Left Axis Deviation in Tricuspid Atresia and Single Ventricle

The Electrocardiogram in 36 Autopsied Cases

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The electrocardiograms from 28 unselected autopsied cases of tricuspid atresia, and eight autopsied cases of single ventricle were analyzed: only those cases of single ventricle which showed left axis deviation were included. Electrocardiographic evidence of left ventricular preponderance in the precordial and unipolar limb leads was found in 27 of the 28 cases of tricuspid atresia, and three of the eight of single ventricle: In only two cases of single ventricle was the electrocardiogram identical with that seen in tricuspid atresia, and in one of them tricuspid stenosis was present. Right auricular hypertrophy as evidenced by high peaked P waves, together with presystolic hepatic pulsations occurred in tricuspid atresia in those instances in which, at autopsy, the interauricular communication was found to be small.

The finding of left axis deviation in a cyanotic child is very suggestive of tricuspid atresia. It is not, however, found in all cases, while other types of cyanotic heart disease may show left axis deviation. The present study was undertaken to analyze the other electrocardiographic findings in tricuspid atresia and to correlate these with the anatomic structure of the heart at autopsy; and also to determine if there were significant differences between these electrocardiograms and those from patients with single ventricle who showed left axis deviation. The autopsy findings on 28 patients with tricuspid atresia and eight with single ventricle were correlated with the clinical and electrocardiographic data. The eight patients with single ventricle included only those who showed left axis deviation in the electrocardiogram: seven of them were cyanotic. Patients with dextrocardia or dextroversion of the heart were excluded, as were those patients with a single ventricle in whom the electrocardiogram did not show left axis deviation.

Anatomic Discussion

Tricuspid Atresia

In this condition, the tricuspid valve is absent and is replaced by a dimple on the lower surface of the right auricle. Blood can leave the right auricle only through a patent foramen ovale or an auricular septal defect. There is but a single auriculoventricular valve, the mitral, which is almost invariably bicuspid, and leads into a hypertrophied left ventricular chamber. The right ventricle, if present, is small and rudimentary and usually communicates with the left ventricle by a septal defect. Rarely, it is a rudimentary blind chamber with no communications. The great vessels may or may not be transposed.

The 28 cases have been analyzed according to the classification of Edwards and Burchell (table 1). This classification assumes that all cases without transposition have pulmonic or subpulmonic stenosis, but one patient (case 21) in the present study, had an increased pulmonic flow and at autopsy the great vessels were seen to be normally placed.

Single Ventricle (Cor Triloculare)

In this condition, there is a single hypertrophied main ventricle and usually a rudimentary outlet chamber. It differs from tri-
Table 1.—Analysis of Autopsy Findings in 28 Cases of Tricuspid Atresia

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age at Death</th>
<th>Vent. Muscle Thickness (cm)</th>
<th>Size ASD (cm)</th>
<th>Size VSD (cm)</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>L.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>F</td>
<td>7 mo.</td>
<td>1.0</td>
<td>Large</td>
<td>0</td>
<td>PDA</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>16 mo.</td>
<td>1.0</td>
<td>Large</td>
<td>0</td>
<td>PDA</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>21 mo.</td>
<td>1.1</td>
<td>0.3</td>
<td>1.5</td>
<td>0 Ductus closing bronchials</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>7 yr.</td>
<td>1.8</td>
<td>0.3</td>
<td>2.0</td>
<td>0.2</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>8 mo.</td>
<td>1.0</td>
<td>0.5</td>
<td>1.5</td>
<td>0.2</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>1 yr.</td>
<td>0.9</td>
<td>Large</td>
<td>0.3</td>
<td>C.S. entered L.A.</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>10 mo.</td>
<td>1.0</td>
<td>1.0</td>
<td>0.4</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>4 mo.</td>
<td>0.7</td>
<td>0.3</td>
<td>2.0</td>
<td>0.4 L.S.V.C.</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>3 mo.</td>
<td></td>
<td>0.7</td>
<td>0.4</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>6 yr.</td>
<td>1.2</td>
<td>0.5</td>
<td>S.A.</td>
<td>0.5</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>4 yr.</td>
<td>1.0</td>
<td></td>
<td>S.A.</td>
<td>0.5</td>
</tr>
<tr>
<td>12</td>
<td>M</td>
<td>19 mo.</td>
<td>1.3</td>
<td>Large</td>
<td>0.5</td>
<td>C.S. entered L.A. and R.A.</td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>7 mo.</td>
<td>0.8</td>
<td>0.4</td>
<td>1.5</td>
<td>0.8 C.S. Stenosis</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>3½ yr.</td>
<td>0.5</td>
<td>0.1</td>
<td>2.0</td>
<td>0.5</td>
</tr>
<tr>
<td>15</td>
<td>M</td>
<td>6½ yr.</td>
<td>0.8</td>
<td>0.1</td>
<td>1.3-1.0</td>
<td>1.0 2 defects in aur. sept.</td>
</tr>
<tr>
<td>16</td>
<td>F</td>
<td>5 mo.</td>
<td>0.8</td>
<td>0.1</td>
<td>3.0</td>
<td>0.1 PDA</td>
</tr>
<tr>
<td>17</td>
<td>M</td>
<td>21 mo.</td>
<td>0.7</td>
<td>0.2</td>
<td>1.0</td>
<td>0.5 PDA</td>
</tr>
<tr>
<td>18</td>
<td>M</td>
<td>15 mo.</td>
<td>1.1</td>
<td>0.5</td>
<td>0.5</td>
<td>0.1 PDA</td>
</tr>
<tr>
<td>19</td>
<td>M</td>
<td>2 mo.</td>
<td></td>
<td>1.2</td>
<td>0.3</td>
<td>Bronchials</td>
</tr>
<tr>
<td>20</td>
<td>F</td>
<td>4 mo.</td>
<td>0.8</td>
<td>0.2</td>
<td>1.0</td>
<td>0.2 PDA</td>
</tr>
<tr>
<td>21</td>
<td>M</td>
<td>18 mo.</td>
<td>1.1</td>
<td>0.3</td>
<td>0.8</td>
<td>3.0 0</td>
</tr>
<tr>
<td>22</td>
<td>F</td>
<td>1½ yr.</td>
<td>1.2</td>
<td>0.7</td>
<td>2.0</td>
<td>2.0 L.S.V.C.</td>
</tr>
<tr>
<td>23</td>
<td>M</td>
<td>2½ yr.</td>
<td>1.0</td>
<td>0.5</td>
<td>0.2</td>
<td>P.V. entered R.A.</td>
</tr>
<tr>
<td>24</td>
<td>M</td>
<td>6 yr.</td>
<td>1.0</td>
<td>0.5</td>
<td>0.5</td>
<td>2.0</td>
</tr>
<tr>
<td>25</td>
<td>F</td>
<td>5 yr.</td>
<td>0.9</td>
<td>1.5</td>
<td>0.5</td>
<td>1.5 Aorta overriding</td>
</tr>
<tr>
<td>26</td>
<td>F</td>
<td>19 mo.</td>
<td>+</td>
<td>0.5</td>
<td>Large</td>
<td>0.5</td>
</tr>
<tr>
<td>27</td>
<td>M</td>
<td>2 mo.</td>
<td>1.1</td>
<td>3.5</td>
<td>0.2</td>
<td>Coarct.</td>
</tr>
<tr>
<td>28</td>
<td>F</td>
<td>3 yr.</td>
<td>1.0</td>
<td>0.5</td>
<td>1.0</td>
<td>2.0 Narrow L.P.V.</td>
</tr>
</tbody>
</table>

Abbreviations: C.S., Coronary sinus. S.A. Single auricle. L.S.V.C., Consistent left superior vena cava entering coronary sinus. P.V., Pulmonary vein. ASD, Auricular septal defect or widely patent foramen ovale.

cuspid atresia in the following features: (1) The right auricle communicates directly with the common ventricular chamber. (2) An interauricular communication is not invariably present. (3) Either one or two auriculoventricular valves may be present; if there is but a single valve, it usually has three cusps. (4) Transposi-
tion of the great vessels is the rule rather than the exception. (5) In most cases, there is a rudimen-
tary chamber lying in the position of, and possibly representing, the infundibular region or outflow tract rather than a separate ventricle. The vessel arising from the rudimentary chamber is usually stenotic but may be of normal
TABLE 2.—Analysis of Autopsy Findings in 8 Cases of Single Ventricle Showing Left Axis Deviation in the Electrocardiogram

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age at Death</th>
<th>Vent. Muscle Thickness (cm)</th>
<th>A V Valves</th>
<th>Position Rudim. Chamber</th>
<th>Size ASD (cm)</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>29</td>
<td>F</td>
<td>8 mo.</td>
<td>0.7</td>
<td>1</td>
<td>Post.</td>
<td>3.5</td>
<td>Hypoplastic aorta</td>
</tr>
<tr>
<td>30</td>
<td>M</td>
<td>4 days</td>
<td>0.5</td>
<td>2</td>
<td>Ant.</td>
<td>1.0</td>
<td>Coarct. (infantile)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group Ib. With Transposition and Pulmonic or Subpulmonic Stenosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>31</td>
<td>M</td>
<td>10 mo.</td>
<td>2</td>
<td>Ant.</td>
<td>None</td>
<td>Pulm. atresia. PDA, overriding aorta</td>
<td></td>
</tr>
</tbody>
</table>

Electrocardiographic Findings and Their Interpretation in Cases of Tricuspid Atresia and Single Ventricle with Left Axis Deviation.

The characteristic electrocardiographic findings in tricuspid atresia are left axis deviation, high peaked P waves, and left ventricular preponderance in the precordial leads1, 2, 10 (fig. 1).

The P Wave

The amplitude of the P wave was measured in lead II, where the deflection usually attains its maximum height and shows least variation. The maximum normal amplitude is 2.5 mm. in infants below the age of one year, and 3 mm. in children over that age (10 mm. deflection equivalent to 1 microvolt).12 Of the patients with tricuspid atresia, 20 had P waves of increased amplitude in lead II, and in the remaining eight the amplitude was normal. In three of these eight cases (cases 4, 8, 22), the P-wave axis, as plotted on the triaxial system of Bayley,12 was far toward the left (less than 30 degrees) and all of these showed increased amplitude of the P wave in V1 or V3. It was thought that in these patients the normal amplitude in lead II was due to the shift of the P-wave axis. In the remaining five patients with tricuspid atresia the P wave was of normal amplitude in all leads.

The P waves were increased in amplitude in four of the patients with single ventricle (table 3).

Correlation of Electrocardiographic and Pathological Evidence of Right Auricular Hypertrophy

At autopsy, the right auricle was enlarged in all cases of tricuspid atresia and was hypertrophied in the majority. Since this hypertrophy is theoretically due to difficulty in the expulsion of blood from the right auricle, it should be less marked in the presence of a large associated defect between the auricles. The electrocardiographic evidence of right auricular hypertrophy (high peaked P waves) and the clinical evidence
were septal pulsations at the margin of the liver) were correlated with the size of the interauricular communications found at autopsy. Hepatic pulsations and P waves of increased amplitude were present in 10 of the 28 patients with tricuspid atresia: in all these patients the interauricular communication was small (1 cm. or less in diameter).

There were 13 patients with P waves of increased amplitude who did not show hepatic pulsations: the interauricular communication was small in four of these, measured between 1 and 3 cm. in five, and appeared large but could not be accurately measured in four. The remaining five patients with tricuspid atresia had P waves of normal amplitude in all leads and no hepatic pulsations. In all five the interauricular communication measured over 1 cm. in diameter.

In this series, the combination of high peaked P waves and hepatic pulsations in tricuspid atresia indicated a small interauricular communication. Tall P waves alone, however, were found in some patients with large auricular septal defects.

The P waves were of normal amplitude in four of the patients with single ventricle and increased in amplitude in four. In two of the latter group (cases 33 and 35), hepatic pulsations were present: at autopsy, a marked tricuspid stenosis was found in one, and subpulmonic stenosis with closed foramen ovale in the other.

The QRS Complex

The electrical axis was deviated to the left (between plus 30 and minus 90 degrees) in 24 of the 28 patients with tricuspid atresia (fig. 2) and lay between 0 degrees and minus 90 degrees in 23. All the patients with single ventricle included in this study showed left axis deviation. One patient (case 30) had a left bundle branch block, and in another, the electrical axis shifted from minus 80 degrees to plus 110 degrees with each inspiration.

The electrical axis as determined between leads I and II differed from that determined between leads I and III by more than 10 degrees in 9 patients (table 3). This finding indicates that the electrical axis is not always accurately projected on the sides of the Einthoven triangle.

Unipolar Limb Leads

Leads aVR, aVL, and aVF were recorded in 12 cases of tricuspid atresia and four of single ventricle. The heart occupied a horizontal position in 13, and a semihorizontal in two, as indicated by a predominantly positive deflection in aVL associated with a less positive or negative deflection in aVR. The findings in aVR are of special interest. The deflection was of the qS type in 9 of the 12 cases of tricuspid atresia, and the R wave present in the remaining three cases measured 1 mm. or less, whereas in two of the four patients with single ventricle, a QR pattern was present and the R wave measured 5 mm. or over. Thus, the presence of an R wave in this lead appears at the present time to be against the diagnosis of tricuspid atresia.

Precordial Leads

A predominantly negative QRS deflection in V1 was present in 19 of the 21 patients with tricuspid atresia on whom precordial leads were obtained, and a predominantly positive QRS
deflection in V5 in 20. These patterns were considered to indicate left ventricular preponderance. The two patients with an RS pattern in V1 showed a left ventricular preponderant pattern in V5.

Precordial leads were available in seven of the eight cases of single ventricle. A pattern of left ventricular preponderance was present in three, equiphasic QRS deflections in V1, V3, and V5 in three, and right ventricular preponderance in one (figs. 2 and 3).

The time of onset of the Intrinsicoid Deflection was measured from the beginning of QRS to the peak of R in V1 and V5. This Q-R interval was
normal or below normal in V1 in all patients with tricuspid atresia. In V5, the time of onset was delayed beyond the average normal for the age in 18 of the 21 patients and beyond the upper limit of normal in eight. In case 28, the prolonged Q-R in V5 was the only evidence of left ventricular pathology. In single ventricle, the onset of the intrinsicoid deflection was delayed in V1 in one patient and normal in six: in V5 it was delayed in three of the seven patients (table 3).

Correlation of Electrocardiographic and Pathological Evidence of Left Ventricular Pathology

Left ventricular hypertrophy was a constant autopsy finding in tricuspid atresia. In the electrocardiogram of a child, the combination of left axis deviation and a horizontal heart strongly suggests left ventricular hypertrophy. Left axis deviation was present in 24 of the 28 cases and a horizontal electric position was recorded in 9 of the 12 with unipolar limb leads. Additional electrocardiographic evidence of left ventricular hypertrophy was also present: T1 was lower than T3 in 26 instances and the T wave in aV1 was inverted in 10 of 12 records. Isoelectric or inverted T waves in V5 or V6 were found in seven patients. A left ventricular preponderant pattern in the precordial leads was present in 20 of the 21 records.

Although all the patients with single ventricle showed left axis deviation, only three had other electrocardiographic evidence of left ventricular preponderance. In two of these (cases 33 and 35), the unipolar limb leads were similar to those seen in tricuspid atresia: at autopsy, one of these patients had a marked tricuspid stenosis, and in the other the pulmonary artery arose from the posterior main chamber and there was a pulmonary subvalvular stenosis. In both these patients, the part of the ventricular myocardium corresponding to the left ventricle was markedly hypertrophied. In the third patient (case 30), the electrocardiogram differed from...
all the records seen in tricuspid atresia by having a qR pattern in aVR. Autopsy revealed marked subpulmonic stenosis and the pulmonary artery arose from a rudimentary anterior chamber.

Other Electrocardiographic Findings

A Q wave was present in one or more of the left ventricular leads (aVL, I, and V5) in 21 patients with tricuspid atresia and three with single ventricle. Such Q waves are believed to originate in the ventricular septum. The presence of these waves in patients with single ventricle, in whom no ventricular septum was present, is of great interest and suggests that the ridge of tissue lying below the rudimentary chamber may contain conduction tissue and represent a rudimentary septum rather than the crista supra ventricularis.

The total electrical systole of the heart, as represented by the Q-Tc, was below the average normal for the age in 20 of the 27 cases of tricuspid atresia: this finding may be associated with a smaller ventricular muscle mass in this condition than in the normal heart. The Q-Tc was normal in the four patients with single ventricle who were not receiving digitalis.

SUMMARY

The electrocardiographic and autopsy findings in 28 selected cases of tricuspid atresia and eight of single ventricle with left axis deviation were analyzed.

Left axis deviation was present in 24 cases of tricuspid atresia: 20 of the 21 patients with precordial leads showed left ventricular preponderance. Precordial leads were available in seven cases of single ventricle. There were large equiphasic QRS complexes from V1 to V5 in three cases, and right ventricular preponderance in one. The pattern of left ventricular preponderance was seen in three patients; in these no constant anatomical pattern was found at autopsy. In one of the three, the electrocardiographic finding of a tall R wave in aVR was a point of differentiation from all the records seen in tricuspid atresia. In the other two electrocardiograms, no clinical or electrocardiographic differentiation from tricuspid atresia seemed possible; at autopsy, tricuspid stenosis was found to be present in one of these.

Electrocardiographic evidence of right auricular hypertrophy was present in 23 cases of tricuspid atresia and four of single ventricle. In tricuspid atresia, patients who showed both high peaked P waves and hepatic pulsations were found at autopsy to have small interauricular communications. High peaked P waves in patients without hepatic pulsations occurred in association with auricular defects of varying size. Patients with P waves of normal amplitude in all leads uniformly had large interauricular communications. In single ventricle, right auricular hypertrophy had no uniform correlation with the size of the interauricular communication found at autopsy. The Q-T waves were found in the left ventricular leads in both groups of patients. It is postulated that the muscle ridge below the rudimentary chamber in single ventricle may represent septal tissue.

The corrected Q-T interval (Q-Tc) was less than the average normal for the age in tricuspid atresia. This suggests that in these patients the excitation wave traverses a smaller bulk of muscle tissue in the normal heart.

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We should like to acknowledge with gratitude the constant help and encouragement of Dr. Helen B. Taussig without whom this study would have been impossible. We should also like to thank Mrs. V. Rogers, Mrs. M. Lindemann, and Mrs. P. Schaff for their technical assistance.

SUMMARIO IN INTERLINGUA

Esseva analysate le electrocardiogrammas disponibile in 28 non-seligite autopsiate casos consecutive de atresia tricuspid e in 8 autopsiate casos de univentricularitate seligite de manera que solo caso exhibiente sinistrotrone deviationes del axe esseva includite. Signos electrocardiographic de preponderantia sinistroventricular in le derivationes extremital precordial e unipolar esseva trovate in 27 inter le 28 casos de atresia tricuspid e in 3 del 8 casos de univentricularitate. In solo 2 del casos de univentricularitate esseva le electrocardiogramma identic con illo vidite in atresia tricuspid; in 1 de iste 2 casos stenosis tricuspid esseva presente. Hypertrophia dexteroauricular, indicate per undas P a alte
piescos, insimul con pulsationes hepatic presystolic occurreva in atresia tricuspid in le casos in le quales le autopsia revelava que le communication interauricular esseva parve.

REFERENCES

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