Surgical Treatment of Ebstein’s Anomaly

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Abstract

Surgical repair of the tricuspid valve is a milestone in the medical history of patients with Ebstein’s anomaly. The timely alleviation of the insufficiency has an important impact on the prognosis. In this review, we describe features of the disease relevant to surgical correction and the evolution of surgical techniques over six decades. We compare the results of different repair and replacement techniques. Additionally, we discuss concomitant antiarrhythmic surgery and bailout strategies for postoperative right ventricular failure. Finally, we review the surgical options in symptomatic neonates with Ebstein’s disease.

Keywords
► heart valve surgery
► congenital heart disease
► CHD
► tricuspid valve

Introduction

With an incidence of 5.2 per 100,000 live births, Ebstein’s anomaly accounts for 1% of all congenital heart disease.1 Its main features are a pathologic configuration and attachment of the tricuspid leaflets that lead to severe insufficiency of the tricuspid valve and a pathologic configuration of the right ventricle. Additionally, an atrial septal defect (ASD) or a persistent foramen ovale (PFO) is present in two-thirds of patients,2 and cyanosis may develop due to interatrial right to left shunting. The onset of symptoms varies according to the severity of the disease. Some patients become symptomatic as neonates, others as young adults, and some remain free of symptoms until seniority.3

The anomaly is named after the pathologist, Wilhelm Ebstein, who first described it in 1866.4 A long time passed until the diagnosis could be made during lifetime in 1951, using cardiac catheterization.5 Since around 1980, echocardiography simplifies detection and has become the standard method for diagnosis. The first surgical procedures in patients with Ebstein’s disease were ASD closures in cyanotic patients, performed in 1956. The mortality of these procedures exceeded 80%.6 Since the results of isolated ASD closure were poor, physicians searched for ways to reduce the insufficiency of the aberrant valve. In 1956, the American surgeons, Lillehei and Hunter, planned to perform an ASD closure combined with tricuspid valve repair in a 10-year-old girl with Ebstein’s anomaly suffering from severe cyanosis.7 Due to the progressed heart disease, the patient died upon the induction of anesthesia. The two surgeons performed an autopsy and developed the first concept to repair the Ebstein valve anomaly. Later, this concept was successfully applied by Hardy, who performed the first successful repair of an Ebstein valve in 1963.8 One year earlier, the first valve replacement in a patient with Ebstein’s disease was performed by Barnard in South Africa.9 Both repair and replacement of the tricuspid valve will be discussed in the section, “Surgical Techniques.”

Pathologic Anatomy and Pathophysiology

Leaflets

The tricuspid valve develops by delamination of the leaflets from the underlying myocardium.10 In a healthy individual, the three leaflets (anterior, posterior, and septal) separate completely from the myocardium and their hinge point is located at the tricuspid annulus. In Ebstein’s anomaly, the delamination process is incomplete and does not reach the tricuspid annulus. As a result, the leaflets are shortened and their hinge point lies inside the ventricle. The three leaflets are not uniformly affected. The septal and the posterior leaflets are displaced, and the highest degree of displacement...
is usually present at the internal crux of the heart, at the commissure between the two leaflets. The septal leaflet is small or even rudimentary. The anterior leaflet is usually not displaced and is enlarged rather than shortened. The displacement of the septal and posterior leaflets results in a rotation of the valve toward the outflow tract\(^1\) (Fig. 1). Various degrees of connections with the myocardium are present at all leaflets, leading to their restriction. Often, there are holes in the leaflets that contribute to the insufficiency. The leaflets may have a free edge and chordae tendineae, but in some severe cases, the edge is directly connected to the myocardium and the papillary muscle.

**Ventricle and Myocardium**

At the atrioventricular groove, the true tricuspid annulus can still be identified.\(^1\) Due to the partial displacement of the valve toward the apex, the part of the right ventricle (RV) between the true and the anatomical tricuspid annulus becomes functionally a part of the atrium and is referred to as the “atrialized ventricle.” The tricuspid annulus is severely enlarged, as well as the right atrium and the RV (Fig. 2). However, the functional RV is small compared with the gigantic right atrium and the atrialized ventricle. The volume of the entire RV, measured in magnetic resonance imaging, is two to three times larger compared with a normal RV.\(^12,13\)

The myocardium is altered at different sides. At the atrialized RV, the myocardium is usually thin or even absent. The wall thickness of the functional RV may be enlarged or thinned.\(^14\) At the left ventricular wall, an increase of interstitial fibrosis may be found.\(^15,16\) It is assumed that these alterations contribute to ventricular dysfunction and heart failure in Ebstein’s disease.

**ASD**

In two-thirds of patients with Ebstein’s anomaly, an ASD II/PFO is present. If the right atrial pressure is elevated, as typically during exercise, a right to left shunt through the ASD II ensues. Oxygen saturation of the arterial blood is usually moderately impaired (80–90% saturation).\(^17\) The right to left shunt on the atrial level may also account for paradoxical emboli. A quarter of patients beyond 40 years of age exhibit a history of an event potentially caused by paradoxical emboli, such as stroke, transient ischemic attack, brain abscess, or myocardial infarction.\(^18\)

**Rhythm**

Patients with Ebstein’s anomaly may present with different forms of arrhythmia. In one-third of the patients with tachycardia, one or more accessory pathways are present, usually at the posterolateral side of the RV.\(^19\) The enlarged atrium is susceptible for atrial flutter or fibrillation. Table 1 provides an overview of the most common rhythm disorder in patients with Ebstein’s anomaly. A potentially dangerous combination is the presence of an accessory pathway and the occurrence of a supraventricular tachycardia. The accessory pathway may conduct the supraventricular tachycardia in a 1:1 ratio to the ventricle. Accordingly, an atrial fibrillation may transform into ventricular fibrillation and result in sudden cardiac death.\(^20\) In patients with mild leaflet displacement, arrhythmia may be the most important symptom, and even sudden cardiac death has been reported in such patients.\(^21\)

**Indication for Surgery**

The natural history of Ebstein’s anomaly depends on the degree of tricuspid valve dysplasia. In 2000, Attie reported on 72 adult patients who had not received operative treatment.\(^22\) After 20 years of follow-up, survival was less than 10% in patients with a severe displacement of the septal leaflet (n = 14), 30% in patients with a mediocre displacement (n = 35), and 90% in patients with only minor displacement (n = 23), respectively.
In contrast, long-term survival 20 years after the surgery is reported to be 70 to 90%. Thus, surgical treatment of the tricuspid valve regurgitation improves long-term outcome. Because both surgical risk and long-term mortality are increased for patients with advanced disease, surgery should not be delayed. Patients with severe regurgitation beyond 40 years of age should be carefully evaluated with regard to long-term sequelae of congestion, in particular renal, pulmonary, and hepatic failure. Severe left ventricular dysfunction is a risk factor for operative mortality, but it is not a contraindication for surgery. After surgery, left ventricular function usually improves.

### Surgical Technique: Repair

**Surgical Access**
The chest is opened through a median sternotomy. Pericardial traction sutures are placed, and after administration of heparin, the aorta and the caval veins are cannulated. The cannula in the superior caval vein should be placed more cranially, because in selected cases, a cavopulmonary connection might be needed to unload the RV permanently. The operation is performed under cardiopulmonary bypass with mild hypothermia on the cardioplegic arrested heart. Several traction sutures are placed at the right atrium to provide a perfect exposure of the valve (Fig. 3).

### Plication Techniques

The understanding of the plication techniques of the ventricular wall is a key point for the understanding of the different repair techniques. The size of the RV is reduced by folding the wall of the atrialized ventricle.

- The horizontal or transversal plication is intended to reduce the true tricuspid annulus.
- The longitudinal plication creates a fold from the tricuspid valve annulus toward the apex, thereby reducing the size of the annulus as well as of the atrialized ventricle.

Ventricular plication bears the risk for coronary artery injury. Therefore, the coronary branches are continuously followed from the epicardium during plication.

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**Table 1** Mechanism of tachycardia in patients with Ebstein’s anomaly

<table>
<thead>
<tr>
<th>Primary Accessory Pathways</th>
<th>Bidirectional (WPW)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Other Accessory Pathways</td>
<td>Mahaim fibers</td>
</tr>
</tbody>
</table>

**Secondary Accessory Pathways**

<table>
<thead>
<tr>
<th>Atrial reentry tachycardia</th>
<th>Origin at atriotomy scar</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial flutter</td>
<td>Cavo-tricuspid isthmus dependent</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Atrial dilatation</th>
<th>Atrial fibrosis</th>
</tr>
</thead>
</table>

Abbreviation: AV, atrioventricular; WPW, Wolff-Parkinson-White Syndrome.

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**Fig. 3** Intraoperative inspection of the tricuspid valve. Green, anterior leaflet; yellow, posterior leaflet; blue, septal leaflet. The septal leaflet is severely displaced from the true annulus, the posterior leaflet is almost immobilized by adhesions toward the posterior wall.
Hunter–Lillehei–Hardy Technique
The first concept to repair the Ebstein valve was developed by Hunter and Lillehei. Their goal was to “reestablish” the physiologic anatomy of the tricuspid valve by bringing the downward displaced septal and posterior leaflet to the “true” annulus without detaching the leaflets. This was achieved by plication of the RV in the horizontal (transverse) plane, i.e., parallel to the tricuspid annulus. Stitches are placed from the base of the septal/posterior leaflet to the corresponding place at the true annulus. After tying the sutures, the atrialized ventricle is excluded.

Danielson Technique
The Danielson technique is a modification of the Hardy technique. Danielson reasonably describes that the septal part of the atrialized ventricle is not suitable for plication, because a plication may affect mitral valve function, and the part of the right ventricle is added. This brings the anterior leaflet toward the septum and may enhance the coaptation. This repair technique results functionally in a monocuspid valve since the posterior leaflet is almost excluded by the posterior ring plication and the septal leaflet remains as a remnant below the coaptation plane.

Carpentier Technique
The plication techniques for the repair of the Ebstein valve are successful valve construction in the majority of patients. However, in our experience, the consequent application of the originally described cone technique alone is sufficient for successful valve construction in the majority of patients. There are numerous modifications of the cone technique, including leaflet augmentation, ring annuloplasty, usage of autologous or artificial chordae, and the Sebening stitch. However, in our experience, the consequent application of the originally described cone technique alone is sufficient for successful valve construction in the majority of patients.

Wu Technique
Mobilization of the leaflets and reinsertion at the tricuspid annulus are performed in the same way as in the Carpentier repair. However, instead of a plication, Wu advocates a resection of a triangular piece of the posterior part of the atrialized ventricle. Wu further describes the use of autologous pericardium if sufficient septal tissue is missing.

Hetzer Technique
Hetzer describes various techniques for Ebstein’s repair. Their key feature is annular plication. The posterior part of the tricuspid annulus is connected to the septal part of the tricuspid annulus. In some cases, Hetzer suggested the creation of a double orifice annulus. By approaching two points at the opposite side of the true annulus (septal and anteposterior), two orifices are created. The RV is not plicated and the thin myocardium of the atrialized ventricle remains in the RV. The leaflets are not mobilized.

Sebening Stitch
The Sebening stitch transfers the chordal attachment of the papillary muscle of the anterior leaflet to the septum close to...
the “true” annulus. This procedure creates a monocusp valve because the posterior and the septal leaflet are both excluded. The anterior leaflet, which is usually fairly mobile, coapts with the septal rim of the “true” annulus. Although the Sebening stitch alone may result in a sufficient tricuspid valve,35,36 nowadays, it is mostly performed as an additional measure in combination with other repair techniques.30,37,38

**Fig. 4 (a–h) The cone procedure, step by step.**

**Surgical Technique: Replacement**

The prosthesis is placed at the height of the true tricuspid annulus. A horizontal plication of the annulus is usually required. To avoid damage to the atrioventricular (AV) node, some surgeons suggest to place the septal suture line in front of the coronary sinus, thus draining the coronary
that intracardial mapping and ablation accessory pathways had a higher success rate than the transcatheter ablation in patients with Ebstein's anomaly.48

In contrast to the abandoned surgical procedures for accessory pathways, the maze procedure for atrial flutter is a current and successful antiarrhythmic surgical procedure. The placement of a set of ablation lines creates a maze for the electrical activation on the atrial wall. Instead of a planar propagation, the electrical activity propagates inside numerous blind-ending paths. Thus a (macro-) electrical circuit resulting in atrial fibrillation of flutter is prevented.49

Both a maze limited to the right side and biatrial maze are applied successfully in patients with Ebstein's anomaly, with a success rate of 93%.50 Some surgeons recommend the addition of a cavotricuspid ablation line even in patients without previous arrhythmia to prevent isthmus-dependent flutter.50

### Bailout in Case of Right Heart Failure
Cardiac decompensation due to impaired RV function is a major concern after tricuspid valve surgery for Ebstein's disease, since afterload may be increased by the competency of the tricuspid valve when the blood is ejected only into the pulmonary artery. Consequently, RV stroke volume and ejection fraction decrease. On the other hand, volume load, i.e., preload of the right ventricle is also reduced and in most cases, may compensate for the increase in the afterload. In the early postoperative phase, beginning with weaning from the heart lung machine, the risk for RV failure is high.13 A temporary solution is to unload the RV by extracorporeal membrane oxygenation (ECMO) in the postoperative phase.13 A permanent solution is the creation of a bidirectional cavopulmonary anastomosis (Glenn anastomosis). This "one and a half ventricle repair" can only be performed if pulmonary artery pressure is not elevated.51,52 In patients with a 1.5 ventricle repair, the flow over the tricuspid valve is diminished, and a mechanical prosthesis should be avoided due to increased risk of thrombosis.53

### Results after Surgery

Table 2 shows the outcomes after different types of surgery in large patient collectives.

<table>
<thead>
<tr>
<th>Publication</th>
<th>Technique</th>
<th>Patients</th>
<th>Mortality</th>
<th>Reoperations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brown et al23</td>
<td>Replacement</td>
<td>378</td>
<td>6% early, 17% after 10 years</td>
<td>41% after 20 years</td>
</tr>
<tr>
<td>Brown et al23</td>
<td>Danielsson</td>
<td>182</td>
<td>5% early, 12% after 10 years</td>
<td>36% after 20 years</td>
</tr>
<tr>
<td>Badiu et al24</td>
<td>Sebening</td>
<td>130</td>
<td>2.6% early, 13% after 10 years</td>
<td>38% after 20 years</td>
</tr>
<tr>
<td>Hetzer et al25</td>
<td>Hetzer</td>
<td>68</td>
<td>2.4% early, 8.7% after 10 years</td>
<td>7.1% after 20 years</td>
</tr>
<tr>
<td>Chavaud et al57</td>
<td>Carpentier</td>
<td>91</td>
<td>9% early, 13% after 10 years</td>
<td>11% after 20 years</td>
</tr>
<tr>
<td>da-Silva et al29</td>
<td>Cone</td>
<td>52</td>
<td>3.8% early, 14% after 7 years</td>
<td>4 reoperations</td>
</tr>
</tbody>
</table>
Repair versus Replacement
In a single-center study from the Mayo Clinic, Brown and coworkers found no difference in survival following replacement and repair (Danielson technique). In patients below 12 years of age, freedom from reoperation was lower after repair than after replacement. There was no difference in freedom from reoperation in patients over 12 year of age (80% after 10 years). Today, repair is generally considered as the first-line treatment.

Replacement: Biological versus Mechanical
In a recent retrospective study, Brown and coworkers reported similar rates of reoperation after implantation of a mechanical or biological prosthesis in patients with Ebstein’s disease. However, survival was longer after the implantation of a biological prosthesis. An important limitation of this study is that age was significantly higher in patients undergoing replacement with a mechanical device. Furthermore, 69% of the mechanical valves were ball and cage prostheses, which meanwhile have been replaced by bileaflet valves. Barlett and coworkers performed a retrospective multicenter study including children below 6 years of age undergoing tricuspid valve repair. While there was no significant difference in survival after replacement using a mechanical or biological prosthesis, the use of a mechanical prosthesis was associated with a higher rate of failure (twofold) and a higher risk for pacemaker implantation (13-fold). Generally, a biological prosthesis should be recommended independent of the age of the patient. These prostheses may become dysfunctional early, especially in patients younger than 16 years of age, but a redooperation can be performed with a low mortality.

Some authors suggest that due to the low opening and low closing pressure at the tricuspid position, porcine bioprostheses are more suitable compared with pericardial bioprostheses because of their thinner leaflet tissue.

Types of Repair
Till date, no comparative study among different types of repair has been performed. Both the Mayo Clinic (2008, Danielson technique) and the German Heart Centre Munich (2010, predominantly Sebening stitch) reported an excellent early and long-term survival after repair but considerable rate of reoperation in the long-term follow-up (36 and 38% reoperation after 20 years). In contrast, the group of the Hospital George Pompidou, Paris (2003, Carpenter technique) reported considerably less reoperations in the long-term follow-up than the other two groups. Likewise, the percentage of repair procedures (98%) was higher compared with the German Heart Centre (90%) or the Mayo Clinic (34%). In 2007, the group of the Hospital Beneficencia Portuguesa, Sao Paolo (cone technique) reported an excellent survival and few need for reoperation in 40 consecutive patients with Ebstein’s anomaly.

In consequence of the excellent results from the group in Sao Paolo with the cone repair, many groups with an extensive experience in the repair of Ebstein’s anomaly such as the Mayo Clinic in Rochester, the German Heart Centre in Munich, the great Ormond Street Hospital for Sick Children in London, and the Children’s Hospital in Boston switched to this technique. Furthermore, the cone technique was adopted by the two highly frequented hospitals—West China Hospital, Chengdu, and the Shanghai Children’s Medical Center. It is noteworthy that the favorable results with regard to mortality, valve competency, and the high percentage of achieved repair could be reproduced by all of these groups. Furthermore, MRI measurements show a major change in RV size and shape after the cone repair. At the German Heart Centre, the preoperatively indexed end-diastolic volume of the functional RV was 191 mL² (normal value: 75 mL/m²) and was reduced to 123 mL/m² 6 months after the repair. The group of Chengdu reported similar results (index preoperative RV end-diastolic volume: 134 mL/m² reduced to 97 mL/m² postoperatively). According to the law of Laplace, a reduction of RV diameter results in a reduction in wall stress. Thus, there is a good reason to assume that the cone repair may prevent a later heart failure. The RV ejection fraction remains impaired in the midterm follow-up, but the regurgitation volume decreases. The net result is an increased flow over the pulmonary artery and an improved left ventricular filling. Cardiopulmonary exercise testing displays an improved functional status in the midterm follow-up after the cone repair.

Special Case: Newborn
Pathology
During the first weeks of life, a newborn with Ebstein’s disease may present with an aggravated physiologic condition. Right ventricular function is impaired by the presence of severe tricuspid valve insufficiency. At the same time, the pulmonary pressure is physiologically elevated, putting additional stress on the RV. In this situation, severe congestive failure may evolve rapidly and the patients may have to be assigned to surgery in the neonatal period. However, in some cases, symptoms may diminish with the physiologic decline in pulmonary artery pressure. The therapeutic goal in severe neonatal Ebstein anomaly is not “earliest surgery,” but medical management including measures to reduce the pulmonary vascular resistance (even with prostaglandin infusion in the beginning). When weaning from prostaglandin E fails, it serves as an indication for an early surgery. Celermajer and coworkers developed an echocardiographic score (Great Ormond Street Echo score, GOSE score) to help the physicians in this situation to decide between an early surgery or medical treatment until later repair (Table 3).

Accordingly, the disease can be graduated, and a mortality risk can be determined.

Palliation: Starnes Procedure
The first series on successful palliative surgery in neonatal patients with Ebstein’s disease was published in 1991 by Starnes. In this procedure, the tricuspid valve orifice is closed with a membrane at the true tricuspid annulus and the pulmonary artery is ligated. An atrioseptectomy and a modified Blalock-Taussig shunt are added.
A certain amount of blood still drains into the RV via the perforant veins. In a later experience of the Starnes group (16 patients), the importance of leaving a small hole in the membrane was emphasized to decompress the RV. In a later experience of the Starnes group (16 patients), the importance of leaving a small hole in the perforant veins. In a later experience of the Starnes group (16 patients), the importance of leaving a small hole in the perforant veins.

**Correction: Knott-Craig**

In 2000, Knott-Craig and coworkers published three cases of neonates with Ebstein's anomaly undergoing repair of the tricuspid valve. GOSE-scores were 1.3, 1.8, and 2.4 with a high probability of death without surgery. The authors created a monocuspid valve consisting of the anterior leaflet only and added a bidirectional cavopulmonary anastomosis. An annular plication was performed by placing a suture with only and added a bidirectional cavopulmonary anastomosis. A further modification, the septal leaflet may be detached at the annulus if the anterior leaflet is adherent and therefore not able to coapt with the septal wall. At the place of the detachment, a patch is inserted to enlarge the anterior leaflet.

In 2011, the group of Knott-Craig reported 32 neonates and young children, of which 90% underwent biventricular repair. GOSE score was $\geq 1$ in all patients and $\geq 1.5$ in 22 patients. Early mortality was 22% and there was one late death. Freedom from reoperation after 15 years was 75% in patients undergoing biventricular repair.

Recently, most centers with extensive experience with the cone procedure have also started to apply this technique in newborn children if there is enough valve tissue for the functional concept and successful definitive repair. In particular, the cone reconstruction shows very promising results and may become the technique of choice for patients with Ebstein's anomaly.

**Conclusion**

Surgical repair of Ebstein's anomaly has been a challenge for decades. Since the first description by Hunter and Lillehei in 1957, many different repair techniques have evolved. Plication procedures alone yielded acceptable results but were characterized by a considerable reoperation rate. With the advent of complex leaflet reconstruction techniques, such as the Carpentier and the cone reconstruction, a more physiologic repair became available. In particular, the cone reconstruction shows very promising results and may become the technique of choice for patients with Ebstein's anomaly.

**Table 3** Survival rates of neonates with Ebstein's anomaly

<table>
<thead>
<tr>
<th>Score</th>
<th>GOSE</th>
<th>Natural course</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;0.5</td>
<td>GOSE I</td>
<td>0% Mortality</td>
</tr>
<tr>
<td>0.5–0.99</td>
<td>GOSE II</td>
<td>10% Mortality</td>
</tr>
<tr>
<td>1–1.49</td>
<td>GOSE III</td>
<td>44% Mortality</td>
</tr>
<tr>
<td>$\geq 1.5$</td>
<td>GOSE IV</td>
<td>100% Mortality</td>
</tr>
</tbody>
</table>

Abbreviation: GOSE, Great Ormond Street Echo.
Barnard CN, Schrire V. Surgical correction of ebsstein’s malformation with prosthetic tricuspid valve. Surgery 1963;54:302–308
39 Baumgartner H, Bonhoeffer P, De Groot NMS, et al; Task Force on the Management of Growth-on Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (APEC); ESC Committee for Practice Guidelines (CPC); ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31(23):2915–2957
51 Liu J, Qiu L, Zhu Z, Chen H, Hong H. Cone reconstruction of the tricuspid valve in Ebstein anomaly with or without one and a half ventricle repair. J Thorac Cardiovasc Surg 2011;141(05):1178–1183