenty-one specimens showed the single ventricle to be of the left ventricular type as defined. The interior of the common ventricle showed two features. On one side were the insertions of the papillary muscles of both atroventricular valves, while the other side was smooth. The smooth side, which lay either to the right or left of the insertions of the papillary muscles, was positioned below the bulboventricular foramen (the opening between the base of the main ventricle and the conus). In the 20 cases with a single conus, the side of the smooth wall showed a particular relationship to the type of transposition. In the nine cases with d-transposition the smooth wall was on the right side of the common ventricle, while in the 11 cases with l-transposition it lay on the left side. The bulboventricular foramen led to a small infundibular chamber from which the aorta arose. Both atroventricular valves opened into the common ventricle and were continuous with each other and with the pulmonary valve. In the twenty-first case of common ventricle of the left ventricular type, a double conus was present. The smooth wall was to the right in this case.

*d-Transposition.* Among the nine cases with d-transposition, obstruction to pulmonary flow was present in three cases.

In one specimen the pulmonary valve was atretic and the matter of pulmonary-mitral valve continuity was impossible to determine because of the pulmonary atresia. Pulmonary blood flow was derived from a patent ductus arteriosus.

Two specimens showed membranous subpulmonary stenosis. In both, the pulmonary valve was bicuspid and in one of these it was stenotic (fig. 1).

Among the remaining six, no obstruction to pulmonary blood flow was present. Nevertheless, anomalies were present in each which obstructed flow to the systemic circulation. Subaortic stenosis resulting from a narrowed bulboventricular foramen was present in all cases. Tubular hypoplasia of the aortic arch and/or coarctation of the aorta was present in four and hypoplasia of the ascending aorta and arch was present in an additional case. The aortic valve was

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**Table 1**

*Classification of Single Ventricle According to the Type of Ventricular Chamber and the State of the Great Vessels*

<table>
<thead>
<tr>
<th>Ventricular type</th>
<th>Left (28)</th>
<th>Primitive (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A)</strong> Normally related great vessels (7)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>i) with pulmonary atresia</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>ii) with pulmonary or subpulmonary stenosis</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>iii) with no pulmonary or subpulmonary stenosis</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td><strong>B)</strong> Transposition of the Great Vessels (25)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1) Single (subaortic) conus (21)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a) noninverted (d-transposition) (10)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>i) with pulmonary atresia</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>ii) with pulmonary or subpulmonary stenosis</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>iii) with no pulmonary or subpulmonary stenosis</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>b) inverted (l-transposition) (11)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>i) with pulmonary atresia</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>ii) with pulmonary or subpulmonary stenosis</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>iii) with no pulmonary or subpulmonary stenosis</td>
<td>8</td>
<td>0</td>
</tr>
<tr>
<td>2) Double Conus (4)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>i) with pulmonary atresia</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>ii) with pulmonary or subpulmonary stenosis</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>iii) with no pulmonary or subpulmonary stenosis</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td><strong>C)</strong> Persistent Truncus Arteriosus (9)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The numbers in parenthesis indicate the number of cases observed among 32 cases studied pathologically. The seven cases with normally related great vessels were described previously.3

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normal in five and bicuspid in one of the cases. The latter was associated with coarctation of the aorta. In one of the cases the mitral valve showed a parachute deformity.

l-Transposition. In the 11 specimens with l-transposition, the aortic valve was located anteriorly and to the left of the pulmonary valve. The ascending aorta swept to the left of the pulmonary trunk (fig. 2).

The pulmonary valve was located at a level lower than the aortic valve and was continuous with both atrioventricular valves. In one the continuity could not be clearly established because of pulmonary atresia.

Obstruction to pulmonary blood flow was present in three instances. In one pulmonary atresia was present, in another the valve was unicommissural and stenotic, and in the third muscular subpulmonary stenosis and a dysplastic pulmonary valve were present.

In the other eight specimens, there was no obstruction to pulmonary blood flow, but in five of these, obstructive anomalies were present in the route of systemic flow. Coarctation of the aorta was present in three cases, interruption of aortic arch in another, and aortic valvular atresia in the fifth case. Three of these five showed subaortic narrowing resulting from a small bulboventricular foramen.

Additional anomalies were present in several patients. The left atrioventricular valve showed a narrowed annulus and a parachute mitral valve in one patient. In another case an atrial septal defect was associated with a hypoplastic right atrioventricular valve and dextrocardia. A bicuspid aortic valve was found in a third case and there was partial anomalous pulmonary venous connection to the superior vena cava in the absence of an atrial septal defect in a fourth case.

Double Conus. In a single case of common ventricle of the left ventricular type, each great vessel arose above a conus so that the semilunar valves were separated from the atrioventricular valves. The outflow area was divided by a muscular ridge into a narrow subaortic conus and a wide subpulmonary conus. The aortic and pulmonary valves lay at the same horizontal plane. The great vessels were parallel to each other and the aorta lay to the right of the pulmonary trunk (fig. 3). The body of the common ventricle had the features of a left ventricle, with the smooth wall being anterior and to the right. Both atrioventricular valves emptied into the chamber, although the mitral valve showed a parachute deformity and was stenotic (fig. 3b).

Single Primitive Ventricle

In four specimens, the single ventricle was of the primitive type.

In one of these four, a single conus with d-transposition was present and in the other three there were double coni.

In the specimen with d-transposition, pulmonary-mitral valvular continuity was present. Associated anomalies included a bicuspid pulmonary valve, persistent left superior vena cava, atrial septal defect, and patent ductus arteriosus.

Figure 2
Left ventricular type of single ventricle with l-transposition. a) External view of the heart and subaortic chamber (between arrows). The aorta (A.) arises anterior and to the left of the pulmonary trunk (P.T.). b) Interior of the single ventricle. The smooth wall lies anterior and to the left (arrow). The mitral valve (probe, M.V.) and the tricuspid valve (T.V.) enter the common ventricle. P.V. = pulmonary valve.

Figure 1
Left ventricular type of single ventricle with d-transposition. a) External view of the heart showing d-transposition. Anterior aorta (A.) obscures a view of the pulmonary trunk. b) Lateral view of the interior of single ventricle of the left ventricular type. Aorta (A.) arising anteriorly from the single conus. Membranous sub-pulmonary stenosis (between arrows) is also present.
In the three cases with subaortic and subpulmonary coni, neither of the semilunar valves was continuous with an atrioventricular valve (fig. 4). A muscular ridge divided the outflow area into subpulmonary and subaortic regions. In one case the subpulmonary area was narrow, and in the other two cases the subaortic area was narrow.

The aortic valve and the proximal aorta lay to the right of the pulmonary valve and pulmonary trunk in each. In one the aorta lay slightly more anterior to the pulmonary trunk, whereas with the other two the aorta lay somewhat more posteriorly.

Coexistent anomalies were present in each case. A parachute mitral valve and a nonobstructive supravalvular ring of the left atrium were found in two and tubular hypoplasia of the aorta with a patent ductus arteriosus was present in the third.

Clinical Features

Clinical data were available for review in 49 patients (17 from the necropsy group; 32 from the clinical group). Each patient had been cyanotic to a variable degree. Cyanosis was present at birth in 30 cases, became apparent by the age of one year in 13 patients, and developed after one year of age in the remaining six. No correlation between the severity of cyanosis and the presence or absence of pulmonary stenosis was found. Thirty-eight of the 49 patients were dyspneic. Six patients gave a history of hypoxic episodes. Five of these had pulmonary stenosis. Squatting was described in two patients, in one of whom pulmonary stenosis was present.

Congestive cardiac failure was present in 18 patients, 16 of whom did not exhibit obstruction to pulmonary blood flow. The other two had pulmonary stenosis but in one complete atrioventricular block was also present.

The first heart sound was normal in each patient. The second sound was described in 44 patients: it was single in 28 and split in 16. In seven of these the pulmonary component of the second sound was described as accentuated. None of the latter group exhibited pulmonary stenosis.

An ejection systolic murmur varying from grade II to V/VI in intensity was present in 46 cases. The maximum intensity of the murmur was along the left sternal border and was associated with a systolic thrill in 16 patients.

A mid-diastolic rumbling murmur was audible in four patients, none of whom had obstruction to pulmonary blood flow. An early diastolic murmur along the left sternal border was found in four patients. In only one was there obstruction to pulmonary blood flow.

Electrocardiographic Findings

Electrocardiograms were available in 43 cases (fig. 5). Eighteen were associated with d-transposition, 20

Figure 3

Left ventricular type of single ventricle with double conus. a) The aorta (A) lies to the right of the pulmonary trunk (P.T.) and arises from a small conus. b) Interior of single ventricle of left ventricular type showing the subpulmonary conus from which the pulmonary trunk (P.T.) arises. The probe lies through the mitral valve.

Figure 4

Primitive type of single ventricle with double conus. a) External view of the heart and great vessels. The small aorta (A) lies to the right and somewhat posterior to the large pulmonary trunk (P.T.). b) Interior of the single ventricle which receives the mitral (M.) and tricuspid (T.) valves. The ventricular internal architecture does not conform to that of a left ventricle and is considered of the primitive type. c) Interior of the single ventricle and the subaortic conus. A = aorta. d) Interior of the single ventricle and the subpulmonary conus, the latter giving rise to the pulmonary trunk (P.T.).

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SINGLE VENTRICLE WITH TRANSPOSITION

The mean QRS axis in the frontal plane of the 43 cases of single ventricle with available electrocardiograms. In the majority of the cases with d- or l-transposition, regardless of the presence or absence of pulmonary stenosis, the QRS axis in the frontal plane lies between +45° and +120°. Four of the five electrocardiograms of cases with double conus showed an axis close to −90°.

with l-transposition and five with double conus. No specific electrocardiographic differences could be identified among those with the left ventricular type of single ventricle as compared to those with the primitive type. The mean QRS axis in the frontal plane of the 43 cases is indicated in figure 5 and the findings summarized in table 2.

Q waves were present in the precordial leads in 27 of 43 electrocardiograms. Ten of the 18 electrocardiograms from patients with d-transposition showed Q waves in some precordial leads as follows: right precordial leads in five cases, precordial lead V6 in four cases, and in each of the precordial leads in the remaining case.

Q waves were present in some precordial leads in 14 of the 20 cases with l-transposition, 11 in the right precordial leads, and three in precordial lead V6.

Of the five electrocardiograms from cases with double conus, Q waves were present in three, one in the right precordial leads, and two in precordial lead V6.

In the remaining electrocardiograms, the QRS morphology showed rS, Rs, RS, and pure R complexes. The distribution in the various precordial leads was similar in each type of great vessel-ventricular relationship.

Vectorcardiographic Findings

Vectorcardiograms were available for review in 20 patients, nine with d-transposition, nine with l-transposition and two with double conus.

Initial QRS vectors and maximum mean QRS axes in the frontal, horizontal and left sagittal planes are shown in figures 6, 7 and 8.

In cases with d- and l-transposition the initial QRS vectors were usually located anteriorly, leftward, and about equally divided between inferiorly and superiorly. The maximum QRS vector in the frontal plane was between 0 and +135° in 14 patients and

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**Table 2**

<p>| Summary of the Electrocardiographic Findings in 43 Cases of Single Ventricle with Transposition According to the Type of Conus and the Type of Transposition |</p>
<table>
<thead>
<tr>
<th>d-transposition (18 cases)</th>
<th>l-transposition (20 cases)</th>
<th>Double conus (5 cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Axis QRS</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right axis</td>
<td>9</td>
<td>15</td>
</tr>
<tr>
<td>Left axis</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Normal axis</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td><strong>Conduction disturbance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>First degree A-V block</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Third degree A-V block</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Complete right bundle branch block</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Incomplete left bundle branch block</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Absent</td>
<td>13</td>
<td>14</td>
</tr>
<tr>
<td><strong>Atrial hypertrophy</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>10</td>
<td>15</td>
</tr>
<tr>
<td>Left</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Combined</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Absent</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td><strong>Ventricular hypertrophy</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Left</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Combined</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>Absent</td>
<td>6</td>
<td>7</td>
</tr>
</tbody>
</table>

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Summary of vectorcardiographic findings in single ventricle with d-transposition.

between $-115^\circ$ and $-170^\circ$ in the other four. In the horizontal plane, it was between $-40^\circ$ and $-150^\circ$ in 12 and between $+80^\circ$ and $+180^\circ$ in the other six. In the horizontal plane the QRS loop was inscribed in a clockwise direction in eight and counterclockwise in four, and two showed a figure-of-eight configuration. Nine patients showed a spatial QRS angle wider than $60^\circ$. The magnitude of the maximum QRS vector was increased in 11 patients, seven of whom had pulmonary stenosis.

In the two cases with double conus the initial QRS forces were directed inferiorly and to the right. The mean QRS vector was directed $+90^\circ$ in one and $-90^\circ$ in the other. The direction of the QRS loop was clockwise in the former and counterclockwise in the latter. A wide spatial QRS-T angle above normal limits was present in each case.

Cardiac Catheterization Findings

Cardiac catheterization had been performed in eight of the necropsy and in 31 of the clinical cases. The pulmonary artery could be catheterized in 12 cases. In ten of these the pulmonary arterial systolic pressure was identical to the ventricular pressure while in the other two cases gradients of 95 mm and 86 mm were present between the ventricle and the pulmonary artery. The catheter entered the aorta from the ventricular chamber in 21 cases, of which five were from the necropsy group.

Of the cases of the necropsy group that were thought to exhibit subaortic stenosis, catheterization data in three confirmed this diagnosis. In only one of these was a pressure gradient (10 mm Hg) present between the ventricle and the aorta.

The systemic arterial oxygen saturation varied from 52 to 92%. The mean systemic arterial saturation was higher in cases with l-transposition than in those with d-transposition. In l-transposition the mean arterial oxygen saturation was 82.5% whereas in d-transposition it was 75%. As part of a preferred streaming phenomenon, saturation in the pulmonary...
artery was higher in cases with d-transposition (mean 85%) than in l-transposition (mean 75%).

Radiologic Findings

Roentgenograms usually were not diagnostic, whereas angiocardiography provided evidence for a firm diagnosis of single ventricle.

Roentgenograms

Roentgenograms showed signs of cardiac enlargement in 36 of the 49 patients reviewed and in 21 of these the enlargement was of moderate to marked degree.

The cardiac configuration was variable. Seventeen cases showed a bulge along the upper left border as in corrected transposition of the great vessels. In 12 of these, there was l-transposition, in four d-transposition and in one a double conus. A left ventricular configuration was present in three other cases and nondiagnostic cardiac configuration was found in the remaining 29 cases.

The pulmonary arterial segment was identified in only four cases, while in the other 45 the pulmonary trunk could not be identified as contributing to the left cardiac border.

The pulmonary vasculature was increased in 28 cases, decreased in 13 cases, and interpreted as normal in the remaining eight cases.

A right aortic arch was identified in two cases. Each was in the clinical group and showed a double conus by angiocardiography.

Angiocardiograms

Angiocardiograms were available for evaluation in 37 cases, five from the necropsy group and 32 from the clinical group.

Single ventricle with d-transposition was identified in 14 cases, single ventricle with l-transposition in 18 and common ventricle with a double conus in five cases (one being from the necropsy group). In the group of common ventricle with d-transposition a smooth, morphologically left ventricle was identified in 13 cases (fig. 9) and there was a coarse, trabeculated, undefined ventricle in one. Mild to moderate enlargement of the single ventricle was outlined in ten, marked enlargement was seen in two, and there was a normal-sized main chamber in the remaining two cases. An outflow chamber was delineated in nine cases; it was wide in eight and narrow in one case (fig. 9b). The outflow chamber was identified anterior and to the right of the main chamber in each case. In the remaining cases the presence or absence of the outflow chamber could not be delineated with certainty.

In all cases with d-transposition, two atrioven-

tricular valves were outlined. Pulmonary-mitral valvular continuity could not be made out in nine cases. The pulmonary trunk was narrower than the aorta in six cases, equal in two and wider than the aorta in six. Pulmonary valvular atresia was outlined in two cases and there was pulmonary valvular stenosis in three cases. Mitral regurgitation was found in one case. A left-sided aortic arch was present in all cases with d-transposition.

Single ventricle with l-transposition was present in 18 cases. In this group a smooth, morphologically left ventricle was outlined in 17 instances (fig. 10) and a coarse, trabeculated, undefined ventricle was outlined in one. Mild to moderate enlargement of the common ventricle was identified in ten and the common ventricle was of normal size in the remaining eight cases. An outflow chamber could be delineated in 15 cases; it was wide in 12 and narrow in three. The connection of the outflow chamber with the common ventricle (bulboventricular foramen) was stenotic in three and nonstenotic in the remaining cases. The outflow chamber was localized anterior and to the left of the main chamber in 17 cases. A discordantly positioned outflow chamber, anterior and to the right of the main chamber, was delineated in the remaining case. Two atrioventricular valves were identified in all cases. Pulmonary-mitral valvular continuity could be made out in 14 cases. The pulmonary trunk was more narrow than the aorta in seven cases, equal in one, and wider than the aorta in the remaining ten cases. Pulmonary valvular stenosis was identified in five cases, pulmonary valvular as well as subpulmonary stenosis in one and only subpulmonary stenosis in two instances. Tricuspid regurgitation was found in one case. A left-sided aortic arch was present in each case.

Figure 9

Angiocardiogram from a case of single ventricle of the left ventricular type and d-transposition. a) Frontal view. The aorta (A.) lies to the right of the pulmonary trunk (P.T.). The configuration of the single ventricle (S.V.) is of the left ventricular type. b) Lateral view. The aorta (A.) arises from a small conus (C.) and courses anteriorly to the pulmonary trunk (P.T.).
A coarse, trabeculated, undefined ventricle was observed in each of the cases with double conus (fig. 11). In this group, the common ventricle was mildly to moderately enlarged in two and of normal size in the remaining three cases. The presence of two atrioventricular valves was evident in four cases. A subpulmonary conus as well as a subaortic conus was identified in each. The semilunar valves were located at the same level in the frontal as in the lateral views, resulting in discontinuity between the semilunar valves and the atrioventricular valves. The pulmonary trunk was narrower than the aorta in three cases, equal in one, and wider in the remaining case. Pulmonary valvar stenosis was outlined in three instances and subpulmonary stenosis in two. The aortic arch was right-sided in two cases and left-sided in the remaining cases.

**Comment**

Our definition of single ventricle includes only cases with two atrioventricular valves opening separately into the common ventricular chamber. This definition excludes cases of mitral or tricuspid atresia and double inlet left ventricle, as well as those with common atrioventricular valve. Atresia of the mitral or tricuspid valve results in specific clinical and anatomic entities. Common atrioventricular valve with single ventricle is frequently associated with splenic anomalies. Because of the associated anomalies and the anomalies of the viscero-atrial situs, cases with splenic anomalies were not included in our study.

We did not designate our cases as double inlet left ventricle. Nevertheless, those examples of the left ventricular type of single ventricle as defined here have been classified as a form of double inlet ventricle by others.

Van Praagh, Ongley, and Swan identified four types of single ventricle according to the following specific features: 1) absence of the right ventricular sinus (Type A); 2) absence of the left ventricular sinus (Type B); 3) absent or rudimentary ventricular septum (Type C); and 4) absence of both right and left ventricular sinuses (Type D). Type A of Van Praagh and associates corresponds to our single ventricle of the left ventricular type, while their Types B and D correspond to our primitive type. It is our opinion that Van Praagh’s Type C corresponds to a huge ventricular septal defect.

Lev et al. identified single (primitive) ventricle as that condition with a common sinus and absence of the ventricular septum, whereas the term common ventricle was used for cases exhibiting a true posterior ventricular septum. Nevertheless, the authors pointed out that the latter cases are really examples of a huge ventricular septal defect and two distinct right and left ventricular sinus components are present. Both the left ventricular and primitive types of our classification correspond to Lev’s single (primitive) ventricle.

In contrast with the foregoing authors, we were able to identify two types of single (common) ventricle. One showed anatomic features of a left ventricle, whereas the other showed no features identifiable as belonging to either definitive ventricle. In both types,
SINGLE VENTRICLE WITH TRANSPOSITION

Clinical, electrocardiographic, and vectorcardiographic findings were not distinctive among cases with morphologically single left ventricle as compared to those with primitive ventricle. Similarly, these parameters exhibited little difference between cases of single ventricle with d-transposition or l-transposition. Except for a higher incidence of left axis deviation of the QRS complex in the frontal plane in cases with double conus, no significant distinctions were found in the electrocardiograms and a similar pattern for ventricular hypertrophy was present for the two types of ventricular structures and for the various arrangements of the great vessels.

Abnormal atrioventricular conduction was found in eight electrocardiograms. Three cases of d-transposition and two of l-transposition presented complete atrioventricular block. The remaining three electrocardiograms with a prolonged PR interval were from cases of single ventricle with l-transposition. The similar incidence of atrioventricular conduction disturbances in our cases of single ventricle with d- or l-transposition are in conformity with the observations of Shaher.11 Factors other than a longer bundle of His postulated for corrected transposition with a well developed ventricular septum should be considered for the development of heart block in single ventricle.

Abnormal initial forces of the QRS loop in the frontal plane were a frequent finding in our cases with d- and l-transposition. A similar experience was reported earlier.12 However, in the two cases with double conus in which a vectorcardiographic study was available, the initial forces of the QRS loops were like that of a normal heart. Whether a different pathway for ventricular conduction is present in the latter group compared with the cases of d-transposition or l-transposition is not known.

Roentgenographically, most of our cases of single ventricle showed some cardiac enlargement with a variable cardiac configuration. Twelve of 17 cases with l-transposition exhibited a bulging left upper border as is seen in cases of corrected transposition of the great vessels with a well developed ventricular septum.

In our study, cardiac catheterization, although nondiagnostic, often provided a suspicion of single ventricle. The presence of a significant left-to-right shunt in a ventricular chamber with systemic pressure, as well as the variable range in oxygen content at different sites in the ventricle were of value.

We found, as did Rahimtoola, Ongley, and Swan,18 that cases with l-transposition presented a favorable streaming of saturated blood from the single ventricle as compared with those with d-transposition.

Selective angiocardiography with injection of con-

no evidence for the sinus portion of a second ventricle was present. The primitive type appears to represent an early arrest in the development of the heart, before features of the definitive left ventricle develop. Seven of our ten cases of the primitive type of single ventricle exhibited a double conus. We believe that this fact favors the presumption that this type of single ventricle is embryologically a more primitive condition than the left ventricular type. In contrast, only one case of the left ventricular type exhibited a double conus, in spite of a significantly higher incidence of this type of chamber in our series.

The internal morphology of single left ventricle with d- or l-transposition was different. With d-transposition, the smooth wall and the bulboventricular foramen were situated anteriorly and to the right, while in l-transposition the smooth wall and bulboventricular foramen were situated anterior and to the left. These two structures suggest differences in the bulboventricular loop. Cases of d-transposition with the smooth wall anterior and to the right suggest an arrest in development of classical complete transposition, whereas l-transposition with the smooth wall anterior and to the left suggests an arrest in development of classical corrected transposition. Rarely, a discordance between the type of cardiac loop and the type of external relationship of the great vessels may occur. This was suspected in one of our clinical cases in which the outlet chamber was anterior and to the right of the main chamber, whereas the great vessel relationship was that of l-transposition. Cases with an l-loop and d-transposition in single left ventricle have been reported and embryologic considerations have been described.7

Obstruction to systemic blood flow was frequently present in the necropsy group and an apparently higher incidence occurred in cases with the primitive type of single ventricle than with the left ventricular type. However, among cases with the left ventricular type of single ventricle no differences with regard to evident systemic arterial obstruction were found between cases with d- and l-transposition.

We do not have any proven cases of single ventricle with truncus arteriosus but it was included in our classification because a few cases have been reported.9,10

In our series, cyanosis was a common finding but no correlation between its severity and the degree of pulmonary stenosis was found. Cardiac failure was present in 18 cases and pulmonary stenosis was present in two of these. From the auscultatory data, it is remarkable that there was a split second sound in 16 instances. An early diastolic murmur was described in four cases, one of which had pulmonary stenosis.
trast material into both atria as well as into the single ventricle allowed demonstration of the patency of both atrioventricular valves, identification of the variable structure of single ventricle, delineation of the subaortic and subpulmonary areas, and associated anomalies.

Single ventricle with transposition of the great vessels may mimic other conditions presenting with cyanosis and decreased pulmonary blood flow such as tetralogy of Fallot or tricuspid atresia, as well as cardiac anomalies with cyanosis and increased pulmonary blood flow such as transposition of the great vessels, persistent truncus arteriosus, double outlet right ventricle, and ventricular septal defect with pulmonary vascular disease. The various conditions named usually can be distinguished only by angiography.

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Single Ventricle with Transposition

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