Tricuspid Valve Replacement for Ebstein's Anomaly in Childhood with a Starr-Edwards Caged-Ball Prosthesis*  

21-year Follow-up  

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A 21-year follow-up after tricuspid valve replacement (TVR) with a Starr-Edwards caged-ball prosthesis in a ten-year-old boy is described. TVR is performed for Ebstein's anomaly, with strict indications in childhood. Despite the current preference for bioprostheses, good performance can be expected from the Starr-Edwards caged-ball valve. (Chest 1988; 94:1096-97)

Tricuspid valve replacement for Ebstein's anomaly was first performed in 1962, and variable results were subsequently reported.

CASE REPORT

A four-year-old boy was admitted in 1960 with a heart murmur and history of one cyanotic spell at age six weeks. The heart was not enlarged. The ECG suggested right ventricular hypertrophy. Cardiac catheterization disclosed significant valvular pulmonic stenosis, patent foramen ovale, and trivial tricuspid regurgitation. Being very young and asymptomatic, the patient was discharged and followed. Progressive exercise intolerance and cardiomegaly mandated a second catheterization in 1962, confirming the previous diagnosis but showing increased tricuspid regurgitation with marked right atrial enlargement. Open pulmonic commissurotomy and closure of the patent foramen ovale were performed. Ebstein’s anomaly, noted at surgery, was not corrected because of the small size of the patient and the feeling that relief of the right ventricular obstruction would decrease the amount of tricuspid regurgitation.

Worsening congestive heart failure from massive tricuspid regurgitation mandated a second operation in 1966. Surgery revealed a septal leaflet that was severely malformed, hypoplastic, and displaced into the right ventricle. The atrialized right ventricle was not distended or paradoxic. Attempts at plastic repair were unsuccessful, and the tricuspid valve was replaced with a 3M Starr-Edwards caged-ball prosthesis. The valve was inserted into the true tricuspid annulus below the coronary sinus orifice. Complete heart block ensued with an escape nodal rhythm at a rate of about 50/min. No pacing was necessary postoperatively and the patient became asymptomatic, tolerating heart block well, with regression of cardiomegaly. In 1967, however, a permanent epicardial pacemaker was implanted. From 1968 through 1979, the patient underwent subsequent operations for pacemaker complications, without problems thereafter. He is a very active man who leads a normal life, hunting, fishing, and bicycling, besides working full time. His only medication is warfarin, which is carefully monitored.

DISCUSSION

Surgery for Ebstein’s anomaly of the tricuspid valve has strict indications in childhood. More than 73 percent of children between 1 and 15 years of age are in New York Heart Association class 1 or 2, and many remain so throughout adolescence; in the same age range surgical mortality is very high, approaching 61%. However, progressive heart failure, paradoxic embolism, cyanosis, or other anomalies requiring correction are indications to operate, even in very young children. Since plastic repair of the valve requires a very propitious anatomy to be successful, and this is not frequently the case, tricuspid valve replacement (TVR) is widely performed.

The longest documented follow-up is 19 years and concerns the first patient operated on. The valve implanted was used only at the University of Cape Town, South Africa. The prosthesis most commonly used thereafter has been the Starr-Edwards caged-ball valve, with favorable results at follow-ups as long as 13 years, sometimes even without anticoagulation. Those patients, however, were mostly adolescents or young adults at the time of the implant, whereas overall results in patients under 15 years of age have not been as favorable.

Many surgeons now prefer bioprostheses because of their better performance in the right side of the heart, despite the problems related to their use in children and young adults. Fission of the atrialized segment is controversial. When surgery is indicated, complete correction should be undertaken regardless of age, provided that a good-sized prosthesis can be inserted. Operating before the atrialized segment becomes hyperdistended and paradoxic provides a better outlook, and our case supports this thesis. Since it is the longest reported follow-up after TVR with a mechanical prosthesis, our report may provide helpful information in reevaluating the possible role of mechanical valves in the controversial therapy of such a polymorphic cardiac malformation.

REFERENCES


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Tricuspid Valve Replacement for Ebsteins Anomaly (Di Lello et al)
AeroBid™ (flunisolide)

The BID Inhaler

For oral inhalation only

INDICATIONS

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CONTRAINDICATIONS

AeroBid Inhaler is contraindicated in patients with a history of or known sensitivity to flunisolide or aerosol products. It is also contraindicated in patients with known sensitivity to nebulizers or their component parts. In addition, it is contraindicated in patients with a history of allergic reactions to nebulizers or their component parts.

WARNINGS

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The most common adverse reactions reported with AeroBid Inhaler are cough, throat irritation, and mouth irritation. Other adverse reactions that have been reported include chest tightness, dyspnea, hoarseness, and sinusitis. In rare cases, anaphylaxis, angioedema, and urticaria have been reported.

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