Electrophysiologic Characteristics of Ebstein's Anomaly of the Tricuspid Valve

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SUMMARY

Electrophysiologic characteristics of five patients with Ebstein's anomaly of the tricuspid valve were defined with studies using luminal intracardiac electrode catheters. The diagnosis was made in each case from clinical data and confirmed at cardiac catheterization by the presence of an atrialized right ventricular chamber with atrial mechanical activity and ventricular electrical activity. In three cases intra-right atrial conduction was prolonged (P-A intervals of 50, 50, and 65 msec), a finding which reflected the presence of a characteristically large right atrium. The bundle of His electrogram was recorded in its usual anatomical location. Atrioventricular nodal conduction was prolonged in only one case. Intra-His delay was observed in two cases (bundle of His duration of 30 and 30 msec). Infranodal conduction was prolonged in four cases with H-V intervals of 60, 65, 65, and 80 msec. The anatomical abnormalities were least severe in the only patient with a normal H-V interval (50 msec). The prolonged H-V interval was thought to result from stretching of the conduction system over the atrialized right ventricle (ARV). The late depolarization during the splintered R' of the electrocardiogram found during intracardiac mapping of the ARV in three patients confirms the theory that the ARV produces the "second QRS" typically seen in this anomaly. The ARV was particularly irritable, and ventricular fibrillation was produced in two patients during catheter manipulation in this area. In one case the ARV had a shorter refractory period than the body of the right ventricle. Reentrant supraventricular tachycardia was induced in the only patient with Wolff-Parkinson-White syndrome.

In addition to the previously recognized electrophysiologic features reconfirmed here, patients with Ebstein's anomaly of the tricuspid valve usually have: normal position of the bundle of His, prolonged intra-right atrial conduction, prolonged infranodal conduction, and irritable ARV with delayed activation.

The essential anatomic fault in Ebstein's anomaly is displacement of fused, malformed portions of tricuspid valvular tissues into the right ventricle. In the region of these displaced leaflets, a third right heart "chamber" is present which is morphologically and electrically a ventricle, but mechanically an atrium. An intracardiac electrode catheter in this location detects a ventricular electrogram and an atrial pressure pulse. In addition to these unique and diagnostically useful findings in the electrode catheter, patients with Ebstein's anomaly display a number of other electrocardiographic abnormalities (table 1).

We recently studied five patients with Ebstein's anomaly of the tricuspid valve with currently refined intracardiac electrophysiologic methods. The purpose of this paper is to report the results of these investigations which shed light on a number of previously unanswered questions about electrophysiology in Ebstein's anomaly.

Methods

The patients were evaluated in the Cardiac Clinical Electrophysiology Laboratory after overnight fasting without sedation. They had previously been informed about the procedure and gave their consent. Venous catheters were introduced either by cut-down or percutaneously into the right basilic vein and a femoral vein and placed within the heart under fluoroscopic control. Recording sites included: the right atrium (RA), atrialized right ventricle (ARV), right ventricular apex (RVA), right ventricular outflow tract (RVOT), and across the tricuspid valve to obtain a His bundle electrogram. Two types of luminal electrode catheters were used: (1) Zucker USCI electrode-luminal catheter with electrodes 1.5 cm apart, and (2) close bipolar (Elecath Corporation) luminal catheter with electrodes separated by 1 mm. Intracardiac pressures were measured with Statham P23Db pressure transducers. Other electrograms were recorded with standard bi-, tri-, or...
Table 1

Clinical Electrocardiographic Abnormalities in Ebstein's Anomaly of the Tricuspid Valve

1) Intra-atrial conduction disturbances (unusual P waves)
2) Prolonged atrioventricular conduction (prolonged P-R intervals)
3) Bizarre form of right bundle branch block with "second" QRS associated to the preceding "normal" complex
4) Type B Wolff-Parkinson-White syndrome
5) Supraventricular tachycardia
6) Atrial fibrillation or flutter

Quadripolar electrode catheters. Electrograms were amplified and filtered with Model EEP (Electronics-for-Medicine) amplifiers with low-frequency cut-off at 40, 120, or 400 Hz and high frequency cut-out set at 500 Hz. Comparison of filter settings was performed by feeding the same signal from a catheter into separate amplifiers with different filter settings.

Intracardiac recordings were simultaneously displayed on a multichannel oscilloscope (Electronics-for-Medicine DR16) with several surface leads. Time lines at 10 and 100 msec were also recorded. Information was stored on a Hewlett-Packard or Honeywell 14 channel Tape System for later analysis.

All measurements were made at a paper speed of 150-200 mm/second. P and QRS durations were taken as the maximum value of several simultaneously recorded surface ECG leads. The normal values used in our laboratory are: A-H interval: 60-140 msec; H-V interval: 35-55 msec; maximum duration of bundle of His deflection: 20 msec; and maximum P-A interval: 45 msec.

Electrical stimulation was conducted with a digital stimulator (Bloom Associates, Ltd., Narberth, Pennsylvania). Refractory periods were determined with the extrastimulus method. Basic cycle length was established in the right atrium (A1-A2) or right ventricle (V1-V2) and premature beats (A3 or V3) were introduced every 8-10 beats. The effective refractory period (ERP) of the atrium or ventricle was defined as the longest interval from the last basic cycle length beat to the test stimulus when capture of the chamber failed. ERP of the A-V node was defined as the longest A1-A2 interval when a bundle of His deflection (Hb) did not appear. In the patient with pre-excitation, the ERP of the bypass tract was defined as the longest A1-A2 interval when antegrade conduction with pre-excitation of the ventricle disappeared.

Patients: Clinical and Electrophysiologic Data

Patient D. G.

This 18-year-old man had had a heart murmur first noted at age four years. Growth and development were normal, and his exercise capacity was only minimally impaired. He was referred because of palpitations associated with light-headedness, pounding in the chest, and syncope. This patient, as well as each of the others, was not cyanotic when admitted for study. Physical findings, X-ray appearance and catheterization data for D. G. and the other patients are summarized in table 2.

Surface Electrocardiogram

Leads I, II, aVF, V1, and V6 for D. G. and the other patients are shown in figure 1, and the electrophysiologic data in table 3. Note the presence of a small delta wave directed to the left, superiorly and posteriorly. During sinus rhythm the ventricles usually appear to be depolarized via the A-V node-His bundle pathway.

Right Heart Chambers

The three chambers of the right heart were identified by their electrophysiologic and hemodynamic properties. At the apex of the right ventricle (RVA) a ventricular pressure pulse was recorded, and the principal deflection in the filtered bipolar electrogram occurred 65 msec after the onset of the delta wave (fig. 2, left panel). Upon withdrawal, the catheter tip then entered the atrialized portion of the right ventricle (ARV). The characteristic RV pressure pulse disappeared, but the electrogram continued to be recorded within the QRS, although now the principal deflection was slightly earlier (fig. 2, right panel). The catheter next entered the right atrium proper (RA) where an atrial electrogram was recorded at the onset of the P waves. The mean pressure was similar to that of the ARV but the phasic deflections were less well formed.

His Bundle Location: Atrioventricular Electrophysiology

The bundle of His deflection was recorded when the catheter tip was at the junction of the RA and the...
ARV and could not be recorded distally within the ARV or at its junction with the mechanical RV.

In table 3, the electrophysiologic measurements have been listed under three conditions: in sinus rhythm (slight pre-excitation), when pre-excitation was enhanced by atrial pacing, and during re-entrant supraventricular tachycardia when no pre-excitation was present. Infra-nodal conduction measured during SVT and intra-His conduction were prolonged. The effective refractory period (ERP) of the bypass tract was 280 msec at an atrial cycle length of 650 msec.

Arrhythmias

SVT was induced with both paced atrial premature depolarizations and by catheter induced ventricular premature beats from the apex of the mechanical RV (fig. 3). During SVT pre-excitation disappeared and right bundle branch block with QRS duration of 110 msec was seen.

During manipulation of the catheter in the ARV many ventricular beats were produced. Ventricular fibrillation developed once and the patient was converted to sinus rhythm with external countershock.

Endocardial Activation

Depolarization of the proximal ARV (in the region of the bundle of His), distal ARV, RVA, and RVOT were determined during sinus rhythm, following premature atrial depolarizations, producing predominant ventricular depolarization via the bypass tract, and in some locations during pure A-V nodal-His conduction in SVT. The data are listed in table 3. The site of ventricular insertion of the bypass tract could not be delineated.

Patient W. M.

A 23-year-old man, who had had a heart murmur since birth, was found to have normal growth and development. There was no history of diminished exercise tolerance, exertional dyspnea, weakness or palpitations (table 2).

Surface Electrocardiogram (fig. 1, W. M.)

The duration of P waves was slightly prolonged and their peaked appearance in V1 suggested right atrial enlargement. The P-R interval was slightly prolonged

Figure 2

Patient D.G. Left Panel) Electrical and hemodynamic events at the right ventricular apex. Displayed are surface leads I, II, and aVF plus bipolar electrogram and pressure records recorded from the same catheter, together with time signals. A ventricular pressure pulse is shown with electrical depolarization 65 msec after beginning of the QRS complex. The patient is in sinus rhythm and a small delta wave due to ventricular pre-excitation is present. Right Panel) Electrical and hemodynamic events in the atrialized right ventricular chamber. Note the different pressure pulse, similar to that recorded from the right atrium. Ventricular electrical activity is still present although the deflection begins earlier in the QRS compared to the recording at the right ventricular apex.

Figure 3

Patient D.G. Supraventricular tachycardia induced by depolarization in the RVA. Displayed are surface leads II and V1, plus a bipolar electrogram from the high right atrium (HRA), a His bundle electrogram (HBE), and time signals. Antegrade conduction occurs via the atrioventricular (A-V) node-His bundle pathway and no pre-excitation is seen. Right bundle branch block of Ebstein’s anomaly is present. Infra-nodal conduction is prolonged (H-V = 60 msec), a finding in four of our five cases. The atria are depolarized retrogradely, as expected in re-entrant supraventricular tachycardia whether the re-entrant pathway courses within the A-V node or over the Kent bundle.
Table 2

Clinical, Hemodynamic, and Angiographic Data in Five Patients with Ebstein’s Anomaly of the Tricuspid Valve

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/Sex</th>
<th>BP</th>
<th>Rhythm</th>
<th>Palpation</th>
<th>Physical findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>D.G.</td>
<td>18/M</td>
<td>140/70</td>
<td>SR; WPW</td>
<td>Normal LV impulse, infundibular impulse 3rd LSB</td>
<td>Widely split, P2 soft PEA</td>
</tr>
<tr>
<td>W.M.</td>
<td>23/M</td>
<td>140/80</td>
<td>SR</td>
<td>No precordial impulses</td>
<td>Single</td>
</tr>
<tr>
<td>M.R.</td>
<td>57/F</td>
<td>150/80</td>
<td>SR; PAF</td>
<td>Hyperkinetic LV impulse in anterior axillary line; sub-xiphoid impulse</td>
<td>Widely split, PEA P2 soft</td>
</tr>
<tr>
<td>W.V.</td>
<td>30/M</td>
<td>90/60</td>
<td>SR; type I A-V block</td>
<td>Normal LV impulse</td>
<td>Split and “scratchy”</td>
</tr>
<tr>
<td>A.E.</td>
<td>19/F</td>
<td>135/70</td>
<td>SR</td>
<td>Normal LV impulse</td>
<td>Split, loud second component</td>
</tr>
</tbody>
</table>

Abbreviations: a = a wave; AR = aortic regurgitation; ASD = atrial septal defect; BP = blood pressure; EF = ejection fraction; F = female; LSB = left sternal border; LV = left ventricle; M = male; MR = mitral regurgitation; PA = pulmonary artery; PAF = paroxysmal atrial fibrillation; PEA = partial expiratory approximation of split of S2; RA = right atrium; RV = right ventricle; S1, S2, S3 = first, second, third and fourth heart sounds; SR = sinus rhythm; SVT = supraventricular tachycardia; TR = tricuspid regurgitation; v = v wave; WPW = Wolff-Parkinson-White syndrome.

at 220 msec. The R’ in lead V1 was wide and notched, a finding characteristic of patients with Ebstein’s anomaly. Pre-excitation was not present.

Right Heart Chambers, His Bundle Location, and Atrioventricular Electrophysiology

The electrode catheter was passed through the femoral vein into the body of the right ventricle where ventricular pressure and a ventricular electrogram were obtained. On withdrawal into the ARV atrial pressure curves appeared with a ventricular electrogram which was followed on slight further withdrawal by a His bundle electrogram (fig 4, table 3). The H-V interval, the His bundle spike duration and the P-A interval were prolonged (table 3).

Arrhythmias

Ventricular fibrillation treated with external countershock developed once during an RV-to-RA pullback. The episode was initiated by mechanical stimulation of the ARV. Neither SVT nor pre-excitation could be produced with premature atrial stimulation or by rapid atrial pacing.

Endocardial Activation

Activation in the right ventricular outflow tract was recorded at 80 msec after onset of QRS, as expected in the presence of right bundle branch block.9

Patient M. R.

The patient was a 57-year-old female factory worker who was said to have been "blue" at birth, but not thereafter. In youth she was “sickly” and unable to keep up with her peers, because of shortness of breath and weakness. Nevertheless, she married and...
gave birth to two children although physicians feared the risk of pregnancy. She led an active life until age 56 when after observing palpitations for several years, she developed paroxysmal atrial fibrillation which converted with digoxin (table 2).

Surface Electrocardiogram

The characteristic spiked R' in lead V1 and bizarre wide S waves were seen (fig. 1, M.R.). P-R prolongation and Wenckebach periods, probably induced by digoxin, were present on admission (not illustrated).

Right Heart Chambers, His Bundle Location and Atrioventricular Electrophysiology

In the apex of the right ventricle electrical depolarization occurred with a typical RV pressure pulse. On slight withdrawal the pressure pulse decreased and its form changed, but a ventricular electrogram was still recorded.

The bundle of His depolarization was easily recorded at the point where the intracardiac electrical signal changed from ventricular to atrial, thus at the junction of ARV and right atrium. Intra-atrial and His-Purkinje conduction times were prolonged while A-V nodal conduction was normal (table 3). However, the ERP of the A-V node was prolonged at 480 msec, which may have been due in part to digoxin. Much ventricular irritability appeared when the catheter was being manipulated in the atrialized RV.

Endocardial Activation

The RVOT was depolarized before the RVA, a

Table 3

Electrophysiologic Measurements in Five Patients with Ebstein's Anomaly of the Tricuspid Valve

<table>
<thead>
<tr>
<th>Pt</th>
<th>P-R</th>
<th>P width</th>
<th>P-A</th>
<th>A-H</th>
<th>H-V</th>
<th>BH duration</th>
<th>QRS</th>
<th>Prox ARV</th>
<th>Dist ARV</th>
<th>RVA</th>
<th>RVOT</th>
<th>LVA</th>
<th>VF</th>
</tr>
</thead>
<tbody>
<tr>
<td>D.G. (NSR)</td>
<td>120</td>
<td>110</td>
<td>35</td>
<td>60</td>
<td>25</td>
<td>30</td>
<td>120</td>
<td>50</td>
<td>40</td>
<td>65</td>
<td>60</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>D.G. (WPW)</td>
<td>165</td>
<td>95</td>
<td>70</td>
<td>60</td>
<td>30</td>
<td>110</td>
<td>35</td>
<td>80</td>
<td>180</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>W.M.</td>
<td>220</td>
<td>140</td>
<td>65</td>
<td>80</td>
<td>65</td>
<td>30</td>
<td>110</td>
<td>60</td>
<td>80</td>
<td>50</td>
<td>No</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>M.R.</td>
<td>180</td>
<td>110</td>
<td>50</td>
<td>95</td>
<td>65</td>
<td>20</td>
<td>165</td>
<td>145</td>
<td>25</td>
<td>70</td>
<td>50</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>W.V.</td>
<td>440</td>
<td>160</td>
<td>50</td>
<td>50</td>
<td>160*</td>
<td>80</td>
<td>20</td>
<td>120</td>
<td>60</td>
<td>75</td>
<td>45</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>A.E.</td>
<td>200</td>
<td>120</td>
<td>40</td>
<td>90</td>
<td>50</td>
<td>15</td>
<td>110</td>
<td>70</td>
<td>80</td>
<td>30</td>
<td>No</td>
<td>No</td>
<td></td>
</tr>
</tbody>
</table>

All numbers are in msec.

*After 1 mg atropine.

Data for D.G. are shown in three different states: in sinus rhythm (NSR) when a slight degree of pre-excitation was present (fig. 1); when pre-excitation had been increased by atrial premature beat coupling of 310 msec to the previous sinus beat (WPW); during re-entrant supraventricular tachycardia (SVT) when antegrade conduction occurred exclusively via the A-V nodal-His system (fig. 3).

Abbreviations of locations where electrograms of ventricular activation were made: ARV = "atrialized" right ventricle; RVA = right ventricular apex; RVOT = right ventricular outflow tract; LVA = left ventricular apex. VF = ventricular fibrillation induced during catherization; BH = bundle of His; Pt = patients; Prox = proximal; Dist = distal.
distinctly unusual finding in patients with right bundle branch block.\textsuperscript{7} Within the ARV, two different activation times were recorded. In the distal ARV early depolarization occurred at 25 msec. However, in the proximal ARV just distal to the location where the bundle of His electrogram was recorded, a very late ventricular signal appeared, at 145 msec, which coincided with the R’ of the surface electrocardiogram (fig. 5).

**Ventricular and Atrial Refractoriness**

The effective refractory period of the RV was 280 msec with the basic cycle length of 650 msec established at the RVA. In the ARV at a cycle length of 625 msec, the ERP was shorter at 230 msec. In both locations the electrode had been placed firmly against the chamber walls by curving the electrode in the ARV and applying it under pressure in the apex, and in each instance the same pacing power was used. The effective refractory period of the right atrium was 300 msec.

**Patient W.V.**

This 30-year-old man was first told of a heart murmur at age 13 years. Growth and development were normal, but syncope occurred from youth until about one year before admission. Edema, fatigability, exertional dyspnea, and palpitations were absent (table 2).

**Surface Electrocardiogram**

The electrocardiogram showed a right ventricular conduction defect with QRS width of 120 msec (fig. 1, W.V.). A delta wave was not seen. Atrioventricular conduction was markedly prolonged with type I second degree A-V block and Wenckebach periods present most of the time. The P wave was broad with duration of 160 msec.

**Right Heart Chamber, His Bundle Location, and Atrioventricular Electrophysiology**

In addition to the surface leads and intracardiac pressure both unipolar (0–500 Hz) and bipolar (120–500 Hz) electrograms were recorded. The right ventricular apex concordant ventricular pressure and ventricular electrogram were found (fig. 6). On withdrawal of the catheter, a sizable chamber in which an atrial pressure and ventricular electrogram were recorded was entered. At the usual location of the tricuspid valve, a bundle of His electrogram appeared with prolonged P-A, A-H, and H-V intervals (table 3).

As had been observed on the surface electrocardiogram, second degree ativoventricular (A-V) nodal block with Wenckebach periods was present. Following 1 mg of atropine intravenously the A-V nodal block changed to first degree with P-R interval of 290 msec and A-H interval of 160 msec. The effective refractory period of the A-V node after atropine was prolonged at 400 msec.

**Endocardial Activation**

Filtered bipolar electrograms in the region of the RVA showed deflections 45 msec after the onset of QRS. At the point where RV pressure disappeared, a

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**Figure 5**

Patient M.R. Late depolarization of the atrialized right ventricle was documented just distal to the tricuspid valve. Shown are leads I, aVF, V\textsubscript{1}, the bipolar electrogram in the atrIALIZED RV, and an atrial pressure curve.

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**Figure 6**

Patient W.V. Withdrawal of the electrode catheter from the body of the right ventricle to the atrialized right ventricle. Surface lead I, unipolar and bipolar electrical and hemodynamic records obtained from the same bipolar luminal catheter are displayed. As the catheter was withdrawn and effective ventricular contraction disappears, electrical depolarization remains within QRS.
broad electrogram was recorded with peak deflections at 20 and 75 msec. Upon slight further withdrawal within the ARV, the 75 msec deflection predominated. The catheter was then withdrawn to the proximal ARV where late ventricular activation at 60 msec was seen.

The left ventricle was entered through the left atrium via an atrial septal defect or foramen ovale. Activation of the LV apex occurred at onset of QRS. In the left atrium activation was recorded 100 msec after the beginning of a broad P wave of 160 msec duration.

Patient A.E.

She is a 19-year-old woman, who was cyanotic during the first week of her life, but not thereafter. A murmur was first heard at birth. At age five years she was catheterized and the diagnosis of Ebstein's anomaly was made. Although avoiding competitive sports, she has been completely asymptomatic and has never had paroxysmal tachycardias (table 2).

Surface Electrocardiogram

The width of the P wave was prolonged at 120 msec, the P-R interval was at the upper limits of normal, QRS (width 110 msec) showed right bundle branch block with notched S wave in V2 (fig. 1, A.E.).

Right Heart Chambers, His Bundle Location, and Atrioventricular Electrophysiology

The electrode catheter passed through the femoral vein recorded normal pressures in the pulmonary artery, right ventricle, and right atrium. Dissociation between electrical and mechanical activity in the ARV was not observed. However, when inserted through the right antecubital vein, the catheter entered a chamber from which both a ventricular electrogram and atrial mechanical activity just proximal to the normally contracting right ventricle were recorded. This was the only patient in whom the intracardiac conduction values were normal (table 3). Right ventricular angiography showed mild tricuspid regurgitation and confirmed the presence of an abnormally small contracting right ventricle and a large right atrium.

Discussion

Atrial Abnormalities

Electrocardiographic abnormalities in Ebstein's anomaly closely relate to the anatomic malformations of the right heart. The enlarged right atrium is recognized by P waves which are characteristically tall, peaked, and occasionally broad. The mean axis of these P waves is usually inferior, anterior, and to the left, and their extraordinarily large amplitude may be maintained in the left precordial leads.2, 8-12

Abnormal P wave morphology was found in all five subjects. The P waves were peaked and the width greater than 100 msec in each case. Although abnormally tall P waves are accepted as evidence of right atrial enlargement, increased duration is usually the hallmark of left atrial enlargement. The increased width of the P wave in Ebstein's anomaly may actually reflect the prolonged time required to traverse the abnormally large right atrium.2, 9, 11 This thesis is supported by the prolonged P-A interval we found in three of the five cases.

Atrioventricular Conduction

Atrioventricular block has been reported to occur in over 25% of cases of Ebstein's anomaly; usually only P-R prolongation is present.2, 8, 10, 11 In our patients the P-R interval was normal in M.R. and A.E., and in D.G., who had ventricular pre-excitation. It was prolonged in W.M. and in W.V., who showed both first degree block and type I second degree A-V block.

With intracardiac electrocardiography we found prolongation of some aspect of atrioventricular conduction in four of our five patients with Ebstein's anomaly although the P-R interval on the surface ECG was normal in three of five cases. Atrioventricular nodal conduction was prolonged in two and infranodal conduction was longer than normal in four. We did not establish the A-V nodal refractory periods in each patient, and a more subtle abnormality of A-V nodal function might have been present.

The prolonged infranodal conduction in patients with Ebstein's anomaly results from either impaired conduction and/or a lengthened, stretched route of conduction within the atrialized RV. Presumably the anatomic abnormality affects the specialized conduction system relatively early, possibly before bifurcation occurs. Otherwise one would have to conclude that the H-V prolongation reflects some associated left bundle branch disease. The single patient without H-V prolongation (A.E.) also had the most nearly normal anatomy. Her atrialized RV was small, and she was clinically without symptoms.

Intraventricular Activation

The pattern of right bundle branch block is found in up to 95% of patients with Ebstein's anomaly.2, 8, 10, 11 The QRS configuration itself may be rather unusual, with relatively low voltage, notching and slurring. Right ventricular conduction disturbances were present in each of our patients with W.M. and W.V. showing a quadriphasic rSr's' pattern. A.E., with the mildest disease, had the least QRS abnormality.

Endocardial mapping helped to define some features of the conduction disturbances within the right bundle branch. Expected endocardial activation times in patients with right bundle branch block are
54 ± 16 msec (± 1 sd) at the right ventricular apex and 78 ± 21 msec at the right ventricular outflow tract. Values obtained in our patients with Ebstein's anomaly in these two locations are within the anticipated ranges except for the unexpected and unexplained reversal in right ventricular apex-right ventricular outflow tract activation sequence in M.R. The short conduction time to the RVA (30 msec) in A.E. begins to approach the value observed in patients without right bundle branch block (18 ± 9 msec) and again reflects the minimal degree of abnormality present.7

Activation within the atrialized portion of the RV in Ebstein's anomaly can occur throughout most of QRS as suggested by the broad range of values obtained from within that 'chamber' (table 3). The data from M.R., W.V., and A.E. support the suggestion that the slurred terminal portion of the QRS (the 'second QRS') is produced by delayed depolarization in some portion of the large, stretched, thin-walled atrialized right ventricle.2, 8, 12

Location and Physiology of Bundle of His

Although tricuspid valve structure is uniformly deformed in Ebstein's anomaly, the normal relationships of the A-V node and bundle of His appear to be preserved. In each instance the His bundle electrogram was easily recorded in the usual location, at the junction of the right atrium and atrialized right ventricle where the tricuspid valve normally arises.

Conduction within the bundle of His, however, was abnormal in two of the five cases, with the duration of the His bundle deflection prolonged to 30 msec in D.G. and W.M. "Split-His" potentials, another indication of intra-His block, were not observed in our patients with Ebstein's anomaly.13

Pre-excitation

It is a clinical maxim that the diagnosis of Ebstein's anomaly should be considered when a cyanotic patient presents with type B Wolff-Parkinson-White (W.P.W.) syndrome.2, 11 This rule has its limitations, since cyanosis is frequently absent and pre-excitation is present only in a minority of cases. Our patient with W.P.W. syndrome demonstrated the type B pattern, which became evident during atrial stimulation.

Insertion of the bypass tract in type B pre-excitation has been demonstrated in the anterior portions of the right ventricle, although variations are quite frequent.14 Although Watson and Lowe have suggested that the site of pre-excitation in patients with Ebstein's anomaly and W.P.W. syndrome is somewhere between the A-V annulus and the apex of the right ventricle, we were unable to map a point within either the atrialized or mechanical ventricle which was depolarized especially early during pre-excitation.15 All right ventricular points except the apex were activated much later during pre-excitation than when A-V nodal-His bundle conduction occurred.

Ventricular Irritability and Refractoriness

We observed, as have others, that the thin-walled atrialized right ventricle was particularly irritable when the catheter was passed into this region. Ventricular fibrillation was provoked in two patients. Because of this possibility, particular care should be exercised when the catheter is manipulated in the ARV.

Assuming unusual electrical characteristics within the ARV, we found it interesting that the effective refractory period of the atrialized RV was shorter than the mechanical RV. Although the basic cycle length was slightly shorter in the atrialized RV the difference in refractoriness was much greater than is usually found from different RV locations at various cycle lengths in patients without Ebstein's anomaly.16

Intracardiac Electrocardiography

The diagnostic value of the intracardiac electrode in patients with Ebstein's anomaly has been recognized for over 15 years.5, 10, 17-22 It is, therefore, not surprising that the first report in the English literature of the human His bundle potential was obtained with an electrode catheter from a 14-year-old girl with Ebstein's anomaly.23 Except in the diagnosis and evaluation of arrhythmias, the electrode catheter has found its most useful clinical application in patients with Ebstein's anomaly.

Prior to the current report, intracardiac electrocardiography in Ebstein's anomaly has been described with the use of unipolar unfiltered leads with the electrode connected to the V lead of the standard electrocardiogram. In our studies the filtered bipolar system has been employed. In such recordings distant electrical events are attenuated, and the locus in the heart where the electrical signal passes the electrodes is emphasized (fig. 6). We found it easier to localize the electrode and to observe activation times with the bipolar filtered system and believe that this technique is diagnostically and technically superior to the other method.

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