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**Introduction**

Ebstein’s anomaly is a rare congenital heart malformation of the right ventricle and tricuspid valve. Failure of delamination of the tricuspid valve, especially of the posterior and septal leaflets, from the underlying myocardium, results in a variable degree of tricuspid valve and right ventricular deformity. The natural history is influenced by a high rate of intrauterine and neonatal death, especially when the anomaly is associated with severe deformity of the tricuspid valve and cyanosis. At the other end of the spectrum, patients with mild dysfunction of the tricuspid valve can remain asymptomatic until late in adult life. Management therefore requires a choice between a conservative approach and operative intervention. Currently available surgical techniques have demonstrated survival and functional benefits to patients with a greater severity of Ebstein’s anomaly.

**Anatomy**

The anomaly is characterized by:

1. **Tricuspid valve malformations**
   a. The septal and posterior leaflets are displaced towards the apex of the right ventricle with a variable degree of adherence to the myocardium (spiral failure of delamination).
   b. The sail-like, redundant anterior leaflet has a normal attachment to the tricuspid valve annulus, but is frequently fenestrated with chordal attachments to the ventricular wall and eventually, as well as linear attachments (fibrous and muscular attachments between the leaflet and myocardium).

2. The right ventricle is malformed and consists of:
   a. A thin-walled “atrialized” component, defined as a segment of the right ventricle, from the level of the “true” tricuspid annulus to the level of displacement of the septal and posterior leaflets.
   b. A thick-walled functional component ("true right ventricular cavity"), consisting of apical and outlet portion, often obstructed by chordal attachments of the anterior leaflet of the tricuspid valve. The inlet portion
is small or nonexistent. The cavity of the functional right ventricle is usually smaller, containing fewer myocytes and more fibrous tissue than a normal right ventricle.

The most commonly associated anomaly is the atrial septal defect or persistent foramen ovale (in about 90% of cases). Survival in symptomatic newborns with severe obstruction to pulmonary outflow depends on the presence of a patent ductus arteriosus. Wolff–Parkinson–White syndrome is present in about 10% of cases. There is rarely association with an atrioventricular septal defect, a ventricular septal defect, tetralogy of Fallot, congenitally corrected transposition of the great arteries, aortic coarctation, or other defects.

**Indication for Surgery**

**Newborns and Young Infants**

Before considering surgery in newborns, all measures are taken to lower pulmonary vascular resistance while preserving duct dependent pulmonary circulation. The critically ill patient requires mechanical ventilation, sedation, paralysis, hyperventilation, correction of metabolic acidosis, appropriate inotropic/volume management, and prostaglandin E₁ infusion. After stabilization, when pulmonary vascular resistance begins to fall, the prostaglandin is stopped to allow ductal constriction. Subsequently, the true functional capability of the right ventricle in providing antegrade flow is evaluated. The indications for surgery are ventilator dependence, severe cardiac failure, and severe tricuspid insufficiency with prostaglandin-dependent circulation.

**Older Patients**

In older patients, surgery is indicated by symptoms of poor growth, decreased exercise tolerance (New York Heart Association functional classes III, IV), cyanosis (oxygen saturation <90%), progressive cardiomegaly (cardiothoracic ratio >0.6), reduced left ventricular function, tachyarrhythmias that are not controllable by medication or amenable to catheter-based intervention and sig-
nificant associated lesions. Timely operation is warranted before cardiomegaly develops and the functional status deteriorates. Currently, the operation can be considered in asymptomatic patients with low normal-exercise tolerance, no right-to-left shunting, and only mild cardiomegaly (cardiothoracic ratio ≤ 0.6). In children with mild symptoms but with significant enlargement of the right atrium due to massive tricuspid insufficiency, the risk of early rhythm disturbances is so high that surgery should be considered.

Approach and Cardiopulmonary Bypass Strategy

The heart is approached through a median sternotomy. The standard technique of cardiopulmonary bypass with mild hypothermia (32°C) is used. A left ventricular vent is inserted through the entrance of the right pulmonary veins.

Anatomical Repair – Cone Reconstruction

The Goal of Surgery

Surgical therapy for Ebstein’s anomaly should restore tricuspid valve competence and it should address the inadequacy of the inefficient right ventricle. Especially in children, the tricuspid valve plasty with biventricular correction should take preference over tricuspid valve replacement or a single-ventricle pathway. Surgical repair consists of (1) reconstruction of the tricuspid valve, (2) consideration of plication or resection of the atrialized right ventricle, especially when it is thin-walled and moves paradoxically, (3) closure of the atrial communication and correction of any associated anomalies, (4) surgery for arrhythmia, and (5) right atrium reduction atroplasty. In special situations, the concept of 1½ ventricular repair or of a single-ventricular pathway should be adopted. Heart transplantation is the therapy of last resort. Reconstructive surgery is based either on the principle of anatomical repair (cone reconstruction) or on monoleaflet repair.

The cone reconstruction moves the new tricuspid valve orifice to the true valve annulus, restoring the normal anatomical location of the tricuspid valve,
resulting in central diastolic blood flow and full coaptation of the leaflets. The normalization of the right ventricular volume and morphology is achieved by longitudinal plication of the atrialized right ventricle.

The important operative steps are as follow:

1. “Correction” of failure of delamination
   The anterior and posterior tricuspid valve leaflets are detached from the hinge-point and mobilized from their anomalous attachments in the right ventricle. Only the normal attachments of the anterior leaflet to the true tricuspid annulus and the proper subvalvular apparatus are left in place.

2. Creating a cone
   The free edge of this valve complex is rotated clockwise for suture to the septal border of the anterior leaflet, thus creating a cone, the vertex of which remains fixed at the right ventricular apex.

3. Exclusion of the atrialized right ventricle
   The atrialized part of the right ventricle is longitudinally plicated to exclude its thin part.

4. Attachment of the tricuspid valve to the annulus

The new valvular annulus is constructed at the anatomically correct level by means of plication of the true tricuspid annulus to match the proximal circumference of the cone-shaped valve. The base of the cone is sutured to the new valve annulus.

   Additionally, the atrial septal defect is closed in a valved fashion. The redundant right atrium is excised from each side of the atriotomy and then the atriotomy is closed.
Cone Reconstruction with Longitudinal Plication of the Atrialized Ventricle in the Newborn

Patient Characteristics

<table>
<thead>
<tr>
<th>Age at operation: 30 days</th>
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<tbody>
<tr>
<td>Diagnosis:</td>
</tr>
<tr>
<td>1. Ebstein’s anomaly</td>
</tr>
<tr>
<td>2. Secundum atrial septal defect</td>
</tr>
<tr>
<td>3. Patent ductus arteriosus</td>
</tr>
<tr>
<td>4. Hydrops fetalis</td>
</tr>
<tr>
<td>5. Supraventricular reentry tachycardia</td>
</tr>
<tr>
<td>6. Bilateral chylothorax</td>
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<tr>
<td>History:</td>
</tr>
<tr>
<td>1. Prenatally diagnosed, born by Cesarean section</td>
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<tr>
<td>2. Intubated and ventilated, administration of prostaglandins, otherwise cyanotic</td>
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<tr>
<td>3. On continuous positive-airway pressure for a short time; placement of peritoneal catheter</td>
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<tr>
<td>4. Reintubated because of circulatory collapse due to supraventricular tachycardia</td>
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<tr>
<td>5. Resuscitated because of ventricular tachycardia, with subsequent placement of temporary intravenous pacemaker wires</td>
</tr>
<tr>
<td>6. Placement of temporary epicardial wires to replace intravenous ones, which were unreliable</td>
</tr>
<tr>
<td>7. Resuscitation and subsequent extracorporeal membrane oxygenation support for 7 days</td>
</tr>
<tr>
<td>8. Operation indicated due to failure to wean patient from extracorporeal membrane oxygenation support.</td>
</tr>
</tbody>
</table>

Procedure:

1. Cone reconstruction with longitudinal plication of the atrialized portion of the right ventricle
2. Partial closure of the atrial septal defect
3. Downsizing of the right atrium
4. Patent ductus arteriosus closure

Specific Steps of Operation

Preoperative findings.
**Clip 2**

Assessment of anatomy.

**Clip 3**

Detachment of the leaflets of the tricuspid valve.
“Correction” of failure of delamination.

Creating a cone.
Ebstein’s Anomaly

**DVD Clip 6**

Exclusion of the atrialized right ventricle.

**DVD Clip 7**

Attachment of the tricuspid valve to the annulus.
**Clip 8**

Plication of the annulus.

**Clip 9**

Final reattachment of the base of the cone. In addition, the atrial septal defect is partially closed, leaving a 3-mm fenestration. The redundant right atrium is excised from each side of the atriotomy, and then the atriotomy is closed.
The echocardiogram before discharge.
Cone Reconstruction with Longitudinal Plication of the Atrialized Ventricle in the Infant

Patient Characteristics

<table>
<thead>
<tr>
<th>Age at operation: 6 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Procedure:</td>
</tr>
<tr>
<td>1. Cone reconstruction with longitudinal plication of the atrialized portion of the right ventricle</td>
</tr>
<tr>
<td>2. Direct closure of the atrial septal defect</td>
</tr>
<tr>
<td>3. Downsizing of the right atrium</td>
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</tbody>
</table>

Diagnosis:
1. Ebstein’s anomaly
2. Secundum atrial septal defect

History:
1. Prenatally diagnosed, postnatal adaptation without difficulties
2. Gradual progression of cyanosis and failure to thrive

Specific Steps of Operation

Fig. 5.1. Preoperative echocardiogram. The preoperative echocardiogram shows the displacement of the septal leaflet and the size and position of the anterior leaflet of the tricuspid valve (a). The Doppler echocardiogram shows moderate tricuspid regurgitation (b). RA right atrium, ARV atrialized right ventricle, AL anterior leaflet, SL septal leaflet
Fig. 5.2. Intracardiac anatomy. There is severe displacement of the posterior and septal leaflets (a). Vessel loops surround the fibrous and muscular attachments of the leading edges of the leaflet to the underlying myocardium. Note the atrialized portion of the right ventricle and position of the right coronary artery (b). 

RCA right coronary artery, AL anterior leaflet, PL posterior leaflet, SL septal leaflet, ARV atrialized right ventricle

Fig. 5.3. Creating a cone. All fibrous and muscular attachments between the body of the leaflets and the right ventricular myocardium are incised. The cone reconstruction is completed by rotating the posterior leaflet clockwise to meet the proximal edge of the septal leaflet. 

AL anterior leaflet, PL posterior leaflet, SL septal leaflet
Cone Reconstruction without Plication of the Atrialized Ventricle

Patient Characteristics

**Age at operation**: 3.5 years  
**Diagnosis**:  
1. Ebstein’s anomaly  
2. Secundum atrial septal defect  
**History**:  
Gradual progression of cyanosis and failure to thrive

**Procedure**:  
1. Cone reconstruction without plication of the atrialized portion of the right ventricle  
2. Direct closure of the atrial septal defect  
3. Downsizing of the right atrium

Specific Steps of Operation

Assessment of the extra- and intracardiac anatomy.
Ebstein’s Anomaly

**Clip 2**
Detachment of the leaflets of the tricuspid valve.

**Clip 3**
Detachment of the linear attachments of the posterior leaflet.
**Clip 4**

Detachment of the septal leaflet.

**Clip 5**

Creating a cone.
**Ebstein’s Anomaly**

**Clip 6**
Attachment of the tricuspid valve to the annulus.

**Clip 7**
Lateral annuloplasty.
**DVD Clip8**

Atrial septal defect closure and reduction atriotomy.

**DVD Clip9**

The echocardiogram before discharge.

**DVD fullversion**

[Image]
Recommended Reading


Ebstein W (1866) Über einen sehr seltenen Fall von Insuffizienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. Arch Anat Physiol 7:238–254


