Common (Single) Ventricle with Normally Related Great Vessels

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SUMMARY
A rare form of common ventricle with normally related great vessels and normal tricuspid and mitral valves should be considered in the differential diagnosis of cardiac anomalies presenting with cyanosis. Fourteen cases of this type of common ventricle were studied; in seven the diagnosis was made at necropsy and in seven it was based on angiographic evidence. That the common ventricle was morphologically a left ventricle was determined in the seven necropsy cases, and angiocardiographic examination confirmed the finding in the others. Continuity of the aortic valve and the mitral valve was observed in all the necropsy cases and in five of the clinical cases. Cyanosis had been observed clinically in 13 of the 14 cases, and pulmonary stenosis was present in four necropsy and three clinical cases. Electrocardiograms showed left axis deviation in nine of the 13 cases studied. Vectorcardiographic findings, though variable and available in only seven cases, tended to show a narrow horizontal plane QRS vector loop in patients with restricted pulmonary blood flow and a wide loop in those with increased pulmonary flow. The clinical, laboratory, and pathologic findings are reported and the differential diagnosis and the profile of the condition are considered.

Additional Indexing Words:
Pulmonary stenosis Cor triloculare biatrium Holmes' heart

COMMON OR SINGLE VENTRICLE is usually associated with transposition of the great vessels. Its incidence has been estimated in large published series of congenital heart disease as 3%. 1, 2 In a rare form of common ventricle, the great vessels are normally related and the tricuspid and mitral valves are normal. Only a limited understanding of this variety can be obtained from the literature because of the few cases reported. It should be considered in the differential diagnosis of cardiac anomalies presenting with cyanosis such as tetralogy of Fallot, tricuspid atresia, ventricular septal defect with pulmonary hypertension, or transposition of the great vessels. Definitive diagnosis is difficult without angiocardiographic studies.

Our purpose is to present the findings in 14 cases of single ventricle with normally related great vessels and two atrioventricular valves. From the clinical and laboratory findings, we will define the clinical profile of this condition and describe the typical angiocardiographic features. The pathologic features will be presented in seven cases.

Patients Studied and Methods
Fourteen cases of single ventricle and normally related great vessels were reviewed (table 1). The diagnosis was confirmed at necropsy in seven cases (necropsy group) and by angiography in another seven cases (clinical group). Cases of common atrioventricular valve, tricuspid atresia, or mitral atresia were not included.

Each of the seven necropsy cases in this study exhibited two atrioventricular valves, mitral and aortic valvular continuity, common (single) ventricle, and normally related great vessels. Three of these cases have been reported earlier. 3

The clinical studies had been done for the purposes of diagnostic evaluation of the patients.

Clinical and radiologic findings were available for study in each of the 14 cases and electrocardiographic findings in 13 cases. Schmitt's corrected orthogonal lead vectorcardiograms were available in two of the
Table 1

Age, Sex and Pulmonary Stenosis in the 14 Cases

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Sex</th>
<th>P.S.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Necropsy Group</td>
</tr>
<tr>
<td>1</td>
<td>10 mo</td>
<td>F</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>1 mo</td>
<td>F</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>3 yr</td>
<td>M</td>
<td>+</td>
</tr>
<tr>
<td>4</td>
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<td>M</td>
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<tr>
<td>5</td>
<td>9 yr</td>
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<tr>
<td>6</td>
<td>23 yr</td>
<td>F</td>
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</tr>
<tr>
<td>7</td>
<td>5 yr</td>
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<td>+</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Clinical Group</td>
</tr>
<tr>
<td>8</td>
<td>7 yr</td>
<td>F</td>
<td>+</td>
</tr>
<tr>
<td>9</td>
<td>7 yr</td>
<td>M</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>3 yr</td>
<td>F</td>
<td>+</td>
</tr>
<tr>
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<td>2 mo</td>
<td>F</td>
<td>0</td>
</tr>
<tr>
<td>12</td>
<td>7 yr</td>
<td>F</td>
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</tr>
<tr>
<td>13</td>
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<td>F</td>
<td>0</td>
</tr>
<tr>
<td>14</td>
<td>17 yr</td>
<td>M</td>
<td>0</td>
</tr>
</tbody>
</table>

Abbreviations: P.S. = pulmonary stenosis; M = male; F = female; + = present; 0 = absent.

necropsied cases and in five of the clinical cases. Selective biplane angiocardiography or forward venous angiography was performed in 12 patients including each of the seven living patients. Cardiac catheterization data were accessible in ten cases. Except for the one case in the clinical group in which cardiac catheterization findings were not available, cardiac catheterization and selective biplane angiocardiography or forward venous cardiology were performed in all of the clinically confirmed cases.

In the present report, the terms "single ventricle" and "common ventricle" are used synonymously.

Observations

The ages of the patients varied from one month to 23 years. Four of them were one year of age or less, four others were two to five years of age, and four from six to ten years. The two remaining patients were 17 and 23 years old. Of the 14 patients, ten were females and four were males (table 1).

Angiographic Basis for Diagnosis in the Clinical Group

The diagnosis was made angiographically in the seven patients of the clinical group. In each, a common ventricle, with morphologic features of a left ventricle was found. Two distinct atrioventricular valves were identified in various ways including the course of the catheter, forward flow of contrast material across the atrioventricular valves, and the appearance of the valvular annuli in ventriculograms. An infundibulum located anteriorly was identified in six of the seven cases, and in these, the pulmonary trunk was shown to arise from the infundibulum, while the aorta arose from the main chamber. The great vessels were normally related. Aortic-mitral valvular continuity was clearly identified in five of the seven cases.

Pathologic Study

Specimens of heart were examined in each of the cases in the necropsy group. One of the specimens was then returned to its original source, leaving six specimens available for re-examination.

The viscera were in situ solitus, and the cardiac apex was directed to the left in each of the seven cases. The systemic and pulmonary venous connections were normal, and the aortic arch was on the left side in each.

Atria

The morphologically right atrium was on the right side and was anterior to the morphologically left atrium in all seven cases, and the coronary sinus entered the right atrium as is normal. The right and left atria were normal to slightly increased in size in all except one case, in which the left atrium was very large owing to mitral insufficiency. In four cases, the atrial septum showed a valvular competent patent foramen ovale. An atrial septal defect at the fossa ovalis, measuring 4 by 2 cm, was present in one case, and the atrial septum was anatomically sealed in the remaining two cases. The atrial appendages were normally positioned in each case.

Atrioventricular Valves

Two separate atrioventricular valves were present. The right atrioventricular valve had the characteristics of a tricuspid valve and the left of a mitral valve. There was continuity of the mitral and tricuspid valves with each other and with the aortic valve in the common ventricular chamber (figs. 1 and 2). The chordae tendineae from each valve frequently inserted into a common papillary muscle. In the case exhibiting a very large left atrium, the association of a cleft in the anterior leaflet of the mitral valve with an accessory commissure in the posterior leaflet had caused severe mitral regurgitation.

The Ventricular Complex

The morphology of the common ventricle was characteristic of a left ventricle in each case corresponding to type A single ventricle of Van Praagh et al.4 The anterior and right walls were
The infundibulum (I.) with pulmonary trunk (P.T.) which arises above it. Jet lesions (arrows) are prominent above the stenotic communication (probe) with the single ventricle. b) Interior of the single ventricle (S.V.) showing the mitral (M.) and tricuspid (T.) valves continuous with each other and with the aortic valve. The aorta (Ao.) arises from the single ventricle as in a normal heart. The communication (probe) with the infundibulum is stenotic.

smooth and resembled the smooth part of the normal ventricular septum. The posterior wall showed fine trabeculation like that in the free wall of the normal left ventricle. The aorta joined the ventricle in a fashion similar to that in a normal heart. An infundibular chamber of variable size, present in six of the seven specimens, was located anteriorly. The long axis of the infundibulum was directed from right to left and resembled the normal right ventricular outflow tract (figs. 1a and 2a and b). The pulmonary trunk arose from this chamber. In the one case in which there was no infundibulum, the communication between the common ventricle and the pulmonary trunk was through a narrow tract located in the anterior wall of the ventricle.

Semilunar Valves

In each case, the aortic valve showed three cusps without evidence of valvular stenosis. The pulmonary valve was tricuspid in three cases and bicuspid in three cases. The number of cusps in one case is not known. Valvular pulmonary stenosis was present in two of the three cases with the bicuspid pulmonary valves.

Anatomic Basis for Pulmonary Stenosis

In three specimens there was no anatomic obstruction to pulmonary blood flow; in one specimen mild obstruction was present; and in three specimens severe obstruction was present. In each of the three specimens without obstruction to pulmonary blood flow, the infundibulum as well as its communication with the common ventricle was wide (fig. 2b).

The infundibulum was normal in size in two of the four specimens with obstruction to pulmonary blood flow. In one of these, a moderate-sized communication between the common ventricle and the infundibulum had resulted in mild subpulmonary stenosis; in the other case, the markedly stenotic communicating tract had resulted in severe subpulmonary stenosis (fig. 1a). In one of the other two specimens, the infundibulum was narrow; in the other it could not be made out. A narrow tract communicating with the ventricle and located in the anterior wall of the common ventricle opened into the infundibulum in the former specimen and at the pulmonary valve in the latter. The pulmonary valve was bicuspid and stenotic with a narrow valvular ring in both of these cases.

Coronary Arterial Pattern

The origin and distribution of the coronary arteries was normal in six of the seven cases in which this point could be evaluated.
Associated Findings

The ductus arteriosus was ligamentous in four cases and patent in three; the ages of the three patients with patent ductus were one month, one year, and three years. One of them showed mild coarctation of the aorta, subaortic stenosis, and supravalvular ring, grade 1, in the left atrium.

Clinical and Laboratory Features

There were no differences in the clinical features in the necropsy and clinical groups. Each of the 14 patients was symptomatic, but the severity of symptoms was variable. Thirteen of the 14 patients were cyanotic. The cyanosis was present at birth in five, became apparent by the age of one year in four, and developed after one year of age in four. One patient appeared acyanotic, while the systemic-arterial oxygen saturation was 92%. There was no clear correlation between the severity of cyanosis and presence or absence of pulmonary stenosis.

Eight of the 14 patients were dyspneic. Two patients gave a history of hypoxic episodes, and one patient squatted. Each of the latter three patients had pulmonary stenosis. Four patients presented with congestive cardiac failure, none of whom had obstruction to pulmonary blood flow.

The first heart sound was normal in each patient. The second heart sound, adequately described in 12 patients, was single in five and split but accentuated in seven.

An ejection systolic murmur, varying from grade II to IV (on the basis of VI) in intensity, was present in each patient and associated with a thrill in three. The maximum intensity of the murmur was along the left sternal border. A holosystolic murmur, grade III in intensity, was present in the patient with a cleft in the anterior leaflet of the mitral valve. A middiastolic, rumbling murmur was audible in four patients. None of the patients with diastolic rumbles exhibited obstruction to pulmonary blood flow.

Radiologic Findings

Cardiac enlargement was noted in eight of the 14 patients. The cardiac configuration was variable (fig. 3). Three patients showed typical coeur en sabot configuration; six others, a left ventricular configuration. One of the necropsied cases, the one with a mitral cleft and mitral insufficiency, showed a globular heart suggestive of biventricular enlargement. In the remaining four patients cardiac configuration was normal.

Each of the patients showed a left aortic arch.

Figure 3

a–d) Variation of cardiac configuration on roentgenograms of four patients with single ventricle.

The pulmonary vasculature was decreased in seven and increased in the other seven.

Electrocardiographic Findings

The electrocardiogram was available for evaluation in 13 cases. Among these the mean frontal plane QRS axis showed a wide distribution (fig. 4). In nine of the 13 cases, however, left axis deviation was present. The axis did not show any correlation with the presence or absence of pulmonary stenosis.

Sinus rhythm was noted in electrocardiograms of 12 cases. Atrial fibrillation was present in the patient with mitral insufficiency and marked left atrial enlargement.

Right atrial hypertrophy was evidenced in electrocardiograms of three patients, combined right and left atrial hypertrophy in seven, and left
atrial hypertrophy in one. Signs of atrial hypertrophy were absent in the remaining patient.

The QRS duration was normal in all except one patient. This exception was a member of the clinical group who showed complete left bundle branch block. Isolated right ventricular hypertrophy was present in three cases, isolated left ventricular hypertrophy in six cases, and combined ventricular hypertrophy was present in two cases (fig. 5 and 6). There was no evidence for ventricular hypertrophy in one patient.

The QRS complex morphology in the precordial leads showed rS, RS, Rs and pure R. The electrocardiograms of only three patients showed q waves; in two of these they were seen in the right precordial leads and one in precordial lead V4.

**Vectorcardiographic Findings**

Vectorcardiograms were available for review in seven cases: two were from the necropsy group and five from the clinical group. The initial QRS vector and maximum mean QRS axis in the frontal, horizontal and left sagittal planes are shown in figure 7, which may be analyzed as follows.

In the frontal plane, the initial QRS vector was directed inferiorly and rightward in four cases,
inferiorly and leftward in two, and superiorly and leftward in one case. The maximum mean QRS axis was between $-10^\circ$ and $-155^\circ$ in six cases and was $+30^\circ$ in one case. The vector loop direction was counterclockwise in five cases. It showed a figure-of-eight configuration in one and clockwise direction in the remaining case.

In the horizontal plane the initial QRS vector was directed anteriorly and to the right in four cases, anteriorly and to the left in two, and posteriorly and to the left in one case. The maximum mean QRS axis was between $-45^\circ$ and $-110^\circ$ in six cases and was $+170^\circ$ in one case. The vector loop direction was counterclockwise in four cases and had a figure-of-eight configuration in three. The magnitude of the mean QRS vector was increased in three cases.

In the left sagittal plane the initial QRS vector was directed anteriorly and inferiorly in six cases and posteriorly and superiorly in one case. The maximum mean QRS axis was between $0^\circ$ and $-120^\circ$ in six cases and was $+20^\circ$ in one case. The vector loop direction was counterclockwise in three, clockwise in one, and a figure-of-eight in three cases.

Cardiac Catheterization Findings

Cardiac catheterization had been performed in four of the necropsy and in six of the clinical cases.

The systemic arterial oxygen saturation varied from 36 to 78% in the necropsy group and from 73 to 92% in the clinical group. The pulmonary artery could be catheterized in five cases. In three cases the pulmonary arterial pressure was identical to the ventricular pressure while in the other two cases gradients of 65 mm and 100 mm Hg were present between the pulmonary artery and ventricle. The catheter entered the aorta in five cases from the common ventricular chamber, and the aortic pressure was identical to the ventricular pressure in each.

Angiographic Findings

Angiograms, performed in 12 cases, were available for re-evaluation in three of the necropsy group and in each of the members of the clinical group.

In the necropsy group, the angiogram correctly identified the presence of a common ventricle with normally related great vessels in all three. The presence of two atrioventricular valves and a subpulmonary outlet chamber was outlined also in two of these three cases. The pulmonary trunk was narrower than the aorta in two cases and equaled it in one case. Mitral regurgitation was marked in one case.

In the clinical group a common ventricle with left ventricular morphology and two atrioventricular valves was identified in each case. A subpulmonary outlet chamber was recognized in six of the seven cases. Aortic valve-mitral valve continuity could be made out in five of the seven cases. The great vessels were normally related in all seven cases (figs. 8 and 9).

The pulmonary trunk was narrower than the aorta in three cases, equal to the aorta in three cases, and larger than it in one case. In three cases the common ventricle appeared enlarged, and in one case mild mitral regurgitation was evident. The aortic arch was to the left side in each.

Discussion

The features of the case described by Holmes include common (single) ventricle, normally related great vessels, and the presence of both atrioventricular valves.

We are aware of 19 previously reported cases. Fifteen of the reported cases were reviewed by Klaus et al., who added one case. In addition, one case was described by Lev et al. and two by Anselmi et al.

Although pulmonary stenosis was absent in the original case, we propose that the term "Holmes' heart" include all cases of common ventricle associated with normally related great vessels and the presence of both atrioventricular valves. The matter of presence or absence of pulmonary stenosis may be used as a basis for dividing the condition known as the Holmes' heart into two subgroups.

It is apparent that cases of common ventricle and normally related great vessels may be subdivided into one of three groups as follows: (1) that with two atrioventricular valves (the Holmes' heart), the 14 cases herein described being examples; (2) that with a common atrioventricular valve; and (3) that with either mitral or tricuspid atresia.

Our description deals with cases of common ventricle and normally related great vessels in which there were two distinct and patent atrioventricular valves. We believe this represents a distinct anatomic condition which may be recognized by appropriate angiographic studies.

The 14 cases described indicate that cyanosis was common, although this process frequently did not appear before the age of one year, and in one

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Figure 8
Angiocardiograms following injection of contrast material into the single ventricle (S.V.) through the tricuspid valve. a) Frontal view. b) Lateral view. The anterior origin of the pulmonary trunk (P.T.) and the subpulmonary stenosis (arrow) is outlined. Ao. = ascending aorta.

Patient, not until the age of nine years. Correlating the intensity of the cyanosis with either the presence or the absence of pulmonary stenosis was difficult. The other features of the history and the auscultatory findings were not sufficiently specific to allow distinction of this anomaly from tetralogy of Fallot or from other types of single ventricle. Likewise, the roentgenographic findings were variable and showed either increased or decreased pulmonary vascularity, depending on the presence or absence of pulmonary stenosis. The cardiac contour was also not specific. Several patients, however, did show cardiomegaly and this tended to exclude conditions such as tetralogy of Fallot.

Electrocardiographic and vectorcardiographic findings were distinctive in that nine of the 13 patients showed left axis deviation. This, plus the coexistence of left ventricular hypertrophy in six of these, forms a pattern which should alert the physician to the presence of Holmes’ heart or tricuspid atresia. Wide variations of vectorcardiographic patterns that are difficult to correlate with the anatomic features of common ventricle have been reported among patients with common ventricle. Such variations were also present in the vectorcardiograms of our patients, as some showed patterns of left ventricular hypertrophy, others of right ventricular hypertrophy, and still others of biventricular hypertrophy. One interesting trend, although based on only seven vectorcardiograms, was that the horizontal plane QRS vector loop tended to be narrow among patients with restricted pulmonary blood flow whereas those with increased pulmonary blood flow showed a wide loop (fig. 10). Similar correlations have been reported by Davachi, Lucas, and Moller in patients with tricuspid atresia. They found that variations in the patterns of the electrocardiogram and vectorcardiogram depended on the volume of pulmonary blood flow.

From clinical data alone, it is not possible to arrive at a diagnosis of common ventricle with normally related great vessels in an individual patient. Two clinical pictures become evident and one of these mimics other conditions with cyanosis and decreased pulmonary blood flow such as tetralogy of Fallot and tricuspid atresia. In six of our seven patients with decreased pulmonary blood flow, the presence of left axis deviation and left ventricular hypertrophy would yield a picture similar to that of tricuspid atresia. The two conditions can be distinguished only by angiographic demonstration. Even after left ventriculography, confusion between Holmes’ heart with pulmonary stenosis and tricuspid atresia can continue. Distinction depends on injection of contrast material into the right atrium and demon-

Figure 9
Angiocardiogram following injection of contrast material into the single ventricle (S.V.) showing left ventricular morphology. a) Frontal view. b) Lateral view. The anteriorly placed wide infundibulum (I.) is well outlined.

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The vectorcardiogram recorded at ½ normal standardization (20) from a patient without pulmonary stenosis. The main QRS forces are directed posteriorly, superiorly, and to the left. The initial QRS forces are directed anteriorly and to the right. The direction of the QRS loop is counterclockwise in the frontal and horizontal planes. The QRS loop is wide. b) The vectorcardiogram recorded at ½ normal standardization in the horizontal plane (20) and at ½ normal standardization (10) in a patient with pulmonary stenosis. The main QRS forces are directed posteriorly and to the left. The initial forces are directed anteriorly and to the right. The loop is inscribed clockwise in the frontal plane and has a figure-of-eight pattern in the horizontal plane. The QRS loop is narrow.

Figure 10

The vectorcardiogram recorded at ½ normal standardization (20) from a patient without pulmonary stenosis. The main QRS forces are directed posteriorly, superiorly, and to the left. The initial QRS forces are directed anteriorly and to the right. The direction of the QRS loop is counterclockwise in the frontal and horizontal planes. The QRS loop is wide. b) The vectorcardiogram recorded at ½ normal standardization in the horizontal plane (20) and at ½ normal standardization (10) in a patient with pulmonary stenosis. The main QRS forces are directed posteriorly and to the left. The initial forces are directed anteriorly and to the right. The loop is inscribed clockwise in the frontal plane and has a figure-of-eight pattern in the horizontal plane. The QRS loop is narrow.

stratation of the patency or atresia of the tricuspid valve.

In our seven patients with a pattern of cyanosis and increased pulmonary blood flow, distinction lies between common ventricle with normally related great vessels and such conditions as ventricular septal defect with pulmonary vascular disease, double outlet right ventricle, transposition of the great vessels without pulmonary stenosis, other forms of single ventricle, and persistent truncus arteriosus. The absence of an ejection click would tend to eliminate truncus arteriosus, but in general, the clinical distinction is difficult without angiography. Once again, the angiographic features of the Holmes’ heart and the finding of normally related great vessels would help solve this problem of differential diagnosis. In our cases the angiocardiograms typically showed a large ventricle with the internal characteristics of a left ventricle. This ventricle was smooth-walled and extended to the left cardiac border. Medially placed was a small infundibulum which opacified after the injection of contrast material into the common ventricle. It can also be visualized after injection of contrast material into the right atrium. It is also important to identify the continuity of the aortic and mitral valves to distinguish this condition from forms of double outlet right ventricle.

The long-term survival of patients with common ventricle with or without normally related great vessels are those with coexistent pulmonary stenosis. The pulmonary stenosis has two beneficial effects. 1) It limits pulmonary blood flow and thus tends to prevent congestive cardiac failure. 2) It protects the pulmonary vasculature from pulmonary hypertension and its organic changes. Among our patients, the two oldest were 17 and 23 years of age. Neither had pulmonary stenosis. The older of these was in the necropsy group. Significant hypertensive pulmonary vascular disease was present. Our 23-year-old patient was the second oldest patient with Holmes’ heart, the oldest being a 26-year-old patient reported by Klaus et al. In the latter case, pulmonary stenosis was present.

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Circulation. 1974;49:565-573
doi: 10.1161/01.CIR.49.3.565
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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