Anatomic Studies of the Cardiac Conduction System in Congenital Malformations of the Heart

By Keith Reemtsma, M.D., and W. M. Copenhaver, Ph.D.

A frequent and serious complication of certain intracardiac procedures is heart block, produced by operative injury to the conduction pathways. The anatomic relationship of the conduction system to cardiac defects has previously been studied only in isolated cases of heart block. This report deals with a study, by means of serial sections and reconstructions, of the relationships of the atrioventricular node and bundle to a variety of congenital defects in the area of the membranous ventricular septum.

STUDIES on the conduction mechanism of the heart began in 1845 with the observations of Purkinje, who described in sheep hearts a peculiar type of muscle fiber that now bears his name. In 1893 His published a description of the atrioventricular bundle, and since that time a considerable body of information has accumulated on the structure and function of this specialized tissue. However, except for isolated studies in cases of congenital heart block, there is scant information on the course of the conduction fibers in congenital malformations of the heart.

The development of intracardiac surgery has aroused renewed interest in the anatomy of congenitally malformed hearts, and repair of certain cardiac defects has demonstrated the susceptibility of the conduction system to injury. In an early series of 20 cases of interventricular septal defect repaired by intracardiac operations, 9 instances of conduction disturbances were recorded. Recently Cooley, Kirklin, and Harshbarger stated "knowledge of the exact anatomic location of the bundle of His in the presence of persistent common atrioventricular canal has long been needed, for an inadvertently placed stitch through or around the bundle may lead to complete heart block with fatal termination."

This present report deals with a study of the anatomic relationship of the cardiac conduction system to various congenital defects in the region of the membranous portion of the interventricular septum and adjacent structures.

MATERIALS AND METHODS

In the initial stages of the investigation, dissection of the conduction system was attempted in fresh and formalin-fixed human hearts after the method described by Widran and Lev. Microdissection techniques were applied in fresh fetal and neonatal hearts. Neither of these methods, however, appeared satisfactory in the presence of intracardiac defects. The method of study finally adopted involved histologic study and visual reconstruction of serial sections. Five hearts, obtained from the Department of Pathology of Babies Hospital, were selected for study. Table 1 summarizes the malformations present in each case.

Following the photographing and sketching of specimens, a block of tissue was removed that included the ostium of the coronary sinus, the inferior portions of the atrial walls and interatrial septum, the superior portions of the ventricular walls and interventricular septum, and the attached fibrous ring of the heart. This block of tissue completely encircled the defects in the region of the membranous portion of the interventricular septum and the atrioventricular septum. The block was then sectioned in a direction corresponding approximately to the anteroposterior plane of the body. The sections were stained with either hematoxylin and eosin or Masson's triple stain. Following histologic study, graphic reconstructions were made from projected sketches of the slides.

OBSERVATIONS

Case 1. This heart showed a defect in the membranous portion of the interventricular septum. The atrioventricular node was identified on the superior (atrial) aspect of the fibrous ring of the heart, adjacent to the ostium of the coronary sinus. The atrioventricular bundle coursed to the left and caudad, penetrating the fibrous ring. The bundle then followed the postero inferior aspect of the
TABLE 1.—Summary of Atrial and Ventricular Defects in Five Cases

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2 months</td>
<td>interventricular septal defect (membranous)</td>
</tr>
<tr>
<td>2</td>
<td>6 days</td>
<td>persistent atrioventricular canal, patent foramen ovale</td>
</tr>
<tr>
<td>3</td>
<td>4 months</td>
<td>interatrial septal defect (primum type)</td>
</tr>
<tr>
<td>4</td>
<td>5 weeks</td>
<td>interventricular septal defect (muscular)</td>
</tr>
<tr>
<td>5</td>
<td>5 weeks</td>
<td>complete transposition without interventricular septal defect, patent foramen ovale</td>
</tr>
</tbody>
</table>

septal defect, lying immediately beneath the endocardium. The right bundle branch could be identified coursing subendocardially down the ventricular septum; the left bundle branch was indistinct (fig. 1, case 1, and fig. 2).

Case 2. This heart demonstrated a persistent atrioventricular canal and a patent foramen ovale. The atrioventricular node lay on the atrial aspect of the fibrous ring, and the atrioventricular bundle penetrated this fibrous ring in a course similar to that described in case 1. The bundle passed around the posterosuperior aspect of the ventricular portion of the atrioventricular defect. As described in case 1, the bundle lay directly beneath the endocardium. At the lowermost portion of the ventricular defect the bundle divided into right and left bundle branches, which then coursed down the ventricular septum (fig. 1, case 2, and fig. 3).

Case 3. This specimen showed a large interatrial septal defect, only a thin rim of rudimentary atrial muscle remaining on the superior aspect of the fibrous ring. The atrioventricular node and bundle lay in the normal position. However, before penetrating the fibrous ring, the node and bundle were separated from the atrial cavity only by endocardium and a narrow rim of atrial muscle (fig. 1, case 3).

Case 4. In this specimen, the interventricular septal defect was high in position; however it lay

Fig. 1. Reconstructions demonstrating relationships of the atrioventricular node and bundle in cases of (1) interventricular septal defect, membranous portion; (2) atrioventricular canal; (3) interatrial septal defect, primum type; and (4) interventricular septal defect, muscular portion. AVN and B—atrioventricular node and bundle; AVC—atrioventricular canal; CS—ostium of coronary sinus; FR—fibrous ring of the heart; IASD—interatrial septal defect; RA—right atrium; RV—right ventricle.
in the muscular septum posterior to the septum membranaceum. The atrioventricular node and bundle lay in the usual position, close to the membranous septum, which was intact in this case. Distal to the bifurcation of the bundle, the right branch was indistinct (fig. 1, case 4).

Case 5. This specimen showed complete transposition of the great vessels, patent foramen ovale, and an intact interventricular septum. In this case the location of the atrioventricular node and the course of the bundle were normal.

**Discussion**

During the past 3 decades several reports have appeared on the histologic study of the conduction system in cases of congenital heart block, associated with intracardiac defects.

In 1925 Wilson and Grant reported a case of partial heart block in an infant who was found to have a large interventricular septal defect. By studying serial sections of the posterior wall of the heart they observed a well-developed atrioventricular node in the usual location; however they could find no definite bundle penetrating the fibrous ring, although several strands of nodal tissue were seen coursing toward the posterior wall of the ventricle.

In 1929 Yater reported the first histologic

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**Figure 2.** Photomicrographs of case 1 demonstrating (A) the location of the atrioventricular bundle in the fibrous ring of the heart, above the ventricular muscle; (B) the bundle lying between ventricular muscle and endocardium, with the membranous septum intact; (C) the bundle in a position similar to (B), beneath the interventricular septal defect. A higher magnification (D) shows conductive tissue between ventricular muscle and endocardium. AVB—atrioventricular bundle; IVSD—interventricular septal defect. Masson’s triple stain; × 30 A, B, and C; × 100 D.
study of the conduction system in a case of complete congenital heart block. This patient
demonstrated transposition of the great vessels
without septal defect, and serial sections dem-
strated that the atroventricular node was
completely separated from the bundle of His
by the central fibrous body. In 1933 and 1934,
Yater and co-workers8, 9 studied an additional
2 cases of complete congenital heart block, both
of which were found to have defects in the
membranous portion of the interventricular
septum. In both cases the atroventricular node
was identified; in one case the bundle of His
was separated from the node, and in the other
case the bundle of His could not be identified.
Similar studies on cases of heart block have
appeared sporadically,10-12 and this material
has been summarized in a review of congenital
heart block by Donoso, Braunwald, Jick, and
Grishman.13

Although the location of the atroventricular
bundle in human beings has received little
attention in the past, it is a subject of current
interest in several laboratories. While our
studies were in progress, we learned of research
being done by Truex and Kain14 on conduction
tissue in human hearts with high interventricu-
lar septal defects. From their demonstration
at the American Association of Anatomists in
1956 it appears that their results are compa-
ramble to ours in case 1.

In the cases included in this report, hearts
with a variety of defects in the region of the
membranous portion of the interventricular
septum were selected. In the case of the simple
defect of the septum membranaceum, the con-
duction bundle was readily identified coursing
along the postero-inferior rim of the defect. A
comparable anatomic relationship was ob-
served in the case of atrioventricular canal,
with the bundle lying along the postero-inferior
rim of the ventricular portion of the defect.
Both these defects are believed to be related to
a failure of fusion of the dorsal endocardial
cushion and the ascending ventricular septum.
Walls15 has observed the atrioventricular
bundle in an 8-mm. embryo, and he describes
its course as "growing forward under the dorsal
endocardial cushion, still not fused with the
ventral, to reach the summit of the interven-

FIG. 3. Low-power photomicrograph of case 2,
showing the patent foramen ovale and the atri-
ventricular canal. The atroventricular bundle
branches lie in the superior portion of the ven-
tricular septum, immediately beneath the ven-
tricular portion
of the defect. AVB—atrioventricular bundle; AVC—
atroventricular canal; AVV—common atrioventric-
ular valve; PFO—patent foramen ovale. H and E
stain; X 15.
antricular septum." It seems reasonable that in this area of embryologic fusion the conduction bundle will occupy a position along the poste-
tero-inferior aspect of the smaller defects, both simple interventricular septal defects and per-
sistent atrioventricular canals.

In the case of a large interatrial septal defect, the atrioventricular bundle lay in close prox-
imity to the atrial cavity, beneath the rim of rudimentary interatrial muscular septum. The position of the bundle might render it suscept-
tible to injury during closure of the interatrial defect.

The atrioventricular node and bundle occupied normal positions in the case of a high interventricular defect in the muscular portion of the septum. The bundle coursed in close proximity to the membranous portion of the septum, which was intact.

In the case of complete transposition of the great vessels, the normal course of the conduc-
tion system perhaps could be predicted. In such an anomaly the embryologic defect involves the spiral septation of the truncus arteriosus, and the fusion of the ventricular septum and dorsal endocardial cushion remains undisturbed.

The course of the conduction system in the very large defects, such as common ventricle, remains unknown. In reporting such a case in 1925, Wilson and Grant4 were unable to demon-
strate a true atrioventricular bundle. Prelimi-
nary studies on similar specimens in our laboratory have also failed to show an atrioven-
tricular bundle comparable to those described in this report.

SUMMARY

Five hearts, demonstrating a variety of congenital defects were studied by serial sections, and the atrioventricular node and bundle were identified in each. In cases of membranous interventricular septal defect and atrioventricu-
lar canal, the atrioventricular bundle coursed along the postero-inferior margin of the ven-
tricular defect in a subendocardial position. In a case of large interatrial septal defect, the atrioventricular node and bundle lay beneath a thin rim of rudimentary interatrial muscle. In cases of complete transposition and high

interventricular muscular septal defect the conduction system lay in the normal anat-
omic position.

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SUMMARIO IN INTERLINGUA

Cinque corde con varie defectos congenite esseva studiate per sectiones serial. In omnes le nodo e le fasce atrioventriculares esseva identifi-
cate. In casos de defecto membranose del septo interventricular e de canal atrioven-
tricular, le fasce atrioventricular curreva al longo del margine postero-inferior del defecto ventricular in position subendocardial. In un caso de grande defecto del septo interatrial, le nodo e le fasce atrioventricular jaceva infra un tenue margine de rudimentari musculo interatra-
rial. In casos de transposition complete e de defecto muscular del septo supero-interven-
tricular, le systema de conduction occupava le position anatomic normal.

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A baby girl born at term and normal except for a loud systolic murmur, had complete heart block with a ventricular rate of 50 per min. This was predicted 4 months before term by the attending obstetrician because the fetal heart rate was about 56 per minute. There had never been any other sign of fetal distress. The child has been followed for 2½ years, and the block remains. Administration of atropine in the neonatal period or at subsequent times failed to affect the block. It is thought that the child has an intraventricular septal defect. The previous literature on congenital heart block, and its usual cause, intraventricular septal defect, is reviewed.

Harvey
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