Tricuspid Atresia with Increased Pulmonary Blood Flow
An Analysis of 13 Cases

By Bertrand A. Marcano, M.D., Thomas A. Riemenschneider, M.D.,
Herbert D. Ruttenberg, M.D., Stanley J. Goldberg, M.D.,
and Michael Gyepes, M.D.

SUMMARY
The pathologic and clinical findings in 13 cases of tricuspid atresia and increased pulmonary blood flow encountered in UCLA Medical Center were reviewed. In seven, the great vessels were normally related (group I), and in six, the great vessels were transposed (group II). In all group II cases, aortic arch anomalies were encountered, a finding not duly emphasized in previous reports. Comparison of the two groups revealed significant clinical and laboratory differences. Group II patients typically presented with severe congestive heart failure and expired within the first 2 months of life. Group I patients usually presented with mild congestive heart failure which responded to therapy. Five of the latter demonstrated progressive change from increased to decreased pulmonary blood flow during their clinical course and eventually required palliative shunt procedures, four prior to the age of 2 years and one at 5 years of age.

While the electrocardiogram demonstrated right atrial enlargement and left ventricular hypertrophy in both groups, the mean frontal plane axis was between $-15^\circ$ and $-100^\circ$ in group I and $-75^\circ$ and $+75^\circ$ in group II. QRS vector loops tended to be superiorly oriented in group I and inferiorly oriented in group II.

Because the natural history and prognosis in these two groups differ significantly, diagnostic efforts should include determination of the relationship of the great vessels. When transposition of the great vessels is demonstrated, aortography should be performed because of the high incidence of aortic arch anomalies.

Additional Indexing Words:
Transposition of great vessels
Left ventricular hypertrophy
Cardiac catheterization
Right atrial hypertrophy
Electrocardiogram
Angiocardiography
Necropsy findings
Aortic arch anomalies

Tricuspid atresia is usually associated with decreased pulmonary blood flow. The association of this entity with increased pulmonary blood flow is not uncommon, although it has been documented infrequently. This report analyzes our experience with 13 children in whom tricuspid atresia was associated with increased pulmonary blood flow. These cases were divided into two categories depending on the relationship of the great vessels. The purpose of this report is to emphasize the distinguishing pathologic anatomy, clinical findings, and laboratory data of these two subgroups, since differentiation of the two groups is important both for optimum clinical management and in the ultimate prognosis.

Methods
The 13 patients with tricuspid atresia and increased pulmonary blood flow were admitted to UCLA Medical Center between 1957 and 1967.
Diagnosis was confirmed at cardiac catheterization in 11 cases and at necropsy in six. Tricuspid atresia is herein defined anatomicall as that condition in which no tricuspid orifice and usually no recognizable tricuspid valvular tissue are present. Instead of a tricuspid orifice, there is a dimple or localized fibrous thickening in the floor of the right atrium at the expected location of the valve. The clinical diagnosis was based on the following: (a) presence of cyanosis except in those cases with increased flow when it may be mild or absent; (b) presence of cardiomegaly, dyspnea, tachypnea, tachycardia, and hepatomegaly indicative of congestive heart failure; (c) presence of systolic and diastolic murmurs; and (d) electrocardiographic evidence of left ventricular hypertrophy, with relative absence of right ventricular forces and left axis deviation in the frontal plane. The diagnosis can be confirmed at cardiac catheterization if repeated attempts to pass a catheter into the right ventricle are unsuccessful, if the natural course of the catheter is from the right atrium to the left atrium and the left ventricle and if angiography reveals that no contrast material traverses the tricuspid valve into the right ventricle. The following clinical criteria were used to establish the diagnosis of tricuspid atresia with increased pulmonary blood flow: (a) evidence of increased vascularity or pulmonary venous congestion on chest roentgenogram, or both; (b) it is important to realize that the occurrence of pulmonary venous congestion and heart failure in an infant with tricuspid atresia, in whom cardiac catheterization and cineangiography fail to reveal any abnormality of the mitral valve, or additional left-sided lesions affirms the presence of increased pulmonary blood flow; (b) cyanosis which is clinically mild or absent; and (c) cardiac catheterization data indicating increased pulmonary-to-systemic flow ratio (> 1:1).*

If clinical findings and cardiac catheterization established a diagnosis of tricuspid atresia and if in addition the foregoing criteria for increased pulmonary blood flow were all satisfied, then the diagnosis of tricuspid atresia with increased pulmonary blood flow was considered beyond reasonable doubt, and all such cases were included in this study.

These patients were followed for varying periods of time, during the course of which some manifested evidence of a change from increased to decreased pulmonary blood flow. Diagnosis of decreased pulmonary blood flow was based on the following criteria: (a) evidence of a decrease in pulmonary vascularity or venous congestion on several chest roentgenograms, or both, and (b) significant increase in clinical cyanosis as determined by repeated clinical evaluations.

The patients were divided into two groups. Group I consisted of seven patients whose present ages are 2 months to 9 years in whom the relationship of the great vessels was normal. Group II consisted of the remaining six patients, aged 3 days to 10 months in whom the great vessels were transposed. Thirteen-lead electrocardiograms were available for all patients. Frank vectorcardiograms were available in four patients of group I and in one patient of group II. Cube vectorcardiograms were available in only two patients, both of whom were in group I. In those cases in which vectorcardiograms were unavailable, horizontal and frontal plane QRS vector loops were plotted from electrocardiographic tracings. While exact vectorcardiograms cannot be constructed by this method, a reasonable approximation of the orientation and rotation of the frontal and horizontal loops can be made. Chest roentgenograms in the frontal and lateral positions were available for review in each patient. Cardiac catheterization with selective angiography was performed in all cases of group I and in four cases of group II. Necropsy specimens were available in a single case in group I and in five cases in group II.

Results

Clinical data will be presented separately for each group, while necropsy findings will be presented together.

Group I: Tricuspid Atresia with Normally Related Great Vessels (Seven Cases)

History

Mild cyanosis was noted within the first 2 months of life in two patients and occurred after 6 months of life in four. Cyanosis was not present in the remaining patient (table 1). In five of the six patients who developed congestive heart failure, symptoms appeared

*Pulmonary blood flow ratio = systemic saturation (left ventricle or aorta) / pulmonary venous saturation — mitral venous saturation.

The pulmonary vein saturation was assumed to be 96%. The pulmonary artery saturation was assumed to be equal to systemic saturation because adequate mixing occurs in the left ventricle.
before the age of 2 months and in the remaining patient at age 15 months. The diagnosis in each case in group I was established by cardiac catheterization. Five patients demonstrated progressive change from increased to decreased pulmonary blood flow during their clinical course, and four eventually required palliative shunt procedures prior to the age of 2 years and the fifth at 5 years of age.

Four of the six patients who manifested congestive heart failure in infancy later had a decrease in pulmonary blood flow as did the single patient in whom congestive failure did not occur. All five patients who required an aortopulmonary shunt procedure demonstrated clinical improvement. Of the two patients who did not require any form of palliative procedure, one died of purulent meningitis at the age of 2 months. The other is alive at 1 year of age and has experienced several episodes of congestive heart failure which have responded to medical therapy.

**Physical Findings**

At the time of their first admission, physical examination revealed parasternal systolic thrills and systolic murmurs in all patients. These were either holosystolic or of short duration, beginning with the first heart sound, maximal at the mid left sternal border and of grade II to III/IV intensity. Apical diastolic murmurs were recorded in three of the seven patients. The second heart sound was closely split in all seven, but in four, the pulmonary component was of increased intensity. Flush blood pressures were equal in the upper and lower extremities. Signs of mild congestive heart failure as evidenced by dyspnea, tachy-

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**Table 1**

**Group I: Clinical Data**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age* (yr)</th>
<th>Sex</th>
<th>Age of onset (mo)</th>
<th>Cyanosis</th>
<th>CHF</th>
<th>Thoracic roentgenograms</th>
<th>Surgical procedure</th>
<th>Present status</th>
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<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Age (mo)</td>
<td>Cardiac size</td>
<td>Age (mo)</td>
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<td>↑</td>
<td>↑</td>
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<td>M</td>
<td>2</td>
<td>1</td>
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<td>M</td>
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<td>↑</td>
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<tr>
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<td>↑</td>
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<td>M</td>
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<tr>
<td>7</td>
<td>9</td>
<td>M</td>
<td>14</td>
<td>2</td>
<td>3</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
</tr>
</tbody>
</table>

* Present age.
† Difficulty controlling congestive heart failure.
‡ Died of purulent meningitis at 2 mo of age.

Abbreviations: CHF = congestive heart failure; ↑ = increased, ↓ = decreased

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**Figure 1**

The mean frontal axis in cases of tricuspid atresia with excessive pulmonary blood flow. In those cases with normally related great arteries, the axis varies from $-15^\circ$ to $-100^\circ$, while in those with transposition of the great vessels, the axis varies from $-75^\circ$ to $+75^\circ$. The symbols represent:

- ● - With normally related great arteries
- ▲ - With transposition of great vessels
pnea, and hepatomegaly were present in six of the seven patients.

Electrocardiographic and Vectorcardiographic (ECG-VCCG) Findings

In six cases, the electrocardiogram revealed right atrial enlargement, while combined atrial enlargement was demonstrated in the remaining case.

The mean electrical QRS axis in the frontal plane varied from $-15^\circ$ to $-100^\circ$ (fig. 1). A counterclockwise rotation and superior orientation of the frontal QRS loop was found in the vectorcardiogram.

In the horizontal plane, the QRS loop rotated in a clockwise direction in three cases. In one case, the initial rotation was clockwise changing to counterclockwise, while in the remaining three cases, the rotation was counterclockwise. The orientation of the QRS loop was mainly posterior in all cases. In three cases, the initial cardiac vectors were anterior and leftward. In two of these, the rotation was clockwise, while in the remaining case, the rotation was counterclockwise. In four cases, the initial vector was anterior and rightward. In two of these, the rotation was counterclockwise, and in the remaining two cases it was clockwise.

The precordial leads demonstrated isolated left ventricular hypertrophy in all cases. All seven cases displayed an r/S pattern in the right precordial leads with a variable pattern in the left precordial leads. The mean RV$_1$ amplitude was 4.8 mm (range, 1 to 7 mm), while the mean SV$_1$ amplitude was 23.4 mm (range, 3 to 64 mm). The mean RV$_6$ amplitude was 16.8 mm (range, 6 to 26 mm).

All seven patients demonstrated upright T waves in V$_1$ while T waves in lead V$_6$ were upright in five cases and inverted in two. One of the latter patients was receiving digitalis at the time of the tracing.

Radiographic Findings

Initial chest roentgenograms demonstrated cardiac enlargement in every case. In four cases, the cardiac configuration suggested right ventricular enlargement with lifted apex. The pulmonary conus was normal in five cases and concave in two. The pulmonary vascular markings were increased in all cases in a manner consistent with increased pulmonary blood flow (fig. 2). The increased pulmonary vascular markings were considered not typical of pulmonary edema or pulmonary venous congestion. In those patients who developed clinical cyanosis later in their clinical course, chest roentgenograms demonstrated a decrease in pulmonary vascular markings when compared to repeated previous evaluations. Cardiomegaly, however, persisted in all cases.

Cardiac Catheterization

Cardiac catheterization was performed in each patient (table 2). Both right and left atrial pressures were increased in four cases. Right atrial pressure exceeded left atrial pressure to a varying degree (1 to 10 mm) in four cases, and atrial pressures were equal in the remaining case. Calculated pulmonary-to-systemic flow ratio was increased in every case (range, 1.5:1 to 5.8:1). Although the calculation of such flow ratios in this anomaly is subject to considerable error, they are presented as an approximation. Systemic desaturation was present in all cases.
Table 2

Group I: Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Case</th>
<th>Age* (mo)</th>
<th>Right atrium</th>
<th>Left atrium</th>
<th>Left ventricle</th>
<th>Ascending aorta</th>
<th>Inferior vena cava</th>
<th>Superior vena cava</th>
<th>Qp/Qs†</th>
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</table>

* Age at catheterization.
† Pulmonary/systemic flow ratio.

Abbreviations: a = atrial a wave; v = atrial v wave; m = mean atrial pressure.

Angiocardiography

The diagnosis was confirmed by angiocardiography in all cases. Following injection of contrast material into the right atrium, the left atrium and left ventricle filled sequentially. No contrast material was observed to enter the right ventricle, thus leaving an unopacified triangular area or window where this structure is normally situated. With left ventricular contraction, the ascending aorta and pulmonary artery filled and contrast material could be seen entering a hypoplastic right ventricular infundibulum through a ventricular septal defect (fig. 3). Left ventricular angiocardiography demonstrated a large posterior chamber from which the ascending aorta arose. In most cases, the left ventricular cavity extended to the anterior border of the heart in the lateral or left anterior oblique projection.

Group II: Tricuspid Atresia with d-Transposition of the Great Vessels (Six Cases)

History

Mild cyanosis was noted in all six patients within the first 3 months of life (table 3). All developed congestive cardiac failure prior to the age of 2 months. Congestive cardiac failure was deemed moderately severe in all patients. The correct diagnosis was established by cardiac catheterization in four patients. In the other two, clinical diagnosis was confirmed at necropsy. Two patients expired shortly after pulmonary artery banding. The remaining four infants died in severe congestive heart failure. Death occurred early in this group:

Figure 3

Antero-posterior angiocardiogram (group I; case 6) after injection into the right atrium (RA). The contrast material has passed through the atrial septal defect into the left atrium (LA), left ventricle (LV), and aorta (Ao). The arrow points to the right ventricular "window." (LAA = left atrial appendage).
five patients died by 2½ months of age and the sixth patient died at 10 months.

**Physical Findings**

At the time of admission, all six patients were in acute distress with severe cardiac decompensation, as evidenced by dyspnea, tachypnea, tachycardia, and marked hepatomegaly. A systolic thrill was felt at the left parasternal area in all six cases. Auscultation revealed a grade II to III/V holosystolic murmur at the upper left sternal border. A short, low-pitched diastolic murmur was heard at the lower left sternal border in two patients. The second sound was narrowly split in two cases and single in four cases. Femoral pulses were considered normal in two cases and weak in the remaining four cases. The flush blood pressure in the upper extremities exceeded that in the lower extremities by more than 20 mm Hg in four of the six cases.

**ECG-VCG Findings**

The electrocardiogram demonstrated right atrial enlargement in all six cases.

The mean electrical axis in the frontal plane varied from $-75^\circ$ to $+75^\circ$ (fig. 1). Leftward axis for age was present in all six cases. Vectorcardiographic data revealed the frontal QRS loop to be a counterclockwise direction in all patients. In the horizontal plane, the QRS loop rotated in a counterclockwise direction in five cases, while in one case it was initially counterclockwise and then clockwise. The QRS loop was mainly posterior in all cases.

The precordial leads in all cases demonstrated isolated left ventricular hypertrophy.

The predominant pattern in the right precordial leads was an r/S configuration present in four cases. In one of the remaining two cases, a qS configuration was present; in the other a qRs pattern was evident. The r wave in V1 averaged 2.5 mm (range, 1 to 6 mm), while the mean SV1 amplitude was 14.6 mm (range, 7 to 32 mm). The mean RV6 amplitude was 24.5 mm (range, 10 to 54 mm). In the left precordial leads, three patients demonstrated a qRS configuration, and one demonstrated a qR pattern; no q wave could be identified in the tracings of the remaining two patients.

In five cases, the electrocardiogram was characterized by a relative lack of right ventricular vectors while in one case, the right ventricular electrical vectors were considered normal. The initial cardiac vectors were normal in all patients.

**Roentgenographic Findings**

Chest roentgenograms were obtained on all six patients and demonstrated cardiac enlargement with a concave pulmonary artery segment and narrow base. The pulmonary vascular markings were increased in all patients (fig. 4).

**Cardiac Catheterization**

Cardiac catheterization was performed on four patients in this group. Right atrial pressure was increased in four and exceeded left atrial pressure in all cases. Oxygen desaturation of systemic blood was mild in

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**Table 3**

**Group II: Clinical Data**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at death (mo)</th>
<th>Sex</th>
<th>Onset of cyanosis (days)</th>
<th>Onset of CHF (days)</th>
<th>Thoracic roentgenogram</th>
<th>Surgical procedure</th>
<th>At age (mo)</th>
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<td>1 ½</td>
<td>M</td>
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<td>↑</td>
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<td>↑</td>
<td>Pulmonary banding</td>
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<td>21</td>
<td>21</td>
<td>↑</td>
<td>↑</td>
<td>Pulmonary banding</td>
</tr>
<tr>
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<td>5/30</td>
<td>F</td>
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<td>2</td>
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<td>↑</td>
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<td>↑</td>
<td>-</td>
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</table>

Abbreviations: CHF = congestive heart failure; ↑ = increased.
The diagnosis was confirmed by angiocardiography in each of the four patients who underwent cardiac catheterization. Following injection of contrast material into the right atrium, the left atrium and left ventricle filled sequentially. No contrast material was seen to enter the area of the right ventricle leaving an unopacified triangular area where this structure is normally situated. With left ventricular contraction, the ascending aorta and pulmonary artery filled, and contrast material could be seen entering a hypoplastic right ventricular chamber through a ventricular septal defect. Left ventricular angiocardiography was performed in all four cases and demonstrated a large chamber from which a markedly dilated pulmonary artery took origin posteriorly. The anteriorly placed small aorta arose from the hypoplastic right ventricle and invariably opacified simultaneously with the pulmonary artery. Angiography from the ascending aorta was performed in three patients in the lateral or left anterior oblique projection and in each case demonstrated coarctation of the descending aorta. Furthermore, in one of these patients, tubular hypoplasia of the ascending aorta was an additional lesion.

**Pathologic Anatomy (Table 5)**

There was one death in group I and six deaths in group II. Necropsy specimens from the single case in group I and five cases in group II were available for review.

**Table 4**

<table>
<thead>
<tr>
<th>Group II: Cardiac Catheterization Data</th>
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<td><strong>Case</strong></td>
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* Age at catheterization.
† Pulmonary/systemic flow ratio.
Abbreviations: a = atrial a wave; v = atrial v wave; m = mean atrial pressure.
### Table 5

**Groups I and II: Pathologic Findings**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at death (mo)</th>
<th>Ventricular hypertrophy pattern</th>
<th>Semilunar valve ring diameter (mm)</th>
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<td>LV</td>
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<td>Hypertrophy</td>
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<td>4</td>
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<td>Hypertrophy</td>
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<td>15</td>
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<td>Hypertrophy</td>
<td>6</td>
<td>12</td>
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<td>Musc. Normal Normal Preductal coarctation of aorta; juxtaposition of atrial appendage</td>
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<td>Hypertrophy</td>
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<td>Hypertrophy</td>
<td>6</td>
<td>12</td>
<td>2</td>
<td>Musc. Normal Normal Interruption of aortic arch</td>
</tr>
</tbody>
</table>

Abbreviation: musc. = muscular.
External Configuration of the Heart and Great Vessels

In both groups, 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. In all cases, the right atrium was enlarged, and in one case of group II there was a juxtaposition of the atrial appendages. In the single case of group I, the great vessels were normally related and the coronary artery patterns were normal. In group II cases, the aorta arose from the right ventricle and was situated to the right and anterior to the pulmonary artery in an oblique fashion. The coronary artery pattern was abnormal in all four cases with the right coronary artery arising from the right posterior aortic sinus and the left coronary sinus originating from the left posterior aortic sinus. The anterior aortic sinus was noncoronary.

Size of Ventricles

In all specimens, the right ventricle was hypoplastic and the left ventricle was enlarged. The right ventricular infundibulum was a long narrow structure in all, and in only one (case 2, group I) did it appear restrictive. The left ventricular outflow tract appeared normal in all cases. The left atrium was enlarged in all cases.

Size and Appearance of Semilunar Valves

The pulmonary and aortic valves appeared normal in both groups. In group II cases, the pulmonary valve ring measured 12 to 15 mm in diameter, whereas the aortic valve ring measured 4 to 6 mm in diameter. Thus, the pulmonary valve ring was 2 to 3.7 times the size of the aortic valve ring in all these cases. In the group I case, the aortic and pulmonary valve rings each measured 10 mm in diameter.

A-V Valves

The tricuspid valve was atretic, and the mitral valve appeared dilated in all of our cases.

Septal Defects

In each case, a large atrial septal defect of the secundum type and a small ventricular septal defect were present. The latter defect ranged in size from 2 to 5 mm in diameter and was encircled by muscular tissue. In each case, the size of the ventricular septal defect appeared restrictive and probably caused obstruction to egress of blood from the left ventricle into the right ventricular outflow tract. The ratio of the diameter of the ventricular septal defect to that of the semilunar valves is shown in table 5. The ratio of the ventricular septal defect to the pulmonary valve ring ranged from 1:6 to 1:2, and that of the ventricular septal defect to the aortic valve, from 1:2 to 5:6.

Associated Anomalies

While the aortic arch was normal in the group I case, aortic arch anomalies were present in all necropsied cases of group II. In two cases, tubular hypoplasia of the aortic arch was associated with preductal coarctation. In two other cases, preductal coarctation alone was encountered, while in the fifth case, complete interruption of the aortic arch was present. A patent ductus arteriosus was present in three cases.

Discussion

Tricuspid atresia has a reported prevalence of 1 to 5% of all cases of congenital cardiac anomalies, studied at necropsy or 2 to 3% of clinical cases. Anatomically, the entity can be classified into two main types: cases of normal relationship of the great vessels and those with transposition of the great vessels. Each type can be further subdivided on the basis of the presence or absence of pulmonary stenosis or pulmonary atresia and the size of the ventricular septal defect. Group I and group II cases described in this report can be classified respectively as type Ic and type IIc according to Edwards and Burchell and as modified by Keith and associates. Type Ic is encountered in approximately 9% of all cases of tricuspid atresia studied at necropsy and accounts for 12% of all cases of tricuspid atresia without transposition of the great vessels. Of 32 cases of tricuspid atresia seen at UCLA Medical Center from 1957 to 1967, seven (22%) exhibited normally related great arteries and increased pulmonary blood flow. Thus, our
prevalence rate is twice the reported incidence. This may be due to the fact that these patients frequently develop decreasing pulmonary blood flow with time, and this may not have been taken into account by some authors.

The reported incidence of type IIc cases is approximately 18% of all postmortem cases of tricuspid atresia. This subtype is most commonly encountered in association with transposition of the great vessels. We have located 32 similar cases in the world literature. The six cases of this report represent the largest group reported in a single series.

Of particular interest in our cases is the presence of aortic arch anomalies in all cases of transposition of the great vessels. The association of arch anomalies with tricuspid atresia has not been emphasized in previous reports. We have found only 15 instances of this entity previously reported in the world literature.

The presence of aortic arch anomalies in all group II cases, and in addition, the presence of patent ductus arteriosus in three cases of this group make assessment of increased pulmonary blood flow more difficult in these patients. However, the pulmonary blood flow is increased in patients with transposition of the great vessels without pulmonary or subpulmonary stenosis and without high pulmonary vascular resistance. The same circumstances apply in tricuspid atresia with transposition of the great vessels. It is possible that some shunting does take place from the aorta via the patent ductus to the pulmonary artery. However, as was stated previously, the ventricular septal defect and the aortic valve ring in every case were much smaller than the pulmonary valve ring. Under these circumstances, it is reasonable to assume that most of the flow from the left ventricle occurred through the pulmonary artery, and a much smaller amount traversed the ventricular septal defect and was ejected into the aorta. On the basis of the anatomic findings, therefore, one can anticipate that the shunt through the patent ductus contributed only a small amount to pulmonary blood flow in the cases included in this report.

Comparison of the two groups reported herein reveals significant clinical and laboratory differences. All infants described in this report demonstrated minimal cyanosis in the first few months of life. At the time of admission to the hospital, however, those patients with tricuspid atresia and transposition of the great vessels (group II) had a rapid onset of severe congestive cardiac failure. This is in contrast to those patients with tricuspid atresia and normally related great arteries (group I) who typically presented with mild or no distress. By 2 months of age, most of the group II infants had expired. The group I patients did relatively well for a few months before increasing cyanosis and signs of respiratory distress developed. Thus, palliative shunt procedures were not required during the first year of life in four of the five patients in group I because of slow progression of symptoms (table I).

Analysis of the ECG-VCG findings in the two groups of patients also suggests certain differences which may be of help in distinguishing these two entities. The mean electrical axis in the frontal plane in group I cases is more leftward than that usually seen in group II cases. Thus, in all of our group I cases, the frontal axis was $-15^\circ$ to $-100^\circ$ whereas in four of our six cases of group II, the axis was $+15^\circ$ to $+75^\circ$. In addition, analysis of the vectorcardiographic data obviously reveals that in all cases of normally related great arteries QRS loops were superiorly oriented in the frontal plane. In the cases of tricuspid atresia and transposition of the great vessels, the QRS loop in the frontal plane was usually inferior and leftward oriented. In the horizontal plane, the QRS loop was usually posterior, but in three cases of group I, abnormalities of the initial cardiac vectors were evident. In two of the three cases, the initial cardiac vectors were anterior and rightward with clockwise rotation, whereas in the remaining case, the initial cardiac vector was anterior and leftward with clockwise rotation. Initial cardiac vectors were normal in group II cases.
changes in the initial vectors have previously been reported to occur in conditions characterized by anomalies of the A-V valves, common ventricle, anomalous left coronary artery, and conditions characterized by overwork and hypertrophy of the left ventricle.  

Thus, in patients with tricuspid atresia and increased pulmonary blood flow, it may be possible to suspect the relationship of the great vessels on the basis of the ECG-VCG findings. As Gamboa and others have pointed out however, there may be significant variability in the electrocardiogram of infants with tricuspid atresia and transposition of the great vessels.

The natural history and prognosis of these two groups of patients differ significantly. In our group II patients with transposed vessels, there has been 100% mortality. The significantly better prognosis demonstrated by group I is inconsistent with the findings of Astley and co-workers, but in agreement with the report of Edwards.

As mentioned previously, patients with normally related great vessels may do well clinically during the first several months of life. Infants who do develop chronic congestive heart failure may later show improvement in clinical status associated with decreasing pulmonary blood flow as demonstrated by chest roentgenograms and suggested by disappearance of the apical diastolic murmur. However, pulmonary blood flow may continue to decrease leading to increasing cyanosis and respiratory difficulty as occurred in five patients in group I (table 1). The change in clinical status of group I children is related to decreasing pulmonary blood flow. The nature of the anatomic changes that produce this decrease in pulmonary blood flow has been the subject of some speculation. The change in condition may be due to a closing ventricular septal defect or progressive infundibular pulmonary stenosis. A closing ventricular septal defect has been documented by several authors and is probably the most common reason for clinical deterioration.

Because the prognosis differs significantly between these two groups of patients, all patients with tricuspid atresia and increased pulmonary blood flow require early evaluation of the associated cardiovascular anomalies. Since the presence of aortic arch anomalies and transposition of the great vessels alters the prognosis significantly, an attempt must be made to determine the relationship of the great vessels. In those cases in which transposition of the great vessels is encountered, retrograde aortography may be necessary to evaluate the aortic arch.

Surgical intervention seems indicated in two specific situations encountered during the clinical course of this entity. When increasing cyanosis and respiratory difficulty are caused by a decrease in pulmonary blood flow, either an aortopulmonary or vena caval-pulmonary artery anastomosis should be performed. Recently, use of a balloon catheter for atrial septostomy at the time of cardiac catheterization followed by anastomosis of the ascending aorta to the right pulmonary artery has been successful. In infants with excessive blood flow, refractory congestive heart failure, and recurrent pneumonia, a pulmonary banding procedure may be of help. Despite loss of two of our patients following this procedure, pulmonary artery banding would appear to be the procedure of choice at the present level of our knowledge. Patients in whom aortic arch anomalies are present may require surgical correction of the coarctation to improve their prognosis.

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Tricuspid Atresia with Increased Pulmonary Blood Flow: An Analysis of 13 Cases
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