Early results after cone reconstruction of the tricuspid valve in Ebstein’s anomaly.

Schrempf J., Sames-Dolzer E., Kreuzer M., Benedikt P., Gitter R., Grohmann E., Tulzer G., Mair R. 
Children’s Heart Center Linz, Austria

Objectives: Patients with Ebstein’s anomaly often suffer from severe tricuspid regurgitation, right ventricular dysfunction or rhythm disturbances. Our study shall evaluate early results after cone reconstruction of the tricuspid valve in pediatric and adult Ebstein patients.

Methods: Our single center retrospective study reports 16 patients with Ebstein’s anomaly corrected by cone reconstruction since 2012. Four of them were infants including two neonates; median patient age was 5.79 years (ranging from 5 days until 56.34 years). Tricuspid reconstruction was done according to da Silva’s method including the septal leaflet in the repair. A longitudinal ventricular plication and an annular plication were done as reported by Carpentier. A 4 mm patent foramen ovale was left open.

Three patients also had a pulmonary atresia, additionally treated with an RVPAC and a modified Blalock-Taussig shunt. The cone procedure was done primarily in one of these patients and at time of conduit change in the other 2 patients.

Results: Both neonates needed ECMO therapy postoperatively, but could be weaned successfully. Unfortunately one of them died despite good cardiac function on the fourteenth postoperative day because of massive cerebral hemorrhage. The other patients are doing well. Postoperative ultrasound showed none or minimal tricuspid insufficiency in 12 patients and moderate tricuspid valve insufficiency in 4 patients. There was no need for reoperation because of tricuspid valve insufficiency or stenosis during follow up. Postoperative rhythm disturbances occurred in 7 cases (4 patients showed episodes of atrial tachycardia, 3 had a temporary AV block grade III without need for pacemaker implantation).

Conclusions: The cone reconstruction can achieve excellent tricuspid valve reconstruction with good right ventricular function. Our study shows a promising option for symptomatic infants with severe Mb. Ebstein. Rhythm disturbances occurred usually self-limiting, but should be treated thoroughly to avoid negative effects on cardiac output.