

Echocardiographic Assessment of the Right Ventricle in Ebstein's Anomaly: Relation to Clinical Outcome

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Two-dimensional echocardiography was performed in 16 patients with Ebstein's anomaly to assess right ventricular anatomy and function in relation to clinical features and prognosis. Measurements of right ventricular anatomy and function were established in 10 normal subjects for comparison. Ten patients were in New York Heart Association functional class I, four in class II and one each in classes III and IV. Right ventricular morphology and the three tricuspid valve leaflets were assessed from right ventricular inflow tract and apical four chamber views. The anterior tricuspid leaflet was abnormal but not displaced in all patients; the septal and posterior leaflets were displaced in 14 (88%) and 11 (69%) patients respectively. The posterior leaflet was best visualized from the right ventricular inflow tract, and in two patients this view was required for the echocardiographic diagnosis of Ebstein's anomaly, based on displacement of the septal tricuspid valve leaflet.

An index of right ventricular function, the fractional area contraction, was defined as the difference between the end-diastolic and the end-systolic area, normalized

to the end-diastolic area. This index was calculated for both the proximal (atrialized) right ventricle and the total right ventricle. Total right ventricular end-diastolic area and fractional area contraction exceeded 95% confidence limits when compared with values in the normal group. During a median follow-up period of 4 years three patients died. They had severe right heart morphologic or functional abnormalities; two were in functional class III or IV and one was asymptomatic. None of the survivors had severe symptoms. Eight survivors had mild morphologic or functional abnormalities of the tricuspid valve or right ventricle, whereas five others including one patient with absent septal leaflet had paradoxical systolic expansion of the proximal right ventricular chamber.

Thus, in these patients with Ebstein's anomaly there was no correlation between clinical status, including the occurrence of arrhythmias or sudden death, and the severity of morphologic and functional abnormalities of the right ventricle. This suggests that other factors, particularly electrical instability, may be important.

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In Ebstein's anomaly the right ventricle is divided into two parts by an abnormal tricuspid valve that separates the proximal or atrialized portion from the distal or functional right ventricle (1-6). The excellent correlation between the echocardiographic description and surgical and postmortem findings has established two-dimensional echocardiography as the technique of choice in the assessment of atrioventricular (AV) valve morphology in congenital heart disease (7-9).

An evaluation of right ventricular function in Ebstein's anomaly, however, has not been undertaken. In this study we reviewed two-dimensional echocardiographic studies from 16 consecutive patients with Ebstein's anomaly and de-

scribed right ventricular morphology and tricuspid valve leaflet anatomy in this group. We then attempted to relate an index of right ventricular function, the fractional area of contraction, to clinical and prognostic features.

Methods

Patients. Sixteen consecutive patients with a diagnosis of Ebstein's anomaly studied in the cardiac ultrasound laboratory were included in this study. Ten were male and six female, with an age range of 1 day to 25 years (median 8 years). Cardiac catheterization and angiography were performed in all patients, and the diagnosis was consistent with Ebstein's anomaly in 15. In the remaining patient, angiography failed to confirm the echocardiographic diagnosis, and the definitive diagnosis of Ebstein's anomaly was established only after a second cardiac catheterization and recording of an intracardiac electrocardiogram.

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The clinical findings and associated anomalies of the 16 patients are summarized in Table 1. Ten patients were in New York Heart Association functional class I, four in class II, one in class III and one, a 1 day old infant, in class IV. None of the patients underwent surgical correction of the tricuspid valve. During a follow-up period of up to 5 years (range 1 day to 5 years, median 4 years), three patients died. A year old girl (Case 6) died suddenly during a swimming lesson. A neonate (Case 10) who had had frequent episodes of ventricular tachycardia during catheterization 2 days before death collapsed during feeding. A third infant (Case 16) had ventricular fibrillation and could not be resuscitated 1 day after cardiac catheterization. Autopsy data were available only in Case 6 and confirmed the echocardiographic diagnosis of the tricuspid valve and associated abnormalities.

Control group. Right ventricular measurements were obtained from 10 normal subjects who were selected on the basis of having good quality echocardiographic images of the right ventricle for comparison.

Two-dimensional echocardiography. Echocardiographic studies were performed using a high resolution ultrasound system (IGE, Slough, Berkshire, UK) or a mechanical sector scanner (Advanced Technology Laboratories Limited), utilizing a 3.3, 3.5 or 5 MHz transducer. For each patient a complete two-dimensional echocardiographic examination was performed (10,11), with particular reference to specific right ventricular views (12). To best visualize

the total right ventricle with proximal and distal segments, right ventricular inflow tract and apical four chamber views were selected as the two standardized orthogonal views that best defined the apical region and the location of the anomalous tricuspid valve leaflet insertions (Fig. 1). Images were recorded on 1/2 inch (1.27 cm) videotape and were subsequently analyzed with an off-line computer system (Microsonics Company). A joystick-operated cursor that minimizes parallax errors was used to measure by planimetry the areas of the right atrium, total right ventricle and proximal right ventricle. The proximal right ventricular chamber was defined as the area enclosed between the true tricuspid annulus (identified by an AV remnant) and the point of insertion of the displaced leaflet (Fig. 2). The area of the distal chamber was obtained by subtraction of the proximal chamber from the total right ventricular area. Measurement of each region was made at end-systole and end-diastole from each of the two orthogonal projections, inflow tract and the apical four chamber views. End-systolic and end-diastolic frames of the right ventricle were identified by the closure and opening of the tricuspid valve and by the simultaneous recording of lead II of the electrocardiogram. Careful identification of endocardial boundaries was facilitated by repeated slow-frame replay.

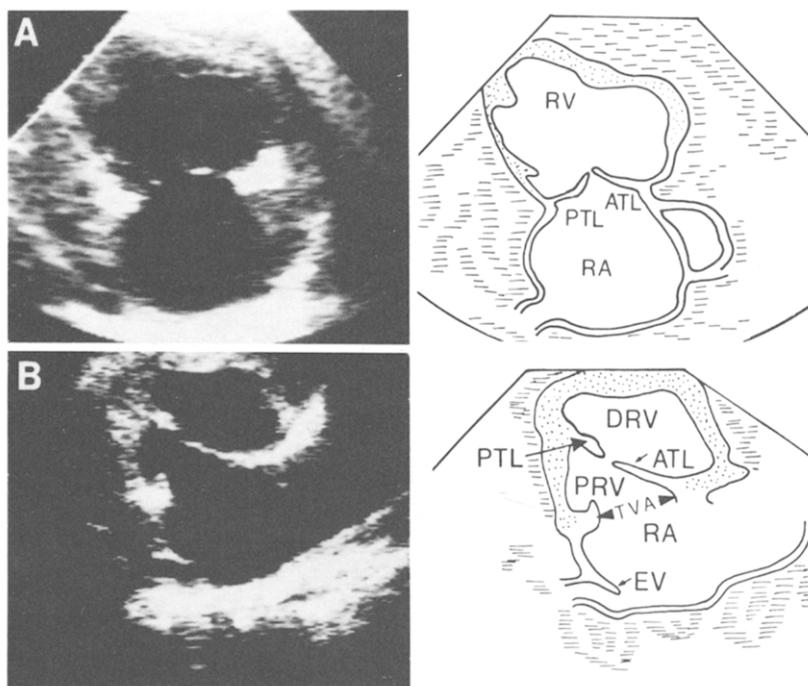
Right ventricular function. The complex geometry of the right ventricle has made accurate volume determination difficult (13). Accepting these constraints, we calculated an index of right ventricular function, the fractional area con-

Table 1. Clinical Features and Associated Anomalies of 16 Patients With Ebstein's Anomaly (at time of diagnosis)

Case	Age (yr)	Sex	NYHA Class	Cyanosis	Standard 12 Lead ECG	Ambulatory ECG Monitoring	Associated Anomalies	Follow-Up (median 4 years)
1	25	M	I	None	RBBB	Normal	Coarctation; PFO	Normal
2	12	F	II	Mild	IRBBB	SVT	None	Normal
3	13	M	II	Mild	IRBBB	Normal	2°ASD; PFO; redundant EV	Normal
4	7	M	II	Mild	IRBBB	Normal	2°ASD	Normal
5	14	M	I	Mild	IRBBB	Normal	PFO	Normal
6	7	F	I	None	IRBBB	...	2°ASD	Sudden death
7	18	M	I	None	WPW	Normal	PFO; Redundant EV	Normal
8	15	F	I	None	RBBB	Normal	PFO	Normal
9	1	F	II	Mild	RBBB	...	VSD	Normal
10	3 days	F	III	Moderate	IRBBB	...	2°ASD; mild PS	Sudden Death
11	10	M	I	None	RBBB	Normal	PFO; PS; redundant EV	Active sports
12	2 days	M	I	Mild	IRBBB	...	2°ASD; VSD	Normal
13	8	M	I	None	WPW	Normal	VSD; PS	Normal
14	6	M	I	Mild	RBBB	Normal	PS; VSD; LSVC; redundant EV	Normal
15	8	M	I	None	RBBB	Normal	PFO	Normal
16	1 day	M	IV	Severe	RBBB	...	ASD; VSD; PV atresia with hypoplastic PA	Myocardial failure, death

ASD = atrial septal defect; ECG = electrocardiogram; EV = eustachian valve abnormality; F = female; IRBBB = incomplete right bundle branch block; LSVC = persistent left superior vena cava; M = male; NYHA Class = New York Heart Association functional class; PA = pulmonary artery; PFO = patent foramen ovale; PS = pulmonary stenosis; PV = pulmonary valve; RBBB = right bundle branch block; SVT = supraventricular tachycardia; 2° = second degree; VSD = ventricular septal defect; WPW = Wolff-Parkinson-White syndrome; ... = not done.

Figure 1. Comparable right ventricular inflow tract view from a normal subject (A) and a patient with Ebstein's anomaly (B). There is a marked displacement of the posterior tricuspid leaflet (PTL) in the patient with Ebstein's anomaly (Case 8), dividing the total right ventricle (RV) into two portions: the proximal (PRV) and distal (DRV) right ventricle. ATL = anterior tricuspid leaflet; EV = eustachian valve; RA = right atrium; TVA = tricuspid valve anulus.

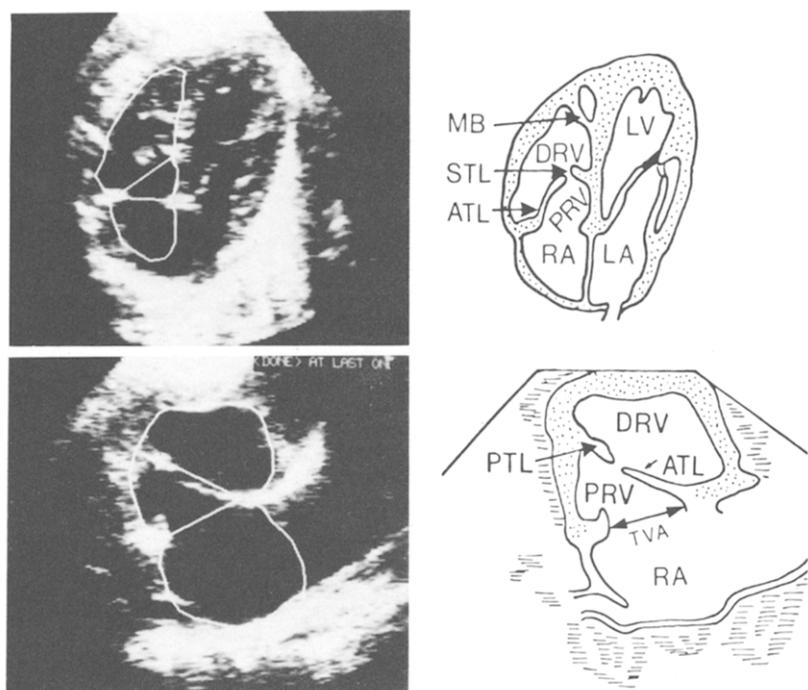


traction, defined as the difference between the end-diastolic and the end-systolic area normalized to the end-diastolic area. This calculation was made in each of the paired echocardiographic views, which, being orthogonal to each other, imaged all four right ventricular walls. From the apical four chamber view, the septal and lateral right ventricular walls, and from the right ventricular inflow tract view, the anterior and posterior (or diaphragmatic) walls are visualized. The fractional area contraction was calculated separately for the

proximal, distal and total right ventricle and for the right atrium from both views (Fig. 3). All measurements were corrected for body surface area.

Tricuspid valve. The insertion of three tricuspid valve leaflets cannot be visualized simultaneously from a single echocardiographic view. The anterior and posterior leaflets were best imaged from the right ventricular inflow tract view and the posterior and septal leaflets from the apical four chamber view, in keeping with previous descriptions

Figure 2. Two-dimensional echocardiographic apical four chamber view (upper panels) (Case 12) and right ventricular inflow tract view (lower panels) (Case 8) demonstrating the areas of the proximal right ventricle PRV, distal right ventricle (DRV) and right atrium (RA) measured by planimetry. Note that the anterior tricuspid leaflet (ATL) is visualized from both views, whereas the septal tricuspid leaflet (STL) is visualized from the apical four chamber view and the posterior tricuspid (PTL) leaflet from the right ventricular inflow tract view. LA = left atrium; LV = left ventricle; MB = moderator band; TVA = tricuspid valve anulus.



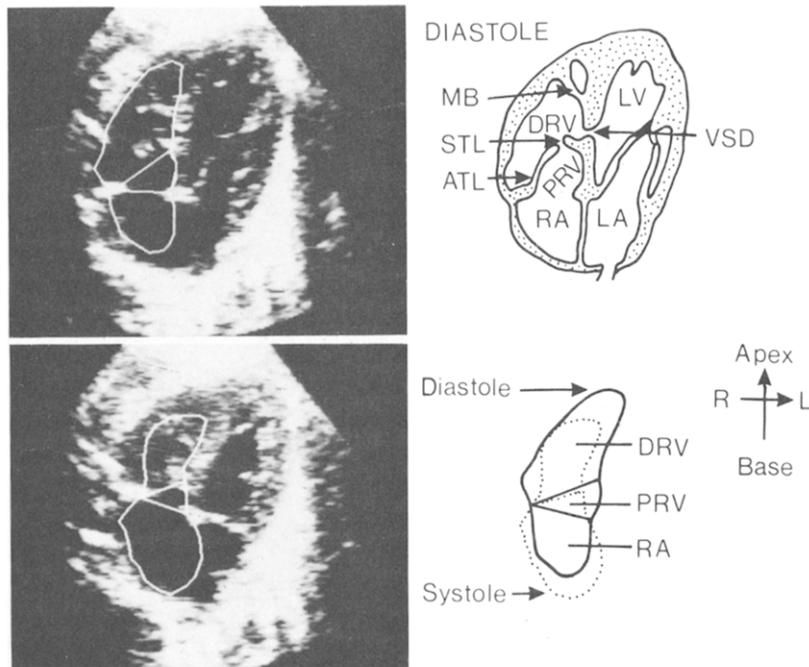


Figure 3. Apical four chamber view (Case 12) at end-diastole (**upper panels**) and end-systole (**lower panels**) with diagrammatic representation of the area changes of the right heart chambers during the cardiac cycle. L = left; R = right; VSD = ventricular septal defect; other abbreviations as in Figures 1 and 2.

(10,11,14,15). Displacement of the septal or the posterior leaflet, or both was measured in systole as the distance between the true tricuspid annulus and the nearest point of insertion of the leaflet to the underlying myocardium.

The morphology of each tricuspid valve leaflet was evaluated against a large experience of reviewing normal right heart echocardiographic studies, and changes in size (hypoplasia or elongation), shape and leaflet motion were noted.

Statistical analysis. The morphologic findings and fractional area contraction were related to symptomatic status, presence or absence of arrhythmias and clinical outcome using a chi-square test. Paired data were analyzed with a two-tailed Student's *t* test; a probability value of less than 0.05 was considered significant.

Intraobserver variability of the measurements was assessed from a reevaluation of the original echocardiographic study performed at least 1 month after the initial study. The end-systolic and end-diastolic areas for the total right ven-

tricle and proximal right ventricular chamber were assessed from both the right ventricular inflow tract and apical four chamber views and expressed as the absolute difference and the percent difference between the two measurements.

Results

Reproducibility. Intraobserver variability was least with the apical four chamber view in which the percent difference between the two measurements was less than 15% (Table 2).

Tricuspid Valve

Anterior leaflet. The anterior leaflet was normally attached at the AV junction in all patients. In 15 (94%), this leaflet appeared grossly elongated and in 11 (69%), it had a characteristic undulating whiplike motion. Tethering to the right ventricular wall was evident in nine patients (Table

Table 2. Intraobserver Variability of Right Ventricular Chamber Dimensions

	Four Chamber View				RV Inflow Tract View			
	RV		PRV		RV		PRV	
	ES (n = 15)	ED (n = 15)	ES (n = 13)	ED (n = 13)	ES (n = 14)	ED (n = 14)	ES (n = 10)	ED (n = 10)
Absolute difference (mean ± SD)	1.3 ± 1	1.3 ± 2.4	1.2 ± 1.4	1 ± 1.2	0.5 ± 1.4	0.2 ± 1	1 ± 1.4	2.2 ± 5
Range of absolute difference	0.2 to 4.6	0.1 to 1.5	0.4	0.3	0.5 to 4.5	0.7 to 8.2	0.1 to 2.2	0.3 to 2.1
Median (cm)	1.1	2	0.4	0.6	1.9	4.2	0.5	0.7
Percent of difference (mean ± SD)	9 ± 12.1	6.6 ± 15	1.9 ± 2.3	3.4 ± 2.7	7.3 ± 21	14 ± 31	5 ± 9	4.6 ± 10.8

ED = end-diastole; ES = end-systole; PRV = proximal right ventricle; RV = right ventricle.

Table 3. Tricuspid Valve Leaflet Description in 16 Patients With Ebstein's Anomaly

Case	Anterior Leaflet			Septal Leaflet			Posterior Leaflet		
	Morphology	Tethering	Mobility	Morphology	Mobility	Displacement (mm/m ²)	Morphology	Mobility	Displacement (mm/m ²)
1	E	+	↑	D	N	30	N	N	0
2	E, T, D	+	Fixed	H	Fixed	22	T	↓	0
3	E	No	↑	D, H	↓	22	N	N	16
4	E	+	↑	D	↓	30	N	N	0
5	E	+	↑	D	↓	19	T	N	17
6	E, T, D	++	Fixed	D	↓	21	N	N	0
7	E	No	↑	H	N	10	N	N	13
8	E	No	↑	D, H	↓	36	N	N	23
9	E	No	↑	D	N	16	N	N	0
10	T, D	++	↓	D	N	33	T	↓	43
11	E	++	↓	Absent			T	↓	50
12	E	No	↑	D	N	28	T	↓	28
13	E	No	↑	D	N	16	N	N	24
14	E, T, D	+	Fixed	N	N	0	N	N	50
15	E	No	↑	H	↓	35	T	↓	52
16	E	+	↑	H	N	36	N	N	60
Mean						25.3			34.2

D = dysplastic (apparently abnormal structure ± thickening); E = elongation; H = hypoplastic; N = normal; T = thickened; ↑ = increased mobility; ↓ = decreased mobility; + = moderate; ‡ = abundant.

3). In four patients the anterior leaflet was thickened and dysplastic.

Septal leaflet. The septal leaflet was identified in 15 patients (94%) and in 14 (88%) it was displaced toward the apex from 10 to 36 mm/m² (mean 25.3) (Table 3). No leaflet tissue was identified in one patient (Case 11) despite excellent echocardiographic images. In another (Case 14), this leaflet was attached to the AV junction at the same level as the mitral valve (Fig. 4); this patient had an associated large ventricular septal defect that involved the inlet septum.

Posterior leaflet. The posterior leaflet was identified in all patients from the right ventricular inflow tract view. It was displaced in 11 patients (69%) from 13 to 60 mm/m² (mean 34.2). Although this leaflet did not appear to vary in length, it was thickened in seven patients (44%) and had a reduced motion, implying tethering, in five (31%) (Table 3).

Right Ventricle

Dimensions. Right ventricular endocardium was clearly identified in all patients from the apical four chamber view, and in 15 from the right ventricular inflow tract view. There was no significant difference between the values for the total right ventricle, proximal and distal right ventricular chamber and right atrial areas obtained from these two views (Fig. 5). The diastolic areas of the proximal right ventricular chamber was smaller than the corresponding end-systolic area in four patients as assessed from the four chamber view, and in two patients (one additional), as assessed from the inflow tract view. Patients with Ebstein's anomaly had sig-

nificantly greater diastolic dimensions than those of the control group ($p < 0.001$) (Fig. 6).

Fractional area contraction. The fractional area contraction for the total right ventricle was similar from both views and in each category of the patient's functional class (Fig. 7). It was always positive for the total right ventricle, whereas it was negative for the proximal right ventricular chamber in the four patients who exhibited systolic expansion from the apical four chamber view and in the additional patient who exhibited this expansion from the right ventricular inflow tract view.

Relation of Right Heart Morphology, Function and Outcome

We failed to define any relation between symptoms, arrhythmia or sudden death and tricuspid valve leaflet morphology, reduction in fractional area contraction of the total right ventricle or paradoxical systolic expansion of the proximal right ventricular chamber (Fig. 7).

Three patients died suddenly. Of these, two had grossly abnormal anterior and septal leaflets (Cases 6 and 10) and the third (Case 16) had septal and posterior leaflets that were displaced but neither thickened nor dysplastic. Only one of the three (Case 6) was asymptomatic and one (Case 16) had severe associated intracardiac abnormalities (Table 1). None of the three patients had severe tricuspid regurgitation and the fractional area contraction in this group showed no difference from that of surviving patients (Fig. 7).

Thirteen patients are alive without severe symptoms

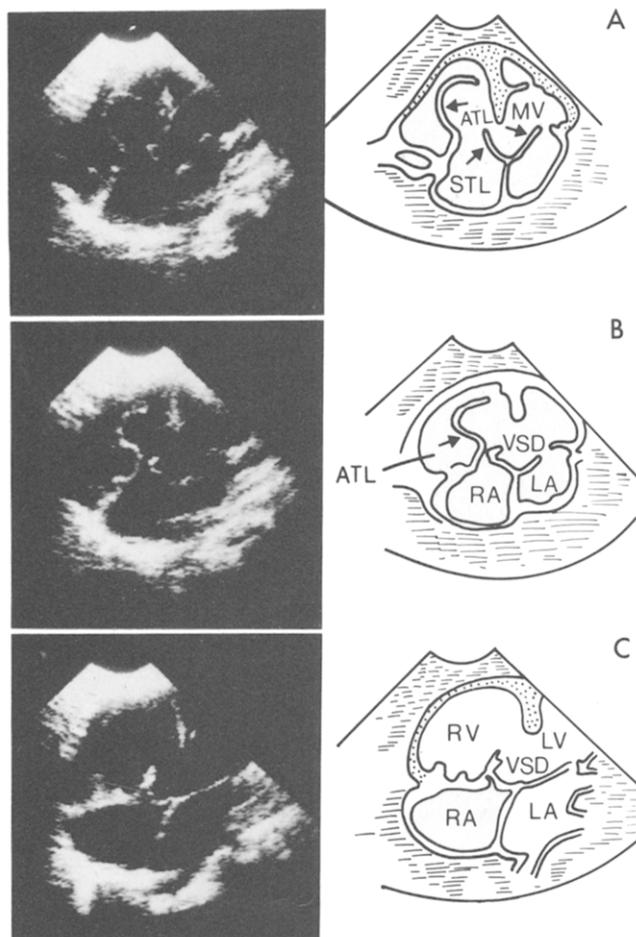


Figure 4. Apical four chamber view (Case 14) showing the equiplanar insertion of the septal tricuspid leaflet (STL) with the mitral valve (MV) in mid-diastole (A), onset of systole (B) and mid-systole (C). Note the large ventricular septal defect involving the inlet septum and the elongated anterior tricuspid leaflet with a characteristic whiplike motion. Abbreviations as in previous figures.

(functional classes I and II) (Table 1). Of these 13, two patients (Cases 2 and 14) had a grossly abnormal anterior tricuspid leaflet and another (Case 11) had no septal leaflet identified but had a very displaced posterior leaflet and moderate tricuspid regurgitation. Five patients had paradoxical systolic expansion of the proximal right ventricular chamber, involving the right ventricular free wall (three patients), the inferior (diaphragmatic) wall (one patient) and both the inferior and free right ventricular free walls (one patient). There was no difference in fractional area contraction between survivors and patients who died. There was no relation between size of the proximal or distal right ventricle and fractional area contraction of the total right ventricle.

A mild to moderate degree of tricuspid regurgitation was present at cardiac catheterization in all patients, but in none was it hemodynamically severe. There was no relation between functional class and the degree of septal or posterior leaflet displacement.

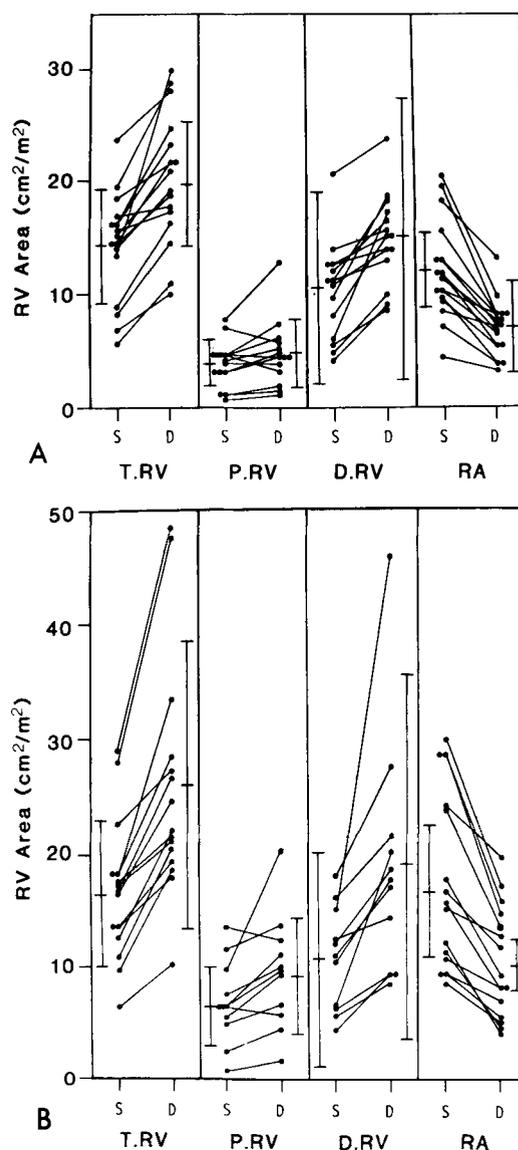


Figure 5. Individual values and means \pm 1 SD for the areas measured by planimetry in systole (S) and diastole (D) of the total right ventricle (T.RV), proximal right ventricle (P.RV), distal right ventricle (D.RV) and right atrium (RA) measured from the apical four chamber (A) and right ventricular (RV) inflow tract (B) views.

Discussion

Echocardiographic diagnosis. In Ebstein's anomaly the echocardiographic description of tricuspid valve abnormalities, in particular of the anterior and septal leaflets, has been found to correlate closely with surgical and postmortem findings (10). The presence of a displaced septal leaflet has been considered to be a highly sensitive echocardiographic feature of Ebstein's anomaly (16-20). In our series, 88% of the patients exhibited this finding. The posterior leaflet, however, is also displaced in the majority of patients (1-6) and can be visualized from the right ventricular inflow tract view (10,11,21). It is particularly important to identify and

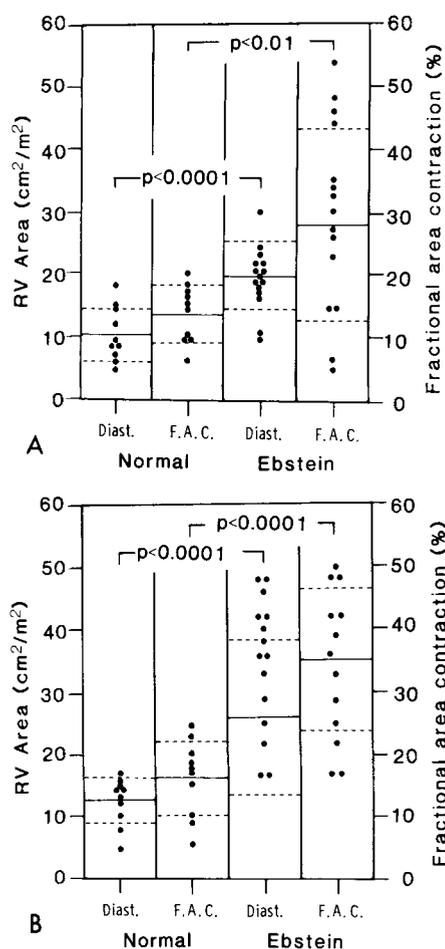


Figure 6. Individual values and mean \pm 1 SD for right ventricular (RV) end-diastolic (Diast.) area measurements and fractional area contraction (F.A.C.) in normal subjects and patients with Ebstein's anomaly evaluated from the apical four chamber (A) and right ventricular inflow tract (B) views.

describe this leaflet in patients whose septal leaflet is absent or not displaced, conditions that may lead to a false negative echocardiographic diagnosis of Ebstein's anomaly. Failure to do so may have accounted for the occasional patient reported by other investigators (5,22) in whom two-dimensional echocardiography failed to diagnose Ebstein's anomaly, and it would have led to a false negative diagnosis in two of our patients, one who had an equiplanar insertion of the septal tricuspid leaflet with the mitral valve (Case 14) and another who showed absence of the septal leaflet (Case 11). These two patients demonstrate the value of orthogonal views in the echocardiographic diagnosis of Ebstein's anomaly.

Right ventricular function. Two-dimensional echocardiography permits examination of the right ventricle from several planes and is useful in detecting wall motion abnormalities (10). Several views and formulas have been proposed (23-28) to assess right ventricular function with echocardiography. These methods generate volume mea-

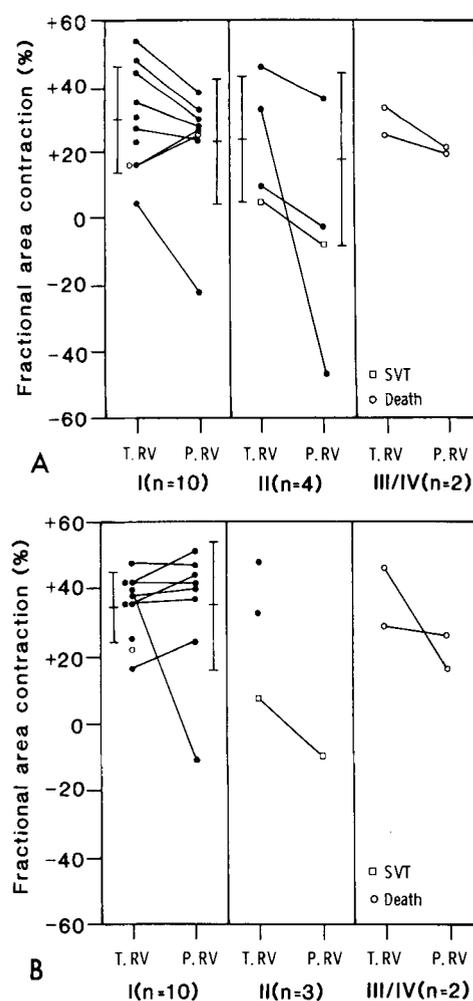


Figure 7. Individual values and means \pm 1 SD for the fractional area contraction, calculated for the total right ventricle (T.RV) and proximal right ventricular (P.RV) chamber, in the patients in functional class I, II and III/IV, obtained from the apical four chamber (A) and right ventricular inflow tract (B) views. Proximal right ventricular measurements were not made in five patients from the inflow tract view and in two patients from the four chamber view because the posterior and the septal leaflet, respectively, were not displaced. SVT = supraventricular tachycardia.

surements but are based on assumptions about right ventricular geometry that may not be valid, particularly in patients with right ventricular volume overload. In the present study we assessed right ventricular function from area measurements derived from orthogonal views. These measurements were reproducible, particularly from the apical four chamber view. Patients with Ebstein's anomaly had significantly higher values for fractional area contraction than those of the control group, from both the right ventricular inflow tract and apical four chamber views (Fig. 6). Measurements of fractional area contraction of the proximal right ventricle were either similar to the fractional area contraction of the total right ventricle (11 patients) or significantly lower (5 patients), but in this latter group the proximal right ven-

tricular chamber was contracting paradoxically and in synchrony with the right atrium, indicating functional atrialization of the proximal right ventricle. In the former group there was no functional evidence of atrialization of the proximal right ventricle.

Right ventricular morphology and proximal right ventricular function did not allow prediction of the total right ventricular fractional area contraction and there was no association with symptoms, prognosis or tricuspid valve morphology. Furthermore, we were unable to correlate the size of the distal right ventricle with overall right ventricular size, overall right ventricular fractional area contraction or symptomatic status of the patients.

Although there was a clear separation of patients with Ebstein's anomaly according to the mode of contraction of the proximal right ventricular chamber, the spectrum of measurements within each group was narrow. This and the fact that 13 of our 16 patients had minimal or no symptoms may explain the lack of correlation with total right ventricular function and symptoms. Our findings are consistent with the echocardiographic study of Gussenhoven et al. (22) but differ from reports of others (7) of a strong correlation between severity of symptoms and morphologic abnormalities; the latter group, however, reported on an older and more symptomatic population (average age 18 versus 8 years) than ours. Age may be an important determinant for the development of significant right ventricular impairment and symptoms, and may account for the discrepancy between our study and previous reports.

Clinical implications. Sudden death is a major risk in patients with Ebstein's anomaly (29-30). In our study three patients died suddenly during 5 years of follow-up. Functional characteristics and a careful search for arrhythmia during repeated ambulatory electrocardiographic monitoring failed to identify those at greatest risk. In addition, we were unable to demonstrate morphologic or right ventricular functional abnormalities that were particular to patients who died suddenly. This suggests that other factors, such as the propensity to electrical instability, may be important causes of these deaths.

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