such patients to determine the best course of future medical management.

Acknowledgment

Appreciation is expressed to Mrs. Pat Parker for her technical assistance.

References


Two-Dimensional Echocardiographic Assessment of Ebstein’s Anomaly

THOMAS A. PORTS, M.D., NORMAN H. SILVERMAN, M.D., AND NELSON B. SCHILLER, M.D.

SUMMARY  Nine patients with Ebstein’s anomaly of the tricuspid valve were studied by two-dimensional echocardiography, using the standard long and short axis views as well as the apex four chamber view. With this latter view, the displacement of the tricuspid valve into the right ventricle was clearly seen in all nine cases of Ebstein’s anomaly and was not noted in a control population. The severity of the tricuspid displacement was assessed by comparing the position of the mitral and tricuspid valves relative to the cardiac apex. The apex four chamber view allowed visualization of the atrioventricular (AV) ring simultaneously with the displaced tricuspid valve, and therefore the size of the “atriIALIZED” right ventricle, true right ventricle and right atrium could be determined. These dimensions compared favorably with angiography.

EBSTEIN’S ANOMALY of the tricuspid valve is a rare congenital heart disorder. It consists of downward displacement of all or part of a malformed tricuspid valve and associated architectural abnormalities of the right ventricle. Recent reviews have stressed the pathologic and clinical spectrum of this disorder. In the newborn it is frequently accompanied by cyanosis and heart failure, and must be differentiated from other forms of cyanotic heart disease. Patients with milder forms may remain asymptomatic, even in adulthood. This varied expression often makes the clinical diagnosis difficult, and a non-invasive means to diagnose and accurately assess the severity of this disorder is important. Phonocardiography, M-mode echocardiography and, more recently, two-dimensional imaging systems have proved useful in the diagnosis of Ebstein’s anomaly, but fail to estimate its severity. We evaluated nine patients with Ebstein’s anomaly, using a wide-angle (80°) phased-array sector scanner, using both conventional precordial images and apex echocardiography to define the anatomic severity of this deformity.

Materials and Methods

Nine patients with Ebstein’s anomaly were studied. They ranged in age from 6 hours to 32 years. Clinical severity was variable, ranging from asymptomatic in four to severely symptomatic with cyanosis and/or congestive heart failure in three. Cardiac catheterization and biplane cineangiographic data were available for all patients. The diagnosis of Ebstein’s anomaly was...
was based on the demonstration of a displaced tricuspid valve on right atrial or ventricular cineangiography in all patients. Simultaneous pressure and pullback intracardiac electrocardiographic recordings from the apex of the right ventricle to right atrium were obtained in eight of nine instances and used for further diagnostic confirmation.

Echocardiographic studies were performed on all patients, using a Smith Kline Echoline 20A ultrasoundoscope interfaced with a strip chart recorder. All patients were studied by two-dimensional echocardiography. The instrument employed was a prototype of the commercially available Varian Associates 32-element phased-array, wide-angle (80°) sector scanner. Two-dimensional images were obtained in the long axis, or sagittal plane, by directing the tomographic plane between the apex and base of the heart. Short axis, or transverse, views were obtained by directing the plane of sweep along a line drawn between the right hip and left shoulder, perpendicular to the long axis of the left ventricle. Apex echocardiography (the apex four chamber view) was performed by the technique established in our laboratory. For this view the transducer was placed over the cardiac apex and angulated in such a way as to visualize simultaneously all four of the cardiac chambers. The two-dimensional echocardiographic images were permanently recorded on videotape for further analysis and displayed on a calibrated screen. The illustrations presented here were obtained from Polaroid photographs of stop-action, single-frame scan images made from the videotape recordings. This process results in a reduction in image quality and a loss of the visual appreciation of motion normally present in these phased-array real-time recordings.

From the 30° right anterior oblique right ventricular cineangiograms, the size of the "atrialized" portion of the right ventricle was determined in diastole by measuring the distance from the AV groove to the displaced tricuspid valve (fig. 1). The AV groove was identified on cinfluoroscopy by the fat in the AV sulcus and on right atrial or ventricular angiography by the notch along the inferior border of the heart created by the junction of the true atrium and atrialized portion of the right ventricle. Similar measurements were made from the apex four chamber view from the two-dimensional echocardiographic images. In diastole the distance from the AV groove to the displaced tricuspid valve at its point of attachment to the interventricular septum was measured. Figure 2 illustrates the dimension used to measure the length of the "atrialized" right ventricle. The AV groove was identified on two-dimensional echocardiography by visualization of the fibrous AV ring which lies in the same plane as the

**Figure 1.** Right atrial angiogram in a 30° RAO projection from patient 3. The dashed line indicates the AV groove, which is much more apparent on the cineangiogram; arrows point to tricuspid valve; ″ defines length of "atrialized" right ventricle.
mitral valve (fig. 2). These echocardiographic and angiographic dimensions were then compared.

From the two-dimensional echocardiograms, using the apex four chamber view, the distance from the mitral valve to the cardiac apex and the distance from the tricuspid valve to the cardiac apex were determined in all nine patients with Ebstein's anomaly and compared to similar measurements made from 25 normal adults and children and 25 patients with right ventricular volume overload of various etiologies. The control patients had undergone cardiac catheterization. A ratio was then obtained, comparing the mitral-to-apex distance to the tricuspid-to-apex distance. Figure 2 illustrates the tricuspid-to-apex and mitral-to-apex distances.

One patient underwent surgical repair of this disorder (annuloplasty) and two patients had palliative shunt procedures. Pre- and postoperative two-dimensional echocardiograms were obtained in each of these cases. Contrast echocardiographic studies were performed in four patients to evaluate left-to-right shunting at the atrial level. The technique employed was the rapid peripheral intravenous injection of 2–5 cc of normal saline at the time the heart was being imaged with the apex four chamber and short axis views. The study was considered to be positive when the passage of air bubbles from the right to left atrium was detected.

Results

Apex View in Normals

A two-dimensional apex four chamber view from a normal patient is shown in figure 3. This view permits visualization of both the right and left ventricles, atria and AV valves side by side. The anterior and septal tricuspid leaflets are seen, as well as both mitral leaflets. Viewed in this manner, in the normal patient the mitral and tricuspid valves lie in approximately the same horizontal plane in their respective AV grooves. The AV valves appear to meet at the interventricular septum at approximately right angles. In the normal heart, viewed in this manner at end-systole, a straight horizontal line will connect the mitral and tricuspid valves.

Apex View in Ebstein's Anomaly

Using the apex four chamber view, we were able to visualize the thickened and deformed tricuspid valve displaced into the right ventricle. This finding, which we consider to be specific for Ebstein's anomaly, was present in all nine patients studied. Figures 2, 4 and 5...
Illustrate this apical tricuspid valve displacement in patients with moderately severe Ebstein's anomaly. In the presence of severe Ebstein's anomaly the massive right atrium and “atrialized” right ventricle make imaging the entire left ventricle difficult within the arc of an 80° sector scanner, as illustrated in figure 4. With the apex four chamber view, the points of attachment of the anterior and septal tricuspid leaflets were visualized in all patients. In addition, with this view the true AV ring could be consistently imaged. The fibrous nature of the AV ring allows it to be readily imaged and discerned from the surrounding atrium by two-dimensional echocardiography. Figure 2 illustrates the AV ring which is in the same plane as the mitral valve. We were thus able to define by two-dimensional echocardiography the “atrialized” portion of the right ventricle, which is that portion of the right ventricle between the AV groove and the displaced tricuspid valve.

The specificity of this tricuspid valve displacement toward the apex on the four chamber view was assessed by comparing the echocardiograms of the nine patients with Ebstein's anomaly to those of 25 normals and 25 patients with right ventricular volume overload. In the apex four chamber view of the normal heart the distances from the mitral valve to the cardiac apex and from the tricuspid valve to the cardiac apex are approximately equal. The ratio comparing the mitral valve-to-apex distance to the tricuspid valve-to-apex distance in 25 normals and 25 cases of right ventricular overload ranged from 1 to 1.2 (mean 1.09). By contrast, patients with Ebstein’s anomaly demonstrated a mitral-to-apex and tricuspid-to-apex ratio ranging from 1.8 to 3.2. Figure 2 illustrates how this ratio was obtained in patient 6. Because of right heart enlargement, it is often difficult in patients with Ebstein's anomaly to image both ventricles in their entirety on a single frame, and different frames may be required to determine the distance from the AV valve-to-apex in each ventricle.

Assessment of Severity

For help in assessing the morphologic severity of Ebstein’s anomaly, the right heart can be functionally and anatomically divided into the right atrium, true right ventricle and “atrialized” right ventricle. Using the apex four chamber view, we were able to define these three divisions in all nine patients with Ebstein’s anomaly and correlate the findings with available angiographic data in five patients. We defined the functional right ventricle as that portion of the cavity which lies apically or inferiorly to the displaced

**Figure 4.** Apex four chamber view from patient 3. Note how the tricuspid valve is displaced toward the apex of the right ventricle away from the plane of the mitral valve, dividing the right heart into a small true right ventricle, “atrialized” right ventricle and large right atrium. The enlargement of the right heart forces part of the left ventricle from the arc of the sector scan. Abbreviations: ARV = “atrialized” right ventricle; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle; MV = mitral valve; A-VR = atroventricular ring; S = septum; TV = tricuspid valve.

**Figure 5.** Systolic stop-frame apex four chamber view from patient 10. Note the tricuspid valve displaced toward the apex. Arrows indicate air bubbles resulting from peripheral intravenous injection of saline to exclude R → L shunting; bubbles are seen in the right sided chambers, but not the left in this negative study for shunt. Abbreviations: LV = left ventricle; MV = mitral valve; RA = right atrium; RV = right ventricle.
tricuspid valve. The area of "atrialized" right ventricle is bounded, or defined, anteriorly by the displaced tricuspid valve, posteriorly by the true AV ring. The right atrium lies superior to the AV ring. Figures 2 and 4 illustrate these anatomic divisions in patients 8 and 6, as visualized by two-dimensional echocardiography.

The dimension from the AV groove to the point of attachment of the displaced tricuspid valve to the interventricular septum defines the length of the "atrialized" right ventricle. Comparison of this dimension obtained from the two-dimensional echocardiogram with that obtained from similar measurements on the anteroposterior cineangiogram in five patients with Ebstein's anomaly demonstrated good correlation, all values agreeing within 1 cm of one another (r = 0.92).

Table 1 summarizes the two-dimensional echocardiographic and angiographic evaluation of the nine patients with Ebstein's anomaly. The patients are listed according to their age.

**Long Axis View in Ebstein's Anomaly**

In mild expressions of this disorder and in those cases of Ebstein's anomaly associated with right ventricular volume overload, the long axis view can generally define the enlarged anterior tricuspid leaflet, the motion and the chordal attachments of which can also be seen. In our patients the septal tricuspid leaflet was only occasionally seen in this view and the degree of tricuspid valve displacement could not be accurately assessed, as the AV ring was not visualized.

**Short Axis in Ebstein's Anomaly**

With the short axis view of the heart at the level of the aortic valve, portions of the right ventricular outflow tract can be visualized. The large excursion of the sail-like anterior tricuspid leaflet can be seen with this view, but its attachment to the AV groove is difficult to define. Figure 6 illustrates the short axis view at the level of the aortic valve obtained from patient 3. Because the right ventricle can be imaged from the right atrium to the pulmonary artery in this axis, a further assessment of right ventricular size and function can be made. In milder forms of this disorder where there was significant right ventricular volume overload, we found that the right ventricular outflow tract was hyperdynamic. Paradoxical septal motion could be seen below the level of the mitral valve in these cases. In the two cases (patients 1 and 4) of Ebstein's anomaly associated with significant hypoplasia of the right ventricle, the right ventricular outflow tract was narrowed and the pulmonary arteries were small. Contrast echocardiography in these patients showed little flow from the right ventricle and a dominant right-to-left shunt.

Figure 7 is an example of Ebstein's malformation of the left sided AV valve in patient 5 with l-transposition with ventricular inversion, demonstrating apical displacement of the tricuspid valve within the

**Table 1. Two-Dimensional Echocardiographic and Angiographic Data**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Associated heart defects</th>
<th>Mitral-to-apex/Tricuspid-to-apex ratio (Normal: 1-1.2/1)</th>
<th>Size of &quot;atrialized&quot; right ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1 day</td>
<td>ASD with R → L shunting</td>
<td>2.0/1</td>
<td>2.0</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>ASD no R → L shunting</td>
<td>1.9/1</td>
<td>3.5</td>
</tr>
<tr>
<td>3</td>
<td>8</td>
<td>ASD with R → L shunting</td>
<td>2.2/1</td>
<td>3.0</td>
</tr>
<tr>
<td>4</td>
<td>13</td>
<td>Corrected transposition</td>
<td>3.0/1</td>
<td>N.A.</td>
</tr>
<tr>
<td>5</td>
<td>17</td>
<td>ventricular inversion</td>
<td></td>
<td>N.A.</td>
</tr>
<tr>
<td>6</td>
<td>19</td>
<td>ASD no R → L shunting</td>
<td>2.9/1</td>
<td>7.0</td>
</tr>
<tr>
<td>7</td>
<td>23</td>
<td>ASD no R → L shunting</td>
<td>3.2/1</td>
<td>2.0</td>
</tr>
<tr>
<td>8</td>
<td>24</td>
<td>ASD no R → L shunting</td>
<td>2.9/1</td>
<td>6.0</td>
</tr>
<tr>
<td>9</td>
<td>32</td>
<td>ASD no R → L shunting</td>
<td>3.0/1</td>
<td>7.0</td>
</tr>
</tbody>
</table>

Abbreviations: R = right; L = left; ASD = atrial septal defect; N.A. = not adequate.
morphologic right ventricle which is in the inverted position.

Technically adequate M-mode echocardiograms were obtained in all patients. Six M-mode echocardiograms were felt to be consistent with Ebstein's anomaly exhibiting increased anterior tricuspid leaflet excursion and delayed tricuspid closure (M-T interval > 0.03 sec). In three patients the M-T interval was normal and the M-mode echocardiogram suggested only right ventricular enlargement. The size of the true right ventricle was grossly overestimated by the M-mode study in two patients (1 and 6) with severe tricuspid displacement.

Discussion

Ebstein's anomaly represents an extremely variable anatomic, pathologic and physiologic spectrum in which the essential anatomic defect is apical displacement of malformed tricuspid valve tissue into the right ventricle. The adherence in varying positions of the septal and posterior tricuspid valve leaflets to the right ventricular wall and septum results in the formation of the so-called "atrialized" portion of the right ventricle. The clinical severity of Ebstein's anomaly is the result of several factors, most important of which are the degree of tricuspid valve displacement and the resulting size of the "atrialized" right ventricle, the size and function of the true right ventricle, the degree of tricuspid regurgitation and the presence of associated defects, in particular, atrial septal defect causing right-to-left shunting. In order for it to be of most value in the evaluation of Ebstein's anomaly, a noninvasive technique should be able not only to diagnose this entity accurately, but also to evaluate each of these morphologic abnormalities which contribute to the severity of clinical expression. M-mode echocardiographic experience with this disorder has shown several frequently associated findings, most of which relate to the large sail-like anterior tricuspid leaflet. These findings include increased excursion of the anterior tricuspid leaflet, delayed tricuspid closure and slowing of the EF slope. Abnormal septal motion and variable enlargement of the right atrium and ventricle have also been seen with M-mode echocardiography. Unfortunately, only the delayed closure of the tricuspid valve seems to be specific for Ebstein's anomaly with M-mode echocardiography, because the other findings can also be produced by conditions of right ventricular volume overload. Furthermore, in mild forms of Ebstein's anomaly, the septal motion, anterior tricuspid valve EF slope and amplitude of valve motion may all be normal. Thus, while many echocardiographic M-mode features have been described which suggest the diagnosis of Ebstein's anomaly, none is definitive and none provides information about the severity. By contrast, two-dimensional imaging techniques would seem to offer valuable and accurate information which can be used not only to diagnose, but also to assess the severity of this disorder. Matsumoto et al., using a mechanical variety of cross-sectional sector scanning device in sagittal and "horizontal" views, were able to visualize portions of the right ventricle, "atrialized" right ventricle and the abnormal tricuspid valve displacement. Hirschklau et al., employing a multocrystal system and using the conventional sagittal (long axis) and transverse (short axis) views, reported finding "... inferior and leftward tricuspid valve displacement" along with "a pattern of multiple [tricuspid valve] echoes" and a "whipping motion" of the leaflets. They did not specify which leaflet of the tricuspid valve was imaged. In addition, because the short and long axis views do not allow visualization of the AV ring, the boundaries of the "atrialized" right ventricle could not be defined. The short and long axis views permit visualization of the enlarged anterior tricuspid leaflet. Although identification of this leaflet is important in recognizing Ebstein's anomaly, this finding is nonspecific because we have occasionally found that the anterior tricuspid valve will appear to be enlarged in right ventricular overload states.

Tricuspid and Right Ventricular Morphology

It is important to recognize that Ebstein's anomaly involves not only enlargement of the anterior tricuspid leaflet, but also distortion of the entire tricuspid apparatus and right ventricular architecture. The enlarged anterior leaflet is usually visualized with both M-mode echocardiography and two-dimensional long
and short axis views, but the rest of the tricuspid valve and is relationship to the often altered right ventricle is difficult, if not impossible, to appreciate with these views. A major advantage of the apex four chamber view is that it allows for easy visualization of the anterior and septal tricuspid leaflets simultaneously with the right atrium, AV groove and sinus portion of the right ventricle. Thus, two important determinants of the severity of Ebstein’s anomaly, the degree of displacement of the tricuspid apparatus and the size of the “atrialized” right ventricle, can be defined.

Immobility of the tricuspid valve can also contribute to the severity of clinical expression. In Ebstein’s anomaly, the leaflets are frequently redundant and adherent to the right ventricular walls and septum. This thickening and immobility, when present, can be easily appreciated with real-time echocardiography.

The apex view was of value in one patient (patient 8) who underwent tricuspid valve annuloplasty in an attempt to correct severe tricuspid incompetence and reduce the size of the “atrialized” right ventricle. Preoperatively, there is severe tricuspid valve displacement into the right ventricle, resulting in a large “atrialized” portion. With annuloplasty, the large anterior tricuspid leaflet was pulled toward the AV groove to reduce the size of “atrialized” right ventricle and improve valve competence. The postoperative echocardiogram showed the reduction of the “atrialized” right ventricle with the anterior leaflet now positioned more toward the base of the heart.

Right ventricular size can be evaluated with both precordial and apical views. For reference, the right ventricle can be compared to the normally larger left ventricle. This comparison is easiest with the apex view in which the right and left ventricles are visualized side by side. Similarly, right atrial size can be compared to the left atrium with this view.

Right ventricular volume overload secondary to tricuspid incompetence is common in Ebstein’s anomaly, and we were able to appreciate this associated finding in each instance in our series by noting an enlarged true right ventricle. Recently, saline contrast two-dimensional echocardiography has been shown to be of value in detecting and quantifying tricuspid regurgitation. This technique may prove useful in assessing tricuspid regurgitation in Ebstein’s anomaly, although in our preliminary experience the turbulent flow in the “atrialized” right ventricle and the presence of atrial septal defects may make interpretation inconclusive.

Although M-mode echocardiography never failed to detect an enlarged right ventricle, we noted two instances in which it overestimated the size of the true right ventricle. This error is understandable, because in the long axis at the level of the anterior tricuspid leaflet, where M-mode echocardiographic dimensions are often measured, the “atrialized” right ventricle may be included in the ventricular measurement and the true right ventricle may not be accurately defined. In one case of Ebstein’s anomaly associated with hypoplastic right ventricle and apparently normal right ventricular dimensions on M-mode echocardiography, it was only the two-dimensional apex image that permitted accurate definition of the size of the true right ventricle. Because the apex four chamber view allows side-by-side visualization of the right and left ventricles, we find it useful in the qualitative assessment of right ventricular contractility and wall motion analysis, by comparison with the left ventricle.

Associated congenital defects, particularly atrial septal defect with right-to-left shunting, frequently contribute to the clinical severity of Ebstein’s anomaly. Two-dimensional echocardiography has a proven role in the detection of atrial septal defect. We routinely employ the use of saline contrast echocardiography to evaluate right-to-left shunting in patients with Ebstein’s anomaly. In the patients in whom we used this technique, no significant right-to-left shunt was missed (table 1), nor were any false positive results obtained. Perhaps the greatest value of this contrast technique is in the detection of unexpected right-to-left shunting in the acyanotic patient.

The sensitivity of two-dimensional echocardiography cannot be assessed from this series, but as we have been able to correctly diagnose Ebstein’s anomaly in all known cases thus far encountered, it should prove to be reliable. The major limitation would seem to be the operator’s skill necessary to obtain adequate precordial and apex images. Obtaining quality images may be difficult in patients with Ebstein’s anomaly with enlargement of the right heart, and requires a careful examination, with particular attention to patient positioning and proper transducer angulation to ensure imaging the entirety of the right ventricle.

In summary, two-dimensional echocardiography using the apex four chamber view permits the detection of tricuspid valve displacement into the right ventricle, a finding which is diagnostic of Ebstein’s anomaly. Because it allows the assessment of tricuspid valve mobility, right ventricular size and the size of the “atrialized” right ventricle, the morphologic severity of this anomaly can also be appreciated.

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Real-time Cross-sectional Echocardiographic Imaging and Measurement of the Patent Ductus Arteriosus in Infants and Children

DAVID J. SAHN, M.D., AND HUGH D. ALLEN, M.D.

SUMMARY While echocardiography has been used to noninvasively document indirect effects on the heart of left-to-right shunting through a patent ductus arteriosus, no noninvasive technique has been developed to image the duct itself. In this study, 35 sequential studies were performed on 28 patients with a mechanical sector scanner to image the distal pulmonary artery and its bifurcation by scanning along the axis of the right ventricular outflow tract. Cross-sectional imaging, just superior to the take-off of the right pulmonary artery, provided visualization of the patent ductus as a distal continuation of the pulmonary artery connecting to the descending aorta. Ductal visualization by cross-sectional echo was validated by saline echo contrast observations of right-to-left and left-to-right shunting through the duct in 14 patients, by surgical observations in 11, angiographic observations in 13 and autopsy observations in three. Angiographic size of the ductus arteriosus, whether constricted or widely patent, tortuous or straight, was predicted correctly and echo/angiographic correlations for smallest inner ductal dimension were excellent (r = 0.97). This study provides a method for and validates the direct imaging of the ductus arteriosus and suggests that cross-sectional echocardiography can accurately predict ductal contour and quantitative ductal cross-sectional size noninvasively.

PATENT DUCTUS ARTERIOSUS, a common condition in pediatric patients, has recently assumed major importance as a life-threatening disease of small premature infants, who now survive due to advances in neonatology.1-4 While in an older child, the classical physical findings of ductus arteriosus and the minimal risk of surgery have at times obviated the need for compulsory catheterization before ductal ligation, atypical physical findings and the presence of concomitant respiratory disease in premature infants have often created problems of ductal identification, as well as problems in determining which infants might benefit by ductal closure.

Though echocardiography has achieved importance in assessing the hemodynamics of ductal shunting by demonstrating quantitative left atrial enlargement and/or left ventricular enlargement in premature infants, and has been most important when serially applied to the same infant throughout his course,5-8 Nonetheless, many of the M-mode echocardiographic features can be mimicked by sepsis,8 hypoglycemia, hypervolemia or severe anemia in premature infants, and are nonspecific for ductal shunting. Recently, concern has been expressed over the failure to identify infants who have ductal shunting without an audibly detectable murmur.9 In order to aid in the detection and serial followup of these infants, our laboratory recently developed an arterial saline echo contrast technique for identification of left-to-right shunting patent ductus arteriosus.90 Unfortunately, this latter technique requires the presence of an umbilical arterial catheter above the level of the diaphragm.
Two-dimensional echocardiographic assessment of Ebstein's anomaly.
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