Electrophysiological Delineation of the Specialized A-V Conduction System in Patients with Corrected Transposition of the Great Vessels and Ventricular Septal Defect

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SUMMARY
The specialized atrioventricular (A-V) conduction system was electrophysiologically delineated during open-heart surgery in four patients with congenitally corrected transposition of the great vessels and associated ventricular septal defect and one patient with single ventricle. Two consistent observations were made: 1) In no case was the specialized A-V conduction system found in the right atrium, whether or not there was a coronary sinus ostium present. 2) Specialized A-V conduction system electrograms were never delineated posterior to the ventricular septal defect, in contradistinction to this usual location in hearts having ventricular septal defects associated with other congenital lesions. In three of five patients, the initial course of the A-V conduction system of the ventricles was delineated between the anterior aspect of the ventricular septal defect and the pulmonary artery. In one patient the proximal portion of the A-V conduction system was delineated on the anterior aspect of the pulmonary conus. The course and extent of the A-V conduction system delineated in the morphological left ventricle suggests it is a left bundle branch. The surgical implications of the ectopic location of the A-V conduction system anterior to the ventricular septal defect, and the variability of the more proximal portion of the A-V conduction system are discussed.

This report describes electrophysiological studies performed during the course of open-heart surgery to delineate the course of the A-V conduction system in four patients with corrected transposition of the great vessels with various types of ventricular septal defects and in one patient with single ventricle, the "right ventricle" to the left ("ventricular inversion" or l-loop). In each instance the A-V conduction system was not where one would expect it to be in normal hearts or in most types of congenital heart disease, with or without ventricular septal defect.

Methods
The patients ranged in age from six to 26 years. We define corrected transposition of the great vessels as a congenital cardiac malformation in which there is a discordant relationship between the atria and ventricles and between the ventricles and the great arteries. This implies two atria, two ventricles, and two atrioventricular valves each emptying into a separate ventricle. We define single ventricle as a malformation in which both A-V valves enter the same or a truly common ventricle. A summary of the pertinent clinical data for each patient is provided in Table I. Four patients had corrected transposition. Two of these also had a large ventricular septal defect. One patient had an A-V canal type ventricular septal defect and valvular pulmonary stenosis. One patient had an associated large ventricular septal defect and both infundibular and valvular pulmonary stenosis. One additional patient had a single ventricle, l-loop, a rudimentary ventricular septum, and subvalvular pulmonary stenosis with the ascending aorta to the left.

All studies were performed at open-heart surgery during cardiopulmonary bypass using standard electrophysiological techniques described previously. The experimental protocol varied somewhat for each patient and was determined by the nature and requirements of the surgical procedure and by the anatomy of the congenital defect. During periods of data collection, arterial perfusate temperature was 32°C in four patients and 28°C in one patient. An electrode plaque was sewn to the right atrium and used to identify atrial activation during spontaneous atrial rhythm or to deliver a stimulus during paced atrial rhythm. An electrode probe was used to record bipolar electrograms from selected portions of the right atrium and the morphologic left ventricle during a spontaneous or paced atrial rhythm. The atrial sites (fig. 1) were low in the right atrium anterior to the coronary sinus ostium (present in three patients) or where the coronary sinus ostium would be expected to be in the cases where none was present (two patients). The ventricular sites (fig. 1) included both the
anterior and posterior aspects of the ventricular septal defect and the region of the pulmonary artery. All sites mapped with the electrode probe in each study are illustrated by the circles in figures 3–7.

Electrograms from the electrode probe, a second electrogram or stimulus artifact from the right atrium, and ECG leads I, II and III were monitored on an Electronics-for-Medicine Model DB 12 switched-beam oscilloscope and recorded simultaneously on photographic paper moving at 100 mm/sec (fig. 2). All records were simultaneously recorded on a Honeywell 5600 Model magnetic tape recorder for later playback and systematic analysis of the data. All leads in contact with the heart were isolated from ground and from the recording apparatus by isolation transformers. The appearance of a deflection in the recorded electrogram during the isoelectric portion of the P-R interval identified electrical activity in the specialized A-V conduction system.

The previously described electrophysiological technique to delineate the A-V conduction system was modified somewhat in that all studies were performed during periods of aortic cross-clamping at which time the coronary arteries were not perfused. The aortic cross-clamping permitted clear visualization of the complicated anatomy by eliminating the venous drainage which would otherwise cover the field, and acted as an additional safety factor in preventing air from escaping into the systemic circulation. A-V conduction therefore continually changed during the period of electrophysiological mapping, precluding meaningful evaluation of conduction intervals. The mapping was continued as long as there was satisfactory A-V conduction (fig. 2). When impaired A-V conduction greater than 2:1 A-V block occurred, the aortic cross-clamp was released, and coronary artery reperfusion continued until 1:1 A-V conduction recurred.

### Results

Figure 3 represents a schematic drawing of the anatomical grid sites from which electrograms were recorded using the electrode probe in case 1. In this and subsequent schematic drawings, open circles represent sites which were mapped but from which no specialized A-V conduction system electrograms were recorded. The filled circles represent the sites from which specialized A-V conduction system electrograms were recorded. Each site represents 5 mm, i.e., the diameter of the electrode probe.

A coronary sinus ostium was present in the right atrium of this patient. Mapping sites along the atrial side of the mitral valve failed to demonstrate any A-V conduction system electrograms, even opposite the ventricular site from which such electrograms were recorded. In the four patients in whom we were able to map from sites in the low right atrium, this was a consistent finding, whether or not a coronary sinus ostium was present. When sites posterior to the ven-
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tricular septal defect were mapped, no A-V conduction system electrograms were recorded in this or any patient studied. This, of course, is in contradistinction to the usual posterior location of the A-V conduction system in most types of ventricular septal defects. However, A-V conduction system electrograms were recorded from sites anterior and inferior to the ventricular septal defect. In the four patients in whom we were able to electrophysiologically identify the A-V conduction system, we were able to trace its course to its peripheral most portion, i.e., to the Purkinje-muscle junction (fig. 2).

This study well illustrated two points. First, as is especially well shown (fig. 3), the A-V conduction system broadened as it coursed distally, thereby resembling the course of the left bundle branch. Second is the surgical problem of obtaining adequate closure of the ventricular septal defect without producing complete heart block, even when the surgeon is aware of the location of the A-V conduction system. This patient (case 1) developed complete heart block shortly after termination of cardiopulmonary bypass. Since the complete heart block persisted intermittently in the postoperative period, a permanent demand pacemaker was placed prior to discharge. In the more than one year since surgery, the patient has remained largely in a sinus rhythm with the P-R interval unchanged from the preoperative P-R interval of 0.20 sec.

Case 2 had no coronary sinus ostium in the right atrium. The A-V conduction system was not delineated low in the right atrium (fig. 4) even from sites opposite the ventricular site from which A-V conduction system electrograms were recorded. Once again, no specialized A-V conduction system electrograms were recorded on the posterior aspect of the

Figure 2

ECGs recorded from the same patient during a paced atrial rhythm with 1:1 A-V conduction and first degree A-V block (Panel A) and during a spontaneous atrial rhythm with 2:1 A-V conduction (Panel B). In each panel, all traces are recorded simultaneously. The top trace of each panel represents the electrogram recorded from the electrode probe (Probe EG) and the bottom trace represents the recorded ECG. Although three ECGs (I, II, III) were recorded, only one is illustrated. In Panel A, the middle trace represents the stimulus artifact recorded during atrial pacing. In Panel B, the middle trace represents an atrial electrogram recorded during spontaneous atrial rhythm. In each panel, an arrow points to the specialized A-V conduction system electrogram which was recorded through the electrode probe. Note in the top trace of Panel B the very short interval from the specialized A-V conduction system electrogram to the onset of ventricular activation. This electrogram was recorded from a peripheral site illustrated in figure 3. Time lines are at an interval of 1 sec.
ventricular septal defect (fig. 4). Rather, they were recorded on its anterior aspect. Note that the A-V conduction system again became broader as it coursed distally (fig. 4). This patient remained in sinus rhythm during the postoperative period and in the long-term (> 1 year) follow-up still remains in sinus rhythm. The postoperative P-R interval is 0.17 sec, unchanged from before surgery.

Case 3 did have a coronary sinus ostium in the right atrium. Recording from atrial sites inferior and anterior to the coronary sinus ostium, and from sites posterior to the ventricular septal defect again failed to demonstrate any A-V conduction system electrograms (fig. 5). These electrograms were recorded from sites anterior to the ventricular septal defect, but with a significant variation. The proximal portion of the A-V conduction system was delineated on the anterior aspect of the pulmonary conus which was present in this patient. Its presence at this latter location and its absence between the anterior aspect of the ventricular septal defect and the pulmonary artery is important for consideration of suture placement when closing the ventricular septal defect and in consideration of any resection for pulmonary stenosis. This patient has remained in sinus rhythm during the year and one-half following surgery. The preoperative and postoperative P-R intervals were 0.26 sec.

Case 4 is the patient with single ventricle. Again, no A-V conduction system electrograms were recorded from sites anterior to the coronary sinus ostium (fig. 6). However, A-V conduction system electrograms were recorded at all sites mapped between the anterior aspect of the ventricular septal defect and the pulmonary artery. This patient developed 2:1 A-V block at the termination of cardiopulmonary bypass and complete heart block transiently in the immediate postoperative period. However, sinus rhythm returned on the sixth postoperative day and the
patient has remained in sinus rhythm during the nine months following surgery. The preoperative and postoperative P-R intervals were 0.14 sec.

Case 5 did not have a coronary sinus ostium in the right atrium. However, it was technically not possible to perform a right atriotomy to map the low right atrial sites because of the presence of dextrocardia, and because of the scarring from previous cardiac surgery (table 1). Electrograms were recorded from all available sites around the periphery of the ventricular septal defect, but no A-V conduction system electrograms were demonstrated (fig. 7). Unfortunately, no electrograms were recorded from sites anterior to the pulmonary artery or from any other ventricular sites. In retrospect, we feel that the A-V conduction system very likely would have been demonstrated by recording electrograms anterior to the sites illustrated in figure 7, particularly sites anterior to the pulmonary outflow tract. This patient remained in sinus rhythm throughout the postoperative period and during the 23 months since operation. The preoperative and postoperative P-R intervals remained identical at 0.16 sec.

Discussion

In our electrophysiological studies in four patients with corrected transposition of the great vessels with associated ventricular septal defects and one patient with single ventricle and 1-loop, two consistent observations were made. First, the specialized A-V conduction system was never delineated low in the right atrium, whether or not there was a coronary sinus ostium present, and even at sites in the atrium opposite the ventricular site from which the A-V conduction system electrograms were recorded. Second, the specialized A-V conduction system was never present posterior to the ventricular septal defect, in contradistinction to its usual location in ventricular septal defects associated with other congenital heart lesions.

Histological studies by Anderson et al. in patients with corrected transposition of the great vessels demonstrated that the A-V conduction system emanated from an A-V node located quite anterior in the A-V junction. These authors also found that the specialized A-V conduction system always coursed on the anterior border of the pulmonary outflow tract and then coursed around the anterolateral margin of the ventricular septal defect, if one were present, in a manner similar to that which we found in case 3 (fig. 5). However, in two of our patients with corrected transposition of the great vessels (cases 1 and 2), and in our case of single ventricle (case 4), the A-V conduction system in the ventricles was delineated posterior to the pulmonary outflow tract between it and the ventricular septal defect, as well as along the anterolateral margin of the ventricular septal defect (figs. 3, 4, 6). This location between the pulmonary outflow tract and the ventricular septal defect of what appears to be the proximal A-V conduction system differs from the description of Anderson. However, in our three cases, sites anterior to the pulmonary outflow tract were not explored with the electrode probe. Therefore, we cannot conclude that A-V conduction tissue was not located in this latter area. Thus, another interpretation of the data in these three cases is that the proximal portion of the A-V con-
duction system lies in the location described by Anderson et al.,15,16 but the specialized A-V conduction system provides a branch which courses between the ventricular septal defect and the pulmonary outflow tract. A clear answer to this question might have been provided if meaningful evaluation of conduction intervals within the A-V conduction system were possible in this study. Kupersmith and his colleagues15 electrophysiologically identified the specialized A-V conduction system anteriorly between the pulmonary outflow tract and the ventricular septal defect in two cases similar to ours.

A posterior A-V node located in the usual position of the normal A-V node did not connect with the A-V conduction system in all but one of the cases of corrected transposition of the great vessels and in none of the cases of single ventricle described by Anderson et al.15,16 This seems the likely explanation for our failure to record any His bundle electrograms low in the atrial septum in the region of the A-V junction. It should be noted that it is not possible to record electrograms from the A-V node with the electrophysiological techniques used in this study.11,12

The anatomic distribution of the A-V conduction system was delineated in the morphologic left ventricle as a rather broad sheet. As this resembled our previous findings for the normal human left bundle branch, it suggested to us that it is a left bundle branch. These electrophysiologic data support the older anatomic observations of Walmsley,13 Uher14 and Aschoff15 and the more recent histological data of Lev16 and Anderson.14 On the basis of the anatomical fanlike appearance of the conduction system in the morphologic left ventricle in cases of corrected transposition of the great vessels, all these authors have suggested that the A-V conduction system in this ventricle was the left bundle branch.

Surgical Considerations

Absence of the specialized conduction system inferior to the ventricular septal defect has a special importance. The fact that a considerable portion of the anteriorly located specialized conduction system may be left bundle branch is less of a hazard should sutures be placed in close proximity to it. It is the most proximal portion of the A-V conduction system which is particularly to be avoided. The fact that in two of our cases the proximal portion of the A-V conduction system was not in relationship to the ventricular septal defect reinforces the usefulness of knowing as precisely as possible the course of the A-V conduction system; in such patients, sutures could be placed along the superior and anterior aspect of the ventricular septal defect without fear of interrupting the A-V conduction system.

The relationship of the proximal portion of the A-V conduction system to the pulmonary outflow tract is an additional factor to consider, particularly in patients with any of the various forms of pulmonary outflow tract obstructions. Surgeons repairing such pulmonary outflow tract obstructions must be aware of the nearness of the proximal A-V conduction system to this outflow tract and therefore consider alternative means to relieve outflow tract obstruction, particularly the pulmonary infundibular or subinfundibular type.

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