

Ebstein`s anomaly: Rhythm disturbances after modified reconstruction of tricuspid valve without ventricle plication

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Introduction: Ebstein`s anomaly (EA) is a rare congenital heart defect of the tricuspid valve in which the hinges of the septal and/or posterior leaflets are displaced downward to the right ventricle. The anterior leaflet is usually not displaced but is enlarged and sail-like and valve closure is likewise displaced downwards.

Methods: Since 1988 we have operated patients with Ebstein`s anomaly (EA) using a modified repair technique of the tricuspid valve. This technique reconstructs the valve mechanism at the level of the true annulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber. Additional attachment of the anterior right ventricle wall to the interventricular septum ("Sebening stich") and if possible, reconstruction of a tricuspid valve as a double orifice valve was performed in a modified technique since 2004.

Preoperative, perioperative and postoperative arrhythmias in 57 consecutive patients who underwent operation for EA were reviewed. There were 22 male and 35 female patients (median age 22 years, range 3 months to 68 years. Median follow-up period was 33 months (range 1 month to 22 years).

Results: Early mortality was 7.1%, late mortality 5.2%, since 2004 no patient died. Re-operation was necessary in 2.3%. Preoperative rhythm disturbances were present in 54% of all patients [n=4 with WPW-syndrome, n=6 with bradycardia and n=21 with supraventricular tachycardia (SVT)]. 35% had intraventricular conduction system disturbances [n=13 with incomplete right bundle branch block (RBBB), n=7 with complete RBBB and no complete atrioventricular block].

Perioperative SVT was reduced from 37% to 12%, and late follow-up demonstrated an incidence of 14% for SVT. Bradycardia was reduced from 11% preoperatively to 3.5% in the late follow-up. No WPW syndrome was documented after the operation. Postoperative complete RBBB increased from 12% to 32% and complete atrioventricular block was observed in 22%. Analysis of the type of severity of EA (due to the Carpentier classification) seems to demonstrate increasing rhythm disturbances in more severe forms of EA.

Conclusions: Rhythm disturbances in patients with EA are common. Reconstruction of the tricuspid valve even without ventricle plication seems to minimize the incidence of supraventricular tachycardia, the incidence of postoperative complete atrioventricular block was significant.