Electrophysiologic Identification of the Specialized Conduction System in Corrected Transposition of the Great Arteries

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Summary
Electrophysiologic identification of the specialized conduction system was performed during open-heart surgery in two cases of corrected transposition of the great arteries (l-TGA) with situs solitus, dextrocardia or dextroversion and membranous ventricular septal defect (VSD). In one case there was accompanying subvalvar pulmonary stenosis and in the other there was an Ebstein-like deformity of the systemic atrioventricular (A-V) valve. Electrograms were recorded throughout the right atrium and morphologic left ventricle in the case with subvalvar pulmonary stenosis and throughout the left atrium and morphologic right ventricle in the case with the Ebstein-like deformity of the systemic A-V valve. Specialized conduction system electrograms were recorded exclusively at sites anterior to the membranous VSD in both cases and at no other sites in the respective atrium or ventricle studied. There was first degree A-V block in both cases and in each the site within the specialized conduction system of this delay was proximal to the specialized fiber electrogram recording sites and thus probably proximal to the bifurcation of the bundle branches. In addition, the Ebstein-like deformity of the systemic A-V valve present in one case was confirmed by directly recording ventricular electrograms in an area of "atrialized-ventricle" above the valve.

Additional Indexing Words:
His bundle
Subvalvar pulmonary stenosis
Bundle branches
First degree atrioventricular block
Ventricular septal defect

Corrected Transposition of the great arteries (l-TGA) is a congenital cardiac abnormality in which there is inversion of the ventricles and transposition of the great arteries. The systemic venous return enters the pulmonary artery via the morphologic left ventricle and the pulmonary venous return enters the aorta via the morphologic right ventricle. Abnormalities of atrioventricular (A-V) conduction, including first, second, and third degree A-V block, are commonly associated with l-TGA.1-10 These conduction abnormalities are important in the prognosis of l-TGA5, 7, 8, 10 and hamper attempts at repair of ventricular septal defect (VSD) and other cardiac malformations that occur in l-TGA.5, 7, 8 For these reasons, the anatomic location of the specialized conduction system in l-TGA is of particular interest and importance.

In the present study, electrophysiologic delineation of the specialized conduction system was performed for the first time during open-heart surgery in two cases of l-TGA. Surgical repair of an associated membranous VSD was performed in both cases. In one case repair of an associated subvalvar pulmonary stenosis by means of a Hancock outflow tract prosthesis was also performed while in the other case replacement of the systemic A-V valve for an Ebstein-like deformity, which was confirmed by the technique of electrophysiologic mapping of the "atrialized-ventricle," was performed. In addition, evidence is presented as to the site of A-V conduction delay in l-TGA.

Methods
Electrophysiologic studies were performed during cardiopulmonary bypass initiated in the course of open-heart surgery in two patients using previously described techniques.11-14 An electrode probe containing 3 silver electrodes 1-2 mm apart was used to record bipolar electrograms simultaneously with 3 standard bipolar ECG leads. Electrograms were carefully recorded throughout the right atrium and morphologic left ventricle in case 1 and the left atrium and morphologic right ventricle in case 2. During data collection the temperature was 32°C in case 1 and 28°C in case 2. Case 1 received no operative premedication.
and curare and morphine sulfate anesthesia during surgery while case 2 received small amounts of morphine sulfate approximately 3 hours prior to surgery and halothane anesthesia during surgery. Previously described characteristics of atrial, specialized conduction system, and ventricular electrograms were used to identify the origin of the electrograms recorded in this study. Intervals were measured from the major deflections of all electrograms recorded from the specialized conduction system to the onset of the earliest QRS deflection in the standard ECG leads.

All leads in contact with the heart were isolated from ground and from the recording apparatus by isolation transformers. The electrograms and ECGs were monitored simultaneously on an Electronics for Medicine DR-12 switched beam oscilloscope and recorded on photographic paper moving at a paper speed of 100 mm/sec. Electrocardiograms, calibrated at 2 cm/mV, were recorded between filter frequencies of 0.1-500 Hz. Electrograms were recorded between 12-500 Hz.

Case Report

The patient was a 13-year-old female who at age 6 months became cyanotic and also had occasional episodes of dyspnea. Physical examination at age 1½ years revealed cyanosis, clubbing of the fingers, a harsh pansystolic murmur heard over the entire precordium that was transmitted to the back, and a single second heart sound. Chest X-ray revealed dextroversion of the heart and clear lung fields; ECG revealed right axis deviation, deep S waves over the right precordium, a normal P wave axis and a P-R interval of 0.14 sec. The patient remained essentially unchanged over the next several years with cyanosis, mild dyspnea, moderate limitation of activity, and slight retardation of growth and development. Cardiac catheterization and angiography at age six years revealed situs solitus, I-TGA, dextroversion, VSD with a right-to-left shunt and subvalvar pulmonic stenosis; the brachial artery oxygen saturation was 77% and the pressure in the morphologic left ventricle was 94/4 mm Hg. At age 12 years, first degree A-V block with a P-R interval of 0.23 sec was first noted, but the ECG (fig. 1) was otherwise similar to her previous ECGs. At age 13 years, open-heart surgery was performed for correction of the subvalvar pulmonic stenosis and closure of the VSD. The surgical findings are described below. The patient’s postoperative course was uneventful, and she was discharged on the 17th postoperative day, acyanotic and in normal sinus rhythm.

Surgical Findings

At surgery, situs solitus, I-TGA, dextroversion, a membranous VSD, and severe subvalvar pulmonary stenosis were found. The atria were normally situated except that the left atrium was rotated slightly anteriorly. The right atrium emptied into the morphologic left ventricle and the left atrium emptied into the morphologic right ventricle. There was dextroversion of the heart with the morphologic right ventricle anterior and slightly to the left of the morphologic left ventricle. The aorta arose from the crista supraventricularis of the morphologic right ventricle and was anterior and to the left of the pulmonary artery, which arose from a markedly stenotic subvalvar region into which the morphologic left ventricle emptied. The VSD measured 1.5 cm in diameter and involved the interventricular portion of the membranous septum and the anterior muscular septum adjacent to it. The surgery performed was closure of the VSD by means of a teflon felt patch, closure of the narrow subvalvar pulmonary stenosis, and placement of a Hancock outflow tract prosthesis (a dacron conduit containing a porcine aortic valve) to restore continuity between the morphologic left ventricle and pulmonary artery.

Electrophysiologic Studies

Specialized fiber electrograms were recorded in the morphologic left ventricle at the superior and anterior margin of the membranous VSD (fig. 2) and also anteriorly and inferiorly from the margin of the VSD for a distance that equalled the diameter of the recording electrode (5 mm). Specialized fiber electrograms were not observed in recordings from any other site in the right atrium or morphologic left ventricle. The rhythm during the mapping was a conducted atrial rhythm produced by pacing the right atrium at a rate of 103 beats/min.

The most proximal site from which we recorded specialized fiber electrograms, i.e., the site with the longest recorded specialized fiber electrogram to QRS interval (41 msec), was at the antero-superior margin of the VSD adjacent to the A-V ring (fig. 2). It is likely
that this was a His bundle rather than a bundle branch recording site since it was located adjacent to the A-V ring and since no specialized fiber electrogram could be recorded at a more proximal site in the atrium. It is significant that the specialized fiber electrogram to QRS interval of 41 msec recorded here was not prolonged for recordings of His bundle electrograms. This indicates that the delay in A-V conduction represented by the prolonged P-R interval in the preoperative ECG (0.23 sec) occurred proximal to our recording sites.

This observation is in part confirmed by the fact that the interval from the onset of the P wave, here represented by the atrial stimulus artifact, to the most proximally recorded specialized fiber electrogram was 254 msec (fig. 2). This interval is greatly prolonged for a 13-year-old patient and is very likely abnormal, although the normal ranges for intervals from the onset of the P wave (or atrial stimulus artifact) to the His bundle electrogram for an atrial paced rhythm are not certain, and there was hypothermia (32°C) during the study. Thus, there appeared to be prolongation of A-V conduction from the atrium to the most proximal site from which we obtained specialized fiber electrograms in case 1, and we will comment on this further in the discussion.

Figure 2

Schematic diagram of the heart of case 1 in cross-section showing the right atrium and morphologic left ventricle. The pulmonary artery arose from the morphologic left ventricle above the membranous VSD, and subvalvar pulmonary stenosis was also present. The circles show the only sites where specialized conduction system electrograms were recorded. The insert shows the most proximally recorded specialized fiber electrogram (HB) which was probably recorded from the His bundle adjacent to the A-V ring. The lower three traces are ECG leads I, II, III. The interval from the atrial stimulus artifact (S) to the specialized fiber electrogram at this site was 254 msec, and the interval from the specialized fiber electrogram to the QRS was 41 msec.

Case 2

Case Report

The patient was a 14-year-old female who shortly after birth was noted to have visceral situs solitus, dextrocardia, a pansystolic murmur, and a single second heart sound. She was not cyanotic, and the diagnosis of corrected transposition of the great arteries and VSD was made. The patient had episodes of left-sided congestive heart failure which regressed at one year of age after which she became asymptomatic. At ages two and 13 years, cardiac catheterization revealed no shunt demonstrable by oxygen saturation studies and normal right-sided pressures; cardiac angiography was consistent with situs solitus, l-TGA, dextrocardia, VSD, and an Ebstein-like deformity with regurgitation of the systemic A-V valve.

At 14 years of age, the patient’s dyspnea recurred. Chest X-ray showed dextrocardia and left-sided congestive heart failure and the electrocardiogram (fig. 3) showed sinus rhythm with first degree A-V block, pattern of right atrial enlargement and a tall R wave in lead V1 which progressively diminished in size from lead V1-V6. This QRS configuration was similar to that previously reported in cases of l-TGA with dextrocardia. Digoxin and diuretics were administered and open-heart surgery was performed; the findings are described below. Postoperatively, the patient improved at first but later congestive heart failure recurred and progressively worsened. Cardiac catheterization on the 31st postoperative day showed a large left-to-right shunt at the ventricular level. A second open heart operation was performed under marked hypothermia (22°C) which made it impossible to record electrograms from the heart. Ventriculotomy of the morphologic left ventricle was performed rather than of the morphologic right ventricle in which the location of the specialized conduction had been previously

Figure 3

ECG in case 2, showing sinus rhythm, first degree A-V block with P-R interval of 0.21 sec, pattern of right atrial enlargement and a tall R wave in lead V1, which progressively diminished from lead V1-V6. This QRS configuration is similar to that previously reported in cases of l-TGA with dextrocardia.
determined by electrophysiologic mapping. A residual VSD was found which required extensive suturing for complete closure. Permanent epicardial pacing wires were inserted, and postoperatively, the patient gradually improved but required permanent ventricular pacing for complete heart block. The patient was discharged on the 42nd postoperative day with minimal symptoms and on digoxin. Two weeks following discharge from the hospital she died suddenly at home. Postmortem examination was not performed.

Surgical Findings

At surgery, l-TGA, dextrocardia, situs solitus, a membranous VSD and an Ebstein-like deformity of the systemic A-V valve were found. The relationships of both ventricles, the aorta, and pulmonary artery were similar to those in case 1 except that the rotation of the heart rightward was greater. Also, the systemic A-V valve in case 2 was abnormal and insufficient, causing left atrial dilatation. The anterior leaflet of this valve was normally positioned, but the remainder of the leaflets, the number of which could not be determined, arose within the morphologic right ventricle 1-2 cm below the annulus fibrosis, creating an area of thin-walled "atrialized-ventricle." The VSD involved the interventricular portion of the membranous septum and the anterior muscular septum adjacent to it. There was an aneurysm of the membranous septum overlying the VSD that protruded into the morphologic left ventricle and in part closed the VSD. Membranous septal tissue was not otherwise seen in the heart. The aorta emerged from the crista supraventricularis above and anterior to the VSD. The surgery performed was closure of the membranous VSD by means of a teflon felt-lined patch and replacement of the systemic A-V valve with a Braunwald-Cutter prosthesis.

Electrophysiologic Findings

The anatomic locations of the sites where specialized conduction system electrograms were recorded are shown in figure 4. Specialized fiber electrograms were recorded at the superior and anterior margin of the VSD and not at any other recording site in the left atrium or morphologic right ventricle. The right atrium and morphologic left ventricle were not exposed during surgery and electrograms were therefore not recorded in these chambers of the heart. Specialized conduction system mapping was performed at a temperature of 28°C. The rhythm during the mapping was atrial fibrillation (fig. 4) in spite of an attempt at electroconversion of the rhythm and in spite of the fact that atrial fibrillation did not occur in this patient before or after surgery. The presence of this arrhythmia during the study would not have interfered with our ability to record His bundle electrograms in the atrium.19

![Schematic diagram of the heart of case 2 in cross-section showing the left atrium and morphologic right ventricle. The aorta arose from the morphologic right ventricle. There was an Ebstein-like deformity of the systemic A-V valve causing the formation of an "atrialized-ventricle." The circles show the only sites where specialized conduction system electrograms were recorded anterior to the membranous VSD. Left Inset: Most proximally recorded specialized conduction system electrogram (SCS) which was probably recorded from the His bundle or proximal portion of the right bundle branch near the A-V ring. The lower trace is ECG lead aVF. The specialized fiber electrogram-to-QRS interval at this site was 49 msec. Upper Right Insert: Atrial electrogram (A), indicating the presence of atrial fibrillation during the study, and ECG lead I. Lower Right Insert: Ventricular electrogram (V) recorded from the "atrialized-ventricle" confirming the presence of an Ebstein-like deformity of the systemic A-V valve, and ECG lead I.

The most proximal site from which we recorded specialized fiber electrograms was at the anteroseptal margin of the VSD, 5 mm from the A-V ring (fig. 4). The specialized fiber electrogram-to-QRS interval at this site was 49 msec. Because its recording site was near the A-V ring, this electrogram was probably recorded from the His bundle or most proximal portion of the bundle branch. A specialized fiber electrogram to QRS interval of 49 msec is not prolonged for the His bundle or most proximal portion of the bundle branch when consideration is given to the severe hypothermia (28°C) present when recordings were made.19, 20, 21 Therefore in case 2, as in case 1, it is likely that there was no intrinsic delay in conduction from the sites where we recorded specialized fiber electrograms to the ventricle. The delay in A-V conduction represented by the prolonged P-R interval (0.21 sec) prior to surgery occurred proximal to these sites.

As mentioned above, the location of the systemic A-V valve in case 2 was abnormal. We recorded ventricular electrograms from an area of the heart above the valve and in this way confirmed the presence of an "atrialized-ventricle" and an Ebstein-like deformity.22

Circulation, Volume 50, October 1974
Examples of atrial and ventricular electrograms recorded from the left atrium and "atrialized-ventricle" respectively are shown in figure 4.

Discussion

In the present study, electrophysiological identification of the specialized conduction system in l-TGA was performed for the first time. Specialized fiber electrograms were recorded in two cases in the morphologic right or left ventricle anterior to the VSD and at no other site in either ventricle or atrium. This location of the specialized conduction system anterior to the membranous VSD differs from that in hearts with membranous VSDs and normally-related ventricles in which the specialized conduction system passes posterior to the VSD and bifurcation of the bundle branches occurs adjacent to the membranous septum at the posterior margin of the VSD.23-25

There are only two previous anatomic studies of the specialized conduction system in cardiac specimens with l-TGA and membranous VSD, one of an individual heart25 and the other of four hearts.27 In these five cardiac specimens in the two studies, the anatomic location of the specialized conduction system was as follows: Two discrete A-V node-like structures were found in the interatrial septum above the VSD, one anterior and the other posterior. In all hearts, the His bundle arose from the anterior A-V node at the A-V ring and passed in the ventricle and bifurcated at the anterior margin of the membranous VSD. In the individual specimen reported, there was, in addition, hemorrhage and fibrosis of the His bundle.26 These studies, showing that in l-TGA the His bundle passes and bifurcates in the ventricle anterior rather than posterior to the membranous VSD, are thus entirely consistent with the present study.

Site of A-V Conduction Block in l-TGA

It is important to determine, if possible, the precise anatomic site in the specialized conduction system of the commonly occurring A-V conduction delays or blocks in l-TGA.1-10 Prior to the time of open-heart surgery, there was first degree A-V block in both cases presented here (figs. 1 and 3). During surgery, there appeared to be no intrinsic conduction delay from the sites from which we recorded specialized fiber electrograms to the ventricles. Further, in case 1, the interval from the atrial stimulus artifact to the most proximally recorded specialized fiber electrogram was greater than should normally occur. (In case 2, the presence of atrial fibrillation during the study precluded determination of a corresponding interval.) Therefore, the site of conduction delay causing first degree A-V block in both cases was proximal to the sites where we recorded specialized fiber electrograms (figs. 2 and 4) and thus, most likely, proximal to the bifurcation of the bundle branches, i.e., in the His bundle or A-V node.

Our observations as to the site of A-V conduction delay are consistent with the anatomic studies of the specialized conduction system in l-TGA with membranous VSD described above. In these studies, there were abnormalities of A-V nodal structure in all hearts26, 27 and in one heart there was in addition, hemorrhage and fibrosis of the His bundle.26 These histologic abnormalities could certainly explain A-V conduction delay or block and in all instances they were situated proximal to the bifurcation of the bundle branches. Furthermore, studies of the specialized conduction system in hearts with l-TGA and no membranous VSD have shown in addition to inversion of the bundle branches: fibrosis of the His bundle,27 complete interruption of the His bundle in the central fibrous body,28 and in one instance, no other abnormality.29 Thus the histological observations are in agreement with our electrophysiologic observations that placed the site of A-V conduction delay or block in l-TGA proximal to the bifurcation of the bundle branches. One previous report of a His bundle electrogram in a patient with l-TGA and both first degree and Type II, second degree A-V block is also in agreement with these observations. In this patient, the delay or block was proximal to the recorded His bundle electrogram and the His bundle electrogram-to-QRS interval was normal.20 Finally, in previous reports of ECGs of patients with l-TGA and complete heart block, the QRS durations were not prolonged,30 which also suggests that the site of block was proximal to the bifurcation of the bundle branches.

Electrophysiologic Confirmation of Ebstein-like Deformity

The use of the electrophysiologic mapping technique to delineate an area of "atrialized-ventricle" in case 2 is of interest. By recording ventricular electrograms above the systemic A-V valve in the patient studied, we confirmed the presence of an Ebstein-like deformity of the valve. Similar malformations of the systemic A-V valve in l-TGA have been reported previously.8, 31 The technique of electrophysiologic mapping of the "atrialized-ventricle" can be helpful in precisely identifying the annulus fibrosis when systemic A-V valve replacement is contemplated in patients with l-TGA.

Importance of Electrophysiologic Mapping in l-TGA

Complete heart block following surgery has been a major problem in VSD repair and systemic A-V valve replacement in patients with l-TGA.5, 7, 8 This complication may be related to the differing location of

Circulation, Volume 50, October 1974
the specialized conduction system anterior to the membranous VSD rather than posterior to the VSD as occurs in hearts with normally-related ventricles. Our identification of the specialized conduction system during the first open-heart surgical procedure in case 2 was of help in preventing injury to the specialized conduction system from either systemic A-V valve replacement or VSD closure. However, in the second open-heart procedure, the VSD repair required extensive suturing and could not be properly performed with avoidance of the specialized conduction system. In case 1, the technique of electrophysiologic mapping was very useful in preventing injury to the specialized conduction system from VSD closure since sutures for VSD closure are commonly placed at sites where we recorded specialized fiber electrograms.

Addendum

Since submission of this manuscript three additional patients with 1-TGA and membranous VSD have been studied. In each the results were similar to those described for the two patients in this study, with the specialized conduction system situated in the ventricule anterior and superior to the membranous VSD.

References

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