Cone reconstruction for Ebstein's anomaly: Patient outcomes, biventricular function, and cardiopulmonary exercise capacity

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Objective: Cone reconstruction is advocated to treat severe tricuspid valve (TV) regurgitation associated with Ebstein's anomaly. Data on postoperative clinical status, ventricular adaptation, and objective cardiopulmonary testing are lacking in these patients.

Methods: The clinical characteristics, echocardiography, magnetic resonance imaging, and exercise data from 27 consecutive cone reconstructions, undertaken from 2009 to 2013, were retrospectively compared between preoperative baseline and follow-up.

Results: There were no deaths. The cone TV functioned well in all but 1 patient with late dehiscence of inferior annuloplasty sutures that were subsequently repaired. Four patients required pacemaker insertion (3 for new complete heart block). At median follow-up of 2.7 ± 1.5 years, tricuspid regurgitation was reduced in all patients, without causing stenosis. Global left ventricle function remained unchanged (pre-operative fraction 60% ± 4% vs postoperative fraction 61% ± 3%; $P = .96$). MRI showed enhanced forward pulmonary flow (pre 26 ± 1 mL/beat vs post 36 ± 10 mL/beat; $P < .005$) and increased left ventricle filling (body surface area-indexed left ventricle end-diastolic volume pre 49 ± 14 mL/m^2 vs post 60 ± 14 mL/m^2; $P < .005$). New York Heart Association functional class improved (pre 2.5 ± 0.6 vs post 1.3 ± 0.6; $P < .0001$) and there was significant improvement in peak oxygen uptake (pre 54% ± 18% vs post 66% ± 22%; $P = .02$).

Conclusions: Cone reconstruction of TV offers an effective repair in patients with severe regurgitation associated with Ebstein's anomaly. The patients' clinical status improved with better left ventricle filling and objective exercise capacity. The durability of repair, and mechanisms by which the ventricles adapt to the new loading conditions, need longer-term study. (J Thorac Cardiovasc Surg 2015;149:1144-50)

CHD

See related commentary on pages 1150-1.

Ebstein's anomaly (EA) is a rare cardiac defect accounting for approximately 1% of all congenital cardiac abnormalities. It is characterized by apical displacement of the septal and inferior leaflets of the tricuspid valve (TV) and consequent atrialization of a portion of the RV.[1] EA may present at any age, with a natural history related to the severity of the lesion.[2]

A number of surgical approaches to repair EA have been described.[3-5] In 2007 da Silva and colleagues[6] described the cone reconstruction technique that involves delamination and rotation of the detached TV anterosuperior leaflet and the use of the remnant of the septal and inferior leaflets to create a cone, the vertex of which faces the RV apex. With sufficient valve tissue, this technique promotes the full coaptation of valve leaflets, with a more physiologic distribution of stresses and the creation of a central bloodstream through the new TV. This recently introduced method consistently improves the functional anatomy of RV inflow and provides a hospital mortality of 2.5% with a reduction in tricuspid regurgitation (TR) grade from 3.6 ± 0.5 preoperatively to 1.2 ± 0.4 postoperatively.[6] These early encouraging results have also been supported by the surgical series from Boston and the Mayo clinic.[7-9]

Despite the good early outcomes, data on postoperative RV and left ventricle (LV) adaptation, and objective cardiopulmonary testing are lacking.

The aims of our study were to describe our results and determine the integrity of the cone reconstruction in patients with symptomatic EA with significant TR, to use echocardiographic and magnetic resonance imaging (MRI) data to compare preoperative and postoperative biventricular size and function, and to objectively compare exercise capacity before and after surgery.

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Abbreviations and Acronyms
CMR = cardiac magnetic resonance
EA = Ebstein's anomaly
LV = left ventricle
MRI = magnetic resonance imaging
RV = right ventricle
TR = tricuspid regurgitation
TV = tricuspid valve

METHODS

Study Design

This was a retrospective observational study using systematically acquired pre- and postoperative data from study participants in the short- to mid-term. Ethical approval for data collection was granted following local ethical review.

Patient Selection

Beginning in 2009, all symptomatic EA patients with moderate or severe TR who were considered for surgery were referred for cross-sectional imaging, involving echocardiography and cardiac magnetic resonance, unless contraindicated. Cardiopulmonary exercise testing was included in the preoperative assessment.

Those considered to have suitable TV anatomy (sufficiently mobile and abundant anterosuperior leaflet for the creation of a cone valve) were offered the cone reconstruction. All patients who underwent a cone reconstruction are included in the analysis (Table 1).

Surgical Procedure

All operations were performed by a single surgeon (V.T.T.) at 2 centers, in which all patients were followed-up.

The procedures were performed under mild hypothermic (34°C) cardiopulmonary bypass with intermittent antegrade cold blood cardioplegia. The cone reconstruction has been described previously,[6] and is challenging surgery in which a number of elements need to be considered, including the heterogeneous TV morphology. Bridging of the often deficient inferior leaflet poses more difficulties than the deviated and deficient septal leaflet.

The anterosuperior leaflet of the TV is detached from the annulus en bloc, and the accessory attachments of the leaflets to the ventricular walls are divided, providing access to the subvalvular apparatus. It is essential to delaminate the deficient septal and inferior leaflets, if present. The free edge of the reconstructed inferior leaflet is rotated clockwise and sutured to the reconstructed septal leaflet edge, forming a new TV surrounded by native leaflet tissue with an apical papillary attachment.

In some cases the inflow orifice toward the infundibulum and any commissural deficiency or leaflet fenestrations may require closure to improve the competency of the reconstructed valve. In valves with extensive muscularization of the leaflets, surgically created fenestrations may be required to offer unrestrictive inflow. If present, an enlarged atrialized RV chamber is reduced using endocardial, longitudinal plicating sutures, without compromising the epicardial branches of the right coronary artery.

The neoannulus is reconstructed by plication of the dilated TV annulus (anteroseptal and inferioseptal annuloplasty) onto which the newly reconstructed cone valve is reattached, nearly restoring its anatomical position. The restored leaflet coaptation is confirmed at surgery by filling the RV with saline through a bulb syringe and by visually inspecting the leaflets. In addition, after weaning from cardiopulmonary bypass, intraoperative transoesophageal echocardiography is used to assess the results of the valve reconstruction. A small atrial communication is often left in situ to assist the early postoperative management of RV functional impairment. Concomitant maze procedure was used for patients with symptomatic preoperative arrhythmias refractory to electrophysiologic ablation.

The postoperative management is focused around the support of RV function with vasodilators and agents to maintain low pulmonary vascular resistance. There is a low threshold to administer prophylactic amiodarone for the management of atrial tachyarrhythmia.

Transthoracic Echocardiography

Transthoracic conventional echocardiographic studies were performed using Vivid 7, Vivid e9 machines (GE, Milwaukee, Wis).[10] Subcostal, apical 4-chamber and parasternal long- and short-axis views were used to obtain images on the morphology of tricuspid valve, atrial and ventricular chambers, and right ventricular outflow tract. All echocardiographic recordings were stored on digital versatile discs for offline analyses with Echopac software (GE). All measurements were supervised by J.M. Measurements were made over 3 cardiac cycles, and the average was used for statistical analysis. Whereas echocardiography was performed on all patients pre- and postoperatively, formal quantitative assessment was available for 24 patients. Data from the latest available echocardiogram was used in this study. Postoperative echocardiographic studies were completed within 6 months.

Cardiac Magnetic Resonance (CMR)

CMR imaging was performed on a 1.5T scanner (Avanto, Siemens Medical Solutions, Erlangen, Germany). Evaluation of ventricular and valve function was achieved using retrospectively gated, breath-held, balanced, steady-state free-precession cine images. The TV leaflet attachments and the margins of right atrium and right ventricle were visualized and analyzed throughout the cardiac cycle in multiple planes. Net flow through the pulmonary valve and aortic valve was measured using electrocardiogram-triggered, breath-hold cine phase contrast flow mapping. Flow image planes were planned perpendicular to the proximal main pulmonary artery and perpendicular to the ascending aorta.

During postprocessing, ventricular volumes were measured by manually tracing the blood-endocardium boundary for each slice at end-diastole and end-systole. The ventricular volume was then determined by summation of the volumes of all slices. From these data ventricular stroke volumes and ejection fraction were calculated. TV regurgitant fraction was calculated by subtracting the pulmonary forward flow stroke volume from the RV stroke volume, and expressing the remainder as a percentage of the RV stroke volume. CMR was available for analysis in 16 patients preoperatively and 15 patients postoperatively. Postoperative CMR was completed within 6 months of surgery.

Cardiopulmonary Exercise Test

Due to the young age of some patients, objective exercise testing was not possible in all patients. Pre- and postoperative objective exercise capacity, using cardiopulmonary testing protocols or the Bruce test, is available for 13 patients. Data are presented as absolute peak oxygen uptake and also as a percentage of predicted peak oxygen uptake. Cardiopulmonary exercise testing was performed on a bicycle ergometer with an initial, unloaded warm-up. Following this, the workload was increased by 10 to 20 W/min. The patients were encouraged to exercise until exhaustion after about 10 minutes of loaded exercise. The tests were considered maximal with a respiratory exchange ratio of $\geq$1.09. The 12-lead electrocardiogram was monitored continuously and blood pressure was recorded every 2 minutes. Breath-by-breath respiratory gas exchange measurements were recorded throughout the test. The peak oxygen uptake was defined as the average of the values obtained in the last 20 seconds of exercise.