Electrophysiologic Studies in Patients with Ventricular Inversion and “Corrected Transposition”

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SUMMARY We evaluated the intracardiac conduction intervals using His bundle recordings in 40 patients with ventricular inversion and l-transposition of the great arteries. Twenty-nine subjects had 1:1 atrioventricular (AV) conduction. In 15 of those with normal PR intervals and QRS durations, the conduction intervals were not different from those of subjects with normal hearts. In the 14 patients with first-degree AV block, the block was located between the sinus node and AV node in four, between the low right atrium and bundle of His in seven, and below the common bundle of His in four.

In 11 subjects with complete AV block, the site of block was above the site of the His potential in four, below in two and within the His bundle in one. In four patients we could not record a His potential and thus could not localize the site of block. Complete block below the His recording site was associated with syncope in one patient and sudden death in another. His bundle recording is a safe technique for studying the conduction system in children with ventricular inversion and l-transposition of the great arteries.

VENTRICULAR INVERSION with l-transposition of the great arteries occurs in 1.4% of congenital heart defects. Since it is physiologically a “corrected transposition,” the associated anomalies and conduction defects determine the prognosis. Ventricular inversion frequently is associated with conduction disturbances ranging from first-degree atrioventricular (AV) block to complete (third-degree) AV block. These blocks may be present at birth, may progress gradually or rapidly, or may occur during surgical treatment of associated defects.

The anatomical course of the conduction system in hearts with ventricular inversion has been delineated by mapping during open heart surgery and by serial histopathologic sections. The usual posterior AV node in these hearts fails to connect to a bundle of His. There is an accessory anterior AV node that gives rise to the bundle of His and courses anteriorly around the pulmonary annulus.

The techniques of intracardiac recording and electrical stimulation during cardiac catheterization have not been applied widely to patients with ventricular inversion, although such patients are frequently subject to dysrhythmias. This may be due in part to the suggestion that it may be impossible to record the bundle of His potentials in this anomaly, or that doing so may be dangerous.

In this study we investigated the feasibility of His bundle recording in patients with ventricular inversion and associated congenital heart defects and conduction disturbances, and report intracardiac conduction times from patients with both normal and abnormal conduction by surface ECG.

Methods

His bundle recording was attempted in 40 subjects during diagnostic cardiac catheterization performed to establish the nature and severity of associated cardiac defects. Each subject had ventricular inversion and l-transposition of the great arteries documented by biplane cineangiography. The patients ranged in age from 17 hours to 29 years. Eleven patients had complete AV block diagnosed by surface ECG before catheterization (only one of which resulted from surgery) and 14 patients had first-degree AV block. All cardioactive drugs were discontinued 36 hours before study. Each patient was premedicated with meperidine 2 mg/kg, chlorpromazine 0.5 mg/kg and promethazine 0.5 mg/kg 30 minutes before study. Informed, written consent was obtained from the patients or their parents. Each electrophysiologic study was carried out before angiography.

The technique for recording the electrogram was the same as that used in subjects with noninverted ventricles (fig. 1). A bi-, tri- or quadripolar electrode catheter with interelectrode distances of 1–10 mm was introduced percutaneously from the femoral vein. In most instances a bipolar catheter with one interelectrode distance of 1 mm and one of 10 mm was used to obtain two bipolar tracings (one 1 mm and one 10 mm). The tip of the catheter for recording the His bundle potential was placed well inside the right-sided, morphological left ventricle, and then withdrawn across the AV valve with clockwise rotation. If necessary, this maneuver was repeated until clear low right atrial (LRA), His bundle, and ventricular (V) inscriptions were seen. In some instances a “left” bundle branch potential also was recorded with the catheter farther into the ventricle. Recordings of the intracardiac potentials, the surface ECG (usually leads I, II and III) and time markings were made simultaneously on photographic recording paper at 100 and 200 mm/sec paper speed. The filter setting for
the intracardiac potentials was 40–500 Hz. Intervals were measured between the first sharp inscription of each complex. Each subject was in a stable hemodynamic condition at the time of electrophysiologic study.

Results

The apparent bundle of His depolarization potential could be recorded in 32 of 40 subjects. Confirmation that these potentials were recorded from the bundle of His, rather than the “left” bundle branch, or an atrial
structure, is based on the following: 1) there was a large LRA potential associated with the His potential, 2) atrial pacing in the six subjects resulted in prolongation of the LRA-His interval, thus separating the His potential from the atrial depolarization, 3) His bundle pacing attempted in the most recent patient was successful, resulting in a normal QRS and a stimulus-to-ventricular interval identical to the H-V interval (fig. 2), and 4) in 10 subjects a separate left bundle potential was recorded with the catheter advanced further into the ventricle.

Tables 1 and 2 are a summary of the electrophysiologic data in patients with and without first-degree and complete AV block. Twenty-nine of the 40 had 1:1 AV conduction. Fifteen of these 29 subjects with 1:1 AV conduction also had PR intervals in the normal range of patients with normal conduction. The intervals for conduction of the sinus impulse from the high (HRA) to low right atrium (HRA-LRA), from the LRA to the bundle of His (LRA-H) and from the bundle of His to the ventricular myocardium (H-V) were within the normal limits for children with normally related ventricles studied in our laboratory. Fourteen of the 29 subjects with 1:1 AV conduction had PR intervals greater than the upper limit of normal for patients with normally related ventricles. Twelve of the 14 had one or more intracardiac intervals that were prolonged compared with patients with normal PR intervals with normally related ventricles studied in our laboratory. The HRA-LRA interval was prolonged (> 40 msec) in four subjects. The LRA-H interval was prolonged (> 120 msec) in
### Table 2. Ventricular Inversion: Patients with Complete Atrioventricular Block

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age</th>
<th>QRS interval (msec)</th>
<th>RR interval (msec)</th>
<th>PP interval (msec)</th>
<th>LRA-H interval (msec)</th>
<th>H-V interval (msec)</th>
<th>&quot;Lb&quot;-V</th>
<th>Site of block</th>
</tr>
</thead>
<tbody>
<tr>
<td>30</td>
<td>2 y</td>
<td>80</td>
<td>875</td>
<td>670</td>
<td>†</td>
<td>30</td>
<td>†</td>
<td>His</td>
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<tr>
<td>31</td>
<td>12 y</td>
<td>120</td>
<td>688</td>
<td>680</td>
<td>†</td>
<td>55</td>
<td>31</td>
<td>His</td>
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<td>80</td>
<td>1250</td>
<td>470</td>
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<tr>
<td>33</td>
<td>7 y</td>
<td>80</td>
<td>1500</td>
<td>715</td>
<td>†</td>
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<tr>
<td>34</td>
<td>13 y</td>
<td>140</td>
<td>1330</td>
<td>650</td>
<td>84</td>
<td>51</td>
<td>†</td>
<td>His</td>
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<tr>
<td>35</td>
<td>7 y</td>
<td>100</td>
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<td>750</td>
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<td>21</td>
<td>*</td>
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<td>36</td>
<td>17 h</td>
<td>100</td>
<td>1090</td>
<td>430</td>
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<td>*</td>
<td>*</td>
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<tr>
<td>37</td>
<td>2 y</td>
<td>90</td>
<td>820</td>
<td>580</td>
<td>†</td>
<td>40</td>
<td>†</td>
<td>† His</td>
</tr>
<tr>
<td>38</td>
<td>19 y</td>
<td>120</td>
<td>610</td>
<td>690</td>
<td>150</td>
<td>70</td>
<td>†</td>
<td>† His</td>
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<tr>
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<td>22 m</td>
<td>90</td>
<td>720</td>
<td>500</td>
<td>150</td>
<td>Blocked</td>
<td>20</td>
<td>† His</td>
</tr>
<tr>
<td>40</td>
<td>29 y</td>
<td>90</td>
<td>950</td>
<td>800</td>
<td>120</td>
<td>70</td>
<td>†</td>
<td>In His</td>
</tr>
</tbody>
</table>

*No His potential recorded.
†No electrogram recorded.

Abbreviations: h = hours; m = months; y = years; LRA-H = low right atrial-His; H-V = His-ventricle;
"Lb"-V = "left" bundle-ventricle; † His = above the His bundle recording site; † His = below the His bundle recording site; In His = two His potentials recorded.

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seven (fig. 2), while the H-V interval was prolonged (> 55 msec)\(^2\) in four (fig. 3).

One patient developed 2:1 AV block during an episode of atrial tachycardia (fig. 3). No other patient had second-degree AV block during the study. Eleven of the 40 subjects had preexisting complete AV block at the time of study. This had been documented by three or more surface ECGs in each subject with a regular RR interval and no evidence of "capture." In four subjects, no His depolarization could be recorded. The block was above the site of recording of the His bundle depolarization in four of the seven in whom it could be recorded (fig. 4), below the His in two (fig. 5) and in the bundle of His in one. This was

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**Figure 3.** Simultaneous recording of surface ECG lead II and a His bundle electrogram (HBE) in patient 2. On the top tracing the patient is in sinus rhythm with a normal low right atrium (LRA) to His (H) interval and a prolonged H-to-ventricle (V) interval. In the second panel the patient is in atrial flutter with 2:1 block in the atrioventricular node, as well as prolongation of the LRA-H interval, the His duration and the H-V interval.
documented by finding a His spike before each QRS, but not after each atrial depolarization in patients with block above the His. (fig. 4). In those with the block below the His there was an H after each atrial depolarization (fig. 5). The one subject with complete block in the bundle of His has been described in detail previously. The QRS duration was prolonged in one subject with block below the H and normal in the other. The QRS was prolonged in two of the four with block above the H and was normal in the patient with intrahisian block. Conduction below the His bundle was normal in three of the four patients with block above the His bundle and prolonged in one. The LRA-H interval was prolonged in one of the two subjects with block below the His bundle.

Two of the patients with complete antegrade block had intact retrograde conduction. In one this was observed during accelerated junctional rhythm and in one during ventricular pacing. We could not determine whether the retrograde conduction was through a bundle of His or an accessory pathway. One of the patients with block distal to the bundle of His demonstrated occasional antegrade conduction to the ventricle with H-V conduction time of 50 msec. Confirmation that these were conducted beats is based on the following: 1) consistent shortening of the RR interval by an amount that varied according to the relationship between P and QRS complexes, 2) a constant H-V interval for these beats, and 3) appearance of the complex only when atrial depolarization occurred.

FiguRE 4. Simultaneous recording of surface ECG leads I, II, and III, with two His bundle electrograms (HBE) in a patient with complete antegrade atioventricular (AV) block in the AV node. On the left panel the catheter is positioned for His bundle recording, while on the right it is farther into the ventricle, recording the right-sided (morphologically left) bundle branch potential. The low right atrial (LRA) depolarization is much smaller when the “left” bundle is being recorded. FAP = femoral artery pressure.

FiguRE 5. Simultaneous tracing of surface ECG leads I, II, and III with a right atrial (HRA) and two His bundle electrograms (HBE) in a patient with complete atrioventricular block distal to the bundle of His. There is a His potential after each atrial depolarization and none preceding the ventricular depolarization. Abbreviations are as in previous figures.
in 310–330 msec after a preceding QRS complex. The other patient with block below the His depolarization also had occasional conducted beats.

Two subjects were studied because of frequent episodes of supraventricular tachycardia that were not responsive to digoxin or a combination of digoxin and propranolol. Supraventricular tachycardia was induced during catheterization by single premature atrial or ventricular stimuli, thus suggesting a reentry mechanism. The atrial activation sequence showed the left atrium (coronary sinus) to be activated first during supraventricular tachycardia and during ventricular pacing, suggesting the presence of a left-sided concealed accessory pathway in each subject. 13

No complications occurred during any of these studies.

Discussion

His bundle recording has improved our understanding and treatment of cardiac dysrhythmias in adults and children. 14 Although the first two reports were in patients with congenital heart disease, the technique has been used most often in patients with structurally normal hearts. 14, 15 Only a few cases have been reported of bundle of His recordings in patients with ventricular inversion and corrected transposition, despite the fact that cardiac dysrhythmias are particularly common in such patients. 6-8, 16-18 This may be because of a feeling that it may be difficult or even dangerous to perform such studies in patients with ventricular inversion.

The present study, together with our previous report and the others in the literature, 6-8, 16-18 make it clear that His bundle recording is both feasible and safe in children and adults with ventricular inversion.

Normal Conduction in Ventricular Inversion

This study reveals that the normal conduction intervals in patients with ventricular inversion are the same as those in hearts with normally related ventricles. 10, 12 Patients with normal conduction on surface ECG uniformly had normal conduction intervals. Thus, the inversion of the conduction system does not, per se, result in any abnormalities of conduction.

Abnormal Conduction

The site of AV block in ventricular inversion may be important in determining the prognosis. The site of first-degree AV block found in this study included the AV node, the bundle of His and the bundle branch system. The patients with the longest PR intervals had multiple sites of block. The sites of complete AV block were also diffuse and, in some cases, multiple. The site of complete AV block in infants with ventricular inversion could not be addressed in this study, since no His potentials could be recorded in these infants. Perhaps the bundle of His is so abnormal that no potential can be recorded.

It is not possible to generalize about prognostic implications because of the small sample of patients in this study. Several features, however, deserve comment. 1) Of the two subjects with complete block below the His potential, one required pacing for syncope attacks and the other died suddenly. Both neonates (in neither of which the His potential could be recorded) also required pacing early in life because of congestive heart failure and have done well. Only one of the subjects without complete AV block at the time of study subsequently required pacing. This patient had first-degree AV block in the AV node at the time of study. He later developed Mobitz type II second-degree AV block and syncope during sinus tachycardia. The site of the type II block was not localized. He also had severe left AV valve regurgitation and ventricular septal defect. Six months after pacemaker insertion, he died in his sleep. We do not know if the death was due to a dysrhythmia or congestive heart failure.

The finding that retrograde conduction was intact in two patients with complete AV block and that intermittent antegrade conduction occurred in two others indicates that complete AV block is not always due to complete anatomic discontinuity of the conduction tissues. Thus, it is probably wise to use QRS-inhibited pacemakers when a pacemaker is necessary. The findings in the patients with supraventricular tachycardia indicate that multiple catheter studies with atrial and ventricular extrastimulus study are useful in patients with ventricular inversion, as they are in other children in establishing the mechanism of complex dysrhythmias. 19 The findings help to determine the optimal plan of management in these patients, such as eventual surgical division of the bundle of Kent at the time of repair of other cardiac defects (e.g., ventricular septal defect).

His bundle recording is possible and usually successful in subjects with ventricular inversion. The techniques and normal values are similar to those in normal hearts. There appears to be no increased risk.

References

Depressed Responsiveness to Vasoconstrictor and Dilator Agents and Baroreflex Sensitivity in Conscious, Newborn Lambs

W. Thomas Manders, B.S., Massimo Pagani, M.D., and Stephen F. Vatner, M.D.

SUMMARY The effects of vasoconstrictors and vasodilators were compared in conscious, newborn lambs and adult sheep instrumented with electromagnetic flow probes on the ascending aorta and catheters in the thoracic aorta. Methoxamine, angiotensin II, norepinephrine, nitroglycerin and isoproterenol were administered intravenously to evaluate their effects on arterial pressure, cardiac output and systemic vascular resistance (SVR). The difference in response between adults and newborns was most apparent with methoxamine. Methoxamine, 400 µg/kg, i.v., which increased mean arterial pressure by 57 ± 6% and SVR by 278 ± 27% in newborn lambs, caused greater increases (p < 0.01) of 81 ± 8% and 1418 ± 141%, respectively, in the adults. Responses also differed significantly between newborn and adult animals to norepinephrine, angiotensin II, nitroglycerin and isoproterenol. In a second group of animals in which smaller amounts of methoxamine and isoproterenol were injected directly into the terminal aorta, changes in arterial flow and resistance were examined. Again, both vasoconstrictor and vasodilator responses were more marked in adults than in newborns. Finally, the sensitivity of the arterial baroreceptor reflex was evaluated by comparing the regression of pulse interval (PI) on systolic arterial pressure (SAP) after an intravenous dose of methoxamine in conscious, adult and newborn animals. The PI/SAP slopes in adult sheep, 45.4 ± 3.5 msec/mm Hg, were significantly greater (p < 0.01) than in newborn lambs, 11.7 ± 2.2 msec/mm Hg.

THE REQUIRED therapeutic dose of digitalis is relatively greater in the newborn patient than in the adult.1-3 While the responsiveness to vasoactive drugs might also demonstrate age-dependent differences, results of prior studies have conflicted. For instance, studies conducted in anesthetized animal preparations4-9 or isolated vessel strips10-12 have shown both an increased8, 10 and decreased4, 9, 11, 12 responsiveness of the neonatal peripheral circulation to vasoactive agents.

In this investigation we compared the peripheral vascular responses to pharmacological agents that constrict or dilate peripheral vessels in conscious newborn lambs and adult sheep, with all control mechanisms intact and the complicating influences of a general anesthetic absent.13-15 Since several reflex adjustments are likely to be induced by the changes in arterial pressure after systemic drug administration, two groups of animals were studied. In one group, drugs were administered i.v. and responses were examined in terms of changes in systemic arterial pressure (SAP), blood flow and systemic vascular resistance (SVR). In a second group smaller amounts of the drugs were injected intra-arterially into the ter-
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