Electrophysiologic Delineation of the Specialized Atrioventricular Conduction System in Two Patients with Corrected Transposition of the Great Arteries in Situs Inversus [I,D,D]

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SUMMARY Electrophysiologic delineation of the atrioventricular conduction system at surgery is described in two patients with corrected transposition of the great arteries in situs inversus. Intra-atrial electrograms were recorded in one patient from sites immediately adjacent to the coronary sinus located in the left-sided right atrium. The intraventricular portion of the atrioventricular conduction system was identified in both patients along the posterior and inferior margin of the ventricular septal defect, in contrast to the superior and anterior location found in corrected transposition of the great arteries in situs solitus.

The course of the conduction system in the hearts of these two patients and a possible relationship to the cardiac loop and dual origin of the atrioventricular node is discussed. These cases illustrate the usefulness of segmental diagnosis of congenital heart disease and of electrophysiologic identification of the specialized atrioventricular conduction system at surgery.

INTRACARDIAC OPERATIONS in patients with corrected transposition of the great arteries and two well developed ventricles are necessitated by symptoms related to associated defects. These most commonly are pulmonary stenosis with ventricular septal defect (VSD) resulting in right-to-left shunt, and regurgitation of the left-sided atrioventricular valve. Recent anatomic studies indicate that the specialized atrioventricular conduction system (AVC) in corrected transposition in situs solitus (L-transposition) emerges inferiorly and to the right of the pulmonary valve. It then proceeds along the superior and anterior margin of the VSD to its distal ramifications within the myocardium. Electrophysiologic delineation during surgery has confirmed this course of the intraventricular AVC relative to the VSD in corrected transposition in situs solitus (S,L,L). 1-4

This paper reports the electrophysiologic identification at surgery of the intra-atrial and intraventricular His conduction system in two patients with situs inversus of the viscera and atria, D-loop, D-transposition of the great arteries II,D,DI and dextrocardia, i.e., corrected transposition in situs inversus.

Methods

Table 1 summarizes the clinical, electrocardiographic, and anatomic features of the two patients. Following palliative surgery (Blalock-Taussig shunts) performed several years previously, both patients underwent correction because of increasing cyanosis and dyspnea on exertion.

Preoperative evaluation, including electrocardiogram, chest radiograph, cardiac catheterization, and biventricular angiography disclosed identical anatomic features: 1) dextrocardia, 2) situs inversus of the viscera and atria, 3) D-loop of the ventricles with atrioventricular discordance, 4) D-transposition of the great arteries with ventriculo-arterial discordance. In addition, hemodynamic data in conjunction with the angiographic findings indicated severe pulmonary stenosis and a ventricular septal defect with right-to-left shunting. Preoperative electrocardiographic data demonstrated a rightward inferior frontal plane P wave axis suggesting atrial inversion in both patients. Both patients had normal PR intervals before and following surgery; patient 2 had transient complete heart block for 17 days following surgery, and prolonged her PR by .04 sec. Both patients developed prolongation of their intraventricular conduction time (QRS interval) postoperatively.

After instituting cardiopulmonary bypass in each patient and prior to cross-clamping the aorta, the left-sided right atrium was incised and the mitral valve exposed. Upon retracting the posterior leaflet of the mitral valve, the entire perimeter of the ventricular septal defect could be seen. Nearly normothermic temperatures (35.5°C in patient 1 and 34.5°C in patient 2) were maintained during the electrophysiologic studies. A simultaneous scalar electrocardiogram (lead II) and a high right atrial (left-sided) bipolar electrogram were recorded in order to monitor atrioventricular conduction. A hand-held probe, with three bipolar pairs of electrodes (1 mm apart) and a diameter of 5 mm, was placed first on the endocardial surface of the right atrium (left-sided) in the area between the coronary sinus and the mitral valve; then within the ventricle it was positioned sequentially around the margin of the ventricular septal defect. 5-9 Each bipolar pair of electrodes was connected to a Hewlett-Packard Bioelectric 8811A amplifier and isolated from both ground and the recording apparatus by an isolation transformer. Electrograms were recorded at frequencies between 15 and 300 Hz. All tracings were monitored on a Hewlett-Packard 1308 A oscilloscope and recorded simultaneously on photographic paper moving at 100 mm/sec. The appearance of a deflection on the intracardiac electrogram during the isoelectric portion of the PR interval identified electrical activity in the specialized AVC system. 10 In one patient intra-atrial and in both patients intraventricular His electrograms were recorded. Following

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TABLE 1. Clinical, Electrophysiologic and Pathologic Data of the Two Patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age</th>
<th>PR interval (sec)</th>
<th>P Axis (deg)</th>
<th>QRS interval (sec)</th>
<th>Atrial situs</th>
<th>Vent. loop</th>
<th>Great vessel relation</th>
<th>Other</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>26 yr</td>
<td>0.17</td>
<td>+120°</td>
<td>0.09</td>
<td>inversus</td>
<td>dextro</td>
<td>D-transposition</td>
<td>PS, VSD</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>15 yr</td>
<td>0.12</td>
<td>+120°</td>
<td>0.09</td>
<td>inversus</td>
<td>dextro</td>
<td>D-transposition</td>
<td>PS, VSD</td>
</tr>
</tbody>
</table>

Abbreviations: ECG = electrocardiogram; PS = pulmonary stenosis; VSD = ventricular septal defect.

Figure 1. Schematic drawing of sagittal section through the left ventricle (left-sided) and right atrium (left-sided). The mitral valve (left atrioventricular) has been removed. The pulmonary stenosis, ventricular septal defect, and coronary sinus can be seen. The circles indicate relative placement of the exploring electrode probe: closed circles indicate the area where specialized atrioventricular conduction (AVC) tissue electrograms were obtained; open circles indicate the areas from which no specialized AVC electrograms could be recorded. Note the intra-atrial recording of specialized AVC tissue electrograms, near the coronary sinus, found in patient 1. Especially note the identification of the intraventricular specialized AVC tissue along the posterior and inferior margin of the ventricular septal defect.

Results

Patient 1 maintained atrioventricular conduction during the electrophysiologic studies. Following surgery, persistent mild interatrial right-to-left shunting (systemic oxygen saturation 85% by postoperative catherization) was noted. On the fourth postoperative day sinus node slowing with junctional escape mechanism and short runs of supraventricular tachyarrhythmia (sick sinus syndrome) occurred. Although atrioventricular conduction was maintained it was thought prudent to insert an epicardial demand pacemaker. Figure 1 illustrates the anatomic findings at surgery in both patients. The coronary sinus was found in the left-sided right atrium in its mirror image position. The left-sided atrioventricular valve was morphologically the mitral valve. The exploring electrode probe located the intra-atrial biphasic (fig. 2) His electrogram immediately medial (right) to the ostium of the coronary sinus. The probe was then placed around the rim of the VSD (fig. 1); AVC electrograms (fig. 3) were recorded along the posterior and inferior margin (fig. 1) of the ventricular septal defect before disappearing at approximately 6 o'clock. The anterior and superior margin of the ventricular septal defect was explored without finding a similar electrogram.

Patient 2 developed atrioventricular dissociation as soon as cardiopulmonary bypass was begun, prior to aortic cross-clamping and mapping. Figure 4 displays the simultaneous ECG and intraventricular electrogram recorded in this patient. The atrial impulse arose ectopically, low in the right atrium (LRA), and was conducted retrogradely to the high right atrium (HRA), as well as antegrade through the His bundle where it was blocked within the His-Purkinje system (HPS). Ventricular excitation was spontaneous and independent of atrial activation. Intra-atrial exploration for specialized AVC tissue proved unsuccessful. Intraventricular specialized conduction tissue electrograms were recorded through the exploring electrode probe at sites similar to those found in patient 1 (fig. 1). Patient 2's transient complete heart block persisted for 17 days before atrioventricular conduction resumed.

Both patients are alive and improved 12 to 18 months following surgery, with intact atrioventricular conduction.

Discussion

Corrected transposition of the great arteries, in contrast to complete transposition of the great arteries, implies not only ventriculo-arterial discordance but also ventriculo-atrial discordance. The morphologic configuration leading to this designation generally is situs solitus of the viscera and atria, L-loop of the ventricles, and L transposition of the great arteries S,L,L. A much less frequent form of corrected transposition of the great arteries is the mirror image of S,L,L and is composed of the following morphologic segments: situs inversus of viscera and atria, D-loop of the ventricles, and D-transposition of the great arteries L,D,D. In this alignment, both the ventriculo-arterial and atrioventricular discordances of corrected transposition are maintained.

The specialized AVC system in the normally developed heart proceeds in a relatively predictable course. Congenital heart defects which are not associated with abnormal intersegmental (atria-ventricles-great arteries) relationships, with the exception of endocardial cushion defects and muscular ventricular septal defects, are not associated with an abnormal course of the specialized AVC.
Corrected transposition of the great arteries of the usual type [S,L,L] may, however, be associated both with interruption of the atrioventricular conduction system resulting in complete heart block (12%), and with alteration in the course of both the intra-atrial and intraventricular portions of the specialized AV system. Corrected transposition of the great arteries of the usual type [S,L,L] may, however, be associated both with interruption of the atrioventricular conduction system resulting in complete heart block (12%), and with alteration in the course of both the intra-atrial and intraventricular portions of the specialized AV system.

The findings in these two patients provide several exceptions to the usual form of corrected transposition. First, the angiograms disclosed the morphologic configuration of situs inversus of the atria, D-loop of the ventricles, and D-transposition of the great arteries [I,D,D], corrected transposition of the great arteries in situs inversus. Second, the intra-operative electrophysiologic study revealed a relatively "normal" course of both the intra-atrial and the intraventricular portions of the specialized conduction system. This conclusion is supported by several obser-
vations. Previous reports of electrophysiological mapping at surgery have failed to demonstrate an intra-atrial specialized AVC electrogram in patients with corrected transposition of the great arteries in situs solitus $\{S,L,L\}$ despite diligent exploration; in contrast one of our patients with corrected transposition in situs inversus demonstrated intra-atrial His electrograms immediately medial to the orifice of the coronary sinus (figs. 1, 2), suggesting that the atrioventricular (His) bundle was located in the left-sided right atrium and arose from an atrioventricular node in the "normal" posterior location. In addition, atrioventricular (His) electrograms were recorded in both patients from the posterior and inferior border of the VSD (figs. 1, 3, 4) in contradistinction to the superior and anterior location found in patients with corrected transposition of the $\{S,L,L\}$ configuration. Finally, the intraventricular electrograms were obtained through the mitral valve from the septal surface of the morphologic left ventricle, and therefore most likely represented recording from both the intraventricular His bundle and left bundle branch of the specialized AVC tissue.

Lev$^2$ and Anderson$^4$ have studied the conduction system in corrected $L$-transposition with an $L$ (inverted) ventricular loop and situs solitus $\{S,L,L\}$. Both investigators found that the major atrioventricular node was along the anterior margin of the right atrioventricular valve in the right atrium, behind the pulmonary valve anulus. In the eleven specimens which Anderson studied,$^4$ a primitive, posterior hypoplastic atrioventricular node was found in its normal location in the right atrium, and in all but one it was isolated from the remainder of the specialized AVC system. The intraventricular His bundle — in contrast to the posterior and inferior location in normally looped hearts with ventricular septal defects — was found to emerge from the anterior node and pass into the anterior aspect of the roof of the pulmonary outflow tract, lying just inferior to the valve ring. It then encircled the right lateral margin of the valve ring to reach the expanded portion of the ventricular septum. In hearts with intact ventricular septa, it was anterior to the region of the membranous septum. In hearts with ventricular septal defects, it was found in the right side of the anterior rim of the defect. The bundle branches were inverted in accord with ventricular inversion. Anderson$^4$ con-

Figure 4. Simultaneous surface electrocardiogram (ECG) (lead II) and three intraventricular electrograms recorded from patient 2. The high right atrial (left-sided) electrograms were recorded through a parallel jaw clip attached to the base of the right atrial appendage (left-sided). Immediately after the beginning of cardiopulmonary bypass, but prior to mapping and aortic cross clamping, atrioventricular dissociation developed, as demonstrated by the independent but nearly isorhythmic atrial and ventricular rates (see ladder diagram). Local intraventricular electrograms were recorded through the HBE exploring probe at sites around the margin of the ventricular septal defect. The small $A$ wave, coincident in time with the $P$ wave, most likely represents recording from a low atrial or junctional focus; this impulse is propagated antegradely to the His system, where it is blocked. In addition the right atrial or junctional focus, $A$, is propagated retrogradely to the HRA. The ventricular mechanism proceeds independently of this mechanism. The significant point, however, is that the His bundle electrogram was identified along the posterior and inferior margin of the VSD, similar to case 1. $P = P$ wave of surface ECG, $R = R$ wave of surface ECG, $V =$ ventricular electrogram.
cluded that the abnormal course of the specialized AVC tissue in corrected transposition of the great arteries in situs solitus left-handed heart position was related to the malalignment of the interatrial and interventricular septa, itself a consequence of discordant bulboventricular looping.

In his anatomic studies of a patient with situs inversus of the viscera and atria, D-bulboventricular loop, and L-malposition of the great arteries, i.e., anatomically corrected malpositions L, D, L,18 Anderson found the atrioventricular node posteriorly, adjacent to the coronary sinus, in the left-sided right atrium. The intra-atrial His bundle emerged from this posterior node, traversed the posterior wall of the right atrium subendocardially to reach the central fibrous body where it joined the common bundle to proceed normally into the ventricles. This disposition of the specialized AVC tissue conforms most closely to that found by the electrophysiologic technique in our two patients. These findings suggest that the course of the specialized AVC tissue is related to the type of ventricular loop: posterior and inferior to the VSD in the D-loop, and superior and anterior to the VSD in the L-loop.

The finding of a left-sided intra-atrial electrogram, adjacent to the coronary sinus in one of our two patients with corrected transposition (with D-loop hearts) and situs inversus differs from the observations in patients with corrected transposition (with L-looped hearts) and situs solitus, and strongly suggests a left-sided His (atrioventricular) bundle and, by inference, a left-sided atrioventricular node. James and Anderson,95 have postulated a dual origin of the atrioventricular node: one origin, an area of specialized tissue near the coronary sinus arising from the left sinus horn (in situs solitus); the other a bundle of cells originating from the atrioventricular canal. This dual origin may account for the two "nodes" found in the L-looped hearts: the anterior node, arising from and communicating with the atrioventricular bundle, and the posterior node, adjacent to the coronary sinus, remaining a hypoplastic remnant that fails to communicate with the atrioventricular (His) bundle.

Taking these observations into consideration one may postulate that, in patients with abnormal intersegmental relationships, as in the transpositions, but with two well developed ventricular sinuses, the cardiac loop may relate both to the location of the ventricular specialized AVC tissue and to the latter's continuity with the atrial specialized AVC tissue. In short, a D-loop will be associated with a normal posterior atrioventricular node continuous with the normal atrioventricular (His) bundle and intraventricular conduction system, whereas an L-loop will be associated with both an anterior node continuous with the inverted and anterior intraventricular system with or without a usually isolated posterior hypoplastic node. This posterior node, whether single (and normal) as in D-loop hearts or in association with the anterior node as in L-looped hearts, will be in the morphologic right atrium which will be right or left-sided depending upon the atrial situs. Concerning this hypothesis it would be of interest to study electrophysiologically the specialized AVC system both in patients with mirror image dextrocardia II, L, L, and in patients with complete transposition in situs inversus II, L, L. We have attempted electrophysiologic mapping in one patient of each type, but have been unsuccessful because of technical difficulties.

In our two patients, the exact site of penetration of the His bundle descending from the left-sided right atrium through the atrioventricular ring to reach the ventricular specialized conduction tissue is unknown; one might postulate, given the above cited studies and electrophysiologic findings at surgery, that the intra-atrial His bundle of the specialized conduction system progresses from the left-sided right atrium, reaches the point of junction (membranous septum) between the ventricular and atrial septa, traverses the atrioventricular ring and joins the common bundle (intraventricular His bundle) to continue normally into the intraventricular bundle branches. Although the patients demonstrate intact atrioventricular conduction postoperatively, the prognosis is unknown.

Acknowledgment

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References

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