Echocardiography in the Diagnosis of Ebstein’s Anomaly*

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This study presents four patients with Ebstein’s malformation of the tricuspid valve, with hemodynamic, phonocardiographic and echocardiographic investigation. The echocardiogram demonstrated right atrial enlargement and an “atrialized” right ventricular area, pronounced anterior movement of the tricuspid valve with low ejection fraction slope values and a greater than normal delayed closure and opening as compared with the mitral valve. Abnormal septal motion was found in all the cases. Echocardiographic studies, when compared with phonocardiographic and catheterization data, show that an early systolic sound occurs when the tricuspid valve reaches the limit of its systolic excursion, after functional closure of the valve. Early diastolic sounds were generally tricuspid opening snaps.

METHODS

Four patients with Ebstein’s anomaly presented at the “La Paz” Hospital in Madrid. The first case was that of an 18-year-old girl with minimal cyanosis and minimal exercise intolerance; the second patient was a 35-year-old man without cyanosis or symptoms; the third patient was a 10-year-old boy who had undergone a Blalock-Taussig anastomosis because of increasing cyanosis and fatigability; the fourth patient was an 11-year-old girl who was asymptomatic. All patients had cardiac catheterization and angiography. The echocardiographic studies were performed with a Unirad 100 machine with a 2 M Hz transducer and repetition rate of 1,000 impulses/sec. All patients were studied in the recumbent position, and the echocardiographic recordings were obtained with the transducer in the fourth or fifth left intercostal space near the sternal edge. The echo from the anterior tricuspid leaflet was obtained with the device in an anteroposterior and slightly medial direction, position 1 (Fig 1). In this position the echoes from the posterior right atrial wall appeared. By tilting the transducer in a lateral position, position 2, an echo from the interventricular septum was obtained and the “atrialized” right ventricle size determined. The interventricular septum was studied at a high level, near the aortic root, at a medium level when mitral valve echo appeared and at a low level when the free wall of the left ventricle was recorded (Fig 2).

Permanent records of these echocardiograms were obtained with a Polaroid camera in M-mode. Simultaneous echocardiographic recording with phonocardiography of either the tricuspid or the mitral valve was obtained by transferring the ultrasonic signal, by means of an analogic gate, to an Elema-Schöneider jet recorder.

RESULTS

Echocardiographic recordings with simultaneous phonocardiograms of mitral and tricuspid movement were obtained in all four patients. In cases 2, 3 and 4 it was possible to record simultaneous tri-

Manuscript received August 26; revision accepted February 26.
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CHEST, 66: 3, SEPTEMBER, 1974
cuspid and mitral valve echoes (Fig 1, 3, 4), but in case 1 separate recording of these echoes was necessary. The mitral valve posterior position coincided with the first component of the first sound in the phonocardiographic recording of the mitral area. The tricuspid echo showed a 4-to-15 msec delay compared to the mitral echo (Fig 5) and its posterior position coincided with the early systolic sound, so-called "sail sound," recorded phonocardiographically at the tricuspid area. Cases 1, 2 and 3 showed a tricuspid opening snap simultaneously with the anterior position of the tricuspid valve echo (Fig 6-7). The tricuspid valve delay versus mitral movement is shown in Table 1, comparing the distance between the Q wave of the electrocardiogram to the anterior and posterior positions of these valves.

Right atrial size varied between 25 and 40 mm, with the "atrialized" right ventricular dimension between 20 and 40 mm. Measurements were made determining the distance between the posterior tricuspid valve position and the right atrial wall echo or the interventricular septum echo. E-F (ejection fraction) slope values, when determined in the tricuspid valve echo, were less than 45 mm. Tricuspid valve amplitudes, obtained in position 1 of the transducer, were between 22 and 40 mm. The mitral valve E-F slope was less than the normal value in all cases except in case 1 (Table 2).

Motion of the interventricular septum, as determined at different levels, was paradoxic, with anterior movement during systole and posterior movement during diastole in all cases.

Comparative studies between right ventricular and right atrial pressures with the tricuspid echo were obtained by comparing cardiac cycles of the same length (Fig 8). The point at which right ventricular pressure rose above right atrial pressure, the beginning of systole, was found to be identical with the fast backward movement of the tricuspid valve and prior to its posterior position. The maximal posterior position of the valve occurred when the initial slowly rising ventricular pressure wave changed to a more abrupt rise. The posterior position of the tricuspid valve coincided in simultaneous phonocardiographic recordings with the "sail-sound" and was also simultaneous with the "C" wave seen in the "atrialized" pressure curve portion of the right ventricle.

**DISCUSSION**

Clinical diagnostic findings and followup of Ebstein's anomaly have been well established in vari-
ous communications. Because of the previously reported high incidence of cardiac arrhythmias during catheterization and angiography, an accurate diagnosis by a noninvasive technique would be useful. In order to obtain exact timing of the early systolic sound (a typical feature in Ebstein's anomaly, previously attributed to tricuspid closure or to pulmonary ejection) echocardiographic and phonocardiographic findings were compared with catheterization data. The posterior position of the tricuspid valve echo coincided with the early systolic sound and with the point of change in pressure development in the right ventricular pulse pressure. The early systolic sound occurred after the tricuspid valve had closed, as determined by the coincidence of atrial and ventricular pulse pressure waves (Fig 8). With the initiation of contraction and pressure rise in the right ventricle, tricuspid valve leaflets are pushed posteriorly toward the atrium. It is at this posterior limit of excursion that the systolic sound occurs. Right ventricular pressure shows an abrupt rise in pressure development at this moment. These two phases in pressure development can also be seen in examining the contour of the pulmonary artery pulse pressure.

Tricuspid valve closure versus mitral valve closure is delayed by 4-15 msec, exceeding the difference found in normal subjects. This finding might be the most specific echo datum in Ebstein's anomaly since it is not in relationship to the right bundle branch block. Tricuspid valve opening is delayed when compared to the mitral opening; this movement coincides in cases 1 and 3, with a diastolic
sound that would correspond to a tricuspid opening snap. Case 2 shows a tricuspid opening snap in cardiac cycles preceded by a shorter diastolic filling period (Fig 7). In these three patients the E-F tricuspid slope is less than 20 mm, indicating that slow posterior movement is present in diastole. At heart catheterization no significant tricuspid stenosis was demonstrated in any case.

The amplitude of movement of the anterior tricuspid leaflet echo has not been found to be of value when compared to the degree of functional cardiac impairment.

When the transducer is pointed to the tricuspid area, position 1, the posterior right atrium measure-

Table 1—Tricuspid Valve Opening Delay vs Mitral Valve Opening

<table>
<thead>
<tr>
<th>Case, No.</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid valve</td>
<td>Q-PM*</td>
<td>0.15</td>
<td>0.17</td>
<td>0.24</td>
</tr>
<tr>
<td></td>
<td>Q-AM**</td>
<td>0.42</td>
<td>0.54</td>
<td>0.55</td>
</tr>
<tr>
<td>Mitral valve</td>
<td>Q-PM*</td>
<td>0.11</td>
<td>0.09</td>
<td>0.09</td>
</tr>
<tr>
<td></td>
<td>Q-AM**</td>
<td>0.36</td>
<td>0.48</td>
<td>0.46</td>
</tr>
</tbody>
</table>

*Distance between Q wave of ECG and posterior position of valve.
**Distance between Q wave and anterior position of valves.

All cases show paradoxic septal motion. Right ventricular conduction abnormalities do not seem to be the explanation for this paradoxic movement.12 Probably this abnormal motion is produced by the inefficient right ventricular contraction and by the presence of some degree of tricuspid regurgitation.

Mitral valve echo E-F slope is less than normal valve in three cases. Abnormal mitral anterior leaflet valvular motion was described in patients with pul-

Table 2—Measurements Obtained with Transducer

<table>
<thead>
<tr>
<th>Case, No.</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrialized right ventricle (mm)</td>
<td>25</td>
<td>20</td>
<td>40</td>
<td>25</td>
</tr>
<tr>
<td>Right atrium (mm)</td>
<td>25</td>
<td>30</td>
<td>40</td>
<td>35</td>
</tr>
<tr>
<td>Tricuspid valve E-F slope (mm/s)</td>
<td>15</td>
<td>45</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid valve Amplitude (mm)</td>
<td>25</td>
<td>40</td>
<td>24</td>
<td>22</td>
</tr>
<tr>
<td>Mitral valve E-F slope (mm/s)</td>
<td>90</td>
<td>45</td>
<td>35</td>
<td>50</td>
</tr>
</tbody>
</table>

FIGURE 7. Phonocardiogram in tricuspid area (PCG TA) with tricuspid echogram in case 2. A, tricuspid opening snap (TOS) appears in cardiac cycles preceded by shorter diastolic filling period.

FIGURE 8. Comparative study in case 2 between tricuspid echogram, right ventricle pressure pulse (RV) and right atrial pressure pulse (RA) by choosing cardiac cycles of same length. Point at which right ventricular pressure rises over right atrial pressure (T,) was in relationship to downward movement of tricuspid valve; posterior position of valve occurs at point at which rate in ventricular pressure development changes (SS) immediately after "c" wave of right atrial pressure pulse.
monary hypertension. In Ebstein’s anomaly there is no right ventricular pressure overload. This deviation from the normal values could be due to the thickening of the mitral valve described in patients with Ebstein’s anomaly of the tricuspid valve.

Right atrial enlargement and “atrialized” right ventricular area are described by means of echocardiography, in Ebstein’s anomaly. A greater than normal anterior movement of the tricuspid valve with low E-F values and important delay of closure and opening as compared to the mitral valve are constant features in these patients. In addition, abnormal septal motion is found. Echocardiographic studies, when compared with phonocardiographic and hemodynamic data, have also shown that the early systolic sound occurs when the large and abnormal tricuspid valve reaches the limit of its systolic excursion and after functional closure of the valve. An early diastolic sound, generally called the “tricuspid opening snap,” was found to coincide with the anterior position of the tricuspid valve echo. Further anatomopathologic studies are needed to investigate the fact that the mitral valvular echo was abnormal in some patients with Ebstein’s anomaly of the tricuspid valve.

ACKNOWLEDGMENTS: We acknowledge the assistance of Dr. Fishleder from the Instituto Nacional de Cardiología de Mexico and Dr. Galimoto from The Texas Heart Institute, Houston, Texas.

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Richard Strauss the Composer (1864-1949)

Following Wagner, Richard Strauss embraced a method of composing with recurrent thematic motives and with an opulent orchestral texture which often stand independently on its own. But Strauss made his operas faster and more concentrated in action. Salome and Elektra (1909) each play in one continuous scene. Der Rosenkavalier, a realistic comedy, takes three long acts but crams them full of detail: and in place of Wagner’s characteristically slow paced delivery we have fast dialogue—in music. Moreover, Strauss—not for nothing a contemporary of Zola, Ibsen and Wilde—made deliberate use of plots intended to deliver a shock in the theater. They duly shocked and succeeded.