Electrophysiological Delineation of the Specialized A-V Conduction System in Patients with Congenital Heart Disease

II. Delineation of the Distal His Bundle and the Right Bundle Branch

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SUMMARY

The course of the distal His bundle and the right bundle branch was electrophysiologically delineated during open heart surgery in nine patients with tetralogy of Fallot and in six other patients with various forms of congenital heart disease. In patients with tetralogy of Fallot, right bundle branch electrograms were usually recorded up to 25 mm from the plane of the tricuspid valve annulus and only rarely beyond this site, indicating that the electrical activity in the right bundle branch was isolated from right ventricular myocardium to a site 25 mm away from the tricuspid annulus in the patients studied. In one patient with right bundle branch block pattern on the electrocardiogram induced by a ventriculotomy, the right bundle branch was traced to the Purkinje fiber-ventricular muscle junction, supporting the observation that a right bundle branch block pattern induced by ventriculotomy does not indicate that injury to the proximal part of the right bundle branch occurred.

In five patients with various forms of congenital heart disease we did not record electrical activity from the distal His and right bundle branch. The anatomic and functional reasons for this failure are discussed. In one patient with a common ventricle, the identification of the specialized atrioventricular (A-V) conduction system allowed for total surgical correction of this anomaly without injury to the conduction system. The electrophysiological delineation of the specialized atrioventricular conduction system is suggested for all patients undergoing open heart surgery who have complicated congenital heart disease on which no data are available regarding the exact location of the specialized atrioventricular conduction system, for patients with unusual ventricular anatomy, and for patients in whom the hemodynamic and angiographic studies do not correlate well with the electrocardiogram.

Additional Indexing Words:
His bundle
Congenital heart disease
Tetralogy of Fallot
Complete heart block
Right bundle branch
His bundle electrogram
Common ventricle

WE HAVE REPORTED on the electrophysiological delineation of the His bundle proximal to the membranous septum during open heart surgery in patients with congenital heart disease. Although complete heart block can occur during surgical manipulation in the atria, during correction of endocardial cushion defects or transposition of the great vessels, it is also likely to occur during surgical repair of a ventricular septal defect. Complete heart block can also occur as a result of surgical intervention in the left ventricle without an intraventricular communication as in patients with idiopathic hypertrophic subaortic stenosis. Delineation of the specialized atrioventricular (A-V) conduction system in the ventricle might therefore facilitate the surgical correction of congenital heart disease which requires intraventricular repair and closure of intraventricular communications. Electrophysiological delineation of the specialized A-V conduction system will also permit the identification of the conduction system in patients with forms of congenital heart disease for which there
is little or no information on the exact location of these structures. Furthermore, the ease, reliability, and feasibility of electrophysiologically delineating the specialized A-V conduction system may permit surgical repair of some congenital heart defects previously thought irreparable, in large part because of the great likelihood that complete heart block would be produced.

The purpose of this study is to report our observations during open heart surgery on the precise location of the intraventricular recording sites from which electrograms were recorded from the specialized conduction system in patients with various types of congenital heart disease.

Material and Methods

Following institution of cardiopulmonary bypass and using our previously described technique, the course of the intraventricular part of the specialized A-V conduction system was electrophysiologically delineated during open heart surgery in 20 patients: nine with tetralogy of Fallot and with 11 other forms of congenital heart disease (table 1). The ages ranged from 3 to 41 years.

Electrograms recorded from the His bundle or right bundle branch were identified by criteria previously described. The appearance of a deflection in the electrograms during the isoelectric portion of the P-R interval delineated conduction in the His bundle or the right bundle branch. Deflections in the specialized conduction system electrograms recording activity occurring simultaneously with the onset or during ventricular activation, i.e., at the beginning or during the QRS complex, were considered to be Purkinje fiber electrograms.

To delineate the specialized A-V conduction system in the right ventricle in patients with diseases which included a ventricular septal defect, a special grid system was used (fig. 1). The first recording site was under the septal leaflet of the tricuspid valve, immediately adjacent to the edge of the ventricular septal defect (fig. 1, site A). The electrode probe was then advanced by increments of 5 mm, i.e., the probe diameter, along the edge of the ventricular septal defect toward the papillary muscle of the conus (muscle of Lencizi) and from there toward the moderator band of the right ventricle. The sites were labeled according to their distance from the tricuspid valve annulus (fig. 1, sites A to A). Recordings were then obtained from a line one probe width (5 mm) from the edge of the ventricular septal defect (sites B-B) and following immediately below the A line. When appropriate, a third line of recordings, the C line, was obtained. The C line followed the B line in the fashion described above. Recordings from the surface of the papillary muscle of the conus or the moderator band were so labeled. Slight modification of this diagrammatic representation was needed for some patients due to differences in intraventricular anatomy, the distance of the papillary muscle of the conus from the tricuspid annulus, or the location or absence of a ventricular septal defect. None of the patients developed A-V conduction disturbances during the delineation of the intraventricular specialized A-V conduction system.

Results

Studies in Patients with Tetralogy of Fallot

Figure 2 presents the intraventricular electrophysiological delineation of the specialized A-V conduction system in a patient with tetralogy of Fallot. In this patient, specialized conduction system electrograms were recorded from sites A through A. Site A was under the septal leaflet of the tricuspid valve immediately adjacent to the tricuspid annulus. The specialized conduction system electrograms recorded from this site were considered to represent activation of the distal His bundle, first, because histological

Table 1

<table>
<thead>
<tr>
<th>Patients Studied</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>9</td>
</tr>
<tr>
<td>Ventricular septal defect, pulmonary stenosis and</td>
<td></td>
</tr>
<tr>
<td>asymmetric septal hypertrophy</td>
<td>1</td>
</tr>
<tr>
<td>Common ventricle</td>
<td>1</td>
</tr>
<tr>
<td>L-Transposition of the great arteries,</td>
<td>1</td>
</tr>
<tr>
<td>complete heart block</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary and aortic stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Double chambered right ventricle</td>
<td>1</td>
</tr>
<tr>
<td>Subvalvar aortic stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Double outlet left ventricle</td>
<td>1</td>
</tr>
<tr>
<td>Ostium primum defect</td>
<td>1</td>
</tr>
<tr>
<td>Isolated dextrocardia, common ventricle</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>20</td>
</tr>
</tbody>
</table>

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studies show that in most cases the distal portion of the His bundle extends a few millimeters inferiorly to the tricuspid valve annulus, and second, because selective pacing of the specialized conduction system at site A1 at threshold level produced a QRS complex indistinguishable from that observed by pacing the atria from the area of the sinus node or selective pacing of the atrial part of the common bundle. Specialized conduction system electrograms recorded at sites A2 to A4 were considered to represent right bundle branch electrograms. Records obtained at sites A5, A6, and the papillary muscle of the conus demonstrated two specialized conduction system electrograms, the first being a right bundle branch electrogram, and the second, a Purkinje fiber electrogram coinciding with the beginning of the QRS complex. The record obtained from the moderator band showed only a Purkinje fiber electrogram. These records were obtained through a right ventriculotomy which prolonged the QRS duration from 95 to 129 msec and changed the scalar ECG configuration to a “right bundle branch block pattern.” Since the course of the right bundle was delineated in this patient to the Purkinje fiber-ventricular muscle junction, i.e., to the point of its penetration into the myocardium, the study provides further support to the observation that an electrocardiographic pattern of right bundle branch block can be seen in the presence of an intact proximal right bundle branch.

Figure 3 summarizes our experience for nine patients with tetralogy of Fallot. In two patients records were obtained only from the papillary muscle of the conus and the moderator band. In all patients, we were able to trace the conduction system from sites A1 to A3, i.e., about 15 mm from the tricuspid valve annulus. From that latter site specialized conduction system electrograms were found in a decreasing order of frequency: 60% at 20 mm from the tricuspid annulus, 50% at 25 mm, and 25% around the papillary muscle of the conus. In no cases were we able to record a right bundle electrogram from or around the moderator band.

Studies in Patients with Other Forms of Congenital Heart Disease

We studied patients with complicated and less common forms of congenital heart disease in which little or no information was available regarding the anatomical course of the specialized A-V conduction system.

In one patient with a ventricular septal defect, pulmonary stenosis, and asymmetrical septal hyper-
trophy we recorded electrograms of the right bundle branch on line B, about 5 mm from the edge of the ventricular septal defect (fig. 4, sites B2 to B4). Therefore, the ventricular septal defect in this patient was repaired with sutures placed above the conduction system instead of the usual site, which in this patient would have included the right bundle branch. The location of recording sites of the intraventricular specialized conduction system in this patient is similar to the location of the specialized conduction system electrograms described previously in one patient with idiopathic hypertrophic subaortic stenosis.13

In one patient with a common ventricle without an outflow tract and with both great vessels in a side-to-side relationship14, 15 (fig. 5), the specialized conduction system electrograms were recorded along a ridge 7 mm in height on the posterior-inferior ventricular wall and could be traced 30 mm away from the plane of the atrioventricular valves. The favorable distribution of the papillary muscles and the chordae tendineae and the presence of two atrioventricular valves allowed for total surgical repair and division of this common ventricle into two functional cavities. The patient had an uneventful recovery and was in normal sinus rhythm when discharged from the hospital.

We have also operated on one patient with dextrocardia, levo-transposition of the great vessels, and a common ventricle. In this patient, the specialized conduction system electrograms were identified along a segment 15 mm in length along the left side of a posterior ventricular ridge close to a large papillary muscle supplying both atrioventricular valves. The proximal intraventricular part of the specialized conduction system from the plane of the A-V valves toward the site of its identification 30 mm from the A-V valves was not delineated. In this patient the arrangement of the papillary muscles and chordae tendineae was such that a patch could not be inserted without extensive damage to the atrioventricular valve apparatus. This anatomic arrangement prohibited total surgical repair.

In one patient with levo-transposition of the great vessels, inversion of the ventricles, a ventricular septal defect, and complete heart block, we were not able to identify any specialized conduction system electrograms from 29 recording sites, inferior, superior, anterior, and posterior to the ventricular septal defect, nor from sites in the atrium, nor from sites which included an extensive search of the right-sided ventricular septum (anatomic left ventricle).

In two patients with isolated pulmonary stenosis and in one patient with isolated pulmonary and aortic stenosis, we were unable to identify specialized A-V conduction system electrograms along the crista supraventricularis. In one patient with a double-chambered right ventricle, records obtained at sites B3, B5, C3, and C4 did not show specialized conduction system electrograms. In one patient with a fibrous subvalvular aortic stenosis and mild right ventricular obstruction, electrograms denoting the specialized conduction system were recorded at sites A3 through A4 and the papillary muscle of the conus.

**Figure 4**

The figure shows sites from which intraventricular specialized conduction system electrograms were recorded in a patient with ventricular septal defect, pulmonary stenosis, and left ventricular obstruction due to asymmetric septal hypertrophy. Sites in which specialized conduction system electrograms were recorded are marked (+) and sites in which they were not recorded are marked (—).

**Figure 5**

The course of the intraventricular part of the specialized A-V conduction system, recorded in a patient with common ventricle. Specialized conduction system electrograms in this patient were recorded from the posterior-inferior ventricular wall along a segment of 30 mm in length. See text for discussion.
In one patient with an ostium primum defect, specialized conduction system electrograms were recorded from an area which extended from a site inferior to the coronary sinus ostium onto the left side of the ventricular septum up to 5 mm from the plane of the mitral valve. No conduction system electrograms were found on the right side of the ventricular septum immediately adjacent to the tricuspid annulus in this patient.

In one patient with double outlet left ventricle and a severely stenotic pulmonary artery arising to the left and posterior to the aorta, specialized conduction system electrograms were recorded at sites B1, B2, and B3. A specialized A-V conduction system electrogram was not recorded at sites A1 and A2 but was recorded at sites A3 and A4, sites which were close to the papillary muscle of the conus in this patient. No specialized conduction system electrograms were recorded in this patient anterior to the ventricular septal defect. Total surgical correction of the lesion in this patient was performed by closure of the ventricular septal defect and homograft insertion from the right ventricle into the pulmonary artery.

**Discussion**

**Electrophysiological and methodological considerations**

In all patients with tetralogy of Fallot, we were able to electrophysiologically delineate the location of the right-sided specialized A-V conduction system. This electrophysiologically-anatomical correlation agrees well with results of previously described histological studies. Our results indicate that with our method, specialized conduction system electrograms in these patients are usually recorded at sites up to 25 mm from the tricuspid valve annulus and only rarely beyond this site. The progression of the specialized conduction system electrograms recorded from the right bundle, as we have shown for sites A1 to A4, prior to the beginning of the QRS complex and prior to the activation of Purkinje fibers at these sites, indicates that the right bundle branch was electrically isolated from the right ventricular myocardium up to 25 mm from the tricuspid valve annulus. This observation is similar to that made by Myerburg et al. for the canine heart and to our findings for the left bundle branch of man.

In five patients we failed to record any specialized conduction system electrograms from sites inside the right ventricular cavity. In the first such patient, one with a double-chambered right ventricle, records were obtained only from lines B and C. It seems likely that in this patient the specialized A-V conduction system was in its usual location along line A, a site from which records were not obtained.

In a second patient with levo-transposition of the great vessels, ventricular septal defect, and complete heart block, an extensive search of the right side of the septal endocardial surface (anatomic left ventricle) failed to demonstrate any specialized conduction system electrograms. In a similar case described by Lev et al., neither the common bundle nor bundle branches could be identified in their usual location. The absence of specialized conduction system electrograms on the right-sided ventricular septum in the case described by us might therefore suggest the absence of specialized conduction tissue from its usual location. However, since electrograms on this patient were recorded during a ventricular rhythm, the possibility exists that specialized conduction system electrograms were not identified due to depolarizations of the specialized conduction system and the ventricular myocardium simultaneously. The specialized conduction system electrograms may then occur within the ventricular electrogram or during the QRS complex preventing their recognition.

In three patients, none of whom had a ventricular septal defect, two with pulmonary stenosis, and one with aortic stenosis and pulmonary stenosis, we also failed to identify the specialized conduction system electrograms distal to the membranous septum. Since the proximal right bundle branch frequently appears on the left side of the ventricular septum, it is likely that we were unable to record specialized A-V conduction system electrograms from some of the patients because the proximal right bundle was located too deeply beneath the right endocardial septal surface. Such an explanation is also supported by finding specialized conduction system electrograms in one patient with ostium primum defect recorded at site A1, on the left side of the ventricular septum and the absence of such an electrogram on the right side of the ventricular septum immediately adjacent to the tricuspid valve leaflet (right-sided A1). Such a leftward shift of the distal portion of the common bundle did not, however, affect our ability to record specialized conduction system electrograms in patients with a ventricular septal defect, such as in patients with tetralogy of Fallot. Further, failure to record specialized conduction system electrograms with our technique under the septal leaflet of the tricuspid valve might be due to the anatomy of the tricuspid septal leaflet and of its attachment by multiple chordae tendineae to the right ventricular septum. Such an arrangement might preclude the proper placement of the electrode probe under the tricuspid leaflet.

Since we were able to localize the distal His bundle and proximal right bundle branch in all patients who had a ventricular septal defect, it seems likely that the
presence of a septal defect might favor the recording from this proximal intraventricular part of the specialized A-V conduction system.

In patients with aortic and pulmonary valvar stenosis who require surgery, a significant septal and infundibular hypertrophy is usually present. This also might affect the position or the depth of the specialized conduction system. The case reported by us in which a significant difference in the location of the specialized conduction system was found in the presence of asymmetric septal hypertrophy, and the case previously described by Coyne,18 support such speculation.

Latham,4 Titus,29 Truex,24 and Anderson29 reported on the presence of the specialized conduction system superior to the septal defect in some patients with various forms of congenital heart disease. In some patients, we attempted to record specialized conduction system electrograms superior to the defect, but in no case were such electrograms recorded in this position.

Figure 6 summarizes some anatomical considerations affecting the recording of intraventricular specialized conduction system electrograms by our method.

Surgical Considerations

The incidence of surgically-induced complete heart block following total correction of congenital heart disease associated with surgical closure of any ventricular septal defect is currently estimated to be 1%.6 This incidence increases in patients who undergo total correction of endocardial cushion defects,27 idiopathic hypertrophic subaortic stenosis,5,6 and in patients undergoing surgical closure of a ventricular septal defect in the presence of levo-transposition of the great vessels. The incidence of complete heart block in patients with a single ventricle is so high as to be considered prohibitive to total surgical repair.

Surgically-induced complete heart block following repair of ventricular septal defect or tetralogy of Fallot can usually be prevented by avoiding the postero-inferior aspect of the septal defect by placing sutures into the base of the tricuspid leaflet and avoiding placing sutures that reach too deep into the ventricular septum, thus avoiding the specialized conduction system.28 The occasional downward displacement in the location of the specialized A-V conduction system, as has been shown by us, might account for the infrequent occurrence of complete heart block following surgery in patients even when the surgeon was careful to avoid the immediate postero-inferior aspect of the defect.

With the improvement of surgical techniques which permit surgical correction of the more complicated and unusual congenital heart disease, it seems prudent to electrophysiological delineate the conduction system in these cases during surgery.1,7,8 This is especially useful for those diseases in which there are no data about the exact location of the specialized A-V conduction system. Our technique
offers hope for future successful surgical correction in patients hitherto considered unoperable.

The electrophysiological delineation of the intraventricular part of the specialized A-V conduction system is, therefore, used by us routinely for all patients with unusual ventricular anatomy, in patients with complicated congenital heart disease, and in patients in whom the hemodynamic and angiographic studies do not correlate well with the electrocardiogram.

References

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