The long-term impact of various techniques for tricuspid repair in Ebstein's anomaly

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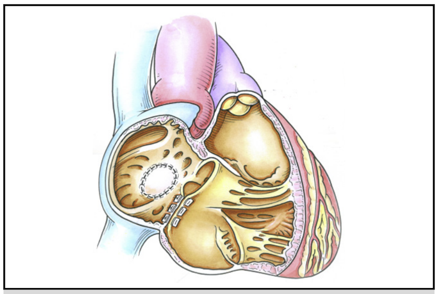
ABSTRACT

Objective: We describe a repertoire of repair techniques according to type of Ebstein's anomaly to correct tricuspid valve (TV) incompetence, and report long-term ventricular function and functional outcomes.

Methods: Sixty-eight patients (mean age, 26.9 ± 7.3 years) with Ebstein's anomaly (type A, n = 21; type B, n = 23; type C, n = 15; type D, n = 9) underwent correction of TV incompetence under normothermic cardiopulmonary bypass. The atrialized ventricle, TV, and subvalvar apparatus were inspected to analyze the precise morphology and determine which leaflet was the most mobile. Various repair strategies (anterior and/or posterior annulorrhapy, Sebening stitich, double-orifice valve technique, with bidirectional Glenn anastomosis if necessary) were used according to the presenting morphology and applied according to the type of Ebstein's anomaly. In all, the atrialized right ventricle (RV) was incorporated into the contractile RV by partial closure of the natural annulus using the most mobile leaflet for valve competence.

Results: The mean duration of follow-up was 13.25 ± 1.3 years (median, 9.34 years; range, 1-24 years). The mean New York Heart Association class improved from 3.4 to 1.3 ($P < .001$). The mean severity of TV incompetence was reduced from 3.2 to 1.3 ($P < .001$). Exercise tolerance tests demonstrated improved maximal oxygen uptake from a mean of 15 ± 7.8 ng/kg/min preoperatively to a mean of 24.9 ± 2.0 ng/kg/min postoperatively ($P < .02$). Displacement tissue Doppler imaging was used to evaluate overall cardiac performance of the RV and left ventricle and interventricular septum. The mean basal, middle, and apical ventricular strain improved significantly from preoperative values of 18.08%, 15.6%, and 13.9%, respectively to postoperative values of 25.7% ($P < .011$), 23.7% ($P < .001$), and 19.36% ($P < .05$), respectively. Freedom from reoperation was 100% at 1 year, 98.3% at 5 years, and 92.9% at 20 years. Early mortality was 2.9%, and late mortality was 5.8%. The overall survival rate was 97.6% at 30 days, 92.7% at 5 years, and 91.26% at 20 years.

Conclusions: The various repair techniques, all of which preserve the atrialized chamber and are used individually according to morphology, provide satisfactory long-term ventricular function and functional outcome even in severe types of Ebstein's anomaly. (J Thorac Cardiovasc Surg 2015;150:1212-9)



Posterior annulorrhaphy at the level of the true tricuspid annulus in Ebstein's anomaly.

Central Message

A repertoire of techniques with an atrialized chamber incorporated into the right ventricle provide satisfactory ventricular function.

Perspective

All types of Ebstein's anomaly are amenable to repair. The repertoire of techniques that incorporate the atrialized chamber, elevate the level of valve closure to the natural annulus, and use the most mobile leaflet achieve valve competence in all types. The satisfactory long-term results suggest that Ebstein's anomaly may be corrected early without undue risks to avoid its natural course of further deterioration.

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Ebstein's anomaly[1] is a congenital defect of the tricuspid valve (TV), in which the origins of the variably deformed septal or posterior leaflets, or both, are displaced downward into the right ventricle (RV), dividing it into a proximal atrialized and distal ventricularized (true or functional ventricle) chambers.[2] These features, in addition to annular dilatation, result in varying degrees of valve incompetence, which exacerbates the abnormalities in structure and function of the RV[3] and less commonly, those of the left ventricle (LV).[4] Pulmonary hypoplasia,[5] a feature that

Abbreviations and Acronyms

ET	= Ventricular ejection
FEV	= Forced expiratory volume
FVC	= Forced vital capacity
IVCT	= Isovolumetric contraction
IVRT	= Isovolumetric relaxation
IVS	= Interventricular septum
LV	= Left ventricle/ventricular
NYHA	= New York Heart Association
RV	= Right ventricle/ventricular
TDI	= Tissue Doppler imaging
TV	= Tricuspid valve
VO_{2max}	= Maximal oxygen consumption

contributes to neonatal death as well as rhythm disturbances,[6] are correlated with the degree of resultant tricuspid regurgitation. If left untreated, heart failure may eventually occur.[7] Thus, numerous techniques,[8-19] both anatomical and functional, have evolved in an attempt to correct the TV incompetence in Ebstein's anomaly and offset the symptoms. Most of these techniques are based on the plication of the tricuspid annulus associated or not associated with plication of the atrialized chamber. Despite reported good mid-term and long-term results of these techniques, the incidence of residual or recurrent TV insufficiency increases and the need for reoperation persists for many patients. Presently, no report exists defining which techniques are suitable for each type of Ebstein's anomaly.

Here we describe a repertoire of repair techniques according to type of Ebstein's anomaly to correct the TV incompetence, and report the long-term ventricular function and functional outcomes in patients undergoing these operations.

PATIENTS AND METHODS

Our hospital's Institutional Review Board approved this retrospective/prospective study and waived the need for patient consent. Between June 1986 and December 2012, 68 patients (mean age, 26.9 ± 7.3 years) with Ebstein's anomaly underwent correction of TV incompetence (Table 1). The TV anatomy was categorized according to Carpentier's classification scheme[12] as type A, B, C, or D:

(1) Type A: a small, contractile atrialized chamber with a large, mobile anterior leaflet and the septal and posterior leaflet origins only moderately displaced into the RV; n = 21 (mean age, 22.89 ± 18.86 years; median, 18.07 years; range, 9 months to 64.8 years)
(2) Type B: large, noncontractile atrialized chamber with a large and mobile anterior leaflet; n = 23 (mean age, 32.04 ± 17.2 years; median, 33.49 years; range, 9.03-59.29 years)
(3) Type C: restricted motion of anterior leaflet, which adheres to the endocardium of the RV wall by fibrous bands or abnormal chordae tendinae; n = 15 (mean age, 32.84 ± 22.71 years; median, 38.57 years; range, 7.9 months to 67.45 years)
(4) Type D: "tricuspid sac" leaflet tissue forming a continuous sac adherent to a dilated RV; n = 9 (mean age, 22.98 ± 19.43 years; median, 17.46 years; range, 2.8 months to 48.03 years).

Cyanosis, defined by a resting systemic oxygen saturation <92% (range, 76%-92%), was present in 33 patients (48.5%). Rhythm disturbances were exhibited by 25 patients (36.7%). Cardiomegaly (cardiothoracic ratio >65%) was found in 51 patients (75%). All patients age 10 years and older (n = 38) were subjected to exercise tolerance tests using the Jones protocol[20] both perioperatively and during regular follow-ups. RV and LV function was evaluated in 19 patients based on standard Doppler echocardiography; since 2000, tissue Doppler imaging (TDI) has been used in all patients, including the surviving 13 patients from the early years, for perioperative and follow-up assessment of ventricular function and myocardial performance.

Functional Evaluation

We used the Jones protocol[20] for exercise tolerance testing. This is a symptom-limited exercise test with a stepwise increase in workload of 16 W/min, starting with unloaded cycling plus the ergometer-related permanent load. Cuff blood pressure measurements and complete 12-lead electrocardiography readings were obtained at 2- to 3-minute intervals during exercise, at peak exercise, and at 1, 3, and 5 minutes after exercise. Expiratory gas analysis was performed using the $CardiO_2$ (MCG Diagnostics, St. Paul, Minn) exercise testing system. Maximal oxygen consumption (VO_{2max}) was defined as the highest VO_2 value detected during the test. Pulse oximetry oxygen saturation was monitored throughout the study. Immediately before each exercise test, spirometric measurements of the patient's forced vital capacity (FVC) and volume of air exhaled in the first second of forced expiration (FEV_1) were also obtained. The time-related change in VO_{2max} was the primary functional outcome for this study.

Echocardiography and TDI

Transthoracic echocardiography was performed with the patient in a left lateral recumbent position, using a transducer individually optimized to body weight (5.0-2.5 MHz) and interfaced with a Vivimed 7 system (GE Healthcare VingMed, Horten, Norway). For each parameter, at least 3 consecutive cardiac cycles were recorded during the respiratory idle state, stored in digital format (Magneto Optical Disc; Sony, Tokyo, Japan), and transferred to a computer workstation for offline analysis with a dedicated software (EchoPac; GE Healthcare). TDI was used to evaluate annular velocity and myocardial strain. Longitudinal performance was evaluated in a standard apical 4-chamber view. Radial function was assessed in a parasternal short-axis view at the LV posterior wall.[21] Two-dimensional and color-sector sizes were minimized, and depth was chosen to achieve the highest frame rate (150-350 frames/second). The angle between myocardial motion and the ultrasound beam was kept at <30°; therefore, each wall—even each segment of interest, if necessary—was recorded separately to obtain an optimal parallel angulation. Gain settings, filters, and pulse repetition frequency (0.16-0.28 per second) were adjusted to optimize color saturation and to avoid aliasing. Peak mean annular velocity profiles were derived at the level of the mitral valve annulus at the interventricular septum (IVS) and LV. Isovolumetric acceleration was derived at the left lateral wall. Isovolumetric contraction (IVCT) and isovolumetric relaxation (IVRT), as well as ventricular ejection (ET) times, were measured in a curved m-mode at the IVS. The Tei index was calculated as TDI-Tei = (IVCT + IVRT)/ET.[22] The longitudinal strain of the LV lateral wall and IVS were examined at their basal, middle, and apical segments, and the mean LV and IVS strain was calculated. Radial LV strain was examined at the LV posterior wall at the level of the papillary muscles in a parasternal short-axis view. A pixel resolution of 6 × 8 was applied, along with a computation distance of 9.7 mm in the longitudinal direction, for a radial strain of 4.7 mm. The angle of the region of interest was adjusted to be parallel to the major direction of myocardial motion.

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