Clinical Study on the Abnormal Flow Patterns in Ebstein's Anomaly Using Pulsed Doppler Echocardiography

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SUMMARY

Pulsed Doppler echocardiography (PDE) was performed in 10 patients with Ebstein's anomaly and 10 cases of tricuspid regurgitation secondary to mitral stenosis.

Distal atrialized right ventricle (ATRV): In all patients with Ebstein's anomaly, tricuspid regurgitant flow was recognized by PDE. In this lesion with moderate tricuspid regurgitation, a widely dispersed dot pattern was recorded during systole. However, in the cases with severe tricuspid regurgitation a relatively smooth dot pattern was recognized. In the cases with marked delay in pressure rise in the right ventricle, PDE showed a bimodal regurgitant flow pattern. The interval between the onset of QRS and that of tricuspid regurgitant flow with right ventricular pressure rise was measured. The interval corrected for heart rate ranged from 0.10 to 0.35 with an average of 0.19±0.08 sec. In the subjects with secondary tricuspid regurgitation, it ranged from 0.07 to 0.11 sec. This interval was significantly prolonged in Ebstein's anomaly as compared to that in secondary tricuspid regurgitation (p<0.001).

Proximal ATRV: Tricuspid regurgitant flow was detected in 6 of 10 patients with Ebstein's anomaly. The disturbed flow was less apparent in the proximal ATRV than in the distal ATRV.

Additional Indexing Words:
Tricuspid regurgitation  Atrialized right ventricle  Functional right ventricle  Time interval histogram

EBSTEIN'S anomaly of the tricuspid valve is one of the rarer congenital malformations. The variation of its clinical manifestations has drawn attention. The basic anomaly is a downward displacement of the tri-
This malformation shows great variations in pathologic anatomy and hemodynamic states. The hemodynamic alteration of this anomaly has been studied by angiocardiography. Recently, a range-gated pulsed Doppler has been combined with M-mode echocardiography to provide intracardiac flow velocity information in a known area of the heart and great vessels. Pulsed Doppler echocardiography (PDE) allows noninvasive determination of the direction of blood flow at the position of sampling volume (SV). The characteristics of blood flow within the SV are also recognized; a smooth flow makes a uniform Doppler shift and the time interval histogram is recorded as a coherent dot pattern, whereas a disturbed flow produces a non-uniform Doppler shift and is represented as a scattered dot pattern.

The purpose of this paper is to study the flow pattern of Ebstein's anomaly especially in the atrialized and functional right ventricles, in comparison with that of tricuspid regurgitation secondary to mitral stenosis.

**MATERIALS AND METHODS**

Pulsed Doppler echocardiography was performed in 10 patients with Ebstein's anomaly, isolated or associated with other heart diseases as shown in Table I.

**Table I. Case Materials and PDE Findings of Ebstein’s Anomaly**

<table>
<thead>
<tr>
<th>Patients</th>
<th>Sex</th>
<th>Age</th>
<th>Diagnosis</th>
<th>Sampling sites</th>
<th>Q-R/√R-R</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Distal ATRV</td>
<td>Proximal ATRV</td>
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<tr>
<td>1. F. A.</td>
<td>M</td>
<td>7</td>
<td>Ebstein</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>2. H. S.</td>
<td>M</td>
<td>35</td>
<td>Ebstein</td>
<td>+</td>
<td>N.D.</td>
</tr>
<tr>
<td>3. A. O.</td>
<td>F</td>
<td>21</td>
<td>Ebstein + WPW</td>
<td>+</td>
<td>N.D.</td>
</tr>
<tr>
<td>4. T. I.</td>
<td>M</td>
<td>29</td>
<td>Ebstein</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>5. R. W.</td>
<td>F</td>
<td>43</td>
<td>Ebstein</td>
<td>+</td>
<td>N.D.</td>
</tr>
<tr>
<td>6. M. Y.</td>
<td>F</td>
<td>19</td>
<td>Ebstein + ASD</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>7. M. T.</td>
<td>F</td>
<td>7</td>
<td>Ebstein + ASD</td>
<td>+</td>
<td>N.D.</td>
</tr>
<tr>
<td>8. K. M.</td>
<td>F</td>
<td>4</td>
<td>Ebstein</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>9. S. Y.</td>
<td>F</td>
<td>30</td>
<td>Ebstein + ASD + VSD</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>10. Y. N.</td>
<td>F</td>
<td>3</td>
<td>Ebstein</td>
<td>+</td>
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</table>

Tricuspid regurgitant flow in the distal ATRV was detected in 10 patients with Ebstein's anomaly. However, in the proximal ATRV, it was recognized in 6 out of 10 patients. The interval between the onset of QRS and that of the regurgitant flow was measured and corrected for R-R interval. Abbreviations: M=male; F=female; ASD=atrial septal defect; VSD=ventricular septal defect; W-P-W=Wolff-Parkinson-White preexcitation syndrome; ATRV=atrialized right ventricle; Q-R=the interval between the onset of QRS and that of regurgitant flow; N.D.=not detected.
They ranged in age from 3 to 43 years. Three were male and the remainders were female. Cardiac catheterization and angiocardiography were performed in all the patients, and the diagnosis of Ebstein's anomaly was based on the demonstration of a displaced tricuspid valve by right atrial or right ventricular angiocardiography. Two patients had undergone cardiac surgery, and 2 of 10 patients showed slight to moderate cyanosis. As the control, 10 patients showing tricuspid regurgitation associated with mitral stenosis were similarly examined.

Prior to pulsed Doppler study, cross-sectional echocardiography was carried out to decide the beam direction and the transducer position where the Doppler tracing was well recorded in the functional right ventricle, and the proximal and distal atrialized right ventricles.

Pulsed Doppler echocardiogram was obtained by the ATL-500A pulsed Doppler system and recorded by Honeywell 1856 strip chart recorder at a paper speed of 100 mm/sec. This instrument registers the time interval histogram. A 3 MHz transducer provided both Doppler and M-mode signals, and pulse repetition frequency was 5.5 KHz; the sample volume of this system was of 2×4 mm tear drop shape.

Cross-sectional echocardiogram was obtained by a commercially available Sonolayergraph of Toshiba SSH-11A with an electronic sector scanner. Its resonant frequency was 2.4 MHz. The scanning speed was 30 cross-sections per second, and one frame was composed of 112 scanning lines.

RESULT

1. Normal tricuspid flow

The PDE in normal subjects showed a bimodal flow pattern representing the flow from the right atrium to the right ventricle during diastole at the tricuspid orifice. In systole, the dot was coherent and fluctuated near zero level. However, in early systole, a short negative deflection is recorded in isovolumetric contraction period, as shown in Fig. 1.

2. Secondary tricuspid regurgitation

Ten patients with tricuspid regurgitation secondary to mitral stenosis were studied by PDE. In all cases, a dispersed dot pattern was recognized during systole in the outflow of the right atrium, representing a disturbed flow due to tricuspid regurgitation. The onset of dot scattering was almost simultaneous with tricuspid closure. Fig. 2 shows a PDE record obtained from a 45-year-old female with secondary tricuspid regurgitation. The interval between the onset of the tricuspid regurgitant flow and that of QRS on a simultaneously recorded electrocardiogram was found to vary individually with heart rate, and it was corrected for the R-R interval. The corrected interval ranged from 0.07 to 0.11 sec in all the 10 patients.

3. Ebstein's anomaly

A: Flow pattern in the distal atrialized right ventricle
Fig. 1. Normal flow pattern in the right atrial outflow. At the top of this tracing is a compressed M-mode recording showing the position of sample volume with respect to identifiable structures. Time interval histogram indicates the flow toward the transducer, whereas the dot below the zero line is the flow away from the transducer. A bimodal filling flow is recorded, and after atrial contraction, a brief negative deflection is recorded. This was thought to be a physiological regurgitant flow during the isovolumetric contraction period.

Fig. 2. PDE finding of tricuspid regurgitation from a 45-year-old female with mitral stenosis and tricuspid regurgitation. The spectral flow tracing shows a widely scattered dot pattern during systole due to a disturbed flow from tricuspid incompetence. The arrow shows the onset of the dot dispersion following a short negative deflection, representing the onset of tricuspid incompetence. The arrow shows the onset of the dot dispersion following a short negative deflection, representing the onset of tricuspid regurgitation. Abbreviations: SV=sample volume; ATL=anterior tricuspid leaflet; RA=right atrium.

Prior to PDE examination, cross-sectional echocardiography was carried out to confirm the beam direction and the transducer position where the Doppler tracing was well recorded. Fig. 3 shows the cross-sectional echocardiogram obtained from a 4-year-old female with Ebstein’s anomaly. The
Fig. 3. Location of sampling site for Doppler echocardiography. Upper panel: Cross-sectional echocardiogram obtained from a 4-year-old female with Ebstein's anomaly. Lower panel: Schematic illustration of the upper panel. A, B, and C show the sampling sites of PDE as follows: A is located in the functional right ventricle, B in the distal atrialized right ventricle, and C in the proximal atrialized right ventricle, respectively. Abbreviations: FRV = functional right ventricle; ATRV = atrialized right ventricle; RA = right atrium; LV = left ventricle; IVS = interventricular septum; IAS = interatrial septum.

The lower panel is a schematic illustration of the upper panel. The line B in the lower panel represents the beam direction of PDE sampling in the distal ATRV. In this cross-section, the septal tricuspid leaflet was displaced into the right ventricle, resulting in the formation of the ATRV and functional right ventricle (FRV).

Fig. 4 shows a PDE tracing from the same patient as in Fig. 3. The sample volume was located in the distal ATRV along beam direction B in Fig. 3. A widely dispersed dot pattern was recorded during systole, showing a disturbed flow of tricuspid regurgitation. The onset of dot scattering was simultaneous with the tricuspid closure. This pattern was relatively similar to that in secondary tricuspid regurgitation.

Fig. 5 is a PDE recording in the distal ATRV from a 35-year-old male with Ebstein’s anomaly. A relatively smooth negative flow was recorded in early systole, whereas the dots were dispersive in mid- to end-systole. The onset of dot dispersion was slightly later than the tricuspid closure on a simultane-
Fig. 4. PDE tracing from the same patient as in Fig. 3. The sample volume was located in the distal ATRV along the beam direction B in Fig. 3. A widely dispersed dot pattern is recorded in systole following an initial smooth negative deflection, indicating a tricuspid regurgitant flow. The arrow shows the onset of the dot dispersion due to the tricuspid disturbed regurgitant flow. Abbreviations: SV=sample volume; ATL=anterior tricuspid leaflet; ATRV=atrialized right ventricle.

Fig. 5. PDE finding in the distal ATRV from a 35-year-old male with Ebstein’s anomaly. A relatively smooth negative flow is recorded in early systole, whereas the dots are dispersed in mid- to late-systole. In Ebstein’s anomaly, the right ventricular pressure rises slowly initially, but rapidly after the tricuspid closure. The right ventricular pressure elevation creates the disturbed flow due to tricuspid regurgitation. The dispersed flow begins slowly later than the tricuspid closure. The arrow marks the onset of tricuspid regurgitation. Abbreviations: SV=sample volume; ATL=anterior tricuspid leaflet; ATRV=atrialized right ventricle.

Fig. 6 shows a PDE in the distal ATRV obtained from a 7-year-old male with Ebstein’s anomaly. The coherent dot pattern during systole oriented away from the transducer represented a severe tricuspid regurgitation, and the notch of the negative deflection was thought to be simultaneous with the onset of the tricuspid regurgitation with pressure rise in the right ventricle.
Fig. 6. PDE in the distal ATRV obtained from a 7-year-old male with Ebstein's anomaly. The coherent dot pattern during systole oriented away from the transducer is thought to represent severe tricuspid regurgitation. The arrow shows the notch of the negative deflection. This notch is thought to represent the onset of tricuspid regurgitation with the right ventricular pressure rise. Abbreviations: SV=sample volume; ATL=anterior tricuspid leaflet; ATRV=atrialized right ventricle.

Fig. 7. PDE record in the distal ATRV from a 21-year-old female with Ebstein's anomaly. This shows a diphasic regurgitant flow pattern; the negative deflection in early systole shows a coherent dot pattern, and the latter shows a scattered dot pattern. The arrow represents the onset of the latter regurgitant flow, and it is the beginning of the tricuspid regurgitation with the right ventricular pressure rise. On catheterization, the right ventricular pressure pulse showed a remarkably slow rise to peak pressure. Abbreviations: SV=sample volume; ATL=anterior tricuspid leaflet; ATRV=atrialized right ventricle.

Fig. 7 shows a PDE record in the distal ATRV from a 21-year-old female with Ebstein’s anomaly. This represents a diphasic regurgitant flow; the negative deflection in early systole showed a coherent dot pattern, whereas the latter regurgitant flow displayed a non-coherent dot pattern beginning simultaneously with tricuspid closure on echocardiogram. The electrocardiogram
of this patient showed complete right bundle branch block pattern and the QRS interval is 0.18 sec. On catheterization, the right ventricular pressure pulse showed a remarkably slow rise to peak pressure.

Tricuspid regurgitant flow was recognized by PDE in all the 10 patients with Ebstein's anomaly in the distal ATRV, as shown in Table I. The regurgitant flow varied in its Doppler configuration from case to case. One extreme was a widely scattered dot pattern representing the disturbed regurgitant flow as shown in Fig. 3, and another was the coherent dot pattern during systole representing the smooth regurgitant flow as shown in Fig. 5. In the anomaly with severe tricuspid regurgitation, the PDE showed a narrow dot pattern during systole. The onset of the negative deflection of the spectral flow in the anomaly was similar to that in the secondary tricuspid regurgitation; however, the smooth negative deflection was more prolonged in the anomaly than in the secondary tricuspid regurgitation.

The interval from the onset of QRS to that of the regurgitant flow was measured and corrected for heart rate. The corrected interval ranged from 0.10 to 0.35 sec with an average of 0.19±0.08 sec (mean±S.D.). It was significantly prolonged over that in the secondary tricuspid regurgitation associated with mitral stenosis (p<0.001). The patient with the anomaly showing the earliest onset of tricuspid regurgitation was associated with Wolff-Parkinson-White preexcitation syndrome, type B.

B: Flow pattern in the proximal atrialized right ventricle
The echo beam direction of the PDE sampling in the proximal ATRV was shown as C in the schema of Fig. 3.

Fig. 8 is a PDE tracing in the proximal ATRV obtained from the same patient as in Fig. 4. A dispersed dot pattern is recorded during systole and is thought to represent a tricuspid regurgitant flow. Abbreviations: SV=sample volume; ATL=anterior tricuspid leaflet; ATRV=atrialized right ventricle.
patient as in Fig. 4 and a widely scattered dot pattern was shown during systole. It was thought to represent the tricuspid regurgitant flow as in the distal ATRV (Fig. 4).

Fig. 9 is a PDE finding in the proximal ATRV from the same patient as in Fig. 5. The tricuspid regurgitant flow was obscure as compared with that in the distal ATRV (Fig. 5).

In 6 out of 10 patients with Ebstein's anomaly, the tricuspid regurgitant flow was recognized in the proximal ATRV, but not in the other 4, as shown in Table I. The PDE finding of tricuspid regurgitation is less apparent in the proximal ATRV than in the distal ATRV.
C: Flow pattern in the functional right ventricle (FRV)

The echo beam direction of PDE was shown as A in the schema of Fig. 3. Fig. 10 is a PDE tracing in the FRV obtained from the same patient as in Figs. 4 and 8. In systole, the dot was narrow and remained slightly below the zero flow level. A relatively smooth flow from the ATRV to the FRV is shown in diastole.

In 7 out of 10 cases with Ebstein's anomaly, the inflow pattern was recorded and an abnormal diastolic PDE pattern was not recognized. On catheterization, a minimal diastolic pressure gradient was found in only 1 patient with the anomaly. However, the PDE in this patient showed no abnormal diastolic pattern.

D: Flow pattern in the right ventricular outflow tract

To obtain the blood flow pattern in the right ventricular outflow tract, the echo beam of PDE was directed to the pulmonary valve in the conventional manner. In only 3 cases with the anomaly was the pulmonary valve recognized in the simultaneously recorded M-mode echocardiogram and the blood flow pattern recorded. No abnormal Doppler tracing was observed in these 3 patients.

DISCUSSION

Since its advent, M-mode echocardiography has served as a useful tool for the diagnosis of Ebstein's anomaly. Crew et al studied the echocardiogram of patients with Ebstein's anomaly and correlated it with simultaneous phonocardiogram, especially with heart sounds, and systolic and diastolic murmurs. Moreover, the displacement of the septal tricuspid leaflet and the presence of the atrialized right ventricle have been recognized by cross-sectional echocardiography.17)-20)

Ebstein's anomaly shows great variations in hemodynamic states according to the degree of the anatomical malformation. These hemodynamic alterations have been studied by angiocardiography.41,43) Takayasu et al classified this anomaly into 3 types: tricuspid stenosis dominant, tricuspid insufficiency dominant, and moderate types.

More recently, a range-gated pulsed Doppler has been combined with M-mode echocardiography to provide intracardiac flow velocity information in known areas of the heart and great vessels.21)-28) PDE has been applied to examine the hemodynamic alteration in various cardiac diseases.

Prior to PDE study, cross-sectional echocardiographic observation was performed to confirm the beam direction and transducer position where the Doppler tracing was well recorded in the FRV, and proximal and distal ATRV.
In cases with the anomaly, the beam direction and transducer position have varied according to the degrees of cardiac enlargement and tricuspid displacement, and the cross-sectional echocardiographic observation was useful to obtain the subsequent Doppler tracing.

In the distal ATRV, the tricuspid regurgitant flow was recognized in all cases with the anomaly, and the configuration and the onset of the regurgitant flow varied. Cases of Ebstein’s anomaly with severe tricuspid regurgitation showed a relatively coherent dot pattern during systole. In the patients with tricuspid insufficiency dominant type, back-and-forth flow of large amount of blood occurs between the right atrium and right ventricle due to the severe hypoplasia of the septal and posterior tricuspid leaflets, with non-turbulent flow, indicated by a smooth dot pattern in systole.

The onset of tricuspid regurgitant flow with the pressure rise delay of the right ventricle on PDE tracing is approximately simultaneous with the tricuspid closure on M-mode echocardiogram. After atrial contraction, in a few patients having the anomaly, a negative smooth flow was recognized similar to the physiological regurgitant flow in normal subjects, whereas in other cases, a negative smooth flow was prolonged.

The onset of tricuspid regurgitant flow due to the right ventricular pressure rise seemed to correspond to the onset of dot dispersion of PDE tracing in the cases with disturbed regurgitant flow, the notch of the negative deflection in those with smooth regurgitant flow and the onset of the latter part of the diphasic regurgitant flow, respectively.

The interval between the onset of QRS and that of the regurgitant flow was greater in patients with Ebstein’s anomaly than in those with the secondary tricuspid regurgitation. The prolonged interval in the anomaly seemed to be related to the delay in rise of pressure in the functional right ventricle and with the elevated pressure in the atrialized right ventricle in systole. Fontana et al29) reported the abnormal right ventricular pulse and prolonged rise to peak pressure and suggested that cause was not a conduction defect alone, but rather an altered pattern of ventricular contraction and abnormal leaflet placement. A delayed tricuspid closure has been reported,11)-16) explained by the right bundle branch block or a mechanical factor directly related to the malformed tricuspid valve with its large anterior leaflet.11),15) Tajik et al have reported a case of Ebstein’s anomaly with Wolff-Parkinson-White preexcitation syndrome, type B, showing tricuspid closure delay and abnormal right ventricular pressure rise. However, only 1 out of 10 patients in our series was associated with W-P-W syndrome, type B, and the patient showed the earliest tricuspid closure on the M-mode echocardiogram. Two cases with Ebstein’s anomaly showed diphasic tricuspid
regurgitant flow and remarkably slow pressure rise in the right ventricle in early to midsystole.

In 6 of 10 patients with the anomaly, a tricuspid regurgitant flow pattern was also noted in the proximal ATRV, but, it was less apparent than in the distal ATRV. Since proximal ATRV was located at some distance from the tricuspid orifice, the energy of disturbance was dissipated by rheologic factors, and the regurgitant flow became more obscure. Also, since proximal ATRV was large, the PDE beam might not have been directed to the regurgitant flow. Due to the contraction of atrialized right ventricle, the regurgitant flow might be modified. It was difficult to assess the severity of tricuspid regurgitation in this anomaly by the depth of regurgitant flow.

The inflow pattern in the functional right ventricle was recognized in 7 patients, but an abnormal dot pattern was not identified. Our cases with Ebstein’s anomaly belonged to the tricuspid insufficiency dominant and moderate types. On catheterization, a minimal diastolic pressure gradient was observed in only 1 subject with this anomaly. Tricuspid stenosis of minimal degree may be difficult to differentiate from relative tricuspid stenosis accompanied by tricuspid regurgitation with PDE examination in the present state of art. We have not performed a PDE study in a case with tricuspid stenosis dominant type. However, Ebstein’s anomaly with tricuspid stenosis dominant type should be diagnosable by PDE examination.

The flow pattern in the right ventricular outflow tract was recorded in 3 patients with the anomaly. Only in these cases was pulmonary valve recognized in the simultaneously recorded M-mode echocardiogram. Due to cardiac enlargement and abnormal position, it was difficult to detect the pulmonary valve in the remainder. The patients with the anomaly showed reduced cardiac output and disturbed flow was not recorded on PDE tracing.

In conclusion, Ebstein’s anomaly showed considerable variations in the flow pattern according to the hemodynamic states. The smooth negative deflection in early systole was prolonged in this anomaly than in the secondary tricuspid regurgitation.

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REFERENCES

1. Am J Cardiol 2: 210, 1958
output from echo-Doppler to detect left-to-right atrial shunts. Circulation 58: 147, 1978


