The Atrioventricular Conduction System in Two Cases of Tricuspid Atresia

By Barbara Guller, M.D., James W. DuShane, M.D., and Jack L. Titus, M.D., Ph.D.

SUMMARY
The morphology of the atrioventricular (A-V) conduction system was studied by serial histologic section in two cases of tricuspid atresia type IB (without transposition of the great arteries, with subpulmonary stenosis). The A-V node was adjacent to the central fibrous body, and its location was indicated by a "dimple" in the floor of the right atrium. The left bundle branches originated unusually close to the nodal-bundle junction. The right bundle branch was markedly elongated in its course to the right ventricular septal endocardium. It was situated along the inferior aspect of the channel-like subpulmonary stenosis5) were chosen for study because the position of the ventricular septal defect and its anatomic relationships in both cases are almost identical to those in examples of this type of tricuspid atresia and because the electrocardiographic pattern considered to be characteristic for tricuspid atresia is most commonly found in this subgroup.1, 6, 7

The electrocardiograms from both cases showed left axis deviation and a counterclockwise rotation of the frontal QRS loop (fig. 1). The gross morphologic findings and the heart sizes were similar in both autopsy specimens, which were from infants aged 25 days (case 1) and 26 days (case 2).

Histologic examination of the conduction system was carried out according to the method described by Titus and associates.8 A rectangular block of tissue from the septum of each heart was serially sectioned. From posterior to anterior, each block extended from the region of the coronary sinus ostium to the pulmonary valve; superiorly each included approximately 0.5 cm of the atrial septum and adjacent aortic root, and inferiorly each included all of the right ventricular aspect of the septum and approximately one half of the left ventricular side of the septum. The blocks measured 27 by 20 by 6 mm in case 1 and 30 by 22 by 6 mm in case 2. Each block was serially sectioned in a plane perpendicular to the

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Ventricular septal defect

In tricuspid atresia, the electrocardiographic finding of a counterclockwise frontal plane loop with left axis deviation cannot be explained by left ventricular hypertrophy alone.2 It has been suggested to result from abnormal impulse propagation due to fibrosis3 or anomalous branching3 of the left bundle branches, which might result in superior and leftward displacement of the activation wave.4 Serial histologic sections of the atrioventricular (A-V) conduction system in tricuspid atresia were studied to determine the anatomic arrangement of the A-V conduction tissue.

Methods
Two specimens of tricuspid atresia type IB (no transposition of the great arteries, subpulmonary...
ventricular septum and to the base of the heart at a thickness of 8 μ. All sections were retained. Every 20th section was stained with hematoxylin and eosin, and each next adjacent section was stained with the Mallory-Heidenhain stain. In some regions, both types of stains were used on every fifth or tenth section in order to elucidate the A-V conduction system better. Approximately 600 histologic sections from each specimen were studied. The positions and lengths of the A-V node, common bundle of His, and proximal portions of right and left bundle branches were determined. Specifically, four distances were measured: (distance 1) length of A-V node, (2) length of common bundle from nodal-bundle junction to first left bundle branch, (3) distance over which left bundle branching occurred, and (4) distance from nodal-bundle junction to point at which right bundle branch was first situated in endocardium of right ventricular septum. The distance from the A-V node to the first left bundle branch (distance 2) was considered to be the minimal anatomic distance over which the activation wave must travel to reach the left ventricle. Similarly, the distance from the nodal-bundle junction to the location of the right bundle junction.
TRICUSPID ATRESIA

Case 1. Schematic representations of A-V conduction system in tricuspid atresia type IB superimposed on block of heart removed for serial histologic sectioning.

(A) Block of tissue viewed from right and anterior aspect of heart. Portions of the opened right atrium (RA) and opened right ventricle (RV) were included in the tissue block, which was approximately 27 by 20 by 6 mm. The atrioventricular node (crosshatched, no. 1) is just anterior to the ostium of the coronary sinus. The short common bundle of His (no. 2) is also crosshatched. The broken lines indicate structures that are situated in the left side of the septum (away from viewer) and the solid lines indicate structures on the right side of the ventricular septum (facing the viewer). The open, broken lines (no. 3) are the portion of the bundle from which left bundle branches (represented by tapering broken lines) originated. No. 4 is the right bundle branch from the nodal-bundle junction to the point at which the right bundle was first located in the endocardium of the right ventricular aspect of the septum. Part of the right bundle, distal to the limiting arrow of no. 4, is shown by the solid lines. The point of the arrow VSD indicates the opening of the ventricular septal defect in the right ventricle.

(B) Left side of block of tissue shown in figure 3A has portion of left atrial septum (LA), part of mitral valve (MV), and part of left ventricle including portion of its outflow tract, the aortic valve, and the aorta (A).

The opening of the ventricular septal defect in the left ventricle is the dark area in the outflow tract. The membranous septum is absent. The A-V conduction system is schematically represented as in figure 3A. The solid lines indicate those portions of no. 2 and no. 3 and the left bundle branches (tapering lines) that are on the left side of the ventricular septum. The dotted lines indicate the course of the right bundle from the left side to the right side of the septum. The right bundle branch is in or near the lower margin of the elongated (from left to right) VSD.

Results

Morphologic Findings

The topographic relationships of the A-V conduction system in tricuspid atresia differed from the normal heart as a result of certain anatomic features of tricuspid atresia. These morphologic features included a minute, endocardial-lined pocket (dimple) in the floor of the right atrium (fig. 2A), which has been considered to indicate the theoretic site of the absent tricuspid valve. The "dimple" was situated just anterior to the coronary sinus ostium, near or in the region in which the A-V node is located in the normal heart. It lay over the central fibrous body. Other morphologic features of importance were the ventricular septal defect, which was a narrow elongated channel\(^5\) that passed through the septum obliquely (fig. 2B), and the absence of the membranous ventricular septum (fig. 3).

The A-V node was situated adjacent to the central fibrous body in the floor of the right atrium. Its location in the intact heart was indicated by the "dimple" (fig. 4A). The most anterior portions of the A-V node and the common bundle of His, which extended anteriorly from the node, penetrated the fibrous ring of the mitral valve in the region of the A-V groove.

The common bundle of His passed through the fibrous A-V ring and was situated in a
Case 1. Serial histologic sections of the A-V conduction system. The abbreviations in figures 4 and 5 are: $N = A-V$ node; $B =$ common bundle of His; $LBB =$ left bundle branches; $RB =$
subendocardial position on the left side of the ventricular septum (fig. 4B). When the common bundle was entirely on the ventricular side of the A-V ring, it was beneath the posterior cusp of the aortic valve.

In both hearts, left bundle branches originated close to the nodal-bundle junction (fig. 4B and C). In the first specimen (case 1), left bundle branches were given off from the common bundle just after the apparent origin of the common bundle from the A-V node. In the second specimen (case 2), left bundle branches separated from the fascicles considered to be a common bundle in the region of junction between nodal and bundle fibers. In both specimens, the left bundle branches consisted of multiple discrete fascicles of similar diameter, which fanned out over the left aspect of the ventricular septum beneath (inferior and apexward) the posterior cusp of the aortic valve. All left bundle branches were situated posterior to the ventricular septal defect, when viewed from the left.

The right bundle branch could be defined as a distinct group of fibers within the common bundle after approximately half of the left bundle branches had been given off. The point at which right bundle-branch fibers could be recognized was the region in which the membranous septum is located in the normal heart. In the first specimen (case 1), the right bundle branch turned to the right and slightly inferiorly immediately after it became identifiable; as a result of this course, the right bundle branch was situated within the ventricular septum and not subendocardially (figs. 4D and 5A). More anteriorly, the right bundle branch again was situated subendocardially in the left ventricle and, in this position, reached the inferior rim of the ventricular septal defect (fig. 5B). In the second case (case 2), the right bundle branch did not have an intramyocardial portion before it became situated beneath the ventricular septal defect. In both hearts, the right bundle branch traversed the septum from left to right, subjacent to the inferior margin of the ventricular septal defect and came to lie in a nearly subendocardial position in the right ventricle just inferior to the ventricular septal defect (fig. 5C). From this point, the right bundle branch descended almost perpendicularly to the apex of the right ventricle in a subendocardial position in the right ventricular aspect of the septum (fig. 5D).

Measurements of the Conduction System

Results of measurements of the different portions of the conduction system were similar (table 1 and fig. 3) in these two examples of tricuspid atresia type IB (no transposition of the great arteries, subpulmonary stenosis).

Discussion

The course and topographic relationships of the A-V conduction system were similar in these two examples of tricuspid atresia without transposition of the great arteries and with
Figure 5

Case 1. (A) Higher power of area in rectangle in figure 4D. Mallory-Heidenhain; X 40. (B) The right bundle is in a subendocardial position on the left side of the septum in the postero-inferior
subpulmonary stenosis. The A-V node was adjacent to the central fibrous body, as in the normal heart8 (fig. 6A and B), and its location was indicated by a “dimple” in the floor of the right atrium. The location of the common bundle (His) on the left side of the ventricular septum below the A-V ring also may occur normally. The course of the bundle branches differed from the normal (fig. 6A and B) in that the left bundle branches originated unusually close to the nodal-bundle junction and the right bundle branch was markedly elongated in its course from the A-V node to the right septal endocardium (figs. 3A and B, and 6B). Although anatomically abnormal, the bundle branches included in this study did not have pathologic lesions in their proximal portions.

The finding of early arborization of left bundle branches in tricuspid atresia is in rim of the VSD. The irregularity of the right bundle is an artifact of sectioning. This histologic section is 0.8 mm anterior to figures 4D and 5A. Mallory-Heidenhain; X 40. (C) The right bundle is on the right side of the ventricular septum and below the VSD. This histologic section is 6.3 mm anterior to that shown in figure 5B. Mallory-Heidenhain; X 6. (D) The right bundle descends apexward in a subendocardial position in the septum of the right ventricle. This histologic section is 0.3 mm anterior to that shown in figure 5C. Mallory-Heidenhain; X 60.
Figure 7

(A) Uncomplicated ventricular septal defect (VSD) of usual variety without electrocardiographic abnormalities of superior and leftward displacement of the main QRS axis. The position and relationship of the A-V node and bundle and the right and left bundle branches are like those in the normal heart (fig. 6A); the same labels are utilized. The membranous septum may be more deficient than shown in this sketch. (Sketch drawn from data reported by Feldt and associates).9

(B) Usual variety of ventricular septal defect (VSD) with electrocardiographic abnormalities of superior and leftward displacement of the main QRS axis. The position, course, and relationships of the A-V conduction system are similar to those shown in figure 6B and are characterized by origin of left bundle branches close to the A-V node and elongation of the right bundle branch. Legends are same as in figures 6A and B and 7A. (Sketch drawn from data reported by Feldt and associates).9

Table 1
Measurement of Different Portions of Conduction System in Two Cases of Tricuspid Atresia of Type IB

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Case 1</th>
<th>Case 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Length of A-V node</td>
<td>2.2 mm</td>
<td>2.0 mm</td>
</tr>
<tr>
<td>2. Length of common bundle from nodal-bundle junction to first left bundle branch</td>
<td>0.5 mm</td>
<td>0.5 mm</td>
</tr>
<tr>
<td>3. Distance over which left bundle branching occurred</td>
<td>2.2 mm</td>
<td>3.9 mm</td>
</tr>
<tr>
<td>4. Distance from nodal-bundle junction to point at which right bundle branch first was situated in endocardium of right ventricular septum</td>
<td>8.9 mm</td>
<td>10.4 mm</td>
</tr>
<tr>
<td>Ratio*</td>
<td>0.06</td>
<td>0.05</td>
</tr>
</tbody>
</table>

* Ratio of "undivided portion" of left bundle branch to "undivided portion" of right bundle branch (ratio of measurement, 2:4).
keeping with previous suggestions\textsuperscript{2,3,10} that morphologic abnormalities of the left bundle might be present. An anomalous course of the left bundle branches as demonstrated herein was suspected by Gamboa and associates\textsuperscript{2} from electrocardiographic studies. Fibrosis of the left bundle branches also has been assumed\textsuperscript{3} but was not present in the characteristic examples of tricuspid atresia we studied. Fibrosis of the left bundle branch has been observed in a patient with pulmonary atresia and stenosis of the tricuspid valve.\textsuperscript{10}

The elongated right bundle branch originated in a subendocardial location on the left side of the ventricular septum. It was situated along the inferior aspect of the channel-like ventricular septal defect\textsuperscript{5} (figs. 3B and 6B) as it traversed the septum to a subendocardial location on the right ventricular septum (fig. 3A). Interestingly, the right bundle branch in case 1 had a subendocardial portion, then a myocardial portion, and then a subendocardial again before it was situated in its usual location on the septum of the right ventricle. These different positions of the right bundle in relation to the endocardium mimic those of the normal heart.\textsuperscript{11}

The ratios of the "undivided portions" of the bundle branches (defined as the distance from the A-V node to the first left bundle branch divided by the distance from the A-V node to the point at which the right bundle branch was first situated in the endocardium of the right ventricle) were similar in both specimens. The smallness of the ratios (0.06 and 0.05) is in contrast to the ratios of approximately 0.3 found in uncomplicated ventricular septal defects with normal anatomic arrangement of the conduction tissue\textsuperscript{9} (fig. 7A)—defects that do not show counterclockwise frontal-plane loops. This contrast suggests that the anatomic differences in the conduction system found in tricuspid atresia might be responsible, in part, for the known electrophysiologic abnormalities.

In tricuspid atresia, the anatomic configuration of the A-V conduction system is characterized by an early origin of the left bundle branches and a markedly elongated course of the right bundle (fig. 6B). Left axis deviation in the frontal plane has been produced experimentally in the primate heart in which an electrocardiographic pattern compatible with that of tricuspid atresia was induced by interruption of the anterior fibers of the left bundle-branch system combined with a block of the right bundle.\textsuperscript{4} It is conceivable that early origin of left bundle branches could affect the spread of the intracardiac wavefront in a manner similar to these experimentally produced lesions of the conduction system.

An abnormal anatomic arrangement of the A-V conduction tissue has been found in other congenital cardiac malformations (fig. 7), which also have electrocardiographic abnormalities of superior and leftward displacement of the main QRS axis, such as persistent common A-V canal\textsuperscript{12} and ventricular septal defect of the A-V canal type.\textsuperscript{13} In both these conditions, elongation of the conduction tissue\textsuperscript{12,13} was reported to be present.

\textbf{References}


Tortured Logic

The people who bind themselves to systems are those who are unable to encompass the whole truth and try to catch it by the tail: a system is like the tail of truth, but truth is like a lizard, it leaves its tail in your fingers and runs away, knowing full well that it will grow a new one in a twinkling.—Henri Troyat: Tolstoy (translated from the French by Nancy Amphoux). New York, Doubleday, 1967.
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