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Outcome in Neonates With Ebstein's Anomaly

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The presentation and outcome of 50 patients with neonatal Ebstein's anomaly seen from 1961 to 1990 were reviewed. The majority (88%) presented in the 1st 3 days of life; cyanosis (80%) was the most common presenting feature. Associated defects, present in 27 infants (54%), included pulmonary stenosis in 11 and atresia in 7. Nine patients (18%) died in the neonatal period; there were 15 late deaths (due to hemodynamic deterioration in 9, sudden death in 5 and a noncardiac cause in 1) at a mean age of 4.5 years (range 4 months to 19 years). Actuarial survival at 10 years was 61%.

A new echocardiographic grade (1 to 4 in order of increasing severity of the defect) was devised with use of the ratio of the area of the right atrium and atrialized right ventricle to the area of the functional right ventricle and left heart chambers. Cardiac death occurred in 6 of 4 infants with grade 1, 1 (10%) of 10 with grade 2, 4 (44%) of 9 with grade 3 and 5 (100%) of 5 with grade 4. In a multivariate analysis of clinical and investigational features at presentation, echocardiographic grade of severity was the best independent predictor of death.

Neonates with Ebstein's anomaly have a high early mortality rate and those surviving the 1st month of life remain at high risk of late hemodynamic deterioration or sudden death. Echocardiographic grading of severity of the defect permits prognostic stratification.

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Ebstein's anomaly has an extremely variable natural history depending on the degree of displacement of the proximal attachments of the tricuspid valve leaflets from the atrioventricular (AV) ring, which may range from mild to severe (1–3). If the deformity of the tricuspid valve is severe, profound congestive heart failure may be present in the neonatal period or may even cause intrauterine death (4). At the other end of the spectrum, patients with a mild degree of tricuspid displacement may remain asymptomatic until late adult life (5).

Early natural history studies, in which detection was based on clinical or angiographic diagnosis, or both, have included mainly older children and adults (6–9). These have an inevitable bias toward factors that favor survival. In a multicenter study (6) of 505 patients, only 35 (7%) were <1 year old at diagnosis. The largest single center review (8) included only 20 of 67 patients (30%) <4 years old at admission (8). In the last decade, two-dimensional echocardiography has facilitated detection of this abnormality in neonates and infants (10,11) and even in prenatal life (12,13), so that the natural history of Ebstein's anomaly seen in the neonatal period can now be defined.

We have therefore reviewed the data of 50 patients with Ebstein's anomaly diagnosed in the neonatal period, defining their clinical presentation and outcome. We aimed to document the early and late survival pattern of neonates with this condition and to identify clinical factors and objective measures of severity of the defect that might be useful for prognostic stratification.

Methods

Study patients. We reviewed the medical records, chest radiographs, electrocardiograms (ECGs), echocardiograms and operative and autopsy reports of 50 consecutive neonates presenting with Ebstein's anomaly of the tricuspid valve between 1961 and 1990 (diagnosed by angiocardiography or two-dimensional echocardiography, or both, during the 1st month of life).

Ebstein's anomaly was defined as any degree of inferior displacement of the proximal attachments of the tricuspid valve leaflets from the AV valve ring (3). Infants with Ebstein's anomaly of the systemic AV valve with congenitally corrected transposition of the great arteries were excluded. Early mortality is defined as death within the 1st month of life and late mortality as death thereafter.

Echocardiographic grading. We devised an echocardiographic grading for severity of Ebstein's anomaly by calcu-

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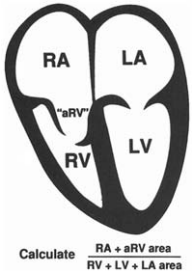


Figure 1. Schematic representation of a four-chamber echocardiographic view of the heart at end-diastole with the ratio used for grading the severity of Ebstein's anomaly (see text). aRV = atrialized portion of the right ventricle; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

lating the ratio of the combined area of the right atrium and atrialized right ventricle to that of the combined area of the functional right ventricle, left atrium and left ventricle in a four-chamber view at end-diastole (Fig. 1). The ratio was used to define four grades of increasing severity: grade 1, ratio <0.5; grade 2, 0.5 to 0.99; grade 3, 1 to 1.49; and grade 4, ≥1.5. This grade was entered as one variable into the multivariate analysis of outcome.

Survival and risk factor analysis. Survival was estimated with use of the Kaplan-Meier method. A Cox regression model was used to estimate relative risks associated with factors measurable at presentation (presence or absence of associated defects, cardiothoracic ratio on chest radiography, presence of pre-excitation on the ECG and echocardiographic grade) in both a univariate and a multivariate analysis (14). Statistical significance was inferred at a p value <0.05.

In the relative risk analysis, echocardiographic grade was considered first as an unordered variable taken on three levels (grades 1 and 2 were combined because the lack of any deaths in grade 1 makes it impossible to estimate the relative risk associated with this grade). Grade was then considered as a continuous variable, with a value of 1 to 4, with the same relative risk associated with each unit increase in grade. The validity of considering echocardiographic grade as a continuous variable was tested by examining the decrease in predictive fit (that is, the change in the likelihood ratio statistic) associated with this simplifying assumption.

Results

Presenting features. There were 22 boys and 28 girls, 6 of whom presented in the 1960s, 13 in the 1970s and 31 in the 1980s. Forty-four (88%) of the 50 patients were seen in the 1st 3 days of life, usually with cyanosis (80%) (Table 1).

Table 1. Presenting Feature of Ebstein's Anomaly in 50 Neonates

	No.
Cyanosis	40
Heart failure	5
Murmur	2
Arrhythmia	2
Cardiac screening*	1

*Baby with Prader-Willi syndrome (thought at birth to have trisomy-18).

Associated cardiac defects (other than patent foramen ovale or atrial septal defect) were present in 27 patients (54%) and included pulmonary stenosis in 11 and pulmonary atresia in 7 (Table 2).

Investigations. Chest radiography demonstrated cardiomegaly in 49 (98%) of the 50 patients. In 3 of the 18 patients whose films were available for review, the cardiothoracic ratio was >90% and all 3 of these patients died in the neonatal period compared with 1 of 15 with a cardiothoracic ratio <90%. The ECG showed evidence of right atrial enlargement in all 50 patients and partial or complete right bundle branch block in 24 (48%). Five patients (10%) had evidence of pre-excitation on the presenting ECG. Diagnosis was made by two-dimensional echocardiography in 33 infants (66%), including all those seen since 1980.

In 28 cases the neonatal echocardiogram was acceptable for calculation of the grade of severity of the defect. Three of four patients with grade 1 Ebstein's anomaly are alive at age 5 to 9 years and one died of a noncardiac cause (biliary atresia). Nine of 10 patients with grade 2 are alive at age 6 months to 16 years and 1 patient died suddenly and unexpectedly at age 4 months. Five of the nine patients with grade 3 Ebstein's anomaly are alive at age 4 months to 9 years and four have died (one early, three late); all 5 with grade 4 died at <18 months of age (Fig. 2, Table 3). All survivors are in New York Heart Association functional class I or II.

Cardiac catheterization was performed in 26 patients at some stage: as the diagnostic procedure in 17, to confirm diagnosis in the early days of echocardiography in 4, for complex associated defects in 4 and in 5 patients who

Table 2. Associated Cardiac Malformations in 50 Patients With Neonatal Ebstein's Anomaly

	No.
Patent foramen ovale or atrial septal defect only	23
Pulmonary stenosis	10
Pulmonary atresia, intact ventricular septum	7
Right ventricular hypoplasia*	3
Ventricular septal defect	2
Pulmonary stenosis, coarctation of aorta	1
Coarctation of aorta	1
Atrioventricular septal defect	1
Tetralogy of Fallot	1
Dysplastic mitral valve	1

*Absence of trabecular portion of the right ventricle.

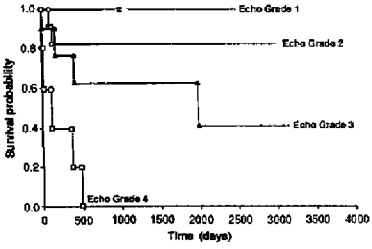


Figure 2. Kaplan-Meier survival analysis for 28 patients who had echocardiographic (Echo) grading of severity of Ebstein's anomaly. Survival probability refers to freedom from cardiac death.

developed late hemodynamic deterioration. Two patients (8%) died during the procedure and nine others (35%) had supraventricular or ventricular tachyarrhythmia that caused hemodynamic compromise.

Surgery. Eighteen patients were operated on (Table 4): the indications for surgery were persistent severe cyanosis with or without congestive cardiac failure in the neonatal period (nine cases) and hemodynamic deterioration or increasing cyanosis leading to functional class III or IV symptoms in older children (nine cases). Of the 18 patients, 12 had palliative surgery for associated defects and 6 had tricuspid valve surgery (repair with creation of a monocusp valve in 4 and valve replacement in 2).

Of the 12 patients who underwent a palliative operation (at age 1 day to 6 years, median 8 days), 1 died in the perioperative period and 2 children died late (1 suddenly and 1 from left ventricular failure, 6 months and 5 years, respectively, after the procedure). Tricuspid valve surgery was performed in six patients aged 14 days to 18 years, all of whom had echocardiographically severe Ebstein's anomaly (grade 3 or 4). Three of these were <2 years old and died perioperatively. Three older patients were aged 12, 14 and 18 years; one died perioperatively, one developed chronic atrial flutter 1 year after operation and died suddenly 1 year later and one is alive and well.

Table 3. Echocardiographic Features in 28 Neonates With Ebstein's Anomaly

Grade	RA + aRV Ratio*	No. of Patients	Cardiac Deaths
1	<0.5	4	0†
2	0.5 to 0.99	10	1 (10%)
3	1 to 1.49	9	4 (44%)
4	≥1.5	5	5 (100%)

*See Methods for explanation. †One patient died of biliary atresia at age 8 months but had no cardiac symptoms or signs of heart failure. All three survivors are asymptomatic. aRV = atrialized portion of the right ventricle; RA = right atrium.

Table 4. Surgery for Ebstein's Anomaly in the Neonatal Period

Operation	No.	Outcome
Aortopulmonary shunt only	6	S (3), ED (1), LD (2)
Pulmonary valvotomy only	2	S
Pulmonary valvotomy, aortopulmonary shunt	2	S
Repair of coarctation of aorta	1	S
Pulmonary valvotomy, repair of coarctation of aorta, ligation of ductus arteriosus	1	S
Tricuspid valve repair	4	PD (2), LD (1), S (1)
Tricuspid valve replacement	2	PD (2)

ED = early death; LD = late death (≥3 months postoperatively); PD = perioperative death; S = surviving.

Outcome

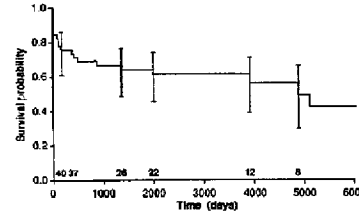
Actuarial survival at 1 year was 76% and at 10 years 61% (Fig. 3).

Early deaths. Nine patients (18%) died in the neonatal period: six with heart failure, two with ventricular arrhythmia and one at the time of tricuspid valve surgery. All nine had morphologically severe Ebstein's anomaly with gross right atrial dilation, as documented at autopsy (six cases) or echocardiography (three cases). The six neonates who died with heart failure were managed medically and did not undergo surgery; the condition of five patients who presented between 1964 and 1979 was considered inoperable and one patient had sustained severe peripartum hypoxic brain damage.

Late deaths. Late death occurred in 15 patients (30%) at a mean age of 4.5 years (median 2.5 years, range 4 months to 19 years) as a result of hemodynamic deterioration in 9, sudden death in 5 or a noncardiac cause (biliary atresia in 1 who died at age 8 months).

Nine deaths occurred in children whose neonatal cyanosis resolved but who later (at age 4 months to 18 years) developed hemodynamic problems with signs of increasing cyanosis or congestive heart failure, or both. Of these, four died at the time of attempted intracardiac repair, three were

Figure 3. Kaplan-Meier survival analysis for 50 neonates with Ebstein's anomaly. Survival probability refers to freedom from cardiac death.



JACC Vol. 19, No. 5
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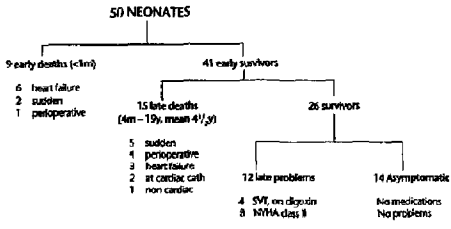

Figure 4. Outcome in 50 neonates with Ebstein's anomaly. cath = catheterization; m = months; NYHA = New York Heart Association; SVT = supraventricular tachycardia; y = years.

treated medically but died and two died at cardiac catheterization. Five of these had evidence of severe left ventricular dysfunction documented at echocardiography or angiography.

Five other deaths occurred suddenly: three in patients aged 4 to 6 months without clinically evident hemodynamic problems, one in a 14-month old boy who had a cardiac arrest on the day of hospital admission for worsening cyanosis and heart failure and one in a 26-year old girl with chronic atrial flutter who died suddenly 2 years after tricuspid valve surgery. Pre-excitation was not associated with late death; four of the five patients with pre-excitation are alive and well (the other died at the time of tricuspid valve replacement).

Survivors. The 26 survivors have been followed up for a mean of 8 ± 1.5 years (range 8 months to 20 years). Twenty-three of the 26 were last seen in the last 12 months and all were seen within the last 3 years. Fourteen survivors are asymptomatic; 12 have developed late complications with supraventricular tachycardia requiring medical therapy in 4 and moderate functional impairment (class II) in 8 (Fig. 4). Of the four surviving patients with ECG evidence of pre-excitation, two have had episodes of palpitation requiring treatment and two have remained asymptomatic.

Risk Factor Identification

Cardiothoracic ratio on chest radiography, presence of pre-excitation by ECG, echocardiographic index, presenting features and presence of associated defects were analyzed as possible predictors for cardiac death (Table 5). Cardiothoracic ratio ≥90%, presence of associated cardiac defects and echocardiographic grade of severity were significantly associated with death in a univariate analysis model.

Multivariate analysis. A multivariate analysis of presenting features as risk factors for cardiac death was performed in 28 cases with echocardiographic severity grading. The chest radiograph variable was not used in the model because all three patients with a cardiothoracic ratio ≥90% also had

Table 5. Risk Factor Identification for 27 Neonates With Ebstein's Anomaly

Factor and Category	Alive	Dead*	Univariate Relative Risk	Multivariate[†] Relative Risk	Multivariate[‡] Relative Risk
Cardiothoracic ratio					
<90%	14	1	†		
≥90%	0	3	7.4 (1.6, 33.6)		
Associated defects					
No	15	9	†		
Yes	12	14	2.3 (1.0, 5.6)	5.0 (1.0, 24.1)	3.9 (0.9, 17.0)
Echocardiographic grade					
1	3	0	†		5.3 (1.9, 15.0) for each grade increase
2	9	1	†		
3	5	4	3.8 (0.7, 20.6)	3.1 (0.6, 17.2)	
4	0	5	16.1 (2.9, 88.5)	28.3 (4.1, 195.7)	
Likelihood ratio statistic				16.6 on 3 df	16.2 on 2 df

*Death from cardiac cause. †Relative risks are the proportionate increases in risk of dying relative to baseline categories. 95% confidence intervals are in parentheses after each relative risk. Echocardiographic grades 1 and 2 were combined as the baseline category because the lack of any deaths in grade 1 made it impossible to estimate the relative risk associated with this grade. Multivariate[†] treats echocardiographic grade as an unordered variable and multivariate[‡] treats it as a continuous variable. The small change in likelihood ratio statistic supports the validity of this approach. df = degrees of freedom.

echocardiographic grade 4. The presence of associated defects had a relative risk of 5.0, echocardiographic grade 3 (compared with grades 1 and 2) a relative risk of 3.1, and grade 4 a relative risk of 28.3. Fitting echocardiographic grade as a continuous variable, adjusting for the presence of associated defects, gave an estimated relative risk of 5.34 for each increase in grade. (The change in the likelihood ratio statistic by treating echocardiographic grade as a continuous variable was only 0.4 for the extra degree of freedom in the unordered model; statistically this finding strongly supports the validity of treating this variable this way).

Discussion

Natural history. With the advent of two-dimensional echocardiography, Ebstein's anomaly is diagnosed more frequently in early life, so that the natural history must now be redefined. Our findings indicate that neonates with this anomaly have both a high early mortality rate and a significant continuing risk of late morbidity or death. These observations contrast with the findings of the only previous report (4), in which 3 of 11 neonates died early but those surviving to 3 months of age had a good long-term outcome.

Early death in our series occurred in children with morphologically severe disease and most of these deaths were due to heart failure. Intrauterine cardiomegaly with consequent pulmonary hypoplasia may have been a contributing factor to the poor outcome in these cases (15). The late and continuing hazard for morbidity and mortality was due to the occurrence of hemodynamic deterioration or sudden death.

Role of echocardiographic grading in defining prognosis. The natural history of Ebstein's anomaly has been obscured by the lack of a system for classifying severity of the lesion. We have therefore classified Ebstein's anomaly into four grades based on an easily measurable echocardiographic ratio: that of the area of the right atrium and atrialized right ventricle compared with the area of the right ventricle, left atrium and left ventricle, measured in a four-chamber apical or subcostal view, at end-diastole. This ratio reflects both the anatomic tricuspid valve displacement (the greater the displacement, the larger the area of the atrialized right ventricle) and the physiologic consequences of tricuspid valve stenosis or regurgitation, or both (the more severe the lesion, the larger the area of the right atrium). Echocardiographic features of severe Ebstein's anomaly in the fetus and neonate have been defined by Roberson and Silverman (4): giant right atrium, distal tethering of the anterior tricuspid valve leaflet, left ventricular compression by the dilated right heart and "right ventricular dysplasia." Not surprisingly, these features coexisted in many of their patients. The distal attachments of the tricuspid valve and features of right ventricular dysplasia may be difficult to assess.

In contrast, our single echocardiographic measure is simple to perform even in the fetal heart and gives important prognostic information, with a 7% incidence rate of cardiac death for grades 1 and 2 compared with 100% for grade 4 in our series. Multivariate analysis confirmed echocardiographic grade of severity as the best independent predictor of mortality; the only other predictor of death was the presence of associated cardiac defects.

Late morbidity and mortality. There was a surprisingly high late hazard for morbidity and mortality. Five neonates whose symptoms had resolved in the 1st month of life later died suddenly. These deaths were not predicted by the presence of pre-excitation and only one of the five infants had any arrhythmia documented previously. More detailed electrophysiologic monitoring may therefore be indicated in patients with Ebstein's anomaly, although this approach has proved disappointing for identifying patients with other congenital heart defects at high risk of sudden death (16,17). The other major late complication was hemodynamic deterioration, with either increasing cyanosis or congestive heart failure, or both. In particular, the presence of late heart failure and severe left ventricular dysfunction presaged a poor outcome; all five patients in whom this occurred died. The association between Ebstein's anomaly and left ventricular abnormalities has been noted previously (18–20).

Left ventricular dysfunction. The cause of left ventricular dysfunction in Ebstein's anomaly is unclear. It has been postulated that chronic cyanosis or chronic right ventricular dilation and septal abnormalities may be responsible (21), but we were unable to identify any presenting features associated with the development of this complication. However, microscopic examination of six of the left ventricles of the neonates who died in the 1st month of life showed markedly increased fibrous tissue content compared with that in a control group, suggesting that damage to the left ventricle may occur very early (22).

Treatment. Many neonates with Ebstein's anomaly improve spontaneously with appropriate medical management. Those who have persistent severe cyanosis or heart failure have either associated cardiac defects or severe Ebstein's anomaly, or both. Palliative surgery for the associated defects can be performed with good results at a risk similar to that of palliative procedures in other complex congenital heart lesions (23).

Treatment is more difficult in those with early clinical deterioration due to severe (echocardiographic grade 3 or 4) disease. In our series young children with Ebstein's anomaly who required tricuspid valve surgery had a very poor outcome. New strategies are therefore required for this age group including consideration of heart transplantation for those with severe symptoms in early life. Encouraging early results were recently reported (24) for conservative surgical treatment consisting of tricuspid valve closure and creation of an aortopulmonary shunt with a view to an eventual Fontan procedure. In older children, tricuspid valve repair (25,26) or replacement (27) can be performed at a much lower risk.

Conclusions. Management of neonates with Ebstein's anomaly may therefore be based on the knowledge of

echocardiographic grade and the presence or absence of associated defects. Most neonates with grade 1 or 2 disease and no associated defect will survive the neonatal period with supportive treatment only and have a good prognosis; those with associated defects may require surgery for these and may also expect a good outcome. Neonates with grade 3 or 4 disease have a much worse outlook; many die in early life, and tricuspid valve surgery may not alter the poor prognosis. Those who do survive the 1st month of life must have careful clinical follow-up, with particular attention to left ventricular function and cardiac rhythm.

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