Transposition of the Great Vessels with Hypoplasia of the Right Ventricle

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SUMMARY

Eight cases are reported in which complete transposition of the great vessels was associated with varying degrees of hypoplasia of the right ventricle. In three, the correct diagnosis was suspected on the basis of certain electrovectorcardiographic findings and confirmed by selective angiography. Quantitative analysis of ventricular volumes and valve circumferences demonstrated that these cases represent a distinct pathological entity.

The diagnosis of complete transposition with hypoplastic right ventricle may be suggested by electrocardiographic evidence of left ventricular hypertrophy, relative absence of right ventricular QRS vectors, and leftward axis for age. The vectorcardiogram reveals a counterclockwise QRS loop in the horizontal plane, situated to the left and posteriorly. These findings occur infrequently in the young infant with complete transposition.

The presence of a hypoplastic right ventricle is confirmed by angiography, which demonstrates a small right ventricular chamber located centrally in the cardiac silhouette, not extending beyond the left border of the thoracic spine.

Additional Indexing Words:
Complete transposition
Hypoplasia of tricuspid valve
Electrocardiographic data
Ventricular wall thickness
Cyanotic congenital heart disease
Vectorcardiographic study
Angiographic diagnosis
Necropsy findings

Recent advances in the surgical treatment of complete transposition of the great vessels have stimulated more precise definition of the pathophysiology of this lesion. Previous reports have emphasized that evaluation of the infant with complete transposition should include determination of: (1) status of pulmonary vasculature,1 (2) location and size of communications between the two circulations,1 and (3) anatomy of the left ventricular outflow tract.2 An additional factor which may be of great importance in the pre-operative assessment of infants with this malformation is the adequacy of ventricular chamber size. We have recently become aware of the importance of this additional factor on the basis of our experience with several unusual cases of complete transposition of the great vessels, in which necropsy revealed a small right ventricular chamber. Seven patients with complete transposition of the great vessels and hypoplastic right ventricle have been encountered at the University of California Medical Center, in Los Angeles, and an additional case was followed by one of us (W.V.) at the University of California Medical Center in San Francisco. In three instances, the correct diagnosis was suspected on the basis of certain clinical and angiographic findings while in the remaining five cases, the presence of hypoplasia of the right ventricle was not appreciated prior to necropsy.

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The purpose of the present report is to describe the pathological anatomy of this malformation and to emphasize those features which may facilitate clinical diagnosis.

Methods

All necropsy-proven cases of complete transposition of the great vessels in patients who were admitted to the UCLA Medical Center between 1956 and 1967 have been reviewed. Complete transposition of the great vessels (hereafter referred to as complete transposition) is defined as that condition in which two ventricles are present and the aorta takes origin from the venous right ventricle while the pulmonary artery takes origin from the arterial left ventricle.

The patients were divided into two groups based upon measurement of ventricular volumes. Group I (study group) consisted of eight patients aged 3 weeks to 3½ months, in whom varying degrees of hypoplasia of the right ventricle were encountered. One of the study group cases was followed at UCSF. To make the groups comparable, group II (control group) was limited to all remaining patients less than 4 months of age at the time of death. Each of the 34 patients in this group had a well-developed right ventricular chamber.

The following observations were recorded for specimens of both groups: (1) diameter and relationship of the great vessels, (2) circumference of the semilunar and atroventricular valves, (3) thickness of the ventricular walls and septum, (4) dimensions and volume of the ventricular chambers, (5) coronary artery patterns, and (6) associated defects. The ventricular free walls and septum were measured at the point of maximum thickness, which was usually located at the midportion of the free wall and the lower portion of the septum. Inflow length of the ventricle was measured from the atroventricular valve annulus to the apex of the chamber, while outflow length was recorded as the distance between the semilunar valve annulus and the apex.

Ventricular volume was measured after each ventricle was filled with mercury (fig. 1). The ventricular cavity was lined with a thin plastic bag into which mercury was injected to the annulus of the semilunar valve. The volume of mercury in the bag was then measured to the nearest 0.1 ml.

Thirteen-lead electrocardiographic tracings were available for review in all cases of the

![Figure 1](http://circ.ahajournals.org/) Roentgenograms of representative necropsy specimens of each group, illustrating the technique used for measurement of ventricular volumes. The ventricles have been filled with mercury, and the differences between right (RV) and left (LV) ventricles of the two groups are apparent.
Table 1

Pertinent Pathological Data

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Ratio of circumferences of valves (tricuspid/mitral)</th>
<th>Ventricular walls RV/LV (mm)</th>
<th>Inflow/outflow RV (mm)</th>
<th>LV (mm)</th>
<th>Ventricular volume ratio (RV/LV)</th>
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<tr>
<td></td>
<td>Study group*</td>
<td></td>
<td></td>
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<td></td>
</tr>
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<td>18/28</td>
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<td>Mean</td>
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<td>31/38</td>
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<td>Control group†: Summary of data</td>
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<td>Mean</td>
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<td>(4-14)/(4-10)</td>
<td>(17-38)/(20-45)</td>
<td>(17-38)/(20-44)</td>
<td>(0.52-2.3)</td>
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</table>

*Case 7 was not available for quantitative analysis.
†Control group consisted of 34 specimens.

Abbreviations: RV = right ventricle; LV = left ventricle.

Study group and in 29 cases of the control group. Vectorcardiograms, recorded by the Frank or cube system, were made in five cases of group I and 10 cases of group II. In all cases of both groups, horizontal and frontal plane QRS vector loops were plotted from electrocardiographic tracings.

Cardiac catheterization with selective angiocardiography was performed in seven cases of the study group and all cases of the control group.

Results

Because this report deals primarily with complete transposition associated with hypoplasia of the right ventricle, data from the control specimens have been summarized (table 1) and will be discussed only to emphasize significant differences between the two groups.

Pathological Anatomy

External Configuration of the Heart and Great Vessels

The interventricular groove was situated to the right of its usual position and divided the anterior cardiac surface into a very small right ventricle and larger left ventricle (fig. 2). The atria were in their usual positions. Enlargement of the right atrium was encountered in four cases.

The great vessels were obliquely related in seven cases, with the aorta anterior and to the right of the pulmonary artery. In the remaining

**Figure 2**

Case 5. External anatomy of the heart. The anterior descending coronary artery is situated to the right (arrow) and divides the heart into a hypoplastic right ventricle (RV) and large left ventricle (LV). Note the indentation at the right mid-cardiac border indicating the lower limit of the right ventricular cavity. The aorta (Ao) and pulmonary artery (PA) are situated side by side.
case, the great vessels were situated side by side with the aorta to the right of the pulmonary artery (fig. 2).

The coronary artery pattern was similar in all specimens. The right coronary artery originated above the posterior aortic sinus and the left, above the left sinus. The left coronary artery divided immediately into a circumflex branch which passed anteriorly and in front of the left ventricular outflow tract, and an anterior descending artery was located in the interventricular groove.

Size and Thickness of Ventricles

The striking anatomic feature in every study group case was marked hypoplasia of the right ventricular chamber (figs. 3 and 4). While mean left ventricular chamber measurements were approximately the same in both groups, mean right ventricular chamber measurements were significantly smaller in the hypoplastic specimens (table 1). Comparison of right ventricular chamber measurements of the two groups revealed that hypoplasia was predominantly due to lack of development of the inflow portion of the ventricle. Despite the small size of the chamber, the right ventricular free wall approximated the thickness of the corresponding left ventricular wall in every study group case, and exceeded the width of the right ventricular wall in most of the control group specimens.

The inferior portion of the ventricular septum, including the cardiac apex, was markedly thickened in all cases. The lower limit of the right ventricular cavity was situated to the right of the thickened cardiac apex. The left ventricle was large and thick-walled in each case.

Quantitative Analysis of Ventricular Volumes

Measurement of ventricular volumes revealed that the hypoplastic right ventricles were approximately one third the size of those in the control group. The right and left ventricular volumes of each specimen were compared by
Case 5. (Left panel) Right anterior oblique view. The lower limit of the small right ventricular chamber is indicated by the dotted line. (Right panel) Lateral view demonstrating the large left ventricle, anterior aorta, and posterior pulmonary artery arising from the left ventricle. Same abbreviations as in figure 3.

Case 2. Posterosuperior view of the opened atria demonstrating the hypoplastic tricuspid valve (TV) and the large mitral valve (MV). The tricuspid leaflets appear to be normally formed.

Means of a ratio (table 1). Volume ratios of the study group specimens varied from 0.16 to 0.26. Volume ratios of the control group specimens varied from 0.52 to 2.3. Statistical analysis by variance of the means revealed that the difference between the mean values of the two groups was highly significant ($P = 0.005$).

Comparison of Valve Sizes

In all cases, the tricuspid and mitral valves emptied directly into the right and left ventricles, respectively. The small right ventricular chamber was associated with hypoplasia of the tricuspid valve in each study group specimen. Despite the small size of the valve, the leaflets were normally formed and the valve was apparently competent. Hypoplastic tricuspid valve circumferences were approximately two thirds those of the mitral valve (fig. 5). In the control group specimens, tricuspid circumferences were equal to, or greater than,
### Table 2

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age at death, sex</th>
<th>Onset of CHF</th>
<th>Onset of cyanosis</th>
<th>Thoracic roentgenogram</th>
<th>Associated defects (mm)</th>
<th>Other</th>
<th>Surgical procedure</th>
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<td>2 wk</td>
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<td>↑</td>
<td>—</td>
<td>(2 x 2) Subaortic stenosis</td>
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<tr>
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<td>3 mo, M</td>
<td>2 wk</td>
<td>5 wk</td>
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<td>↑</td>
<td>(6)</td>
<td>(2 x 2) Muscular</td>
</tr>
<tr>
<td>3</td>
<td>1 mo, M</td>
<td>Birth</td>
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<td>↑</td>
<td>↓</td>
<td>—</td>
<td>(6 x 4) Muscular aortic arch</td>
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<td>↑</td>
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<td>(1 x 8), surg. ASD</td>
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<tr>
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<td>2 wk</td>
<td>4 wk</td>
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<td>↓</td>
<td>—</td>
<td>(1 x 2), surg. ASD</td>
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<tr>
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<td>Birth</td>
<td>2 wk</td>
<td>↑</td>
<td>↑</td>
<td>—</td>
<td>(2 x 2) Muscular</td>
</tr>
<tr>
<td>7</td>
<td>3½ mo, F</td>
<td>2 da</td>
<td>4 da</td>
<td>↑</td>
<td>↓</td>
<td>—</td>
<td>Surg. ASD</td>
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<tr>
<td>8</td>
<td>2½ mo, M</td>
<td>Birth</td>
<td>2 da</td>
<td>↑</td>
<td>↑</td>
<td>—</td>
<td>(1 x 2) Muscular</td>
</tr>
</tbody>
</table>

Abbreviations: CHF = congestive heart failure; ↑ = increased; ↓ = decreased; PDA = patent ductus arteriosus; ASD = atrial septal defect; VSD = ventricular septal defect; memb. = membranous; musc. = muscular; PA = pulmonary artery.
Figure 6
Mean QRS axis in the frontal plane. While all control group tracings displayed a QRS axis of more than +105°, tracings in five cases of hypoplastic right ventricle demonstrated leftward axis for age.

those of the mitral valve. Study group valve ratios varied from 0.5 to 0.8, while control group ratios varied from 0.97 to 1.8. Statistical analysis revealed that the difference between the mean values of the two groups was highly significant (P = 0.01).

Associated Defects
An atrial septal defect was present in each case. In seven of the eight cases a ventricular septal defect was present. It was located in the membranous septum in four cases, and in the muscular septum in one; in two cases, defects were present in both portions of the septum. Additional lesions included tubular hypoplasia of the aortic arch associated with patent ductus arteriosus (one case), mild subaortic stenosis due to hypertrophy of the crista supraventricularis (two cases), and subpulmonic obstruction secondary to muscular hypertrophy (one case). In another case, subpulmonic obstruction was due to prolapse of tricuspid valvular tissue through a septal defect into the left ventricular outflow tract.

Associated defects encountered in the control group consisted of: patent ductus arteriosus (23 cases), patent foramen ovale (17 cases), membranous (eight cases) and muscular (three cases) ventricular septal defects, subaortic stenosis (two cases), subpulmonic stenosis (two cases), and partial anomalous venous return to the right atrium (one case).

Clinical Data
History
Cyanosis was noted in the first two weeks of life in all patients (table 2). Seven patients developed congestive failure prior to the age of 5 weeks. The diagnosis of complete transposition was established by cardiac catheterization in seven cases. In three instances, hypoplasia of the right ventricle was suspected clinically, and the diagnosis was confirmed by selective angiography. Two patients demonstrated significant clinical improvement following atrial septostomy. Six patients, including the two previously mentioned, expired following pulmonary artery banding or Blalock-Hanlon procedures. The seventh infant died after cardiac catheterization, while the remaining patient expired in severe congestive failure, prior to a planned surgical procedure.

Physical Findings
At the time of admission, all patients were acutely ill and demonstrated varying degrees
**Table 3**

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age at ECG</th>
<th>Electrical axis (°)</th>
<th>P waves</th>
<th>Precordial leads</th>
<th>Hypertrophy pattern</th>
<th>T waves</th>
<th>Inscription of QRS Loop</th>
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<tr>
<td>1</td>
<td>10 da</td>
<td>+10</td>
<td>N</td>
<td>r/s—V3R, V1-V6</td>
<td>LVH</td>
<td>↑ V1-6</td>
<td>CCW CCW CCW CCW</td>
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<tr>
<td>2</td>
<td>10 wk</td>
<td>+120</td>
<td>CAE</td>
<td>r/s—V3R, V1</td>
<td>LVH</td>
<td>↑ V1-5</td>
<td>— — CW Fig. 8</td>
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<tr>
<td>3</td>
<td>2 wk</td>
<td>-5</td>
<td>N</td>
<td>r/s—V3R, V1</td>
<td>LVH Flat V1</td>
<td>↓ V6</td>
<td>CCW CCW CCW CCW</td>
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<td>4</td>
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<td>+75</td>
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<td>LVH</td>
<td>↑ V1-2</td>
<td>— — CW CCW</td>
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<td>qR—V3-6</td>
<td>NL</td>
<td>Bi V1</td>
<td>↑ V2-6 CCW</td>
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<tr>
<td>6</td>
<td>1 da</td>
<td>+75</td>
<td>RAE</td>
<td>r/s—V3R, V1-3</td>
<td>LVH</td>
<td>↑ V1-3</td>
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<td>N</td>
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<td>flat V1</td>
<td>↓ V4-6</td>
<td>CW CCW CCW CCW</td>
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<tr>
<td>8</td>
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<td>+50</td>
<td>N</td>
<td>r/s—V3R, V1-4</td>
<td>LVH</td>
<td>↑ V1-6</td>
<td>CW CCW CCW CCW</td>
</tr>
</tbody>
</table>

Abbreviations: N = normal; CAE = combined atrial enlargement; RAE = right atrial enlargement; LVH = left ventricular hypertrophy; ↑ = upright; ↓ = inverted; F = frontal; H = horizontal; CCW = counterclockwise; CW = clockwise; fig. 8 = figure-of-eight.
of cyanosis, hepatomegaly, and respiratory distress. Auscultation revealed a grade II to III/VI systolic ejection murmur at the second or third left intercostal space accompanied by a single second heart sound. Diastolic murmurs were not recorded.

Electrovectorcardiographic (ECG-VCG) Findings

Study Group (Table 3)

P Wave and P-R Interval. The P-R interval was within normal limits in every case. Right atrial enlargement was present in two cases, and combined atrial enlargement in two cases. Four tracings failed to demonstrate atrial enlargement.

Mean Electrical Axis. The mean electrical axis in the frontal plane varied from $-10^\circ$ to $+125^\circ$ (fig. 6). Leftward axis for age was present in five cases. The electrical axis was considered normal for age in the remaining three cases. Review of vectorcardiograms (five cases) and vectorial analysis (three cases) revealed the rotation of the frontal QRS loop to be in a clockwise direction in five cases and counterclockwise in three cases.

In the horizontal plane, the initial vectors were oriented anteriorly and to the right in a normal fashion. The QRS loop rotated in a counterclockwise fashion in seven cases. In the remaining case, the initial vectors were counterclockwise, but the configuration of the loop was figure-of-eight. The loop was situated to the left in three cases and to the left and posteriorly in five cases.

Precordial Leads. Two basic patterns were encountered in the precordial leads (figs. 7 and 8). In five cases, tracings were characterized by prominent posterior vectors in the

![Figure 7](image_url)

Case 1. (Left panel) Electrocardiogram (of patient at 10 days of age) demonstrating left axis deviation for age, large S waves across the precordium, and a tall R wave in V6 which suggest the interpretation of left ventricular hypertrophy. (Right panel) Cube vectorcardiogram shows counterclockwise rotation of the horizontal and frontal loops, which are oriented posteriorly and to the left. F = frontal; R.S. = right sagittal; H = horizontal. Arrows denote direction of rotation.
mid precordial leads with a tall R in V_{5,6}. Three cases displayed an r/S pattern across the precordium. The R composed 50% or more of the RS in V_6 in six tracings. A Q wave was present in the left precordial leads in four cases.

In lead V_1, six cases displayed an r/S or r/s pattern, while an rSr' configuration was encountered in two cases. The average height of the R wave in V_1 was 0.6 mV. The R composed less than 50% of RS in V_1 in five of the eight cases.

**Hypertrophy Patterns.** In all cases the electrocardiogram was characterized by a relative lack of the right ventricular predominance usually seen during infancy. Six cases demonstrated isolated left ventricular hypertrophy, while tracings were interpreted as probably normal for age in two cases.

**T Waves.** The T waves in lead V_1 were upright in five cases, flat in two, and biphasic in the remaining case. Inversion of T waves in the left precordial leads occurred in four tracings, suggesting the presence of severe left ventricular hypertrophy. Two patients with inversion of T waves in V_6 were receiving digitalis at the time of the tracing.

**Control Group**

The mean electrical axis in the frontal plane varied from +105° to +180° (fig. 6). Review of vectorcardiograms (10 cases) and vectorial analysis (19 cases) demonstrated a clockwise rotation of the frontal plane QRS loop in 28 cases, and a counterclockwise loop in a single case. In the horizontal plane, the QRS loop was rotated clockwise in 26 cases, in a figure-of-eight in two cases, and counterclockwise in a single case. The loop was situated to the right and anteriorly in 26 cases, to the right and posteriorly in one case, and to the left and anteriorly in two cases.

A large R wave was frequently encountered in lead V_1 (25 cases), associated with a large S wave in V_6 (20 cases). Reversal of the adult R/S progression was present in 24 patients. The mean height of the R in V_1 was 1.2 mV. A Q wave was present in V_6 in only four cases.

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**Figure 8**

Case 8. (Left panel) Electrocardiogram (of patient at 1 day of age) demonstrating leftward axis for age, right atrial enlargement, and isolated left ventricular hypertrophy. (Right panel) Frank vectorcardiogram reveals a counterclockwise horizontal loop situated to the left and posteriorly. F = frontal; L.S. = left sagittal; H = horizontal plane. Arrows denote direction of rotation.
### Table 4

**Cardiac Catheterization Data**

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No catheterization was performed in case 2.
Abbreviations: a = atrial a wave; m = mean atrial pressure; % O₂ sat. = % oxygen saturation.
Figure 9
Case 1. Angiogram from the right ventricle demonstrating the hypoplastic chamber (RV) during systole (left panel) and diastole (right panel). The catheter tip is situated close to the ventricular septal defect permitting contrast material to enter both the aorta (Ao) and pulmonary artery (PA). The small chamber is located over the thoracic spine and does not extend laterally beyond the left border of the spine.

Figure 10
Case 5. (Left panel) Right ventricular cineangiogram demonstrating the small ventricular chamber (RV) situated centrally in the cardiac silhouette and giving rise to the ascending aorta (Ao). (Right panel) Diagrammatic representation of angiogram shown in left panel.

Right ventricular hypertrophy was noted in 22 cases and combined ventricular hypertrophy in three. The tracings were considered normal for age in four patients.

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Radiographic Findings
Chest radiographs of the study group cases demonstrated cardiac enlargement with a narrow base and a concave pulmonary artery segment. The interstitial markings of the lung were increased in five cases and decreased in the remaining three. The increased interstitial pattern suggested increased flow in five cases, with additional congestive failure in two cases. Radiographic findings in the control group were not significantly different.

Cardiac Catheterization
Cardiac catheterization was performed in seven study group cases (table 4). While right atrial pressures were increased, in no case was a pressure gradient recorded across the hypoplastic tricuspid valve. Both pressures and oxygen saturations in the left atrium exceeded those in the right atrium. Marked systemic arterial desaturation was present in all cases.

Angiocardiography
Right ventricular angiocardiography was performed in five cases in the study group. In each, a small thick-walled ventricle was demonstrated, which was located centrally in the cardiac silhouette and situated directly over the thoracic spine (figs. 9 and 10). The lateral view disclosed a small crescent-shaped chamber occupying only a small portion of the cardiac diameter. The aorta originated from the right ventricle and was situated in a high anterior position. Left ventricular angiocardiography was performed in six cases, and in each demonstrated a large chamber. In comparison, control group angiocardiograms revealed right ventricular chambers which were equal to, or larger in size than, the corresponding left ventricle.

Discussion
Hypoplasia of the right ventricle has been described both as an isolated lesion and in association with other malformations such as atrial septal defect, ventricular septal defect, tricuspid stenosis, and pulmonic stenosis. A small right ventricular chamber is usually present in tricuspid and pulmonary atresia. Absence of the right ventricular myocardium is associated with a hypoplastic right ventricle in Uhl's anomaly. Hypoplasia of the right ventricle is rarely associated with tricuspid stenosis and complete transposition. To our knowledge, the association of complete transposition and hypoplasia of the right ventricle without tricuspid stenosis has been documented in only five cases, and these accounts consist primarily of postmortem descriptions. This is the first report of complete transposition with hypoplasia of the right ventricle in which a premortem diagnosis has been made based on specific electrovectorcardiographic and angiocardiographic findings. The frequency with which complete transposition with hypoplastic right ventricle has been encountered (17% of the necropsied specimens of this series) is striking and suggests that awareness of this lesion will lead to more frequent diagnosis.

Quantitative analysis of ventricular volumes and atrioventricular valve circumferences indicates that the hypoplastic specimens comprise a separate and distinct form of complete transposition. Comparison of absolute volume measurements between the study and control groups was felt to be of limited significance.

Figure 11
Comparison of volume and valve ratios for both groups. Ventricular volume ratios as determined on necropsy specimens by means of mercury-filled bags are plotted on the horizontal axis. The atrioventricular valve ratios are plotted on the vertical axis. The hypoplastic specimens are closely grouped and clearly separated from the specimens of the control group (P=0.005 for the volume ratios; P=0.01 for the valve ratios).

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because of differences in age and body size of patients, as well as in the degree of cardiac dilatation and postmortem changes. To minimize the effects of these variables, ratios comparing right and left ventricular volumes and tricuspid and mitral valve circumferences were calculated for each specimen (table 1). The atroventricular valve ratios were then plotted against the corresponding ventricular volume ratios (fig. 11). Despite the limitations of this method, a clear separation of the two groups was obtained, confirming our impression of the discrete identity of the hypoplastic specimens.

Despite varying degrees of hypoplasia of the tricuspid valve in each of the study group specimens, no obstruction to blood flow was present. In cases of tricuspid stenosis, valve cusps are described as thickened and often fused, while papillary muscles are short and stubby.8 These findings are in distinct contrast to the appearance of the tricuspid valves in the study group specimens, in each of which individual valve leaflets were normally formed and discrete, despite the small size of the annulus. Cardiac catheterization failed to demonstrate a significant end-diastolic gradient across the tricuspid valve, supporting anatomic evidence that tricuspid stenosis was not present in these cases.

Prominent atrial a waves and marked elevation of right ventricular end-diastolic pressures, which have been described as characteristic of hypoplastic right ventricle,6 were recorded only in case 5 in which the ventricular septum was intact. The course of the cardiac catheter may suggest the presence of a hypoplastic right ventricle. As it passes through the chamber, the catheter occupies a medial position and cannot be advanced laterally toward the cardiac apex.

The history, physical findings, and chest x-rays were not of help in distinguishing the cases of hypoplastic right ventricle from the more common forms of complete transposition. The distinguishing clinical features were encountered in the electrocardiogram, vectorcardiogram, and ventricular angiogram.

Comparison of ECG-VCG data for the two groups revealed significant differences between the control and study group tracings. The electrocardiogram in the infant with complete transposition, as documented in table 5 for the control group and reported by others,18

### Table 5

<table>
<thead>
<tr>
<th>Total cases</th>
<th>Study group</th>
<th>Control group</th>
</tr>
</thead>
<tbody>
<tr>
<td>R, % of total RS in V1</td>
<td>R &gt; 75</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>R &gt; 50</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>R &lt; 50</td>
<td>5</td>
</tr>
<tr>
<td>R, % of total RS in V6</td>
<td>R &gt; 75</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>R &gt; 50</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>R &lt; 50</td>
<td>2</td>
</tr>
<tr>
<td>Ventricular hypertrophy</td>
<td>RVH</td>
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</tr>
<tr>
<td></td>
<td>LVH</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>CVH</td>
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</tr>
<tr>
<td></td>
<td>N</td>
<td>2</td>
</tr>
<tr>
<td>QRS rotation</td>
<td>CW</td>
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<tr>
<td>In frontal plane</td>
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<td>3</td>
</tr>
<tr>
<td></td>
<td>Fig. 8</td>
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</tr>
<tr>
<td>In horizontal plane</td>
<td>CW</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>CCW</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>Fig. 8</td>
<td>1</td>
</tr>
</tbody>
</table>

**Abbreviations:** As in table 3.
usually consists of prominent anterior QRS vectors, suggesting right ventricular hypertrophy, and a mean frontal plane axis greater than +90°. Combined ventricular hypertrophy occurred infrequently in the control group, but has been reported in patients with large communications.\textsuperscript{13-15} In contrast, the study group electrocardiograms demonstrated a relative lack of anterior QRS vectors, which was frequently associated with left ventricular hypertrophy and leftward axis for age. In five cases, the precordial leads revealed prominent leftward and posterior QRS vectors while, in the remaining three instances, QRS vectors were predominantly posterior.

Left ventricular hypertrophy and leftward axis for age are unusual findings in the infant less than 4 months old who has complete transposition. Left axis deviation has been reported in a small infant with complete transposition and an associated left ventricular-right atrial communication.\textsuperscript{13} However, the electrocardiogram in this patient demonstrated combined and predominantly right ventricular hypertrophy.

To our knowledge, isolated left ventricular hypertrophy has been documented in only six small infants with complete transposition. Two of these patients demonstrated ventricular septal defects\textsuperscript{16,17} while three had multiple communications.\textsuperscript{18} In these five cases, left ventricular hypertrophy was associated with a frontal axis of more than +90°. The remaining case, that of a 1-month-old infant with complete transposition and coarctation of the aorta, demonstrated left ventricular hypertrophy and leftward axis for age. However, no assessment of right ventricular volume was documented in any of these patients. We have been unable to find additional reports of complete transposition in an infant less than 4 months of age in whom the electrocardiogram demonstrated both isolated left ventricular hypertrophy and leftward axis for age.

The vectorcardiogram in the infant less than 4 months of age with complete transposition usually demonstrates a clockwise rotation of the horizontal plane QRS loop which is located anteriorly and to the right. This was the finding in 20 of the 29 control group cases (table 5). In older infants with systemic left ventricular pressure, the horizontal plane QRS loop may display counterclockwise rotation to the left with additional large terminal rightward QRS vectors, indicative of combined ventricular hypertrophy.\textsuperscript{17} In seven of the study group cases, QRS loops were inscribed in a counterclockwise direction in the horizontal plane and were situated almost entirely to the left and posteriorly.

Correlation of right ventricular volume and free wall thickness with the magnitude of anterior QRS vectors (table 6) supports previous reports by Gamboa,\textsuperscript{19} Gersony,\textsuperscript{20} and Cole\textsuperscript{21} and their associates. These authors have suggested that the volume of the ventricular cavity is an important factor in the production of right ventricular potentials. Despite thick ventricular walls, a relative lack of anterior QRS vectors was encountered in the study group cases with very small right ventricular volumes.

The measurements of ventricular volume correlated well with the angiographic demonstration of a small right ventricle. In the study group, right ventricular angiography

\begin{table}[h]
\centering
\caption{Correlation of Right Ventricular Volume and Wall Thickness with Electrocardiogram}
\begin{tabular}{|l|l|l|l|l|}
\hline
\textbf{Group} & \textbf{ECG} & \textbf{Cases} & \textbf{Mean volume (cc)} & \textbf{Mean wall (mm)} \\
\hline
Control & RVH & 25 & 1.8 & 0.4 \\
Control & N & 4 & 1.0 & 6.0 \\
Hypoplastic & ↓ RVV, LVH & 6 & 0.7 & 8.0 \\
\hline
\end{tabular}

\textbf{Abbreviations:} RVH = right ventricular hypertrophy; N = normal for age; LVH = left ventricular hypertrophy; ↓ RVV = relative lack of right ventricular QRS vectors.
\end{table}
revealed a small chamber which was situated centrally in the cardiac silhouette and did not extend laterally beyond the left border of the thoracic spine. This configuration is characteristic of hypoplastic right ventricle and may be easily differentiated from the larger right ventricular cavity in patients with more typical forms of complete transposition. While the difference between the two groups is apparent in both frontal and lateral views, the hypoplastic chamber was best demonstrated in the frontal projection.

The following conditions must be considered in the differential diagnosis of complete transposition with hypoplastic right ventricle: tricuspid stenosis or atresia with or without complete transposition, pulmonary atresia with intact septum, and common ventricle. The electrocardiogram in tricuspid or pulmonary atresia may be identical with the pattern encountered in five of the study group cases, with an adult-type RS progression across the precordium and a tall R wave in lead V6 (fig. 8). In the remaining three study group cases, the electrocardiogram was similar to that which may be encountered in common ventricle, with an r/S pattern across the precordium. While associated clinical findings may help to limit the differential diagnosis, in the final analysis cardiac catheterization and angiography must be performed to rule out other conditions. When cardiac catheterization confirms the diagnosis of complete transposition and the ECG-VCG findings suggest the presence of a hypoplastic right ventricle, right ventricular angiography should be performed. Furthermore, adequacy of the right ventricle should be evaluated in any patient with complete transposition in whom complete correction is contemplated.

Early management of cases of complete transposition and hypoplastic right ventricle is the same as that in typical cases of complete transposition; that is, creation of an atrial septal defect by surgical intervention or by balloon septostomy. The prognosis in cases of complete transposition is apparently less favorable when hypoplasia of the right ventricle is present. The early demise of each of the study group patients suggests that the hypoplastic right ventricle may be unable to function as a systemic chamber for extended periods of time.

References


Transposition of the Great Vessels with Hypoplasia of the Right Ventricle

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