Electrophysiologic Delineation of the Intraventricular His Bundle in Two Patients with Endocardial Cushion Type of Ventricular Septal Defect

MACDONALD DICK, M.D., DOUGLAS M. BEHRENDT, M.D.,
KENNETH E. JOCHIM, PH.D., AND ALDO R. CASTANEDA, M.D.

SUMMARY Two patients who had an endocardial cushion type of ventricular septal defect underwent electrophysiologic studies for detection of specialized conduction tissue during operative repair. In one patient, with an inferior leftward frontal plane QRS axis on the ECG, we recorded an intraventricular His bundle electrogram anterior and superior to the ventricular septal defect. In the other patient, with a mean QRS axis perpendicular to the frontal plane and a normal PR interval, we obtained intraventricular His bundle electrograms from both the anterosuperior and posteroinferior margins of the defect, suggesting dual atrioventricular conduction tracts (branching intraventricular His bundle). These anatomic and electrophysiologic findings may account for the more normally oriented QRS frontal plane axis on the surface ECG of both of these patients and support the hypothesis that the changes observed on the ECGs of patients with the various forms of endocardial cushion defect can be explained by alterations in the anatomic configuration of the specialized atrioventricular conduction tissue.

THE RELATIONSHIP between the intraventricular His bundle and defects of the ventricular septum has been delineated by both pathologic and electrophysiologic techniques.1-7 In patients with either membranous ventricular septal defects or complete endocardial cushion defects, the His bundle parallels the posteroinferior margin of the defect, deviating 1–3 mm from the margin of the defect to adjacent sites on either side.8 In contrast, the intraventricular His bundle in the majority of patients with defects of the muscular ventricular septum proceeds normally, unrelated to the defect.8 In patients with the endocardial cushion type of ventricular septal defect, the intraventricular His bundle parallels the posteroinferior margin of the defect, similar to the course found in patients with more typical forms of both partial and complete endocardial cushion defects. To determine the location of the intraventricular portion of the His bundle in two patients with an endocardial cushion type of ventricular septal defect, we recorded intraventricular His bundle electrograms during surgery. The findings differ in part from pathologic descriptions of the relationship between the endocardial cushion type of ventricular septal defect and the intraventricular specialized conduction tissue.8

Materials and Methods

The clinical, electrocardiographic and angiographic data of both patients are summarized in table 1.

Patient 1, included in a review of intraoperative recording of the His bundle electrogram,6 underwent pulmonary artery banding in the first year of life for pulmonary artery hypertension and congestive heart failure before complete repair at age 11 years. The second patient underwent, at 11 months of age, ventricular septal defect closure because of a large left–right shunt, pulmonary arterial hypertension, and congestive heart failure. After surgery, a residual ventricular septal defect was suspected. One year later, catheterization and reoperation showed a left ventricular–right atrial shunt. Left ventricular biplane cineangiograms disclosed a large ventricular septal defect in each patient, but without a goose-neck deformity of the left ventricular outflow tract. Patient 1’s ECG demonstrated an inferior and leftward (+80°) QRS axis in the frontal plane. Patient 2’s ECG revealed a QRS axis perpendicular to the frontal plane (fig. 1). After the first operation, right bundle branch block and a leftward superiorly oriented frontal plane axis developed and persisted through the second operation (fig. 2). M-mode echocardiograms in both patients demonstrated a dilated left ventricle, but did not suggest an endocardial cushion type of defect.6

The diagnosis of an endocardial cushion type of ventricular septal defect was made at surgery because in both patients a large posteroinferior defect was present behind the posterior and septal leaflets of the tricuspid valve, with no intervening muscle between the defect and the atrioventricular groove where the tricuspid valve inserted. In addition, in patient 2 chordae from the posterior leaflet crossed the ventricular septum through the septal defect to insert in the left ventricle, indicating overriding of the tricuspid valve (fig. 3). Patients 1 and 2 are alive and well 4 and 2 years, respectively, after surgery, with intact atrioventricular conduction.

Recording of the intracardiac specialized conduction tissue electrograms was performed at each operation (one in patient 1 and two in patient 2) in the manner previously described.7 After institution of car-
diopulmonary bypass the area suspected of containing atrioventricular conduction tissue was explored with a hand-held probe 5 mm in diameter, with three bipolar pairs of electrodes (1 mm apart). Each patient's esophageal temperature was maintained at 30–37°C. Each bipolar pair of electrodes was connected to a Hewlett Packard high-impedance differential amplifier (MN8811A) or Electronics for Medicine ECG amplifier (VET) and was isolated from both ground and the recording apparatus by an isolation transformer. Electrograms were recorded at a bandwidth of 15–300 Hz. The tracings were monitored on a Hewlett Packard 1308A oscilloscope or Electronics for Medicine DR12 switched-beam oscilloscope and recorded simultaneously on photographic paper moving at 100 mm/sec. During the mapping procedure atrioventricular conduction was maintained by normal sinus rhythm in patient 1. In patient 2 atrioventricular dissociation developed with the appearance of an escape His bundle rhythm (fig. 3).

Results

In the first patient, atrial His bundle electrograms were located in the usual position, anterior and medial to the orifice of the coronary sinus. Intraventricular specialized conduction tissue electrograms were then recorded parallel to the anterosuperior margin of the ventricular septal defect. No electrograms were found along the posteroinferior margin of the defect. In the second patient (fig. 3), intraventricular specialized conduction tissue electrograms were recorded along the superior margin of the ventricular septum; in addition, as the probe was placed sequentially in 5-mm steps along the septal crest, intraventricular electrograms were again recorded at approximately 5 o'clock on the ventricular septum. No electrograms were recorded along the intervening ventricular crest (fig. 3). The posterior margin of the defect beneath the posterior leaflet was not explored. At the second operation we explored previously examined sites for intraventricular His electrograms; positive recordings were obtained only along the inferior margin of the ventricular septal defect (fig. 3, site B), and not along the anterosuperior margin.

Discussion

These electrophysiologic findings indicate an unexpected and previously unreported location of the in-

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<tr>
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<td>PAB (1 year old)</td>
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<td></td>
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<td>Axis + 80°</td>
<td>No goose-neck deformity</td>
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<td>PR 0.14 second</td>
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<td>QRS 0.06 second</td>
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<tr>
<td>2</td>
<td>11 months</td>
<td>Perpendicular to frontal plane</td>
<td>Large VSD (Qp/Qs &gt; 2:1)</td>
<td>PAH, CHF, OTV</td>
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<td>PR 0.12 second</td>
<td>No goose-neck deformity</td>
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<td>QRS 0.06 second</td>
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Abbreviations: CHF = congestive heart failure; OTV = overriding tricuspid valve; Qp/Qs = pulmonary blood flow/systemic blood flow ratio; PAB = pulmonary artery band; PAH = pulmonary artery hypertension; PR = PR interval; VSD = ventricular septal defect; QRS = QRS duration.

**Figure 1.** Preoperative 12-lead ECG in patient 2. The mean QRS is perpendicular to the frontal plane. The initial forces are rightward and inferior and are followed by more leftward superiorly directed forces. There is no preexcitation. This ECG configuration is compatible with an endocardial cushion type of ventricular septal defect, but not pathognomonic.
traventricular His bundle in patients with endocardial cushion type of ventricular septal defect. The diagnosis of this type of defect is often difficult. A superior leftward mean frontal plane QRS axis suggests a defect involving the atrioventricular canal region; however, 16% of membranous ventricular septal defects may have a similarly directed axis; in addition, the absence of a goose-neck deformity of the left ventricular outflow tract on the cineangiogram does not exclude this diagnosis. One’s suspicion is usually aroused, as was the case in these patients, by a cineangiogram that reveals a large defect that is not in the usual subaortic membranous septum. Two-dimensional echocardiography, unavailable for these

\[ \text{FIGURE 2. Selective leads from ECGs obtained after both operations in patient 2. There is no difference between the two postoperative ECGs. Compared with the preoperative ECG (fig. 1), there is now right bundle branch block and a leftward superiorly oriented QRS axis in the frontal plane.} \]

\[ \text{FIGURE 3. (center) Artist’s depiction of the surgical anatomy of the endocardial cushion type of ventricular septal defect in patient 2. The right atrial anterior wall has been removed; the retractor is placed within the anterior margin of the tricuspid valve. The ventricular septum can be seen; the chordae of the posterior leaflet of the tricuspid valve can also be seen traversing the defect to insert within the left ventricle. Panels A and B are recordings of intracardiac electrograms (top tracing) and simultaneous ECG (bottom tracing) obtained from two different sites (A and B in center panel). There is atrioventricular dissociation with a His bundle escape rhythm. No electrograms were recorded from intervening sites (ventricular septum). The lack of anatomic contiguity of the two recorded electrograms suggests two separate tracts of specialized atrioventricular conduction tissue. Dotted and dashed lines indicate the intra-atrial location (black dots and dashes) and the intraventricular location (white dots and dashes) of conduction tracts confirmed by His bundle recordings. The black and white dots indicate the anterosuperior tract in both patients, whereas the black and white dashes indicate the posteroinferior tract in patient 2. The specialized conduction tissue electrogram was generated by a three-channel summing amplifier from the bipolar signals that were recorded by the tripolar probe and modified by a full wave rectifier circuit. } \]

\[ \text{AV = atrioventricular node; } H = \text{His bundle electrogram; IVC = inferior vena cava; } \ldots = \text{posteroinferior tract; } \ldots = \text{anterosuperior tract; } P = \text{P wave; SVC = superior vena cava; } V = \text{ventricular electrogram.} \]
two patients, would further enhance the accuracy of the preoperative diagnosis by demonstrating a defect in the posterior ventricular septum. The diagnosis of an endocardial cushion type of ventricular septal defect in these two patients was confirmed at surgery by two observations: (1) the large posterior ventricular septal defect with absence of intervening muscle between the posterior margin of the defect and the insertion of posterior leaflet of the tricuspid valve along the atroventricular groove, and (2) the associated finding of an overriding tricuspid valve in patient 2.

Endocardial cushion defects are associated with a superior leftward frontal plane QRS axis. In pathologic specimens of endocardial cushion defects, Lev and Feldt et al. described a posteroinferior displacement of the atroventricular node and the penetrating and branching portions (intraventricular) of the His bundle, as well as hypoplasia of the anterior fibers, and relatively early origin of the left bundle fibers from the common bundle. Feldt and associates have postulated that this anatomic configuration results in early activation of the inferior and posterobasal portion of the left ventricle and later activation of the superior anterior and more leftward portions of the left ventricle, resulting in a leftward and superiorly oriented frontal plane vector. Studies of canine endocardial activation and both canine and human epicardial activation in the presence of incomplete endocardial cushion defects (ostium primum) demonstrated earlier-than-expected activation of the posteroinferior left ventricle and later-than-expected activation of the anterosuperior left ventricle. In our two patients the absence of a leftward superior frontal plane axis suggests a normally activated left ventricle, and leads us to postulate that the anteriorly—superiorly located intraventricular His bundle lies in a relatively normal location, in contrast to the usual posteroinferior displacement found in the patient with endocardial cushion defect, and accounts, at least in part, for the normally oriented frontal plane axis.

We cannot explain the variation in the location of the specialized atrioventricular conduction tissue in these patients. One of the criteria recently advanced for formation of normally located atrioventricular conduction tissue is extension of the posterior (and inferior) muscular septum, involving the endocardial cushions, to the crux of the heart, establishing union posteriorly between atrially and ventricularly located conduction tissue. Despite being hypoplastic and "scooped out," the muscular septum in patients with endocardial cushion defects has been shown to reach the crux, and thereby establish sufficient contact for development of relatively normally located (posteroinferior) conduction tissue. In view of this observation and our electrophysiologic findings, one can postulate that in patient 1, in the absence of an intratral component to the defect, the atroventricular node was normally placed, as was demonstrated. However, the muscular ventricular septum might have failed to reach the crux posteriorly, thereby interrupting the normal union of the penetrating and branch-

ing His bundle. Accessory conduction tissue crossed the atroventricular junction at a more anterosuperior site. In patient 2, the findings of two intraventricular electrograms dissociated in space on the ventricular septum suggests a dual His bundle; one His bundle crosses, as in patient 1, the atrioventricular junction at the anterosuperior margin (anterior septal commissure of the tricuspid valve) of the ventricular septal defect; this was interrupted at the first surgery. The second His bundle, in a manner similar to that found in patients with complete endocardial cushion defects, and perhaps because of extension of the posterior muscular septum to the crux, parallels the posteroinferior margin of the defect, and remains intact after the second operation. The posteroinferior His bundle electrogram at the second operation and the absence of the anterosuperior His electrogram recorded at the first operation strongly suggest that postoperative atrioventricular conduction is dependent on the posterior His bundle, and that the QRS frontal plane leftward superior shift is related to the loss of early anterior activation of the left ventricle previously excited via the anterior tract and its radiations.

The clinical importance of these findings is apparent. To avoid interruption of atrioventricular conduction, repair of a ventricular septal defect of any type requires careful suture placement, especially near suspected sites of specialized atrioventricular conduction tissue. Endocardial cushion type of ventricular septal defects may be associated with unexpected and atypical sites of atrioventricular conduction tissue. We therefore recommend electrophysiologic identification of atrioventricular specialized conduction tissue in patients with this form of ventricular septal defect.

**Acknowledgment**

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M Dick, D M Behrendt, K E Jochim and A R Castaneda

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