Echocardiography in the Diagnosis of Ebstein's Anomaly of the Tricuspid Valve

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SUMMARY

Nineteen patients aged between 4 days and 40 years with Ebstein's anomaly of the tricuspid valve have been examined with echocardiography. The diagnosis was verified with angiocardiography in all but one patient. The results at echocardiography are compared with those found in some other patients where an echo from the anterior tricuspid leaflet could be obtained (atrial septal defect, total anomalous pulmonary venous return, pulmonary hypertension, and one patient with congenital tricuspid stenosis).

An echo from the anterior tricuspid leaflet could be obtained in all patients with Ebstein's anomaly. This echo had an abnormal pattern of movement with an abnormally anterior position during the entire diastole. A late tricuspid opening was found in most patients. A late tricuspid closure compared with the time of mitral closure was a constant finding. It is suggested that this late tricuspid closure is mainly caused by mechanical factors related to the abnormal, large anterior tricuspid leaflet. The abnormal pattern of movement of the echo from the anterior tricuspid leaflet with a late tricuspid closure has not been found in any other patient examined.

Additional Indexing Words: Congenital heart defects, diagnosis Congenital tricuspid stenosis Ultrasonics, diagnostic use

THE DIAGNOSIS of Ebstein's anomaly of the tricuspid valve is, in typical cases, easily suspected on the physical findings, together with the findings at electrocardiography and roentgen examination. In mild forms and especially in infants, the diagnosis may, however, be difficult without additional investigations such as angiocardiography.

This malformation shows great variations both in pathologic anatomy and clinical manifestations.

The possibility of using echocardiography for the study of the movement of the anterior tricuspid leaflet has been realized for many years but it has been used mainly in the diagnosis of acquired tricuspid stenosis. In 1969 it was first shown that echocardiography could be used in the diagnosis of Ebstein's anomaly. It was later suggested that the abnormal movement of the anterior tricuspid leaflet observed in patients with this malformation could explain some of the auscultatory findings in these patients. These observations have been confirmed by other authors.

The aim of the present investigation was to extend the use of echocardiography in the diagnosis of Ebstein's anomaly to a larger number of patients of different ages. In this way the use of echocardiography in the diagnosis of this malformation could be evaluated.

Materials and Methods

Ebstein's Anomaly of the Tricuspid Valve

All patients observed since 1967 with a diagnosis of Ebstein's anomaly of the tricuspid valve were examined with echocardiography (table 1, cases 1-10). In addition, nine other patients with this anomaly from two other hospitals had echocardiographic examinations performed on them by the author on one occasion (cases 11-19).

The diagnosis was confirmed by heart catheterization and angiocardiography in all cases except one (case 10). The diagnosis in this patient, originally made at another hospital, was based on quite typical physical findings, and typical findings on the electrocardiogram and at roentgen examination of the heart. None of the patients has been operated upon, and all except one are still alive. Except for an atrial septal defect, associated cardiovascular malformations were not encountered in any of the cases. Some clinical data including functional classification are given in table 1. In the three youngest patients (cases 1, 2, and 3) a functional...
Table 1
Survey of Material and some Electrocardiographic Data

<table>
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<tr>
<th>Case, sex</th>
<th>Age at exam</th>
<th>Cyanosis</th>
<th>Functional class</th>
<th>ECG duration or QRS (sec)</th>
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<td>1 F</td>
<td>4 days</td>
<td>+</td>
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<td>I</td>
<td>0.14</td>
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<td>0.08</td>
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<td>14 years</td>
<td>+</td>
<td>II</td>
<td>0.10</td>
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<td>9 F</td>
<td>17 years</td>
<td>—</td>
<td>I</td>
<td>0.11</td>
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<td>0.09</td>
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<tr>
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<td>40 years</td>
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classification has not been made. These three patients had signs of congestive heart failure and cyanosis during the first weeks of life. Since them, the condition of two of them (cases 1 and 2) has improved considerably, and now aged 6 months and 1 year, respectively, they are almost symptom-free. The third infant (case 3) recently died suddenly at home. The diagnosis was confirmed at autopsy. The anterior tricuspid leaflet consisted of a large "sail-like" structure, and the posterior and septal leaflets were rudimentary.

The electrocardiogram showed signs of right bundle-branch block of various degrees in all cases except two (cases 2 and 3). In one patient a type B ventricular preexcitation was persistently found (case 2). The durations of the QRS complexes are given in table 1.

Patients with Conditions other than Ebstein's Anomaly
Some other patients examined with echocardiography of the tricuspid valve are included in this study for comparison.

One group comprised patients without pulmonary artery hypertension (pressure in the pulmonary artery below 40 mm Hg systolic). This group included: three patients with complete right bundle-branch block and normal hemodynamic findings; 16 patients with atrial septal defects of secundum type; and three patients with total anomalous pulmonary venous return. The age of these patients ranged from 3 months to 16 years. This group also included a 16-year-old patient with congenital tricuspid stenosis. She had a rather severe degree of tricuspid stenosis and a small hypoplastic valve as determined by heart catheterization, angiography, and at operation.

The second group consisted of five patients, aged 3% to 16 years, with severe pulmonary hypertension. Two of these patients had primary hypertension and three patients a ventricular septal defect with small mixed shunts.

Echocardiographic Examinations
These examinations were performed with the technic described in a previous paper.16 A commercially available ultrasonoscope (Smith-Kline Eskoline 20), with a repetition rate of 1000 pulses/sec, and a 2.25-MHz transducer of 1.9-cm (0.75-in) diameter were used. The registrations were made on Polaroid film from the ultrasonoscope screen.

The patients were examined in supine position during normal respiration and without premedication. A water-soluble gel was used to obtain airless contact between the transducer and the skin.

An echo from the anterior tricuspid leaflet can be obtained in most infants and children. In individuals without dilatation of the right ventricle the echo is, however, most often fragmentary. Two different transducer positions have been suggested for obtaining an echo from the anterior tricuspid leaflet.16,20 In the first (used in this study) the transducer is placed in the fourth left intercostal space at the sternal border, and the transducer is angulated 30° of 45° in the frontal plane. The second transducer position is in the third left intercostal space at the sternal border, and the transducer is angulated in the medial and caudal direction.

An echo from the anterior tricuspid leaflet is obtained more easily in patients with dilatation of the right ventricle. The echo is then often complete and can be observed with the transducer in anteroposterior direction or angulated only slightly in the frontal plane.

In the patients with Ebstein's anomaly the echo from the anterior tricuspid leaflet was obtained with the transducer in the fourth left intercostal space, 1–4 cm from the left sternal border and in the anteroposterior direction or angulated slightly (15°) in the frontal plane. In some of these patients the echo from the anterior tricuspid leaflet could be obtained with the transducer placed even in the third left intercostal space and sometimes as far laterally as the left midclavicular line.

An echo from the interventricular septum is often obtained posterior to the echo from the anterior tricuspid leaflet. A registration of an echo from the interventricular septum can at least be achieved if the transducer is angulated slightly in the frontal plane. An echo from the posterior or septal tricuspid leaflet is rarely recorded.

The echo from the anterior tricuspid leaflet is characterized by a slow anterior movement during ventricular systole (fig. 1A). At the beginning of diastole a rapid anterior opening movement occurs. As the fully open position is reached the echo immediately moves in a posterior direction toward a semiopen position. The atrial systole causes a new opening movement in the anterior direction, and this is then interrupted by a posterior movement toward the fully closed position at the beginning of ventricular systole.

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Registations of an echocardiogram from the anterior mitral leaflet were obtained in all patients with the technic described earlier. A simultaneous recording of the echo from the anterior tricuspid and the anterior mitral leaflets was made in 12 of the cases with Ebstein's anomaly.

Measurements of time intervals from the Q wave in a simultaneously recorded electrocardiogram to the time of opening and closure of the anterior tricuspid (fig. 1B) and the anterior mitral leaflets were made on all patients. Measurements of these time intervals in the individual case were made on heart cycles of equal lengths. In the 12 cases with Ebstein's anomaly where a simultaneous recording of the echo from the anterior tricuspid and mitral leaflets were obtained, the time interval between mitral and tricuspid closure was measured on these recordings.

Results

Ebstein's Anomaly of the Tricuspid Valve
Echocardiographic Findings

Anterior Tricuspid Leaflet. An echo supposed to come from the anterior tricuspid leaflet was obtained in every case. It has previously been shown that in subjects without malformation of the tricuspid valve an echo obtained in the region anterior to the echo from the interventricular septum, and with a rapid anterior movement in the beginning of diastole, originates from the anterior tricuspid leaflet. In order to verify that the echo supposed to come from the anterior tricuspid leaflet in the cases reported here really did originate

from the tricuspid valve, one patient (case 1) was examined during heart catheterization with the contrast echo method described by Gramiak et al. Indocyanine green, which gives dense intracardiac ultrasound echoes when injected rapidly, was injected into the right atrium (fig. 2). Dense echoes were first seen posterior to the echo, supposed to originate from the anterior tricuspid leaflet. At the next diastole the dense echoes appeared anterior to this echo. During the following heart cycles the density of the echoes diminished in the posterior and increased in the anterior region. These findings agree with earlier observations and confirm that the echo described did originate from the tricuspid valve. The rapid anterior movement in the beginning of diastole identifies the echo-giving structure as the anterior tricuspid leaflet.

The echo from the anterior tricuspid leaflet in the patients with Ebstein's anomaly differed mainly from that obtained from a normal tricuspid leaflet in its pattern of movement during diastole (fig. 1B). In the beginning of diastole a rapid anterior opening movement occurred toward a fully open position. During the entire diastole the echo remained in an abnormally anterior position with only a slow posterior movement (fig. 1B, white arrow). After the QRS complex in the electrocardiogram a rapid posterior movement toward a fully closed position was noted. Due to the abnormally
anterior position of the echo during the entire diastole the atrial systole caused only a small further opening movement (1–3 mm). During diastole the echo from the anterior tricuspid leaflet was close to the echo from the anterior heart wall. In five patients (cases 13, 14, 17–19) it was only possible to show that the echo was in an abnormally anterior position during the entire diastole, but not to study it in detail. All these patients belonged to the category examined with echocardiography on only one occasion.

The results of measurements of the total amplitude of movement, the amplitude of opening movement in diastole, and the speed of posterior movement in the early part of diastole (fig. 1B, white arrow) are given in table 2. In four patients (cases 1, 3, 4, and 11) the heart frequency was too high to permit a measurement of the speed of posterior movement in the early part of diastole, before atrial systole. Measurements of the time of complete tricuspid opening (fig. 1B, T_o) and the time of complete tricuspid closure (fig. 1B, T_c) related to the Q wave in a simultaneously recorded electrocardiogram were made, and the results are presented in table 2.

Interventricular Septum. An echo from the interventricular septum was found in all cases. Diamond et al. have found that the echo from the interventricular septum in normal subjects moves in a posterior direction during ventricular systole (type N). In cases with volume overload of the right ventricle they found that the movement of the septal echo during ventricular systole was parallel to the movement of the echo from the posterior heart wall (type A) or that the septal echo showed hardly any movement at all during ventricular systole (type B). In the patients described here a normal movement of the echo from the interventricular septum was observed in only one patient (case 6). A type A movement was found in five patients (case 7, 11, 12, 15, and 19). The other patients showed the pattern of movement described as type B (fig. 3). The patient with a normal movement of the septal echo had a tricuspid regurgitation like the other patients with Ebstein's anomaly.

Anterior Mitral Leaflet. An echo from the anterior mitral leaflet was obtained in all cases. The amplitude of movement was normal or slightly below normal for age. The speed of posterior movement in the early part of diastole was normal (more than 90 mm/sec) or slightly below normal (70–90 mm/sec) in most patients. In two patients (cases 14 and 18) the speed of movement was.

Figure 2

Echocardiogram of the anterior tricuspid leaflet in a patient with Ebstein's anomaly (case 1) recorded during heart catheterization. The arrow points to the time of injection of indocyanine green into the right atrium. For further explanation see text.
Table 2

Ebstein's Anomaly of the Tricuspid Valve: Echocardiographic Findings

<table>
<thead>
<tr>
<th>Case</th>
<th>Echo from anterior tricuspid leaflet</th>
<th>Cycle length and time intervals from Q in simultaneously recorded ECGs (sec)</th>
<th>Time intervals between movements recorded simultaneously (sec)</th>
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<td>Total ampl (mm)</td>
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<td>NM</td>
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Abbreviations: Mc = mitral closure; Mo = mitral opening; Tc = tricuspid closure; To = tricuspid opening; NM = not measurable.

Figure 3

Registration of simultaneously recorded echoes from the anterior tricuspid leaflet, the interventricular septum, and the anterior mitral leaflet in two patients with Ebstein's anomaly of the tricuspid valve. (A) A 2-week-old girl (case 3). (B) A 6-year-old boy (case 6). The time of mitral (Mc) and tricuspid (Tc) closure is indicated by arrows.

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however, reduced to 46 mm/sec and 51 mm/sec, respectively.

Comparison between Echoes from the Anterior Tricuspid and Anterior Mitral Leaflets

A comparison between the amplitude of movement of the two echoes in the same patient showed a great variability. In 12 patients the amplitude of movement was greater for the echo from the tricuspid leaflet (1-12 mm); in five patients they were of equal magnitude; and in two patients the echo from the mitral leaflet was greater in amplitude of movement (2-4 mm).

The relation between the time of complete opening of the anterior tricuspid and the anterior mitral leaflet was measured on the echocardiogram, and the results are given in table 2. The same comparison was made for tricuspid and mitral closure, and the results are also given in table 2. Mitral opening was simultaneous with tricuspid opening in six patients, and occurred before tricuspid opening in 13 patients. The tricuspid closure always occurred after mitral closure (0.06 sec or more). In 12 patients it was possible to make these measurements on simultaneously recorded echoes from the anterior tricuspid and mitral leaflets (fig. 3, table 2). The measurements of these time relations on simultaneous and nonsimultaneous recordings of echocardiograms from tricuspid and mitral leaflets agree quite well (difference not more than 0.02 sec.).

Conditions other than Ebstein's Anomaly

Echocardiographic Findings

In all patients except the one with tricuspid stenosis, an echo from the anterior tricuspid leaflet could be obtained with the transducer in the fourth left intercostal space, close to the left sternal border or sometimes 1-2 cm more to the left. The transducer was positioned in the anteroposterior direction or up to 30° in the medial direction. It was possible to get a simultaneous recording of an echo from the anterior tricuspid and the anterior mitral leaflet in four patients.

In patients with complete right bundle-branch block, atrial septal defect, and total anomalous pulmonary venous return, the echo from the anterior tricuspid leaflet had a normal pattern of movement. The speed of movement in the posterior direction during the early part of diastole was in all cases above 75 mm/sec (range 75-146 mm/sec). Three of the patients with pulmonary hypertension also showed a normal pattern of movement and a speed of movement above 75 mm/sec. Two patients with pulmonary hypertension showed, however, a slow speed of movement of the echo from the anterior tricuspid leaflet during diastole (24 and 35 mm/sec, respectively).

An echo from the anterior tricuspid leaflet was very difficult to obtain in the patient with congenital tricuspid stenosis. An echo supposed to originate from the anterior tricuspid leaflet was observed with the transducer in the fourth right intercostal space close to the sternal border and angulated slightly in the cranial direction. This echo had a small amplitude of movement (15 mm) with its main movement during ventricular systole and only a small anterior movement (2-3 mm) in the beginning of diastole, suggesting that it originated from an atrioventricular valve. Details about its further movement during diastole could not be obtained. This patient has a deformation of the thoracic cage making the echocardiographic examination difficult to perform.

Comparison between Echocardiographic Findings in Patients with Ebstein's Anomaly and the Other Patients Examined

Five patients with Ebstein's anomaly were compared with five patients with atrial septal defects. The children in each pair were of about the same age and of roughly the same height and weight. The total amplitude of movement and the amplitude of opening movement of the echo from the anterior tricuspid leaflet were greater in the patient with Ebstein's anomaly in one of the pairs (9 mm), of equal magnitude in one pair, and had a greater amplitude in the patients with atrial septal defect in three of the pairs (3-7 mm).

The time interval between mitral closure and tricuspid closure related to the duration of the QRS complex in the electrocardiogram was compared in all patients, except the one with congenital tricuspid stenosis (fig. 4). The patient with congenital tricuspid stenosis was omitted because it was impossible to define exactly the time of tricuspid closure on the echocardiogram. It can be seen in figure 4 that the time interval between mitral and tricuspid closure is greater in the patients with Ebstein's anomaly than in the other patients examined. Within the group of patients with Ebstein's anomaly, the values were rather varied. This cannot be explained by the severity of the cardiac lesion based upon the functional classification indicated in table 1. This discrepancy will be discussed below.
ECHOCARDIOGRAPHY IN EBSTEIN'S ANOMALY

Discussion

The possibility of obtaining an ultrasound echo from the tricuspid valve more to the left on the precordium in patients with Ebstein's anomaly than in other patients is not surprising considering the well-known downward displacement of the tricuspid valve in this anomaly. It is perhaps more astonishing that an echo was obtained in all the patients examined considering the great variability in the malformation of the tricuspid valve in this anomaly. It has, however, been pointed out by many authors\(^1\)–\(^5\) that one of the most constant findings in Ebstein's anomaly is that the anterior tricuspid leaflet is the largest and best preserved. This has been most extensively described by Pechstein.\(^1\) His study on the configuration of the tricuspid valve tissue in 48 cases of Ebstein's anomaly reveals that in 46 cases the anterior tricuspid leaflet was larger than normal. Only in two patients was the entire valve tissue extremely underdeveloped as well as the outflow tract of the right ventricle, indicating a severely disturbed function of the right ventricle. A large “sail-like” anterior tricuspid leaflet was found in the only patient in the present material on whom an autopsy was performed. Since in echocardiography an echo is most easily obtained from the anterior tricuspid leaflet, the anatomic findings in Ebstein's anomaly referred to above can explain why such an echo was observed in all the patients examined in this study. A few severely disabled patients with Ebstein's anomaly will presumably be encountered in the future where it will be difficult to obtain any echo at all from the anterior tricuspid leaflet.

The amplitude of movement of the echo from the anterior tricuspid leaflet has not been found to be of special value in establishing the diagnosis in the present material. A severely reduced amplitude of movement can, however, be assumed to be found in a few patients with this anomaly, though it was not encountered in any of the patients examined here.

The pattern of movement of the echo from the anterior tricuspid leaflet has been found to be
clearly abnormal. The abnormal anterior position of this echo during the entire diastole resembles the pattern of movement described in adult patients with acquired tricuspid stenosis.\textsuperscript{12, 13} The observed slow closing movement in diastole in these patients has been explained by the tricuspid stenosis. In the present material the findings at heart catheterization did not indicate a significant tricuspid stenosis in any of the cases. The same pattern of movement of the echo from the anterior tricuspid leaflet has been found in a case with severe pulmonary valvular stenosis\textsuperscript{20} and in two patients in the present material with severe pulmonary hypertension. It can in these cases be explained by a reduced compliance of the right ventricle, reducing the speed of diastolic filling of the right ventricle and thereby the tricuspid leaflet movement. The reason for the abnormal pattern of movement of the echo from the anterior tricuspid leaflet in patients with Ebstein’s anomaly could also be a reduced compliance of the right ventricle due to the small size of the normally functioning part of the right ventricle, or to some extent be explained by mechanical factors related to the abnormal, large anterior tricuspid leaflet. The findings in the present material in a case of congenital tricuspid stenosis were entirely different from those seen in Ebstein’s anomaly or acquired tricuspid stenosis. The severely reduced amplitude of movement of the echo from the anterior tricuspid leaflet in this case could have been caused by the severely hypoplastic funnel-shaped tricuspid valve found at operation.

In most patients with Ebstein’s anomaly, the tricuspid opening was found to occur later than the mitral opening. Normally the tricuspid valve opening occurs shortly before or sometimes soon after the mitral valve opening.\textsuperscript{24} There were, however, great differences between different patients with Ebstein’s anomaly, and the diagnostic significance of this observation is therefore limited. The tricuspid closure did occur later than the mitral closure (0.06 sec or more) in all patients with Ebstein’s anomaly. This exceeds the difference found in normal subjects (less than 0.03 sec).\textsuperscript{24, 25} A comparison between the time interval between mitral and tricuspid closure related to the duration of the QRS complex (Fig. 4) in different subjects shows that the asynchrony between mitral and tricuspid closure for any given QRS duration is greater for the patients with Ebstein’s anomaly than for any other patients studied. The late tricuspid closure in the patients with Ebstein’s anomaly can therefore not at all, or only to a small part, be explained by the right bundle-branch block present in these patients. The finding of a delayed tricuspid closure even in the patient with type B ventricular preexcitation (case 2), where the early depolarization is supposed to be in the right ventricle,\textsuperscript{26} also suggests that some factor other than a delayed depolarization of the right ventricle can be responsible for the delayed tricuspid closure. Presumably a mechanical factor directly related to the malformed tricuspid valve with its large anterior leaflet is therefore mainly responsible for the delayed tricuspid closure.

In the published examples of abnormal patterns of movement of the echo from the anterior tricuspid leaflet in acquired tricuspid stenosis\textsuperscript{12, 13} or severe pulmonary stenosis,\textsuperscript{20} the time of tricuspid closure has not been found to be severely delayed. The finding of an echo from the anterior tricuspid leaflet with an abnormal pattern of movement of the type described in this material and with a delayed tricuspid closure seems thus to be an echocardiographic finding presumably specific for Ebstein’s anomaly. In may also be mentioned that in four of the patients in this material (cases 1, 2, 3, and 9) the diagnosis was first suggested by the echocardiographic findings, before any further investigations were made. It has, however, not been found possible to evaluate the degree of functional impairment of the tricuspid valve with this technic.

It is obvious that factors other than the late tricuspid closure are mainly responsible for the functional cardiac impairment in these patients. Two such factors are apparent, the tricuspid regurgitation and impaired function of the right ventricle because a large part of it is atrialized.

The echocardiographic findings from the interventricular septum have shown that, in most cases with Ebstein’s anomaly, the interventricular septum is not moving normally,\textsuperscript{22} but this fact has not contributed to the diagnosis.

The echocardiographic findings from the anterior mitral leaflet are, in most cases, normal or show a slight deviation from the normal. This small deviation can be explained by an abnormal positioning of the anterior mitral leaflet in relation to the external thoracic wall caused by the considerable enlargement of the right atrium. In two patients the speed of movement of the echo from the anterior mitral leaflet was clearly reduced but not to the extent seen in patients with mitral stenosis with hemodynamic consequences.\textsuperscript{27} The reason for this is not clear. It could, however, be due to pathologic-anatomic changes of the mitral
valve of the kind described by Lev et al.\textsuperscript{2} in patients with Ebstein’s anomaly of the tricuspid valve. They stated that the mitral valve is often greatly thickened, and thought that this could be the result of a hemodynamic alteration caused by the deviation of the septum to the left.

Among the auscultatory findings in Ebstein's anomaly systolic and diastolic sounds have been described by most authors.\textsuperscript{3,5-7,9,10} It has been suggested that the systolic sound is related to the closure of the large anterior tricuspid leaflet\textsuperscript{28} and that this could be the most specific auscultatory event in Ebstein's anomaly. The findings in the material presented here of a late tricuspid closure prompted a comparison between echocardiography and phonocardiography in Ebstein's anomaly. The results of this comparison will be published separately but it is evident that there is a very close time relation between the systolic sound and tricuspid closure.

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Echocardiography in the Diagnosis of Ebstein's Anomaly of the Tricuspid Valve
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