EDITORIAL

Management of Ebstein's anomaly

THE EXCELLENT review of Ebstein's anomaly by Tuzcu et al in this issue of the *Cleveland Clinic Journal of Medicine* re-emphasizes the wide spectrum of clinical significance that this lesion encompasses. On one hand the malformation may be so severe as to cause profound congestive heart failure and neonatal, or even intrauterine, death. On the other hand, in a well-documented case of this lesion seen at our institution, the patient had no symptoms until age 79 and lived to age 85.[1] The majority of cases of this deformity lie between these extremes but, as studies like that of Tuzcu et al point out, sudden death is not uncommon even in patients thought to be minimally symptomatic. In patients who become symptomatic, the severity of their disability may increase rapidly even though the onset of significant clinical problems may not have occurred until adolescence or adulthood.

■ *See Tuzcu et al (pp 614–618)*

Clinical symptoms in these patients are caused by disorders of cardiac rhythm, alterations in hemodynamics, or both. Paroxysmal supraventricular tachycardia is by far the most frequent rhythm disorder and is thought to occur in approximately 20% of cases. It is often the result of an accessory conduction pathway (Wolff-Parkinson-White syndrome) and successful medical control in such instances may be very difficult to achieve. Of 122 patients with Ebstein's anomaly treated surgically by Danielson at the Mayo Clinic 16 have undergone successful division of the anomalous conduction pathway as part of their operative treatment. This approach merits strong consideration in any patient whose supraventricular tachycardia is refractory to medical control, even if no hemodynamic symptoms are occurring.

The primary hemodynamic abnormality producing symptoms in Ebstein's malformation is tricuspid insufficiency, which is only rarely accompanied by tricuspid stenosis. Symptoms are generally related to the degree of tricuspid regurgitation. In addition, in our series, as in that of Tuzcu et al, an atrial septal defect was present in the majority of patients; right-to-left shunting through this defect, accentuated by increasing tricuspid insufficiency, 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. Our experience with this, like that reported by Tuzcu et al, also indicated that this could be done at very low risk, which was opposed to published reports in earlier literature. However, with the advent of two-dimensional echocardiography, and more recently Doppler technology, we have found that invasive study of these patients is very rarely necessary and we feel that pathologic anatomy of the tricuspid valve is better demonstrated utilizing these newer techniques than it had been with angiocardiography.[2,3]

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 the symptomatic patient or even the asymptomatic patient if cardiomegaly is increasing significantly.

Early attempts at surgical treatment of this malformation all involved valve replacement and most were unsuccessful. However, in the 1970s a new surgical approach, utilizing reconstruction of the patient's natural valve, was pioneered by Danielson and Fuster[4] and this technique, applicable in approximately 75% of patients, has produced excellent results. The operation involves plication of the free wall of the right ventricle, posterior tricuspid annuloplasty, and reduction in right atrial size. It is based on the concept of establishing a competent monocusp valve by utilizing the anterior tricuspid leaf-

let, 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.[3]

Between January 1972 and March 1987 Danielson at the Mayo Clinic operated upon 122 patients with Ebstein's malformation. Ages ranged from 11 months to 64 years; 77% of the patients were age 10 or older. In 90 patients (74%) valve reconstruction was possible and in 28 (23%) a bioprosthesis was inserted. Four patients (3%) underwent plication and a modified Fontan procedure. There were six operative deaths (4.9%). Follow-up of patients for more than two postoperative years revealed 88% to be in New York Heart Association Class I or Class II. There were two late sudden deaths, presumably secondary to cardiac dysrhythmia. Exercise testing in a limited number of patients preoperatively and again late postoperatively revealed a significant improvement in performance; maximal oxygen consumption increased from a mean of 50% of predicted value before operation to a mean of approximately 80% of predicted value after surgery. Postoperative echo-Doppler assessment in patients having annuloplasty has revealed excellent tricuspid valve function with no or minimal incompetence.

The current low operative mortality, the fact that plastic reconstruction of the valve is possible in approximately three-fourths of cases, and the most encouraging late results now lead us to recommend operation for all patients with Ebstein's anomaly whose condition, despite medical therapy, has deteriorated into New York Heart Association Class III or beyond. In addition, because the operative deaths and the few poor late results in our series have occurred in patients with severe preoperative cardiomegaly, we now also recommend operation in less symptomatic patients who exhibit progressive cardiac enlargement; we feel that surgery should be carried out before the cardiothoracic ratio exceeds 0.65. Because the natural history of this deformity has been established to be poor once patients become significantly symptomatic or exhibit severe cardiomegaly, we feel justified in advocating this aggressive surgical approach, and have been extremely pleased to date at the improvement in quality of life it has brought to the vast majority of these patients.

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