VENTRICULARIZATION OF THE ATRIALIZED CHAMBER: A CONCEPT OF EBSTEIN’S ANOMALY REPAIR

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Background. We report results of a technique of Ebstein’s anomaly repair by creating a predominantly monocuspid valve with simultaneous ventricularization of the atrialized right ventricular (aRV) chamber.

Methods. Between March 1993 and April 2003, Ebstein’s anomaly repair by valvuloplasty with combined ventricularization was performed in 23 patients aged 13.6 (4.1–52.6) years presenting with tricuspid valve regurgitation (TVR) (I°, n = 1; II°, n = 3; III°, n = 13; IV°, n = 6). Valvuloplasty consisted of creating a predominantly monocuspid valve at the level of the anatomical atrioventricular junction resulting in a ventricularization of the atrialized chamber. Postoperatively all survivors were examined regularly with an actual prospective evaluation.

Results. One early death (4.4%) occurred and was caused by right heart failure. Follow-up was 4.6 (0.5–10.9) years. Important recurrent ativoventricular valve regurgitation caused by rupture of fixation sutures occurred in 3 patients (13%), necessitating reintervention at 3 (0.03–4) months (revalvuloplasty, n = 2; TV replacement, n = 1). One patient presenting with hypoplastic right ventricle with consecutive right heart failure underwent creation of a total cavopulmonary connection at 10 months. At present all patients are doing well. Actual echocardiographic examination revealed significant improvement of right atrioventricular valve regurgitation (p < 0.0001) and favorable restoration of RV geometry and function.

Conclusions. This technique of Ebstein’s anomaly repair with ventricularization of the atrialized chamber provides excellent results regarding right ativoventricular valve function and leads to a favorable restoration of RV geometry and function.

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Ebstein’s anomaly, described in 1866 by Wilhelm Ebstein [1], is a rare complex congenital defect of the tricuspid valve (TV) and the right ventricle (RV) [2, 3]. It is characterized by a downward displacement of the dysplastic septal tricuspid leaflet (STL) and posterior tricuspid leaflet (PTL) into the RV leading to a division of the RV in a so-called “atrialized” chamber and a small functional RV [2–4]. An interatrial communication is usually present, accompanied by right-to-left shunt and systemic arterial desaturation and cyanosis [5].

Standard valvuloplasty of Ebstein’s anomaly included numerous forms of annuloplasty usually with the exclusion of the atrialized right ventricular (aRV) chamber [4, 6–12]. Two groups [13, 14] proposed a modified repair technique without ventricular plication with excellent results.

We present a technique that is characterized by reintegration of the atrialized chamber into the RV cavity (termed “ventricularization”). Ventricularization was obtained by orthotopic transposition of the detached STL and PTL. The reimplanted STL was then serving as an opposing structure for coaptation of the reconstructed ativoventricular valve. Valvuloplasty consisted of creating a predominantly monocuspid valve at the level of the anatomical ativoventricular junction resulting in a restoration of the valve mechanism at the level of the true tricuspid annulus.

Our concept was based on the assumption that ventricularization would result not only in a favorable restoration of RV geometry but also in improved RV function. We report our experience with this method, which has been our standard surgical repair technique for patients with Ebstein’s anomaly since its introduction at our institution in 1993.

Patients and Methods

Patients

Between March 1993 and April 2003, 23 (males, n = 7; females, n = 16) out of 29 patients presenting with Ebstein’s anomaly underwent surgical repair at our institution using our standard repair technique by valvuloplasty with combined ventricularization. According to Carpentier’s classification [10], type A was present in 2 patients (8.7%), type B was present in 14 patients (60.9%), and type C was present in 7 patients (30.4%). Age was 13.6 (4.1–52.6) years at the time of Ebstein’s anomaly repair. Preoperative investigation included echocardiography and cardiac catheterization. The degree of preoperative TV regurgitation (TVR) was as follows: I°, n = 1
forming an atrIALIZED chamber of various sizes (Fig 1).

...the STLs and PTLs into the RV cavity toward the apex. In 5 patients (21.7%), however, the PTL was quite large and thick-walled. In about half of the patients inspection of the situs revealed clefts and fenestrations mainly of the PTL and less of the ATL, which were closed by single sutures. Additional procedures performed at the time of Ebstein's anomaly repair included direct closure of a patent foramen ovale (n = 8), a secundum atrial septal defect (ASD, n = 9), or ASD patch closure (n = 2), respectively.

Table 1. Ebstein's Anomaly Repair: Preoperative and Actual Follow-Up Echocardiographic Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Degree of TVR</th>
<th>RV Function</th>
<th>RVEDD (mm)</th>
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<td>9*</td>
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<td>23</td>
<td>III*</td>
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* Adjusted to body surface [16]. *a Echocardiographic findings before total cavopulmonary connection. *b Early postoperative death due to right ventricular failure.

n.a. = not available; RV = right ventricular; RVEDD = right ventricular end-diastolic diameter; TVR = tricuspid valve regurgitation.

Operative Procedures and Intraoperative Findings

Operations were performed through median sternotomy with standard cardiopulmonary bypass using bicaval cannulation, moderate hypothermia (26°–30°C) and antegrade single-dose cold crystalloid cardioplegia. After incision of the right atrium (RA) the situs was inspected. The dystopic tricuspid annulus caused displacement of the STLs and PTLs into the RV cavity toward the apex forming an atrialized chamber of various sizes (Fig 1). The downward displacement was of varying degrees with the distance measured from the atroventricular junction to the most distal displacement of the effective valvular orifice ranging from approximately 2–5 cm. The anterior tricuspid leaflet (ATL) was enlarged and sail-like in all patients. It revealed marked variations in its distal attachment ranging from normal in most patients to localized or widespread chordal attachment to the RV free wall restricting leaflet mobility in 7 patients (30.4%). The PTL was adherent to the RV wall and displaced toward the apex. In 5 patients (21.7%), however, the PTL was quite large and thick-walled. In about half of the patients inspection of the situs revealed clefts and fenestrations mainly of the PTL and less of the ATL, which were closed by single sutures. Additional procedures performed at the time of Ebstein’s anomaly repair included direct closure of a patent foramen ovale (n = 8), a secundum atrial septal defect (ASD, n = 9), or ASD patch closure (n = 2), respectively.

Repair Technique

All Ebstein’s anomaly repairs were performed by the same surgeon (SH). The basic principle of our repair technique was the ventricularization of the former atrioventricular chamber by detachment and reimplantation of the STL and PTL at the level of the atrioventricular junction (Fig 2). Detachment of the STL and PTL consisted of an extensive dissection of all muscular adhesions between the leaflets and the septum or posterior RV wall, respectively, including dissection of all chordae attachments causing restricted leaflet mobility. After translocation the detached leaflets were reimplanted at the anatomical tricuspid annulus using a superficial running suture taking care of the adjacent conducting tissue (atrioventricular node and bundle of His). A couple of single pledgeted sutures were positioned to reinforce the running fixation suture of the transposed STLs and PTLs (Figs 2–5). Then a monocuspid valve was created by

(4.3%); II*, n = 3 (13%); III*, n = 13 (56.5%); IV*, n = 6 (26.1%, Table 1). Indication for operation was congestive heart failure of various degrees in all patients. Prosthetic TV replacement had to be in 5 patients presenting with Ebstein’s anomaly type D preventing valvuloplasty and 1 patient underwent implantation of a central prosthetic aortopulmonary shunt. These 6 patients were excluded from analysis.
Fig 1. Intraoperative situs (top) photograph, (middle) schematic illustration of (top) from the surgeon’s view, and schematic cross-sectional view (bottom, posterior tricuspid leaflet not indicated) indicating the typical features of Ebstein’s anomaly in a 37-year-old patient with downward displacement of the septal tricuspid leaflet (top, middle, bottom) and posterior tricuspid leaflet (top, middle), forming the atrialized ventricle (middle, arrow) and partial adherences of the anterior tricuspid leaflet (bottom). (PA = pulmonary artery; RA = right atrium.)

Fig 2. Same patient illustrated in Fig 1. Intraoperative situs (top) photograph, (middle) schematic illustration of (top) from the surgeon’s view, and schematic cross-sectional view (bottom) indicating ventricularization by reimplantation of the detached septal tricuspid leaflet (STL) (top, middle, bottom, arrow on the left side) and posterior tricuspid leaflet (PTL) (top, middle) at the atrioventricular junction (suture lines [middle] and detachment of adherences of the anterior tricuspid leaflet (ATL) [bottom, two arrows on the right side]). (CS = coronary sinus.)
suturaing together the PTL mainly with the ATL and in parts with the STL at their free edges. The reimplanted STL was serving as an opposing structure for coaptation with the newly created monocusp resulting in restoration of the valve mechanism at the level of the original TV annulus (Figs 2–5). In case of restricted leaflet mobility, mobilization of the ATL and/or PTL was achieved by dissection of all abnormal muscular adhesions from the leaflet tissue to the RV wall (Fig 2). In 5 patients (21.7%) a quite substantial PTL was observed, which was mobilized and connected partially to the free edge of the STL creating a more bicuspid atrioventricular valve (Fig 4). We did not perform reduction of the tricuspid annulus as a part of our standard technique, apart from 1 patient presenting with massive dilatation of the tricuspid annulus. Dissection and/or transposition of a papillary muscle was performed in 13 patients (56.5%) to assure satisfactory mobilization of the ATL according to the technique elaborated by Sebening [13, 15]. To restore normal volume of the RV cavity an extensive resection of restrictive trabecula and muscle bundles was performed in almost all patients. After evaluation of a satisfactory functional result of the repair by repeated filling of the RV with saline solution (Figs 3–5) the atriotomy was closed, reducing the volume of the enlarged RA. Cardiopulmonary bypass time was 107 (70–194) minutes and aortic cross-clamp time was 78 (43–129) minutes.

Data Collection, Follow-Up, and Statistical Analysis
Preoperative and perioperative data were collected upon retrospective review of patient records. All patients have been prospectively followed-up by regular investigations including clinical examination, electrocardiography, and transthoracic echocardiography (Table 1) in our outpatient clinic at least once a year. Quantitative and semi-quantitative analysis in the form of two-dimensional (2D) and Doppler echocardiography was performed to determine right atrioventricular valve regurgitation, RV end-diastolic diameter, and RV function [16]. Specific software (SPSS for Windows, Rel. 11.01. 2001; SPSS Inc., Chicago, IL) was used for statistical analysis. Data are expressed as median. Probability (p) less than 0.05 was considered statistically significant.

Results
There was one early death (4.4%) at the eighth postoperative day caused by low cardiac output in a 45-year-old patient presenting preoperatively with marked cardiomegaly, impaired RV function, severe TVR (IV°), and cyanosis at exercise. This patient presented with early rupture of valvuloplasty fixation sutures causing recurrent important right atrioventricular valve regurgitation necessitating prosthetic TV replacement at the second postoperative day. One patient had to be reoperated for bleeding on the first postoperative day. The 22 survivors were examined on a regular basis and complete follow-up data were available on all patients with a median follow-up time of 4.6 (0.5–10.9) years. There were no late deaths. Late revalvuloplasty was performed in 2 patients
at 3 and 4 months, respectively, because of rupture of valvuloplasty fixation sutures causing recurrent important right atrioventricular valve regurgitation. In 1 patient presenting with hypoplastic RV and consecutive right heart failure a total cavopulmonary connection was performed at 10 months. During follow-up there were no late complications or rhythm disturbances observed. At the last follow-up echocardiographic examination, right atrioventricular valve regurgitation was estimated as follows: I°, n = 14 (66.7%); II°, n = 5 (23.8%); III°, n = 2 (9.5%); IV°, n = 0, reflecting a significant improvement of TV function compared with preoperative findings (p < 0.0001, Table 1 and Figs 6–8). In almost all patients actual 2D echocardiography revealed a favorable restoration of RV geometry and function (Table 1). All patients were in good health and had resumed normal physical activity.

According to the functional classification of the New York Heart Association (NYHA), 19 patients (86.4%) were designated as NYHA class I and 3 patients (13.6%) were designated as NYHA class II.

Comment

Ebstein’s anomaly is a complex congenital pathology not only of the TV but also of the RV. Downward displacement of the STL and PTL leads to TVR and to loss of contractile myocardium by partial atrialization of the RV causing impaired RV function.
Restoration of TV competence is the primary goal of reconstructive surgery of Ebstein’s anomaly. The basic principle of the standard repair technique elaborated by Hunter [6], Lillehei [7], and Hardy [4] consisted of transposition of the STL and PTL to the level of the true tricuspid annulus resulting in a transverse plication of the atrialized chamber. Posterior tricuspid annuloplasty and RA reduction was added to this procedure by Danielson and colleagues [9]. Carpentier and coworkers [10] applied a tricuspid annuloplasty by longitudinal plication of the atrialized chamber. A similar technique without reinforcement by a prosthetic ring was proposed by Quaegebeur and colleagues [11].

All these techniques have two features in common: the exclusion of the atrioventricular chamber by plication and the use of the sail-like ATL as a monocuspid atrioventricular valve with the septum serving as an opposing structure for coaptation. In Ebstein’s anomaly the usually enlarged and mobile ATL is generally attached at the anatomical tricuspid annulus. Carpentier and colleagues [10, 12] emphasized the importance of adequate mobilization of the ATL when creating a competent monocuspid valve. In case of restricted mobility of the ATL we achieved mobilization not by complete detachment and reimplantation as they proposed but by dissection of adherences and chordal attachments. To assure satisfactory mobilization we applied the Sebening technique [13, 15] of septal transposition of papillary muscles and/or transposition of the base of the ATL chordal attachment in 13 (56.5%) of our patients. However the ATL could always been used for valve closure without any need for transection of its annular attachment. Using our technique the reimplanted STL and parts of the PTL were serving as an opposing structure for coaptation.

The secondary goal in Ebstein’s anomaly repair is the restoration of normal size and function of the RV. The enlarged thin-walled atrialized chamber was compared with a ventricular aneurysm that might exhibit a negative effect on ventricular energy economics when exposed to RV pressure and that paradoxical contraction of this aneurysmic ventricle might promote clot formation with the risk of embolization [4, 8–12]. Therefore almost all repair techniques included exclusion of the right atrialized chamber by transverse [4, 8, 9] or longitudinal plication [10–12] of this compartment.

Despite various comments in the literature, there is no evidence of a benefit regarding improvement of RV function after exclusion of the atrialized chamber. There are two groups reporting excellent results regarding restoration of RV function after creation of a monocuspid valve without plication of the atrialized chamber [13–15]. Ebstein’s anomaly repair by TV replacement without plication or exclusion of the atrialized ventricle was performed without negative sequelae [7, 17–20]. Postoperative cardiac catheterization [7] and cineangiographic examination [17] demonstrated regained strength and contractility of the RV with continuous global contraction in these patients. Histologic study of the atrialized portion of the RV indicated normal structured muscle cells forming the thin ventricular wall that in time will assume more normal thickness and function when normal work is imposed [7].

It should also be noted that the left ventricle (LV) is not normal in Ebstein’s anomaly. There is a bulging of the muscular septum toward the left that could be deteriorated by ventricular plication causing left ventricular (LV) outflow tract obstruction.

Based on this morphologic and functional findings we think that exclusion of the atrialized ventricle during Ebstein’s anomaly repair by ventricular plication is not necessary at all. Reintegration of the atrialized compartment led to favorable restoration of RV geometry and function and a marked improvement of the right atrioventricular valve function compared with preoperative findings revealed by regular follow-up echocardiographic examinations. Using our technique of ventricularization we have not observed any negative effects regarding RV function or intracardiac clot formation.

In our series we unfortunately lost a 45-year-old patient presented with Ebstein’s anomaly type C, marked cardiomegaly with impaired RV function and severe TVR. Despite an enormous dilatation of the RA and ventricle we attempted a repair by ventricularization and valvuloplasty reducing the dilated tricuspid annulus and RA to normal size. At the second postoperative day important recurrent right atrioventricular valve regurgitation occurred because of the rupture of valvuloplasty fixation suture, probably caused by increased tension of the tricuspid annulus when normal work was imposed. In case of marked dilatation of the true tricuspid annulus we therefore recommend a prosthetic valve replacement as primary intervention rather than our presented repair technique, which we recommend for all patients presenting with Ebstein’s anomaly early in their course.

When employing this repair technique it is very important to perform an adequate mobilization of the STLs and PTLs by extended dissection of all muscular adherions and chordae attachments causing restricted mobility. Additional dissection and translocation of the papillary muscle connected to the ATL had to be performed in nearly two-thirds of the cases to assure satisfactory
mobilization of the ATL. To restore normal RV volume resection of hypertrophied trabecula was necessary in almost all patients. In the presence of a thick-walled PTL we created a bicuspid atrioventricular valve providing favorable functional results as well.

Rupture of the running fixation suture of the STLs and PTLs was a major complication and cause of reintervention after valvuloplasty at the beginning of our series, probably caused by increased tension of the tricuspid annulus when normal work was imposed. After changing our technique by performing an additional reinforcement of the running suture using some single pledgeted sutures we did not observe any recurrent right atrioventricular valve regurgitation caused by the rupture of the running suture.

Certain limitations of the presented study have to be mentioned: larger cohorts are required and assessment of exercise capacity would have been helpful to objectify postoperative functional status of our patients. We did not perform magnetic resonance angiography in our patients, which would allow determining RV volumes and ejection fraction after surgical repair. Additionally the presented method of Ebstein’s anomaly repair has its technical limitation in the presence of a small ATL and the presented method of Ebstein’s anomaly: a functional concept and successful definite repair. J Thorac Cardiovasc Surg 1964;48:927–40.


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