Surgery For Ebstein Anomaly

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• No disclosures
Ebstein Anomaly

- Morphology first described by Ebstein in 1866
- TV leaflets variably adherent to RV myocardium
- Spectrum of disease
- Clinically documented by Taussig in 1950
- Rare
- Bimodal age at presentation
Neonatal Ebstein’s: Predictors of death

- Cardiothoracic ratio greater than 0.85 (100% fatal)
- Echocardiography score grade 4/4 (>1.5:1; 100% fatal)
- Echocardiography score grade 3/4 (>1.1:1) and cyanosis (100% fatal)
- Severe tricuspid regurgitation (mostly fatal)
- Echocardiography score grade 3/4 (>1.1:1; 45% fatal in infancy)

Pavlova, M. Am Heart J 1998;135:1081-1085
Yetman, AT. Am J Cardiol 1998;81:749-754
MRI evaluation

Improved functional assessment
RV and RA volumes
RV function
Delayed Enhancement

Objectively assessment over time
Indications for repair

• Symptoms
• Deteriorating exercise capacity
• Heart failure (NYHA class III-IV)
• Cyanosis
• Paradoxical embolism
• Progressive RV/RA enlargement
• New onset arrhythmias
Carpentier classification

Anatomy is highly variable
Anatomic variability continues to be a challenge for the surgeon
Ventricular dysfunction

- Ineffective LV filling
  - due to “to and fro” flow into the right ventricle / right atrium
  - Increase right atrial capacitance
- “Pancaked” Left ventricle

RV myopathy
Thinned out and fibrotic muscle
STS database: common interventions
Center Experience

- 82 centers
  - Median annual experience 1 case/yr
    - (IQR 0.5-1.8)
  - Highest volume center: 8.3 cases/yr

- Neonates + infants
  - 63 centers
  - Median annual experience 0.5 cases/yr
    - (IQR 0.1-1.0, maximum 5.2)
Arrhythmias are common among adult Ebstein anomaly

<table>
<thead>
<tr>
<th>Preoperative Risk Factor</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any Preoperative Risk Factor</td>
<td>29/89 (32.6%)</td>
</tr>
<tr>
<td>Acidosis</td>
<td>0/89 (0.0%)</td>
</tr>
<tr>
<td><strong>Arrhythmia</strong></td>
<td><strong>19/89 (21.4%)</strong></td>
</tr>
<tr>
<td>Shock</td>
<td>0/89 (0.0%)</td>
</tr>
<tr>
<td>Mechanical Ventilation</td>
<td>0/89 (0.0%)</td>
</tr>
<tr>
<td>Previous Cardiac Procedure</td>
<td>32/93 (34.4%)</td>
</tr>
</tbody>
</table>

Surgical considerations

• Mechanism of TR, # of jets, location, valve morphology
• Delamination and tethering of each leaflet
• Leaflet edges, fenestrations
• Annular dilatation
• Size and function of the RV
• Ventricular septum and LV function
• Age of the patient
Danielson repair

1972-1982
42 pts
Monocusp
Horizontal plication
No sutures on the IVS
81% repair
2 pts (5%) had a Glenn
7.1% mortality
Carpentier Technique

reconstruct a normal shaped RV
longitudinal plication of atrialized RV

reposition the TV at the normal level

Monocuspid or bicuspid
Surgery for Ebstein's Anomaly: The Clinical and Echocardiographic Evaluation of a New Technique

JAN M. QUAEGEBEUR, MD,* NARAYANSWAMI SREERAM, MRCP, ALAN G. FRASER, MRCP, AD J.J.C. BOGERS, MD, OLIVER F. W. STUMPER, MD, JOHN HESS, MD, EGBERT BOS, MD, GEORGE R. SUTHERLAND, FRCP

Rotterdam, The Netherlands

1988-1990
10 pts (4-44 yrs)
9 repairs
Bi leaflet valve
Vertical plication
No atrial reduction
No annuloplasty ring
No transection of PM
No heart block
No mortality
f/u 11.7 mo
Common elements of TV repair

Basic strategy

• Monocusp repair (allows coaptation against the ventricular septum)
  • based on anterior leaflet
  • degree of a RV and annular dilatation
  • tethering of the anterior leaflet

• Sebening stitch

Failures

• Due to focus on the annulus not on delamination
• Attempts to repair with poor valve anatomy
  • marked linear attachment
  • marked leaflet muscularization
  • complete absence of septal leaflet.

• Massive annular dilatation
• Older age
• Pulmonary HTN
The cone reconstruction of the tricuspid valve in Ebstein’s anomaly. The operation: early and midterm results

José Pedro da Silva, MD\textsuperscript{a,1}, José Francisco Baumgratz, MD\textsuperscript{b}, Luciana da Fonseca, MD\textsuperscript{b}, Sônia Meiken Franchi, MD\textsuperscript{a}, Lilian Maria Lopes, MD\textsuperscript{b}, Gláucia Maria P. Tavares, MD\textsuperscript{a}, Andressa Mussi Soares, MD\textsuperscript{a}, Luiz Felipe Moreira, MD\textsuperscript{a}, Miguel Barbero-Marcial, MD\textsuperscript{a}
One (2.5%) hospital death and 1 late death

↓ in TR grade from 3.6 ± 0.5 to 1.2 ± 0.5 ($P < .0001$).

At a mean follow-up of 4 years:
Functional class (NYHA) improved from 2.6 ± 0.7 to 1.2 ± 0.4 ($P < .0001$).
Valve inspection
Leaflet detachment
Mobilization of all leaflet tissue (septal)
Annular reduction
Reimplantation at the annular level
Incorporation of septal leaflet
Leaflet augmentation
Creation of neochordae (fenestrations)
Annuloplasty
Circumferential leaflet coverage leads to improved competency.
Surgical technique

- Circumferential leaflet tissue repair (360)
- Valve anchored at the true annulus
- Ringed annuloplasty
- Leaflet augmentation
- Autologous chordae
- Bidirectional Glenn
• Commonly annular reduction is necessary/Use pledgeted suture

• RCA runs in the right AV groove. Avoid kinking or occlusion of the RCA

• Liberal use of the Sebening stitch. Avoid dimple on the RV free wall (patch augmentation of the anterior leaflet)

• Plication of the RV should be confined to the smooth non trabeculated inferior wall. Avoid the IVS (PDArt)

• Decision about the feasibility of a good durable repair should be made early.

• Repair rate for adults is ~80%
Important considerations

• Aortic occlusion and cardioplegic arrest for precise suture placement, avoidance of coronary or conduction tissue injury, and minimizing motion trauma during suture placement.

• The time required to perform EA repair can be lengthy, even in experienced hands, and the consequences of prolonged operation in a patient with depressed ventricular function may not be prudent.
Cone reconstruction for Ebstein’s anomaly: Patient outcomes, biventricular function, and cardiopulmonary exercise capacity

Michael Ibrahim, MD, PhD, Victor T. Tsung, MD, FRCS, Maryanne Caruana, MD, Marina L. Hughes, DPhil, FRACP, Synetta Jenkyns, BD, Elodie Perdrea, MD, Alessandro Giardini, MD, and Jan Marek, MD, PhD.

TABLE 2. Cardiac magnetic resonance imaging analysis of effect of cone reconstruction on right ventricle (RV) and left ventricle (LV) volumes

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV end-systolic volume</td>
<td>112.1 ± 80.6</td>
<td>91.0 ± 45.3</td>
<td>.54</td>
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<tr>
<td>RV end-diastolic volume</td>
<td>166 ± 66.3</td>
<td>145.9 ± 56.1</td>
<td>.24</td>
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<tr>
<td>LV end-systolic volume</td>
<td>19.8 ± 8.9</td>
<td>22.4 ± 10.4</td>
<td>.25</td>
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<tr>
<td>LV end-diastolic volume</td>
<td>49.4 ± 14.4</td>
<td>60.14 ± 14.5</td>
<td>.006</td>
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</tbody>
</table>

All values are indexed for body surface area. Boldface type indicates statistical significance. RV, Right ventricle; LV, left ventricle.

NYHA

<table>
<thead>
<tr>
<th>NYHA</th>
<th>Number of patients</th>
<th>Pre-Op</th>
<th>Post-Op</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>15</td>
<td></td>
<td></td>
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<tr>
<td>3</td>
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<td></td>
</tr>
<tr>
<td>4</td>
<td>5</td>
<td></td>
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Indexed Forward Flow in MPA

p < 0.01
Summary

- Ebstein anomaly exhibits a broad range of anatomic features commonly leading to severe TR
- Due to morphologic variability, no perfect repair for all patients with EA
- The Cone reconstruction is the only technique that provides circumferential leaflet coverage and is highly effective to restore valve competency.
- Reduced right ventricular function continues to be a challenge, as is the need for reoperation for recurrent tricuspid regurgitation
- Steep learning curve requires institutional and surgeon experience for optimal outcomes
- Improved results should influence timing of repair.
- When severe RV dysfunction is present, TVR should be considered early.
Muchas Gracias!
Ebstein anomaly accounts for 1% of all congenital heart disease. It is a right ventricular myopathy with failure of tricuspid valve delamination and highly variable tricuspid valve morphology that usually results in severe regurgitation. It is the only congenital heart lesion that has a range of clinical presentations, from the severely symptomatic neonate to an asymptomatic adult. Neonatal operation has high operative mortality, whereas operation performed beyond infancy and into adulthood has low operative mortality. Late survival and quality of life for hospital survivors are excellent for the majority of patients in all age brackets. Atrial tachyarrhythmias are the most common late complication. There have been more techniques of tricuspid repair reported in the literature than any other congenital or acquired cardiac lesion. This is largely due to the infinite anatomic variability encountered with this anomaly. The cone reconstruction of Ebstein anomaly can achieve near anatomic restoration of the tricuspid valve anatomy. Early and intermediate results with these repairs are promising. Reduced right ventricular function continues to be a challenge for some patients, as is the need for reoperation for recurrent tricuspid regurgitation. The purpose of this article is to outline the current standard of care for diagnosis and treatment of Ebstein anomaly and describe innovative strategies to address poor right ventricular function and associated right-sided heart failure.
Subtitle

• Bullet number one
• Bullet number two
  • Sub-bullet number three
  • Sub-bullet number four
• Bullet number five

Description of Graph or Table

- 1st Qtr
- 2nd Qtr
- 3rd Qtr
- 4th Qtr
SLIDE TITLE HEADLINE

PLACEHOLDER
Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein’s anomaly: benefits in high risk patients

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• RV unloading, decreases RV dilatation
• Improves LV/RV interaction
• Increases effective LV filling (Cardiac output)
• Allows tighter annuloplasty
• Not feasible in neonates and youg infants

Chaveaud et al. EJCTS 1998;13: 514-19