Echocardiogram in Ebstein's Anomaly with Wolff-Parkinson-White Preexcitation Syndrome, Type B

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SUMMARY
The echocardiographic features of Ebstein's anomaly include a large anterior chamber, paradoxical ventricular septal motion (type B), increased velocity and amplitude of anterior tricuspid leaflet (ATL) motion, and delayed closure of the ATL. This delayed closure has been thought to be secondary to right bundle-branch block (RBBB). We have studied two patients by simultaneous echocardiography and phonocardiography. The findings in both patients were similar. One patient had RBBB and therefore resembles the previously reported patients. However, the other patient had W-P-W preexcitation syndrome, type B, and is reported here in more detail. The ATL closure of this patient was markedly delayed in spite of right ventricular preexcitation. This finding gives further insight into the altered pathophysiology in Ebstein's anomaly and implies that the delayed ATL closure is probably not due to RBBB and may be a specific feature of this anomaly.

Additional Indexing Words:
Ultrasound
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In Ebstein's anomaly, abnormal tricuspid valve and ventricular septal motion with a large anterior chamber have been described. Simultaneous echocardiographic and phonocardiographic recordings have led to some pathophysiologic correlations. Because of its associated accelerated A-V conduction, the present case suggests new correlations for these observed abnormalities, confirms what would appear to be characteristic findings in Ebstein's anomaly, and suggests that delayed closure of the ATL is not due to RBBB but rather is a specific abnormality found in this syndrome.

Case Report
The patient was a 20-year-old white woman who was referred to the Mayo Clinic for cardiac evaluation. A diagnosis of Ebstein's anomaly had been made at age 10 years at another institution. She had remained asymptomatic until 3 years ago, when she began to have frequent episodes of tachycardia, weight loss, cyanosis of digits, swelling of ankles, and loss of appetite.

At examination here, height was 64 inches, weight 104 pounds, temperature 98°F, and pulse rate was 56 beats/min. Blood pressure was 90/66 mm Hg. Cyanosis of the nail beds without clubbing was noted. Malar flush and hyperemia of the hands and feet were present. The jugular vein pressure was normal. The apex beat was in the fifth left intercostal space midway between the midsclavicular and anterior axillary lines. The first heart sound was widely split and had a loud second component. The second heart sound was normal. A third heart sound was also heard. A grade 2/6 mid-systolic murmur was best heard between the left sternal border and the apex.

Basic laboratory tests all gave normal results. A roentgenogram of the chest revealed gross cardiomegaly with normal pulmonary vascular markings. The electrocardiogram revealed a normal sinus rhythm (rate, 50 beats/min) and features of Wolff-Parkinson-White (W-P-W) preexcitation syndrome, type B (fig. 1, top). The delta vector was directed posteriorly and to the left. An echocardiogram was performed (described in following section). The diagnosis of Ebstein's anomaly was confirmed by cardiac catheterization and selective right ventricular angiography. In view of the marked recent deterioration in this patient's clinical status, surgical treatment was recommended.

At operation, the anterior leaflet of the tricuspid valve was noted to have several small perforations, the posterior leaflet was nearly atretic, and no septal leaflet was visualized. Precordial mapping revealed an area of preexcitation at the acute margin of the right ventricle. The atrialized portion of the right ventricle and the area
Electrocardiograms and vectorcardiograms. (Top) Preoperative tracings showing short P-R, prolonged QRS, and delta wave. Delta vector is directed posteriorly and to the left. Repolarization changes are present. (Bottom) Postoperative tracings showing normal P-R and QRS duration and no delta wave. Right-axis deviation and right ventricular conduction delay have now appeared. Minor repolarization changes are also evident.

of preexcitation were obliterated. The tricuspid leaflet was approximated back to the annulus, and a posterior annuloplasty was also performed. Remapping revealed no areas of preexcitation. The immediate postoperative course was complicated by excessive bleeding and cardiac tamponade which were successfully managed. The subsequent course was uncomplicated. A postoperative electrocardiogram (fig. 1, bottom) revealed no evidence of the W-P-W syndrome. An echocardiogram was performed on postoperative day 14.
ECHOCARDIOGRAM IN EBSTEIN'S ANOMALY

Preoperative echocardiograms. CW = chest wall; AW = anterior wall; TV = anterior tricuspid leaflet; IVS = interventricular septum; LVC = left ventricular cavity; LPW = left ventricular posterior wall.

(Left) Pattern shows large anterior cavity, small left ventricular cavity, and paradoxic (type B) septal motion. (Right) Two consecutive echocardiograms joined together. In addition to above features, anterior tricuspid leaflet motion is seen with markedly delayed systolic closure of this leaflet.

Echocardiography

Preoperative

A large anteriorly located chamber was noted, which was bounded anteriorly by the anterior wall and posteriorly by the right side of the interventricular septum (fig. 2, left). It measured 4.3 cm anteroposteriorly at end-diastole. This cavity was believed to represent the atrialized portion of the right ventricle as well as the true right ventricular cavity.

The motion of the interventricular septum was clearly abnormal. After the inscription of the P wave of the electrocardiogram, the septal echo moved slightly posteriorly. Then, after the QRS (during ventricular systole), anterior motion occurred with flattening of the septal echo. This was followed in sequence by a rapid posterior motion during early diastole and then a rapid anterior motion.

The most characteristic abnormal feature was the motion of the anterior leaflet of the tricuspid valve (fig. 2, right). The excursion of this leaflet was 2.0 cm, and it approached the anterior wall echo. The diastolic closure slope was normal. However, the systolic closure of this leaflet was markedly delayed, occurring 0.09 sec after closure of the mitral valve.

The left ventricular cavity (the vertical distance between the left side of the interventricular septum and the endocardial surface of the left ventricular posterior wall measured at end-diastole) was small (2.2 cm; the normal value in adults is 3.5-5.3 cm).3

The mitral valve motion, although essentially normal, included a peculiar anterior motion during the early part of diastole between the E-F slope. The systolic closure occurred at the normal time (immediately after QRS).

In figure 3, the first heart sound is shown to be widely split on the phonocardiogram. The loud second component of this sound (T1) occurred 0.09 sec after the first component (M1). The "Q-1" interval is prolonged to 0.10 sec (normal, 0.03-0.07 sec). This has been previously reported both in Ebstein's anomaly and in W-P-W syndrome.4 An early diastolic sound is also clearly visible. On correlating these phonocardiographic events with the simultaneously recorded echocardiogram, it is noted that the closure of anterior mitral leaflet coincides with the first component of the first heart sound (M1), and the second loud component of the

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first heart sound (T₁) occurs exactly at the time of closure of the anterior tricuspid leaflet. These observations confirm that the two components of the widely split first heart sound in Ebstein's anomaly are related to mitral and tricuspid valve closures, respectively. The early diastolic sound coincides with the point of maximal diastolic excursion of the anterior tricuspid leaflet and therefore most probably represents tricuspid opening snap and not a third heart sound (ventricular gallop).

**Postoperative**

The anterior chamber decreased to 3.7 cm while the left ventricular cavity size increased to 2.7 cm (fig. 4). The ventricular septal motion remained abnormal; that is, both the left ventricular posterior wall echo and the septal echo moved anteriorly.

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**Figure 4**

*Postoperative echocardiograms. Notations as before. (Left) Persistence of delayed tricuspid valve closure as compared to mitral valve. The septal motion is paradoxic (type A). (Right) Posteriorly located pericardial fluid (PF) is evident.*
ECHOCARDIOGRAM IN EBSTEIN'S ANOMALY

We believe that the abnormal delay in closure of the anterior tricuspid leaflet noted preoperatively, persisted postoperatively with approximately the same duration. The amplitude of the ATL motion remained essentially unchanged postoperatively. Another feature which was not present previously was an echo-free space posterior to the left ventricular posterior wall. This indicated the presence of residual pericardial effusion.

Discussion

The preoperative echocardiogram of this patient was distinctly abnormal, and similar echocardiographic features have been described recently by Kotler and Tabatznik in three patients with Ebstein's anomaly. The delayed closure of the anterior tricuspid leaflet (ATL) in their patients occurred 0.04-0.10 sec after the onset of the first heart sound. Moreover, in all three patients, the excursion of ATL was large and approached the chest wall echoes. This latter feature, however, was not present in our patient. The excursion of the ATL was also noted to be normal in one additional patient with Ebstein's anomaly who had right ventricular conduction delay. In this additional patient all other findings were as described above. We believe that the difference noted in ATL excursion in our patients as compared to those of Kotler and Tabatznik can be explained as a result of anatomic variation in the ATL in this syndrome. It is recognized that transducer position location could also account for this difference.

Crews et al. also studied the echocardiograms of three patients with Ebstein's anomaly and correlated these with simultaneous phonocardiograms. They, too, consistently observed a loud high-frequency sound occurring 0.04-0.10 sec after the onset of the first heart sound (S1). This sound coincided exactly with the closing movement of ATL on the simultaneously recorded echocardiogram. They attributed this delayed closure of ATL (resulting in widely split S1) to be a result of the right bundle-branch block (RBBB) which was present in all of their patients. However, they made no comment in their abstract regarding excursion of ATL or ventricular septal motion.

Review of these features has led us to believe that delayed closure of ATL might be the most specific echocardiographic finding in Ebstein's anomaly. This belief is further strengthened by the fact that, unlike Crews and associates' patients, our patient did not have RBBB to account for the delayed ATL closure. On the contrary, she had W-P-W preexcitation syndrome, type B. This leads to earlier excitation of right ventricle, which would be expected to result in earlier closure of ATL. Therefore, the fact that ATL closure was markedly delayed is consistent with the finding of a slow conduction type of atrioventricular (AV) block.

Figure 5

Cardiac catheterization record, showing abnormal right ventricular pressure pulse contour. Arrows mark points corresponding to anterior tricuspid leaflet closure. Vertical lines are 1 sec apart.
delayed in our patient, even in the presence of accelerated conduction, makes it a unique feature of this anomaly.

The delayed closure has been attributed to an altered pattern of ventricular contraction and abnormal leaflet placement. Fontana and Wooley performed simultaneous intracardiac sound and pressure studies in three patients with Ebstein’s anomaly and discovered an abnormal right ventricular pressure pulse contour. This consisted of an initial slow delta-wave configuration followed by a more rapid pressure increase. They noted that the loud early systolic sound (“sail sound”) occurred at the point where the slow portion of right ventricular pressure pulse gave rise to a rapid upstroke. Similar abnormality of right ventricular pressure pulse was also noted in our patient (fig. 5), and the junction point of slow and rapid components roughly correlated with the timing of the loud systolic sound (closure of ATL). The data in this case confirm that abnormal ATL placement with altered hemodynamics independent of the mode of ventricular excitation accounts for the delayed ATL closure.

Another interesting echocardiographic feature of this case was the abnormal ventricular septal motion. In normal subjects, the septal echo and the left ventricular posterior wall echo move in opposite directions during each cardiac cycle. Two types of abnormal ventricular septal motions were described first by Popp et al. and subsequently by Diamond et al. In type A, the left ventricular posterior wall echo and the septal echo both move in the same direction, that is, anteriorly during systole and posteriorly during diastole (paradoxic motion). In type B, instead of anterior motion, the septal echoes become flattened during systole. Conditions which result in paradoxic septal motion include atrial septal defect, partial or total anomalous pulmonary venous connection, partial atroventricular canal, tricuspid insufficiency, and Ebstein’s anomaly. With the exception of Ebstein’s anomaly, the majority of these conditions demonstrate type A paradoxic septal motion, with type B being relatively rare. The abnormal septal motion in our patient and in all three patients described by Kotler and Tabatznik resembled type B motion. The significance of this motion is not clear. However, all patients with Ebstein’s anomaly reported to date have demonstrated type B septal motion, which well may be a specific echocardiographic feature of this anomaly. It is interesting, however, that this septal motion became type A paradoxic after obliteration of the atrialized portion of the right ventricle in our patient.

Addendum

Since submission of this manuscript, another patient with Ebstein’s anomaly has been examined; the electrocardiogram revealed low voltage with minor repolarization changes only. In particular, there was no RV conduction delay. The echocardiogram revealed all of the above-described features of Ebstein’s anomaly with ATL closure occurring 0.05 sec after mitral valve closure. This case further substantiates our belief that the delayed ATL closure in this condition is not secondary to RV conduction delay.

References

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