The Surgical Repair of Neonatal Ebstein’s

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Wilhelm Ebstein

Born Nov. 27, 1836; Jauer, Silezia, Poland
Disciple of Rudolf Virchow
Jul. 11, 1859: Medical degree
Career briefly interrupted by Franco-Prussian War
July 9, 1864: Autopsy on Joseph Prescher

The tricuspid valve was extremely abnormal in appearance. A membrane originated from a normally developed annulus fibrosus. Fifteen millimeters below the annulus fibrosus a malformed leaflet originated from the endocardium.
Ebstein’s Anomaly of the Tricuspid Valve

- Congenital anomaly of the heart
- Characterized by an inferiorly displaced septal leaflet of the tricuspid valve
- Enlarged right atrium, composed of the “true” right atrium and an “atrialized” portion of the right ventricle
- Varying degrees of tricuspid regurgitation and pulmonary obstruction

Clinical Features of Ebstein’s Anomaly

- Cyanosis from R to L shunting at the atrial level
- CXR: Pronounced cardiomegaly secondary to right atrial enlargement
- EKG: R axis deviation and R atrial enlargement – WPW in 25%

CXR of Ebstein’s

EKG-Ebsteins anomaly
Echocardiogram of Ebstein’s:
apical 4-chamber view

Echocardiogram of Ebstein’s:
color doppler

Echocardiogram of Ebstein’s:
parasternal short axis view

**The Big Picture**

- Over 90 neonates with Ebstein’s Anomaly
  born each year in the USA**
- Early mortality = 25%
- Conservatively managed neonates have
  22% early mortality

**Management Algorithm**

**Birth**
- Stable
- Critical unstable
  - Supplemental oxygen for adequate cardiac output (SPO2)
  - Intubate, paralysis, inotropic support, NO, PGE1
  - As PVR ↓, remove support and follow closely for adequate cardiac output ± PGE1
  - Continue observation:
    - Frequent echocardiogram
    - End-organ function evaluation
    - Early surgery

**Symptomatic Neonates with EA**

**Medical Management**
- Adequate CO
- Adequate SaO2
- Worse T waveform
- Low CO
- Severe cyanosis

**Continued observation**
- Cardiac output ± PGE1
- Surgical evaluation
- Cardiopulmonary bypass

**GOSE Score**

Ratio of areas of the cardiac chambers translates to mortality risk

<table>
<thead>
<tr>
<th>Area of (RA + aRV)</th>
<th>Area of (RV + LV + LA)</th>
<th>GOSE Ratio</th>
<th>Mortality Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>I</td>
<td>&lt;0.5</td>
<td>8%</td>
</tr>
<tr>
<td>II</td>
<td>II</td>
<td>0.5-1.0</td>
<td>8%</td>
</tr>
<tr>
<td>III (acyanotic)</td>
<td>III</td>
<td>1.1-1.4</td>
<td>10% early, 45% late</td>
</tr>
<tr>
<td>III (cyanotic)</td>
<td>III</td>
<td>1.1-1.4</td>
<td>100%</td>
</tr>
<tr>
<td>IV</td>
<td>IV</td>
<td>&gt;1.5</td>
<td>100%</td>
</tr>
</tbody>
</table>

**Who needs early operation?**

- Neonates with EA and:
  - Anatomic pulmonary atresia
  - GOSE III and IV
  - Severe tricuspid regurgitation (grade 3-4/4)
  - Ventilator dependency
  - Failed medical therapy
  - GOSE II (acyanotic)
  - Increasing cyanosis
**Which operation to do?**

**Anatomic Pulmonary Atresia**
- Diminutive RV with Mild TR
- Small RV ± reduction aortoplasty
- Small RV ± sildenafil
- Normal LV Size ± early BDG

**Small RV**
- Shunt palliation
- Biventricular Repair with RV-PA conduit

**Normal LV Size**
- Biventricular Repair with RV-PA conduit
- BTS ± reduction aortoplasty

**Transplant**

**Functional Pulmonary Atresia**
- Small LV Size

**Small Left Ventricle**

**Which operation to do?**

**Functional Pulmonary Atresia**
- Small LV Size
- Small LV Size ± early BDG
- Small LV Size ± late BDG

**Transplant**
Apical 4-Chamber Images within 1st Week of Life:
Severe TV Displacement and Mild TR; intubated on iNO

Short Axis Images within 1st Week of Life:
Severe TV Displacement and Mild TR; intubated on iNO

**WHICH OPERATION TO DO?**

- Functional Pulmonary Atresia
  - Normal LV Size
  - Small LV Size
  - Transplant
  - Emergent ligation of MPA
  - Starnes palliation
  - BTS or Hybrid
  - Complete Biventricular Repair

**SEVERE TRICUSPID REGURGITATION**
Neonates with EA 1994-2010 (n=44)

- Medical management (n=15)
- Medical management incomplete (n=4)
- Complete repair in infancy (9/15 alive)
- Partial TET + Pulmonary Atresia (n=2)
- BTS + Pulmonary valvotomy (n=2)

- Neonatal surgical intervention (n=23)
- Starnes palliation (n=7)
- Partial TET (n=1)
- Complete Repair (n=20)
- No Pulmonary Atresia (9/10 alive)
- Minimal TR, Pulmonary Atresia (n=2)

- Died before intervention (n=2)
- Complete repair in infancy (9/15 alive)

Mean wt: 3.9 ± 2.0 kg (1.9 – 8.6 kg)
Mean follow-up: 5.8 ± 4.5 yrs (0.2-16 yrs)

The First Neonate: 16 Years Later

Apical 4-Chamber Images October, 2009 (original surgery in 1994)