Observations on Changing Hemodynamics in Tricuspid Atresia Without Associated Transposition of the Great Vessels

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SUMMARY
Three patients are discussed who, in early infancy, presented with mild cyanosis, cardiac decompensation, failure to thrive, and pulmonary congestion. These manifestations were associated with tricuspid atresia without transposition of the great vessels. The clinical course of these children, who in varying periods of time became progressively more cyanotic, with hearts becoming smaller, and with decreasing pulmonary blood flow, is unique. Cardiac catheterization and angiocardiographic studies were obtained at each end of the clinical spectrum in all of the children. Although a closing ventricular septal defect is thought to be the cause of the decreasing pulmonary flow, the studies were not conclusive. In all three patients, palliative systemic venous-to-pulmonary arterial anastomosis (Glenn procedure) relieved the symptoms of hypoxia.

Additional Indexing Words:
Cyanoses Pulmonary congestion Cardiac decompensation
Congenital heart disease Glenn procedure

Tricuspid atresia has been classified into two anatomic groups: type I, with normal position of the great arteries and type II, with transposed great arteries.1 Each type has been further subdivided by Keith and associates2 into three subgroups according to the degrees of pulmonary stenosis and the size of the ventricular septal defect. Type Ic, defined as tricuspid atresia with a large ventricular septal defect and normal or large pulmonary artery, is the only category without transposition of the great vessels in which increased pulmonary blood flow occurs.

Although type Ic accounts for about 12% of the reported cases of tricuspid atresia without associated transposition, it rarely has been reported as a cause of massive cardiac enlargement, pulmonary congestion, and mild, if any, apparent cyanosis.3,4 Over the past 5 years, four infants of a total of 18 known to have tricuspid atresia without transposition have presented in this manner. Of greater interest, however, is the change in these clinical manifestations over varying periods to those more characteristic of tricuspid atresia with a normal-sized heart, decreased pulmonary blood flow, and deep cyanosis in three of the four children. No previous report could be found of tricuspid atresia with such marked changes in clinical status.

Reports of Cases
Case A
J. W. was admitted to Children's Hospital of Los Angeles for the first time on March 2, 1960 at 2% months of age because of increasing dyspnea and cough unaccompanied by fever. Previous to this he had been asymptomatic. A heart murmur had been noted at 1 month. Cyanosis had not been noted by family or physician.

The infant was in moderate respiratory distress with tachycardia. A systolic thrill with grade III, harsh pansystolic murmur along the left sternal border and hepatomegaly were noted. Cyanosis was not apparent. The hemoglobin was 14.5 gm%. The electrocardiogram showed left axis deviation and left ventricular hypertrophy (fig. 1). The roentgenogram showed a grossly enlarged heart and pulmonary vascular engorge-
Figure 1

Electrocardiograms of the three patients.

Figure 2

Chest roentgenograms of patient A. (Left) First admission. (Right) Before operation.
Table 1

Summary of Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>SVC</th>
<th>RA</th>
<th>LA</th>
<th>LV</th>
<th>Sys. A</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>O₂ saturation (%)</td>
<td>Pressure (mm Hg)</td>
<td>O₂ saturation (%)</td>
<td>Pressure (mm Hg)</td>
<td>O₂ saturation (%)</td>
</tr>
<tr>
<td>A</td>
<td>2½ mo</td>
<td>60</td>
<td>—</td>
<td>66</td>
<td>10</td>
<td>94</td>
</tr>
<tr>
<td></td>
<td>3⅓ yr</td>
<td>59</td>
<td>—</td>
<td>63</td>
<td>4</td>
<td>72</td>
</tr>
<tr>
<td></td>
<td>3⅓ mo</td>
<td>36</td>
<td>—</td>
<td>45</td>
<td>12</td>
<td>68</td>
</tr>
<tr>
<td>B</td>
<td>14 mo</td>
<td>53</td>
<td>—</td>
<td>53</td>
<td>10</td>
<td>68</td>
</tr>
<tr>
<td></td>
<td>2 wk</td>
<td>68</td>
<td>—</td>
<td>77</td>
<td>5</td>
<td>88</td>
</tr>
<tr>
<td>C</td>
<td>4 mo</td>
<td>44</td>
<td>—</td>
<td>47</td>
<td>4</td>
<td>81</td>
</tr>
<tr>
<td></td>
<td>18 mo**</td>
<td>—</td>
<td>—</td>
<td>60</td>
<td>3</td>
<td>71</td>
</tr>
</tbody>
</table>

*Mean pressure. †Probable pulmonary vein. §Thought to be right ventricle. §Prolonged crying.

**12 months following the Glenn procedure.
Abbreviations: SVC = superior vena cava; RA = right atrium; LA = left atrium; LV = left ventricle; and Sys. A = systemic artery.

Case B

T. C., a boy, was admitted at 3½ months of age on August 10, 1964, because of a 3-day history of vomiting and rapid breathing. Cyanosis was noticed by family physicians except for a questionable thickness of the skin. The baby was admitted to the hospital for a hemocrit of 46%. The electrocardiogram was normal, and the heart rate was sinusoidal. A thrill and grade IV murmur were noted. A loud, systolic murmur and liver were not enlarged. The electrocardiogram was normal in size, with normal right axis deviation. The electrogram showed a heart rate of 148 bpm.

The patient was admitted on January 1, 1965, because of palpitations and cyanosis. A thrill and grade IV murmur were noted. A loud, systolic murmur and liver were not enlarged. The electrocardiogram was normal in size, with normal right axis deviation. The electrogram showed a heart rate of 148 bpm.

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A clinical diagnosis of tricuspid atresia probably with transposition of the great vessels and congestive cardiac failure was made and appropriate treatment was instituted. Cardiac catheterization with cineangiography was done on August 11, 1964 (table 1). The diagnosis of tricuspid atresia with a ventricular septal defect was confirmed. Transposition of the great vessels was not present (fig. 4).

At 14 months of age on July 6, 1965, the child was readmitted for repeat cardiac catheterization studies because of increasing cyanosis and dyspnea on exertion. Generalized cyanosis was obvious. There was no evidence of congestive...
failure and the thrill and systolic murmur were unchanged. His hemoglobin was now 18.7 gm% with a hematocrit of 56%. The chest roentgenogram showed a heart of normal size with diminished pulmonary vasculature (fig. 3). The repeat catheterization studies included an arterial oxygen saturation of 55% (table 1).

On July 6, 1965, a Glenn procedure was performed and the atrial septal defect was enlarged. One month following surgery left chylothorax was diagnosed. This was extensive enough to produce marked respiratory difficulty. The left thorax was drained and the chyle did not re-accumulate. The youngster has since been free of symptoms.

Case C

T. M. was admitted at 15 days of age on December 16, 1964 because of vomiting, poor feeding ability, and a heart murmur.

Slight cyanosis was noted only with crying. A systolic thrill and loud harsh pansystolic murmur
were present maximally at the fourth left intercostal space. No diastolic murmur was noted. There was minimal hepatomegaly. The child was irritable. The hemoglobin was 17.9 gm% with a hematocrit of 52%. The electrocardiogram (fig. 1) showed left axis deviation with left ventricular hypertrophy. The roentgenogram showed a grossly enlarged heart with bilateral pulmonary vascular engorgement.

On the basis of the experience of the preceding two cases, a clinical diagnosis of tricuspid atresia was made. On December 17, 1964, he underwent cardiac catheterization and cineangiographic studies (table 1). The diagnosis of tricuspid atresia without transposition of the great vessels was confirmed (fig. 5). The resting arterial oxygen saturation was 93%. The pulmonary artery was large as was the ventricular septal defect.

Because of increasing cyanosis and a marked decrease in the size of the heart and pulmonary vascular markings (fig. 6), repeat cardiac catheterization and cineangiography were undertaken on April 7, 1965 at 4 months of age (table 1).

An attempt was made to delay surgery but at 5½ months of age a severe hypoxic episode resulting in unconsciousness necessitated readmission. A Glenn procedure was performed. Two months following surgery, he was found to have a massive chyloous pericardial effusion. Pericardial taps for relief of symptoms were necessary three times over a 2-month period before resolution occurred.

The child did well for the following year. However, because of an apparent increase in cyanosis with crying and marked decrease in the intensity of his systolic heart murmur, a third cardiac catheterization was done at 18 months of age (table 1). Dye-dilution curves showed no apparent left-to-right shunt at the ventricular level. Cineangiographic studies showed marked diminution in the size of the ventricular septal defect.

**Discussion**

The clinical course observed in these three youngsters was remarkably similar except for the timing of the hemodynamic changes (table 2). All under 4 months of age were admitted to the hospital in congestive cardiac failure. Cyanosis rather than being the predominant sign was recognized only in patient B. A similar clinical picture associated with tricuspid atresia without transposition of the great vessels could be found in only two previous reports. Findings in our patients are in striking contrast to the statements of Keith and associates, Nadas, and Taussig that gross cardiomegaly or massive pulmonary vascular engorgement or both do not occur in tricuspid atresia without associated transposition of the great vessels.

The electrocardiograms (fig. 1) of these infants warrant little comment. All showed the left axis deviation and left ventricular hypertrophy characteristic of tricuspid atresia; p-pulmonale was not impressive.

All three of the infants had a systolic thrill and a loud harsh pansystolic murmur maximal at the lower left sternal border. In addition, the second sound was definitely but not

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**Table 2**

**Summary of Hemodynamic Changes in Patients**

<table>
<thead>
<tr>
<th>Patient A</th>
<th>Patient B</th>
<th>Patient C</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>2.5 mo</td>
<td>3.5 yr</td>
</tr>
<tr>
<td><strong>Cyanosis</strong></td>
<td>0</td>
<td>++</td>
</tr>
<tr>
<td><strong>Failure</strong></td>
<td>YES</td>
<td>NO</td>
</tr>
<tr>
<td><strong>ECG</strong></td>
<td>LVH</td>
<td>LVH</td>
</tr>
<tr>
<td><strong>Syst. m.</strong></td>
<td>+++</td>
<td>+++</td>
</tr>
<tr>
<td><strong>Heart size</strong></td>
<td>+++</td>
<td>WNL</td>
</tr>
<tr>
<td><strong>Pul. vasc.</strong></td>
<td>+++</td>
<td>WNL</td>
</tr>
</tbody>
</table>

*One year prior to surgery.

Abbreviations: LVH = left ventricular hypertrophy; WNL = within normal limits; pul. vasc. = pulmonary vasculature; syst. m. = systolic murmur; and + to +++ = degree of severity.
abnormally split in all. The pulmonic component was diminished. Though characteristic of the murmur created by a ventricular septal defect, the murmur in these cases was not unlike that produced by infundibular pulmonary stenosis.

The initial cardiac catheterizations confirmed the excessive pulmonary flow because high systemic arterial oxygen saturations were obtained in all. In addition, angiocardiograms in the first and cineangiograms in the latter two confirmed the absence of transposition of the great vessels. A large ventricular septal defect and normal-sized main pulmonary artery were clearly seen in patients B and C in addition to a small right ventricular chamber (figs. 4 and 5).

Over a 4-year period patient A had fewer respiratory infections and became better nourished. This same change in clinical status occurred more rapidly in the other two infants—over a 10-month period in patient B, and over a 4-month period in patient C. Hypoxic spells occurred in all three. The electrocardiograms and the auscultatory findings remained constant. The changes in the chest roentgenograms, however, were as dramatic as the changes in symptomatology. The heart size significantly decreased as did the pulmonary vascular markings.

It is not possible to do more than speculate as to the exact nature of the anatomic changes which produced such a marked decrease in pulmonary blood flow. Several possibilities may be eliminated from consideration. A closing patent ductus arteriosus is unlikely both by the unchanging nature of the systolic murmur and by failure to opacify a ductus on the initial angiographic studies. Progressive peripheral pulmonary arteriolar obstruction must be considered as a possible cause of decreasing pulmonary flow. If such were the case, however, one would expect the loud, harsh pansystolic murmur to diminish in intensity and the pulmonary component of the second heart sound to become accentuated. Neither of these changes occurred. The murmur and second heart sound were unchanged. Additional evidence is the fact that a superior caval, right pulmonary arterial anastomosis significantly reduced symptoms in all three patients. If peripheral pulmonary vascular obstruction were present, adequate flow from the superior vena cava to the pulmonary artery could not be expected.

Two possible causes for the diminishing pulmonary blood flow deserve consideration: progressive infundibular pulmonary stenosis or a closing ventricular septal defect. Our studies do not conclusively establish which of these was the case. A closing ventricular septal defect seems the most likely explanation and supportive evidence may be found in the literature. Roberts and associates described the heart of a 27-year-old man dying of hypoxia with tricuspid atresia. Though the right ventricle was hypoplastic, the outflow tract, pulmonary valve, and pulmonary artery were only slightly smaller than normal. There was definite evidence of a ventricular septal defect which had closed. Brock has also described two hearts with tricuspid atresia and normal-sized pulmonary arteries with very small ventricular septal defects producing death from hypoxia. In addition, it may be remembered that, although approximately 12% of the patients with tricuspid atresia without transposition have normal-sized pulmonary arteries and pulmonary valves, only the cases reported by Astley and associates and Carey and Edwards could be found in which the patients died in heart failure with slight or no cyanosis, gross cardiomegaly, and pulmonary congestion. In both of these instances the ventricular septal defect was described as large.

The two studies on each of our patients at different periods of time confirmed that arterial oxygen saturation was decreasing. The arterial saturation dropped from 83 to 73% in patient A and the second study in his case was 1 year prior to onset of hypoxic spells and palliative surgery; from 75 to 55% in patient B; and from 93 to 55%, while crying, in patient C. Neither the right ventricle nor pulmonary artery were entered for measurement in any study. Comparative angiographic studies with injection of contrast medium into
the left ventricular chambers with the patients in the lateral position were equivocal (figs. 4 and 5). The pulmonary artery definitely remained large and no infundibular constriction could be outlined in the comparable studies. Only patient C has had cardiac catheterization following surgery (table 1). This was indicated because his clinical status had changed; in addition to cyanosis, more marked with crying being noted, his systolic murmur had definitely diminished. No left-to-right shunt at the ventricular level was demonstrated with dye-dilution curves and only minimal amounts of contrast could be seen passing through the ventricular septal defect (fig. 5). In patient C, at least, it seems apparent that a closing ventricular septal defect is the cause of the progressive decrease in pulmonary blood flow.

References

From the First Year of CIRCULATION—1950

The stenosis of the outflow tract was of such proportion as to create sufficient resistance for the right ventricle to supply blood to the aorta but at the same time wide enough to allow an adequate flow of blood to the lungs without stress on the intrapulmonary arteries. . . . The eventual appearance of detrimental intimal changes in the intrapulmonary arteries can possibly be prevented and adequate pulmonary blood flow maintained by surgical creation of an appropriate degree of stenosis of the right ventricular outflow tract or pulmonary trunk.—W. Harold Civin and Jesse E. Edwards: Pathology of the Pulmonary Vascular Tree. Circulation 2: 550, 1950.
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