Surgery for Ebstein's Anomaly: The Clinical and Echocardiographic Evaluation of a New Technique

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Ten consecutive patients (age range 4 to 44 years, mean 22) underwent surgical repair of Ebstein's anomaly by vertical plication of the right ventricle and reimplantation of the tricuspid valve leaflets. No patient died during or after operation. Intraoperative postbypass echocardiography documented a good result in nine patients but severe tricuspid regurgitation in one patient, who then underwent prosthetic valve replacement during a second period of cardiopulmonary bypass. Two of four patients who had right ventricular papillary muscle dysfunction in the early postoperative period showed improved papillary muscle function with concomitant reduction of tricuspid regurgitation 6 months later.

All patients were evaluated clinically and by echocardiography 2 to 23 months later. All patients showed clinical improvement, seven by one functional class and three by two classes. All were in sinus rhythm. The mean cardiothoracic ratio decreased by 6% (p < 0.05). On bicycle ergometry performed in six patients, peak oxygen consumption exceeded 20 ml/kg per min in five. Tricuspid regurgitation diminished in eight patients (by three grades in two patients, by two grades in five and by one grade in one patient); it remained unchanged in two. Comparison of preoperative and postoperative pulsed Doppler flow velocities across the pulmonary valve showed an increase in the peak velocity of flow across the valve (mean 83 ± 14 versus 97 ± 11 cm/s, p < 0.005) and a decrease in the time to peak velocity (mean 130 ± 16 versus 91 ± 23 ms, p < 0.05).

The initial evaluation of patients treated by this new operation for Ebstein’s anomaly demonstrates significant clinical improvement in all patients with a reduction in the severity of tricuspid regurgitation and improved forward flow after the creation of a functional right ventricle.

Methods

Study patients. Between January 1988 and February 1990, 10 patients (aged 4 to 44 years, mean 22) underwent surgical repair of Ebstein’s anomaly. Five patients were in New York Heart Association class II and five were in class III at the time of surgery. The reason for surgical correction was progressively increasing disability. Eight patients had associated defects (atrial septal defect in eight patients and ventricular septal defect in one patient). Two patients had
undergone previous surgical closure of a secundum atrial septal defect. The preoperative cardiothoracic ratio ranged from 0.45 to 0.79 (mean 0.62). All patients were in sinus rhythm, but three had recurrent episodes of supraventricular tachycardia requiring drug treatment.

Based on the morphologic characteristics of the tricuspid valve and right ventricle on the preoperative echocardiogram and at operation, patients were classified into one of four grades of increasing severity (A through D), after the method of Carpentier et al. (10). Grade A was characterized by a mobile anterior tricuspid valve leaflet with a small contractile atrialized chamber, grade B by a large, noncontractile atrialized chamber but mobile anterior leaflet, grade C by tethering of the distal attachments of the anterior leaflet associated with a large noncontractile atrialized chamber and grade D by the leaflet tissue forming continuous sac adherent to the right ventricle. Seven patients were in grade B, two were in grade C and one patient was in grade D. All patients gave informed consent for intraoperative echocardiography and for follow-up studies.

Surgical procedure. After institution of cardiopulmonary bypass with moderate hypothermia (28°C), the aorta was cross-clamped and cardioplegic arrest induced. The right atrium was opened parallel to the atrioventricular groove (Fig. 1A). The anterior and posterior tricuspid leaflets were detached in one piece from the tricuspid anulus, starting at the anteroseptal commissure (leaving the attachment of the anterior leaflet at the level of the commissure untouched), and proceeding along the anulus until the posterior leaflet was completely detached (Fig. 1B). Mobilization of the detached leaflets was achieved by cutting the abnormal fibrous bands between the leaflets and the ventricular wall, and dissecting the papillary muscle attachments from the ventricular wall as far as the apical part of the right ventricle.

The atrialized right ventricle was plicated longitudinally with continuous 5-0 Prolene (Fig. 1C). One line of suture insertion points ran approximately along a line from the apex of the atrialized portion of the right ventricle to the coronary

Figure 1. The operative technique, modelled after Carpentier et al. A, Surgeon’s view after opening the right atrium. a = anterior leaflet of the tricuspid valve; ac = atrialized ventricular chamber; p = posterior leaflet. B, Detachment of the anterior and posterior tricuspid valve leaflets and their chordal attachments to the ventricular wall. The dashed lines denote the suture insertion points. C, Longitudinal plication of the atrialized portion of the right ventricle. D, Clockwise spreadout of the anterior and posterior leaflets on the newly created tricuspid valve anulus, and direct closure of the atrial septal defects without right atrial reduction.
sinus and included the septal leaflet attachment. The septal leaflet or its remnants were left untouched. The second line of suture insertion points was started at the distal end (toward the apex of the ventricle) of the first line, and ascended along the diaphragmatic endocardial surface of the atrialized chamber to include the original attachments of the posterior leaflet. Care was taken not to place the suture lines transmurally, to avoid the posterior descending coronary artery. The atrial plication was completed caudal to the coronary sinus. The anterior and posterior leaflets were reattached to the newly created functional tricuspid anulus using a clockwise rotation to cover the circumference of the orifice (Fig. 1D). No prosthetic rings were used to reinforce the anulus. During cardiopulmonary bypass, the competence of the tricuspid valve was checked by injection of saline solution into the right ventricle. After completion of additional procedures (closure of atrial or ventricular septal defect) the right atrium was closed.

**Intraoperative echocardiography.** After median sternotomy, and before initiation of cardiopulmonary bypass, all patients underwent epicardial echocardiography using 5-MHz and 3.75-MHz probes connected to a Toshiba SSH 160A ultrasound system. With the transducer placed directly on the right atrial or right ventricular surface and appropriate transducer angulation, long-axis and short-axis views, a modified four chamber view and right ventricular inlet view of the heart were obtained (12). The proximal attachments of all three tricuspid leaflets were sought, and their mobility was assessed. Tricuspid regurgitation was assessed with color flow mapping (see later) using the 3.75 MHz transducer, and pulsed Doppler recordings of flow in the pulmonary artery were obtained. At the end of the procedure, after cardiopulmonary bypass had been discontinued, the quality of the repair was assessed with color flow mapping of the atrial septum and tricuspid valve. A pulsed Doppler recording of forward flow in the pulmonary artery was also obtained. All epicardial echocardiographic studies were recorded on videotape for subsequent analysis.

**Follow-up clinical assessment.** All patients were evaluated clinically over a period of 2 to 23 months (mean 11.7). Six of the 10 patients (aged 13 to 35 years) underwent symptom-limited exercise testing by upright bicycle ergometry (mean interval from operation 10 months), using a standard protocol with 20 W increments of the work load every 2 min and a constant pedalling speed of 60 rpm. During exercise the heart rate, blood pressure and 12 lead electrocardiogram (ECG) were monitored continuously, and minute ventilation and oxygen uptake were derived at 60 s intervals using an Oxycon 4 gas analyzer.

**Follow-up echocardiography.** Five of the 10 patients were examined within 1 week of surgery; subsequently serial follow-up two-dimensional and Doppler echocardiographic studies were performed in all patients. Pulsed Doppler sampling of flow within the superior vena cava, hepatic veins and across the tricuspid and pulmonary valves was performed, using a combination of subcostal, precordial and suprasternal transducer positions. Four patients (age range 13 to 34 years) were also studied by outpatient transesophageal echocardiography using a standard transesophageal probe connected to a Toshiba SSH 160A or Hewlett-Packard Sonos 1000 ultrasound system.

**Data analysis.** The peak Doppler velocity of tricuspid inflow, the peak systolic velocity across the pulmonary valve and the time to this peak velocity and the presence of systolic flow reversal in the hepatic veins were documented. These data were compared with similar data from preoperative studies, which were available in seven patients. Tricuspid regurgitation (before, during and after operation) was graded from the color flow map in the apical or parasternal four chamber view into one of four grades, using a modification of the method described by Omoto and Suzuki et al. (13,14): grade 1—trivial regurgitation, extending to just above the anulus of the tricuspid valve (as defined by the attachment of the anterior leaflet to the ventricular wall in the four chamber plane of the preoperative echocardiograms); grade 2—the regurgitant jet extended to less than half the length of the right atrium; grade 3—the jet reached half the length of the right atrium; and grade 4—the jet extended to the roof of the atrium.

**Statistics.** Statistical analysis of changes in functional class after surgery and in grades of tricuspid regurgitation was performed using a Wilcoxon rank test. Preoperative and postoperative cardiothoracic ratios on chest X-ray film and pulsed Doppler data were compared with use of paired t tests.

**Results**

**Intraoperative echocardiography.** Before cardiopulmonary bypass, the anterior leaflet of the tricuspid valve was enlarged in all 10 patients and appeared freely mobile. The septal leaflet was displaced to a varying degree (1.8 to 4.8 cm from the tricuspid anulus and tethered to the septum in nine patients; it was absent in one patient). Although the posterior leaflet was seen, its attachment to the ventricular wall could not be definitely identified. Patient 9, who had grade D Ebstein’s malformation and subsequently required valve replacement, the body of the anterior leaflet had appeared mobile on prior precordial and intraoperative echocardiographic studies. At the time of operation, all leaflets were tethered to the ventricular wall, and there was a pinhole orifice between the atrialized portion and the functional right ventricle. Retrospective analysis of the intraoperative recording confirmed tethering of the edges of all leaflets, with only the midportion of the anterior leaflet being mobile.

**Color flow imaging** did not suggest tricuspid stenosis in any patient, although three patients had a pattern of turbulent inflow within the right ventricular cavity due to intraventricular obstruction to flow by papillary muscle or chordae. Tricuspid regurgitation was present in all patients. It was grade 4 in five patients with a turbulent regurgitant jet,
Table 1. Comparison of Tricuspid Regurgitation Grades Before and After Surgery

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*Decrease in tricuspid regurgitation on serial studies with improved leaflet coaptation; † = no decrease of tricuspid regurgitation on serial studies. Patient 9 had tricuspid valve replacement. Intraop = intraoperative; No grading = grading not possible because of poor image quality after cardiopulmonary bypass; Preop = preoperative.

grade 3 in four patients and grade 1 in one patient with a low velocity laminar regurgitation.

On postbypass echocardiography, the orifice of the valve was displaced medially toward the ventricular septum in nine patients in whom a plastic repair was possible. One patient had grade 3 tricuspid regurgitation, three patients had grade 2, and four had grade 1. No regurgitation was detected in one patient, and residual regurgitation could not be assessed in one patient because of poor image quality. These findings correlated well with late follow-up studies (Table 1). Three patients had diastolic inflow turbulence across the tricuspid valve. In one patient with type D disease (Patient 9), in whom repair of the tricuspid valve was attempted, the initial postbypass study showed a prolapsing anterior leaflet associated with severe tricuspid regurgitation. This was confirmed by pressure measurements (right atrial systolic pressure 20 mm Hg); the valve was therefore excised and a 23 mm St. Jude prosthetic valve was inserted during a second period of cardiopulmonary bypass.

Postoperative clinical status. After surgery, eight patients were in functional class I and two patients in class II. Seven patients had improvement by one functional class and three patients by two classes (p < 0.05) (Fig. 2). On auscultation, 5 of the 10 patients had a soft systolic murmur of tricuspid regurgitation. All patients were in sinus rhythm, and a new right bundle branch block pattern was evident on the ECG in three patients. Two of the three patients with Wolff-Parkinson-White syndrome and documented supraventricular tachycardia preoperatively had brief episodes of tachyarrhythmia on postoperative ambulatory ECG (Holter) monitoring (but without symptoms). The cardiothoracic ratio on chest X-ray film had decreased for the whole group (mean 0.62 ± 0.11 preoperatively versus 0.57 ± 0.08 postoperatively, p < 0.05). Although two patients were in functional class II postoperatively, both had a reduced cardiothoracic ratio after surgery (from 0.74 to 0.6 and 0.68 to 0.64, respectively).

Follow-up echocardiography. In two of five patients examined within 1 week of surgery, a mass was noted in the apical and lateral portion of the right ventricle (Fig. 3), but this was not found on subsequent examination 6 months later. In four of the five patients, the initial postoperative echocardiogram showed absent coaptation of the tricuspid valve leaflets and an immobile right ventricular papillary muscle. This was associated with grade 3 (three patients) or grade 2 (one patient) tricuspid regurgitation. At restudy, leaflet coaptation had normalized in two patients (Fig. 4) in association with a recovery of papillary muscle function and reduction of tricuspid regurgitation (from grade 3 to grade 1 in both patients). In the other two, incomplete leaflet coaptation and poor papillary muscle function persisted, and no reduction in tricuspid regurgitation was seen.

Figure 2. Comparison of the preoperative (pre) and postoperative (post) functional class of all 10 patients in this series. NYHA = New York Heart Association.

Figure 3. Precordial echocardiographic four-chamber view in a patient 4 days after surgical repair, showing a mass within the right ventricle (arrow). The mass could not be detected on subsequent follow-up examination 6 months later. la = left atrium; lv = left ventricle; ra = right atrium.
Figure 4. Apical four chamber view (systolic frame) from a patient after surgical repair. Recovery of papillary muscle function was associated with good coaptation of the tricuspid valve leaflets (arrows). RV = right ventricle; other abbreviations as in Figure 3.

At the last follow-up echocardiographic examination, the right ventricle appeared dilated in five patients; paradoxic septal motion noted preoperatively remained unchanged in two patients, but had resolved in another patient after operation. In the five other patients the right ventricle was comparable in size to the left ventricle. Residual tricuspid regurgitation was grade 3 in two patients, grade 2 in one patient and grade 1 in seven patients (Fig. 5). The appearance of the regurgitant jet on color flow imaging also changed. In nine patients, the regurgitant jet appeared turbulent (compared with a laminar appearance preoperatively in five patients).

In four patients outpatient transesophageal echocardiography was performed immediately after precordial echocardiography. It enabled a more accurate grading of tricuspid regurgitation. Grade 1 regurgitation was detected by transesophageal color flow imaging in two patients, in whom precordial echocardiography failed to detect regurgitation. In two other patients with grade 3 and grade 1 regurgitation, respectively, there was no difference in grading by either imaging technique. A better visualization of the plication, of leaflet mobility and coaptation and of the medially shifted tricuspid valve orifice was achieved by transesophageal imaging in two patients (Fig. 6).

Comparison of preoperative and postoperative pulsed Doppler data revealed significant changes in peak velocity of tricuspid inflow (mean 68 ± 14 versus 103 ± 15 cm/s, p < 0.001), peak systolic velocity of flow (mean 83 ± 14 versus 97 ± 11 cm/s, p < 0.005) and time to peak velocity in the pulmonary artery (mean 130 ± 16 versus 91 ± 23 ms, p < 0.05). In one patient, a biphasic pattern of forward flow in the pulmonary artery was seen on the initial study within 1 week of surgery, with the first peak coinciding with atrial systole. The early peak disappeared on subsequent follow-up. Systolic flow reversal in the hepatic vein was recorded in only one patient (with grade 3 regurgitation on color flow imaging) postoperatively, compared with six patients preoperatively.

Exercise testing. Five patients in functional class I and one patient in class II underwent upright bicycle ergometry. The end point was fatigue in five patients (all in class I) and dyspnea in one patient. Exercise duration varied from 3 min 56 s (one patient in class II) to 8 min 13 s, and peak work capacity from 80 to 180 W (mean 82% of predicted). Maximal oxygen uptake exceeded 20 ml/kg per min in the five patients in class I (range 20 to 27.7 ml/kg per min) and was 14.8 ml/kg per min in the patient in class II. Apart from occasional
bigeminy in one patient, which became less frequent with exercise, there were no arrhythmias or pathologic ST segment changes.

**Discussion**

**Previous surgical results.** After the 1st year of life, a large proportion of patients with Ebstein's anomaly are relatively free of symptoms for long periods. There is, however, a significant mortality rate in later years, which is related to the development of congestive heart failure or cardiac arrhythmias (15,16). In addition, when compared with normal persons, unoperated patients have a limited exercise tolerance and reduced work capacity, exercise time and peak oxygen consumption (17). Thus, surgery may be beneficial in some patients. The results of operations for Ebstein's anomaly have improved since repair of the tricuspid valve as reported by Danielson et al. (8,9) was performed in preference to valve replacement. The survivors of corrective surgery show a definite improvement in clinical status and exercise tolerance, even if they were in functional class I or II preoperatively (3,18). Because the malformation is relatively rare, and previous surgery gave poor results, few large surgical series have been reported. The present series of 10 patients is the largest yet reported of patients who have been treated with the new operation first described by Carpentier et al. (10,11).

**Surgical technique.** The technique used differs from that described by Danielson et al. (8,9) in some important ways. First, the atrialized portion of the right ventricle is plicated vertically, to restore the height of the right ventricle. The tricuspid valve anulus is therefore restored to its appropriate position at the AV junction. Second, a bileaflet valve is created, whereas in the Danielson technique the anterior leaflet functions as a monocusp valve. Finally, because the atrialized portion of the right ventricle is effectively incorporated into the functional right ventricle, no additional atrial reduction procedure is required. Although our technique was modeled on that described by Carpentier et al. (10), several modifications of their procedure were undertaken. Thus, the reconstructed tricuspid anulus was not reinforced with a prosthetic ring in any patient. Transection and reimplantation of the papillary muscles of the tricuspid valve were also not required, and mobilization of the anterior and posterior leaflets was achieved in all cases by freeing the attachments of these leaflets to the ventricular walls.

Although a murmur of tricuspid regurgitation could be heard in five patients postoperatively, regurgitation had been reduced by at least two grades in seven of the 10 patients. Although direct comparisons with preoperative exercise tolerance or maximal oxygen consumption could not be made, comparison with reported values in adult patients suggested good results. Five of six patients who underwent bicycle ergometry postoperatively were categorized into functional class I on the basis of maximal oxygen consumption, and one patient was considered to be in class II (19,20).

**Echocardiography.** Intraoperative echocardiography was a valuable technique for the assessment of surgical repair, and residual tricuspid regurgitation graded after cardiopulmonary bypass correlated well with subsequent follow-up echocardiography. On serial precordial ultrasound examination, several findings of interest were noted. Two patients had the echocardiographic appearances of a mass in the right ventricular cavity, which disappeared without intervention. Although neither patient had clinical episodes compatible with pulmonary embolism, in retrospect these masses may have been right ventricular thrombi related to dissection of the papillary muscles and chordae during mobilization of the tricuspid valve leaflet and exposure of the myocardium. Patients who require such extensive dissection may require anticoagulant therapy. These patients and two other patients also had poor coaptation of the valve leaflets associated with evidence of papillary muscle dysfunction. This impairment of papillary muscle function in the early postoperative period may have been the result of ischemic damage, with subsequent recovery of function and improved leaflet coaptation in two patients. In the other two, papillary muscle dysfunction and incomplete leaflet coaptation persisted, with no change in tricuspid regurgitation. The peak velocity of tricuspid inflow increased after surgery, reflecting a smaller valve orifice size. The character of residual tricuspid regurgitation on Doppler color flow mapping also changed and the regurgitant jet appeared turbulent, suggesting a higher peak velocity of regurgitation.

An important reason for the surgical modifications proposed by Carpentier et al. (10) is restoration of normal volume and shape of the right ventricle by longitudinal plication of the atrialized chamber. Right ventricular function, however, was not evaluated in their series (11). In a study (21) of right ventricular performance after intra-atrial repair procedures for transposition of the great arteries, flow acceleration variables in the aorta correlated well with right ventricular ejection fraction. In that series, pulsed Doppler recordings of forward flow in the pulmonary artery revealed two important differences from preoperative findings: the peak velocity of systolic flow increased and the time to peak velocity decreased significantly. These changes were observed, although to a lesser degree, in our Patient 6, who had persistent paradoxical septal motion after surgical repair. These changes in pulmonary artery flow may reflect improved systolic performance of the right ventricle after surgery. Both indexes are relatively easily obtained, and serial measurement may enable monitoring of changes in ventricular function. Patient 6 also had a biphasic pattern of pulmonary artery flow (presumably the result of impaired diastolic compliance of the right ventricle) in the early postoperative period. The early peak, which coincided with atrial systole, disappeared on serial follow-up.

Although all patients were in sinus rhythm at follow-up, two of three patients with supraventricular tachycardia requiring drug treatment preoperatively had brief episodes of tachyarrhythmia postoperatively. They had not been studied.
electrophysiologically and no attempt had been made to ablate an accessory pathway, if present in the series of patients reported on by Oh et al. (22), a large proportion with supraventricular tachycardia had recurrent symptomatic tachyarrhythmias after surgery. Intraoperative electrophysiologic mapping and ablation of the accessory pathway, if it exists, may therefore be required in some patients.

**Conclusion.** Follow-up of patients treated by this new operative technique for Ebstein's anomaly shows significant clinical improvement in all patients, with a reduction in the severity of tricuspid regurgitation and improved forward flow after the creation of a functional right ventricle. Both intraoperative epicardial and follow-up precordial echocardiography were useful in assessing the results of surgery and documenting changes in tricuspid regurgitation and right ventricular function. Longer follow-up of these patients and a larger surgical experience are required before elective surgery for patients with milder forms of Ebstein's anomaly can be recommended.

**References**