Early Closure of the Tricuspid Valve in a Case of Ebstein’s Anomaly with Type B Wolff-Parkinson-White Syndrome

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SUMMARY Echocardiographic studies of a 19-year-old female with Ebstein’s anomaly and type B Wolff-Parkinson-White syndrome showed an earlier closure of the anterior tricuspid leaflet (ATL) than of the anterior mitral leaflet (AML), contrary to the previous reports. The interval between the closure of the ATL and the AML was 20 msec and 30 msec, respectively, before and after administration of edrophonium chloride. However, closure of the AML preceded that of the ATL after administration of atropine sulfate and during supraventricular tachycardia by 10 msec and 60 msec, respectively, concomitant with the shortening of the duration of the QRS complex. We conclude that early closure of the ATL may depend on preexcitation of the right ventricle.

ECHOCARDIOGRAPHIC FEATURES of Ebstein’s anomaly include: 1) abnormally delayed closure of the anterior tricuspid leaflet (ATL); 2) increased amplitude of the ATL with abnormal E-F slope; 3) dilated right atrium; 4) ability to record the ATL over most of the left precordium; and 5) abnormal ventricular septal motion.1-6 The delayed closure of the ATL is the only feature specific for this anomaly, because the other findings may be present in other diseases that produce right-sided volume overload.6-8 Usually, the interval between the closure of the anterior mitral leaflet (AML) and the ATL in Ebstein’s anomaly is greater than 30 msec.1-9 The abnormally delayed closure of the ATL is not considered due to right bundle branch block (RBBB) frequently present in this anomaly; it is considered a specific feature of the anomaly.3

We report a case of Ebstein’s anomaly with the early closure of the ATL and investigate the etiology of this finding.

Case Report

A 19-year-old female was referred to our clinic for cardiac evaluation. She had been faintly cyanotic since infancy. She had had episodes of tachycardia approximately 10 times a year, but no episodes of heart failure.

At admission, height was 156 cm, weight was 50 kg, body temperature was 36.5°C, pulse rate was 60 beats/min, and blood pressure was 116/74 mm Hg. Cyanosis of nail beds with faint clubbing and malar flush were noted. Jugular veins were not distended. The apex beat was in the left fifth intercostal space midway between the midclavicular and anterior axillary line. Two components of the first heart sound and a single second heart sound were confirmed by phonocardiogram. Third and fourth heart sounds were also recorded. A grade 3/6 early systolic murmur was best heard in the fourth intercostal space at the left sternal border. A chest roentgenogram revealed normal pulmonary markings, a marked convexity of the
cardiac shadow to the right of the sternum, an elevation of the apex, a normal-sized aortic knob and a hidden pulmonary knob. The resting ECG (fig. 1A) revealed normal sinus rhythm (67 beats/min), a left-axis deviation and type B Wolff-Parkinson-White (WPW) syndrome (duration of the QRS complex 260 msec). Ten minutes after administration of edrophonium chloride (10 mg i.v.), the ECG (fig. 1B) revealed sinus bradycardia (56 beats/min), a left-axis deviation and type B WPW syndrome (duration of the QRS complex 300 msec). Five minutes after administration of atropine sulfate (1.5 mg i.v.), the ECG (fig. 1C) demonstrated sinus tachycardia (107 beats/min), a right-axis deviation and type B WPW syndrome (duration of the QRS complex 220 msec). During spontaneous tachycardia, the ECG (fig. 1D) revealed supraventricular tachycardia (SVT) (130 beats/min) with a right-axis deviation and the shortening of the QRS complex (duration of the QRS complex 160 msec), and suggested RBBB.

A cardiac catheterization was performed; the pressure data are shown in table 1. A selective right atrial angiogram confirmed the presence of Ebstein's anomaly as follows (fig. 2): In the posterior-anterior view, we noted a marked enlargement of the right atrium which extended leftward beyond its usual

TABLE 1. Pressure Data

<table>
<thead>
<tr>
<th>Location</th>
<th>Pressure (mean) (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAW</td>
<td>a,c = 7, x = 5, v = 6, y = 5</td>
</tr>
<tr>
<td>PA</td>
<td>13/8</td>
</tr>
<tr>
<td>RV</td>
<td>14/2</td>
</tr>
<tr>
<td>RA</td>
<td>a,c = 5, x = 2, v = 3, y = 1</td>
</tr>
<tr>
<td>LA</td>
<td>a,c = 7, x = 4, v = 5, y = 3</td>
</tr>
<tr>
<td>LV</td>
<td>100/2</td>
</tr>
<tr>
<td>Ao</td>
<td>100/70</td>
</tr>
</tbody>
</table>

Abbreviations: PAW = pulmonary arterial wedge; PA = pulmonary artery; RV = right ventricle; RA = right atrium; LA = left atrium; LV = left ventricle; Ao = ascending aorta.

FIGURE 1. A) The ECG at rest shows normal sinus rhythm, left-axis deviation and type B Wolff-Parkinson-White (WPW) syndrome. Duration of the QRS is 260 msec. B) The ECG after administration of edrophonium shows sinus bradycardia, left-axis deviation and type B WPW syndrome. Duration of the QRS is 300 msec. C) The ECG after administration of atropine shows sinus tachycardia, right-axis deviation and type B WPW syndrome. Duration of the QRS complex is 220 msec. D) The ECG shows supraventricular tachycardia, right-axis deviation and suggestive right bundle branch block and the disappearance of the Δ wave.

FIGURE 2. Selective right atrial angiogram, posterior-anterior (left) and lateral view (right) of the present case. A voluminous right atrium extends leftward beyond the usual boundary. A notch is apparent at the site of the anomalous origin of the tricuspid valve. The right ventricle and pulmonary artery are slightly small. In the lateral view, the abnormal origin of the anterior tricuspid leaflet is located far anteriorly.
boundary. A notch was apparent at the site of the anomalous origin of the tricuspid valve. The right ventricle and the pulmonary artery were slightly small. In the lateral view, we saw the abnormally anteriorly located origin of the ATL. These angiographic findings suggested displacement of the tricuspid leaflets and the atrialization of the right ventricle.

The echocardiographic examinations were performed using a linear array real-time scanning system (Toshiba, SSL-53H), which uses a 2.4 MHz multi-transducer array interfaced to a strip chart recorder (Toshiba, FR-06A), or a single-crystal Echocardiograph (Aloka, SSD-110) with a 2.25 MHz transducer. The tracings were recorded on photographic paper at a speed of 100 mm/sec or on Polaroid film at a speed of 50 mm/sec. The closure of the ATL and the AML was defined as the "c" points when they came to the most posterior positions or when the anterior and posterior leaflets came together at the onset of systole. The opening of the ATL and the AML was defined as the "d" points when the slow motions of the ATL and the AML changed to rapid opening motions.

The ATL and the AML were easily traced simultaneously farther to the left of the left sternal border compared to patients with other diseases. Even when the transducers were moved to the left of the left midclavicular line, the ATL could still be recorded. Both the echocardiogram and the ECG were recorded at the same time at rest, after the administration of drugs and during spontaneous SVT. In each record, we saw abnormal interventricular septal motion (fig. 3). After the inscription of the P wave, the septal echoes moved posteriorly and then moved anteriorly after inscription of the QRS complex. These were followed in sequence by flattened septal echoes during late systole.

The most characteristic abnormal feature in this case was the motion of the ATL. The closure of the ATL was earlier than that of the AML by 20 msec before drug and by 30 msec after administration of edrophonium chloride. However, the delayed closure of the ATL compared with that of the AML was observed after administration of atropine sulfate by 10 msec and during SVT by 60 msec. The opening of the ATL was earlier than that of the AML, respectively, by 140 msec before drug, by 145 msec after administration of edrophonium chloride and by 70 msec after administration of atropine sulfate. The opening of the ATL was delayed to that of the AML by 30 msec during SVT.
Discussion

According to previous reports, the delayed closure of the ATL compared with that of the AML by 30 msec or 60 msec is the most characteristic feature of Ebstein's anomaly. Tajik et al. reported an adult patient with right ventricular preexcitation without RBBB who received surgical treatment for the obliteration of the preexcitation and tricuspid annuloplasty. Abnormally delayed closure of the ATL, noted preoperatively, persisted postoperatively with the same duration. They concluded that the delayed closure of the ATL in Ebstein's anomaly is primarily due to the displacement of the ATL and is independent of the mode of the ventricular preexcitation, although RBBB was frequently observed in this anomaly, probably by stretching the conduction system over the atrialized ventricle. Farooki et al. compared the intervals between the closure of the AML and that of the ATL (Mc-Tc interval) in Ebstein's anomaly with the Mc-Tc intervals of other congenital heart diseases, and found that in Ebstein's anomaly, the Mc-Tc intervals exceeded 30 msec, except in rare cases. In our previously reported five cases and in two additional unpublished cases, the delayed closure of the ATL exceeded 40 msec.

The early closure of the ATL may have occurred when the mechanical events had been earlier in the right-sided heart than in the left or contrary to it, when that had been later in the left than in the right. The preexcitation of the right ventricle and/or severe pulmonary insufficiency (PI) belong to the former, and left bundle branch block (LBBB) and/or severe mitral stenosis (MS) to the latter. PI and MS were ruled out by the cardiac catheterization. LBBB could explain the behavior of the ATL, but this is unlikely, since there was no LBBB in this case (fig. 1). The most plausible etiology of the unusual findings through the maneuvers is the contribution of the preexcitation. The ECG at rest before drug administration (fig. 1A, table 2) demonstrated type B WPW syndrome, so the preexcitation pathway is probably located in the anterior wall of the right ventricle; therefore, the right ventricular electrical excitation was premature and occurred before left ventricular electrical excitation. In this situation, the closure of the ATL was 20 msec earlier than that of the AML. After administration of edrophonium chloride (fig. 1B, table 2), the duration of the QRS complex increased and the preexcitation of the right ventricle was more dominant because of the suppression of the normal atrioventricular nodal conduction, and the interval between the closure of the ATL and that of the AML increased to 30 msec. However, after administration of atropine sulfate (fig. 1C, table 2), the transmission of the electrical excitation to the ventricles was relatively more dominant through the normal pathway by the acceleration of the normal atrioventricular conduction, as the duration of the QRS complex was shortened. In this situation, the closure of the ATL was 10 msec later than that of the AML. Finally, during SVT (fig. 1D, table 2), the QRS morphology changed and the Δ wave was no longer present. The reentry circuit was probably from the normal atrioventricular nodal pathway to the accessory pathway, and the pattern of the QRS complexes suggested RBBB. In this case, the closure of the ATL was 60 msec later than that of the AML.

From these findings, we assume that the early right ventricular pressure rise exceeding the right atrial pressure, concomitant with the large Δ wave and the wide QRS complex may result in no reentry, and/or the atrioventricular interval; therefore, the closure of the ATL was 20 msec earlier than that of the AML. Finally, during SVT (fig. 1D, table 2), the QRS morphology changed and the Δ wave was no longer present. The reentry circuit was probably from the normal atrioventricular nodal pathway to the accessory pathway, and the pattern of the QRS complexes suggested RBBB. In this case, the closure of the ATL was 60 msec later than that of the AML.

This explanation is supported by previous reports. March et al. studied the mechanical consequences of 12 cases with WPW syndrome and reported that in one case with intermittent WPW, the right ventricular pressure rise occurred earlier in the WPW beat than in the normal beats. Zuberbuhler et al. reported that premature right ventricular mechanical events could occur in some cases with type B WPW syndrome.

Table 2. Comparative Analysis of ECG and Echocardiogram Before and After Drug and During Supraventricular Tachycardia

<table>
<thead>
<tr>
<th></th>
<th>Before drug (control)</th>
<th>After edrophonium</th>
<th>After atropine</th>
<th>During SVT</th>
</tr>
</thead>
<tbody>
<tr>
<td>HR (beats/min)</td>
<td>67</td>
<td>56</td>
<td>107</td>
<td>130</td>
</tr>
<tr>
<td>PQ (Δ) (msec)</td>
<td>80</td>
<td>80</td>
<td>100</td>
<td>*</td>
</tr>
<tr>
<td>QRS (msec)</td>
<td>260</td>
<td>300</td>
<td>220</td>
<td>160</td>
</tr>
<tr>
<td>Q-Tc (msec)</td>
<td>260</td>
<td>235</td>
<td>220</td>
<td>180</td>
</tr>
<tr>
<td>Q-Mc (msec)</td>
<td>280</td>
<td>265</td>
<td>210</td>
<td>120</td>
</tr>
<tr>
<td>Q-Td (msec)</td>
<td>530</td>
<td>535</td>
<td>450</td>
<td>310</td>
</tr>
<tr>
<td>Q-Md (msec)</td>
<td>670</td>
<td>680</td>
<td>520</td>
<td>340</td>
</tr>
<tr>
<td>Tc-Mc (msec)</td>
<td>20</td>
<td>30</td>
<td>-10</td>
<td>-60</td>
</tr>
<tr>
<td>Td-Md (msec)</td>
<td>140</td>
<td>145</td>
<td>70</td>
<td>-30</td>
</tr>
</tbody>
</table>

Abbreviations: SVT = supraventricular tachycardia; HR = heart rate; PQ(Δ) = PQ (Δ) interval; * = unmeasurable; QRS = duration of QRS complex; Q-Tc = interval between Q wave and c point of anterior tricuspid leaflet (ATL); Q-Mc = interval between Q wave and c point of anterior mitral leaflet (AML); Q-Td = interval between Q wave and d point of ATL; Q-Md = interval between Q wave and d point of AML; Tc-Mc = interval between c points of ATL and AML; Td-Md = interval between d points of ATL and AML.
Pocock et al.\textsuperscript{16} reported the clinical feature of nine cases of Ebstein's anomaly. In a case of theirs with type B WPW syndrome (case 2), the mechanical events occurred earlier in the right-sided heart than in the left.

The delayed closure of the ATL after administration of atropine sulfate and during SVT could be explained by the abnormality of the tricuspid valve itself and/or the potential contribution of the atrialized ventricle, as seen in common Ebstein's anomaly. The increasing heart rate, plus the abnormal anatomy, might have accentuated the delay of the closure of the ATL, because the atrium with the atrialized ventricle might have induced further incompetence of the emptying as the diastolic filling time got shorter. These factors — the abnormal anatomy and the increasing heart rate — may cause the delayed closure of the ATL, but not the early closure we observed in this case.

Although the abnormally delayed closure of the ATL compared with that of the AML is believed to be the most characteristic finding of Ebstein’s anomaly, there are exceptions; and when the delayed closure of the ATL is not observed in the echocardiographic examination of patients with congenital heart diseases, this anomaly cannot be ruled out. We believe that other cases of Ebstein’s anomaly with type B WPW syndrome and with the early closure of the ATL similar to our case will be found.

Acknowledgments

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References

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