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Editorial Comment

Ebstein's Anomaly: Natural History And Management*

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The present study. Perhaps no other congenital heart lesion encompasses as broad a spectrum of clinical significance as does Ebstein's malformation. In an excellent study in this issue of the Journal, Celermajer et al. (1) outline the natural history of this malformation in patients whose condition was diagnosed during the neonatal period. Their data indicate that patients with a severe form of this lesion may demonstrate profound congestive heart failure and die early in infancy or even during fetal life. In contrast, patients with a mildly deformed tricuspid valve may remain asymptomatic well into adulthood: one patient from our institution had no symptoms until age 79 and lived until age 85 years. In the majority of patients the malformation lies between these extremes but, as studies like those of Celermajer et al. (1) point out, sudden death is not uncommon even in patients believed to be asymptomatic. Once symptoms develop, the severity of the disability may increase rapidly even in patients who had no significant clinical problems until adolescence or adulthood.

Associated arrhythmia. Clinical symptoms in these patients are caused by disorders of cardiac rhythm or hemodynamic alterations, or both. Paroxysmal supraventricular tachycardia, by far the most frequent rhythm disorder, is thought to occur in approximately 15% of patients. It is often the result of an accessory conduction pathway (Wolff-Parkinson-White syndrome) and, in such instances, successful medical control may be difficult to achieve. Of 179 patients with Ebstein's anomaly treated surgically by Danielson (2) at the Mayo Clinic between 1972 and June 1990, 21 underwent successful division of the accessory conduction pathway as part of their operative treatment. This operative approach merits consideration in any patient whose supraventricular tachycardia is refractory to medical control even if no hemodynamic symptoms are present.

Tricuspid regurgitation. The primary hemodynamic abnormality producing symptoms in Ebstein's malformation is tricuspid regurgitation. Symptoms are generally related to the degree of regurgitation. In addition, if an atrial septal defect is present, right to left shunting through this defect, accentuated by increasing tricuspid regurgitation, may produce additional symptoms as a result of arterial hypoxemia and secondary polycythemia. Until the late 1970s cardiac catheterization was performed in cases of symptomatic Ebstein's deformity. However, with the advent of two-dimensional echocardiography and Doppler technology, we have found invasive study of these patients is rarely necessary (3) and we believe that the pathologic anatomy of the tricuspid valve is better demonstrated with these newer techniques than with angiocardiography.

Prognosis. As Celermajer et al. (1) point out, in previous studies of the natural history of Ebstein's malformation, the majority of patients did not have their condition diagnosed until well after early childhood. These studies therefore contain a bias toward factors favoring survival. The great value of the Great Ormond Street study (1) is that it documents conclusively for the first time the poor prognosis of patients with Ebstein's anomaly who are symptomatic or have significant cardiac enlargement during the neonatal period. The innovative echocardiographic grading scheme they propose as an index to prognosis is a refinement of the previous knowledge that, whether they are treated medically or surgically, patients with a cardiothoracic ratio >0.65 on the standard chest radiograph have a much worse prognosis than do those with a lower ratio (4).

Surgery. Surgical treatment of this deformity has progressed rapidly during the past decade and surgical intervention on the tricuspid valve, with concomitant atrial septal defect closure if necessary, should now be strongly considered for symptomatic patients or even asymptomatic patients if cardiomegaly is increasing significantly. A new surgical approach pioneered by Danielson (4) in the early 1970s, utilizing reconstruction of the patient's natural tricuspid valve, is applicable in many patients and has produced excellent results. The operation involves plication of the free wall of the right ventricle, posterior tricuspid annuloplasty and a reduction in right atrial size. It is based on the concept of creating a competent monocusp valve by utilizing the anterior tricuspid leaflet, which is usually large. For the repair to be successful, this leaflet must be of sufficient size and the free edge cannot be tethered to the endocardial surface. Two-dimensional echocardiography has been highly reliable in demonstrating the pathologic anatomy of the tricuspid valve and predicting whether the anterior leaflet will lend itself to valve reconstruction (5).

Between 1972 and June 1990 at the Mayo Clinic, Danielson (2) performed surgical repair in 179 patients with Ebstein's anomaly. Patient age ranged from 11 months to 64 years. In 72% tricuspid valve reconstruction was possible; in 26% a prosthetic valve, usually a bioprosthesis, was inserted. Plication and a modified Fontan procedure were performed in 2%. There were 12 hospital deaths (6.7%). At follow-up >2 years postoperatively, 92% are in New York Heart Association functional class I or II. There were six

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late deaths, most of them sudden and presumably secondary to a cardiac arrhythmia. Only two patients required reoperation for valve replacement after valve reconstruction (1 and 7 years later, respectively). Exercise testing in patients preoperatively and again late postoperatively has shown a significant improvement in performance. Postoperative echocardiographic and Doppler assessment showed excellent tricuspid valve function in most patients who underwent valve reconstruction.

Indications for surgery. The current low operative mortality rate, the ability to perform plastic reconstruction of the valve in most cases and the encouraging late results now lead us to recommend operation for all patients with Ebstein's anomaly whose condition, despite medical therapy, has deteriorated to functional class III or IV. In addition, because the operative deaths and the few poor late results in the Danielson series occurred in patients with severe preoperative cardiomegaly, we now also recommend operation in less symptomatic patients who exhibit progressive cardiac enlargement. We believe that surgery should be performed before the cardiothoracic ratio is >0.65.

Symptomatic infants with severe cardiomegaly (Celermajer echocardiographic grade 3 and grade 4 [1]) remain a major challenge. The Danielson surgical series is clearly skewed toward older patients; his youngest patient was 11 months old at operation. Although increasing operative experience and refinement of surgical techniques have permitted valve reconstruction to be applied successfully at ever younger ages, infants with an echocardiographic grade 3 or 4 anomaly often have a degree of tricuspid valve and right ventricular dysplasia that will preclude this approach. In such patients, Starnes et al. (6) have advocated conversion to a situation simulating tricuspid atresia, with establishment of an aortic to pulmonary shunt, in the hope that the Fontan approach might be utilized when these patients are older. Because of the devastating natural history documented by Celermajer et al. (1), it seems justified to attempt this approach in such patients, recognizing that it has significant associated risks and uncertainties.

Finally, cardiac transplantation during early infancy remains an option for patients with an echocardiographic grade 3 or 4 lesion. It may ultimately provide the best hope for prolonged survival if present problems relating to donor procurement and late complications including rejection, accelerated coronary atherosclerosis and susceptibility to infection can be overcome.

References

1. Celermajer DS, Cullen S, Sullivan ID, Spiegelhalter DJ, Wyse RKH, Deanfield JE. Outcome in neonates with Ebstein's anomaly. J Am Coll Cardiol 1992;19:1041–6.
2. Danielson GK. Operative treatment of Ebstein's anomaly. J Thorac Cardiovasc Surg (in press).
3. Shiina A, Seward JB, Edwards WD, Hagler DJ, Tajik AJ. Two-dimensional echocardiographic spectrum of Ebstein's anomaly: detailed anatomic assessment. J Am Coll Cardiol 1984;3:356–70.
4. Mair DD, Seward JB, Driscoll DJ, Danielson GK. Surgical repair of Ebstein's anomaly: selection of patients and early and late operative results. Circulation 1985;72:70–6.
5. Shiina A, Seward JB, Tajik AJ, Hagler DJ, Danielson GK. Two-dimensional echocardiographic surgical correlation in Ebstein's anomaly: preoperative determination of patients requiring tricuspid valve plication vs replacement. Circulation 1983;68:534–44.
6. Starnes VA, Pitlick PT, Bernstein D, Griffin MG, Choy M, Shumway NE. Ebstein's anomaly appearing in the neonate: a new surgical approach. J Thorac Cardiovasc Surg 1991;101:1082–7.