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Outcome in Patients Undergoing Glenn Operation in Association with Tricuspid Valve Repair for Ebstein Anomalyu and Failing Right Ventricle

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Objectives:

Repair of **Ebstein** anomaly and impaired right ventricular function pose challenges for the cardiac surgeon especially in adult patients. We reviewed our experience with the 1.5-ventricle repair in this patient population.

Materials and Methods:

Between 2000 and 2012, 10 patients underwent 1.5 ventricle repair for **Ebstein** anomaly. All of them had a bidirectional cavopulmonary shunt constructed. The median age at operation was 15 years (55 months-27.8 years). All of the patients had severe **Ebstein** anomaly with dilated right-sided chambers and/or right ventricular dysfunction. The mean left ventricular ejection fraction was 55%

Results:

Procedures included bidirectional cavopulmonary shunting (10), tricuspid valve replacement (1), tricuspid valve repair (9), and right ventricular plication according to Danielson 4 and according to Carpentier in 6 patients, ASD closure in 3. Shunting was planned preoperatively in 8 patients; the indication in 2 other patients was hemodynamic instability after separation from cardiopulmonary bypass. The azygos vein was left open in all patients to prevent severe postoperative central venous hypertension. One patient underwent Fontan operation 24 hours after the first procedure. This patient died during the second procedure due to low cardiac output. At follow-up, tricuspid incompetence of 8 survived patients with bidirectional cavopulmonary shunt undergoing tricuspid repair was mild in 6 and moderate in 2. One of the patient undergoing repair at 19 years old, became a mother 3 years later, with an excellent pregnancy period.

Conclusions:

The 1.5-ventricle repair in association with tricuspid valve repair can be employed with excellent outcome in patients with severe **Ebstein** anomaly and impaired right ventricular function who are at high risk for surgical treatment. We believe the bidirectional cavopulmonary shunt may be considered as a planned procedure, as an intraoperative salvage maneuver, or as an alternative to cardiac transplantation in selected patients.