Ebstein Anomaly
Spectrum of Disease and
the Role of Imaging Modalities

Department of Pediatrics
Chonnam National University Medical School
Young Kuk Cho
Overview

- Ebstein anomaly def.
- Classification
  - by Echo.
  - Carpentier
  - Celermajer index
  - anatomic findings at surgery
- Imaging modality
  - Chest X-ray
  - ECG
  - Echocardiography
  - Cardiac MRI
Ebstein anomaly

Ebstein anomaly


Marked cardiomegaly

Severe Ebstein’s malformation

Alain Carpentier, et al. Carpentier's Reconstructive Valve Surgery: From Valve Analysis to Valve Reconstruction CHAPTER 24, 247-257

AV valve formation

Failure of delamination of endocardial cushion tissue

http://www.studyblue.com/notes/note/n/heart-embryo/deck/4384504
Apical displacement

Class. by Echo.

Simple
mild, moderate, or severe

- extent of apical displacement of vv leaflets
- degree of TR
- degree of RV dilatation & dysfunction

## Carpentier class.

<table>
<thead>
<tr>
<th>SL, PL displace.</th>
<th>+</th>
<th>+ +</th>
<th>+ + +</th>
<th>“SAC”</th>
</tr>
</thead>
<tbody>
<tr>
<td>AL</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Morphology</td>
<td>Normal</td>
<td>Abnormal chordae</td>
<td>Partial adhesions</td>
<td>Extensive adhesions</td>
</tr>
<tr>
<td>Mobility</td>
<td>Normal</td>
<td>Normal</td>
<td>Restricted</td>
<td>Absent</td>
</tr>
<tr>
<td>a RV</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Size</td>
<td>Small</td>
<td>Large</td>
<td>Large</td>
<td>SAC</td>
</tr>
<tr>
<td>Contractility</td>
<td>Preserved</td>
<td>Reduced</td>
<td>Minimal</td>
<td>Absent</td>
</tr>
<tr>
<td>RV</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Size</td>
<td>Normal</td>
<td>Reduced</td>
<td>Small</td>
<td>Infundibulum</td>
</tr>
<tr>
<td>Contractility</td>
<td>Preserved</td>
<td>Reduced /reduced</td>
<td>Reduced</td>
<td>Severely depressed</td>
</tr>
</tbody>
</table>
Class. by anatomic findings at surgery

<table>
<thead>
<tr>
<th>Type</th>
<th>Anterior Leaflet</th>
<th>Posterior Leaflet</th>
<th>Septal Leaflet</th>
<th>Atrialized RV Chamber Size</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Larger</td>
<td>Mobile</td>
<td>Apically displaced, dysplastic, or absent.</td>
<td>Varies from relatively small to large.</td>
</tr>
<tr>
<td>II</td>
<td>Relatively small and displaced in a spiral fashion toward the apex.</td>
<td></td>
<td></td>
<td>Moderately large.</td>
</tr>
<tr>
<td>III</td>
<td>Restricted motion Shortened, fused, and tethered chordae. Direct insertion of papillary muscles into the anterior leaflet is frequently present.</td>
<td>Displaced, dysplastic, and usually not reconstructible.</td>
<td></td>
<td>Large</td>
</tr>
<tr>
<td>IV</td>
<td>Severely deformed Few or no chordae. Direct insertion of the papillary muscles into the leading edge of the valve is common.</td>
<td>Typically dysplastic or absent</td>
<td>Represented by a ridge of fibrous material descending apically from the membranous septum.</td>
<td>Nearly the entire RV cavity is atrialized. TV tissue is displaced into the RVOT and may cause obstruction of blood flow (functional tricuspid stenosis).</td>
</tr>
</tbody>
</table>

Celermajer index score

In neonate.

\[
\frac{\text{RA area} + \text{aRV area}}{\text{fRV area} + \text{LA area} + \text{LV area}}
\]

<table>
<thead>
<tr>
<th>Grade</th>
<th>ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>&lt; 0.5</td>
</tr>
<tr>
<td>2</td>
<td>0.5 ~ 0.99</td>
</tr>
<tr>
<td>3</td>
<td>1 ~ 1.49</td>
</tr>
<tr>
<td>4</td>
<td>&gt; 1.5</td>
</tr>
</tbody>
</table>

# Great Ormond Street Echocardiography (GOSE) score

<table>
<thead>
<tr>
<th>GOSE score</th>
<th>ratio</th>
<th>Mortality (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-2</td>
<td>&lt; 1.0</td>
<td>8</td>
</tr>
<tr>
<td>3 (acyanotic)</td>
<td>1.1 ~ 1.4</td>
<td>10 (early)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>45 (late)</td>
</tr>
<tr>
<td>3 (cyanotic)</td>
<td>1.1 ~ 1.4</td>
<td>100</td>
</tr>
<tr>
<td>4</td>
<td>&gt; 1.5</td>
<td>100</td>
</tr>
</tbody>
</table>

Chest X-ray

Electrocardiography

Electrocardiography

Moss & Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult, 8th edit. 889-912
Echocardiography

Armstrong et al, Feigenbaum’s Echocardiography, Lippincott Williams & Wilkins
Echocardiography

Armstrong et al, Feigenbaum’s Echocardiography, Lippincott Williams & Wilkins
Echocardiography
Fetal Echocardiography

Szwast, Anita - Fetal Cardiovascular Imaging: A Disease-Based Approach, 287-296
Fetal Echocardiography

Szwast, Anita - Fetal Cardiovascular Imaging: A Disease-Based Approach, 287-296
Fetal Echocardiography
Angiocardiogram

Perloff, Joseph K., MD - Clinical Recognition of Congenital Heart Disease, 176-195
Cardiac CT

Cardiac MRI

Dimopoulous, Konstantinos - Cases in Adult Congenital Heart Disease, 167-172
Cardiac MRI

Cardiac Magnetic Resonance Imaging and the Assessment of Ebstein Anomaly in Adults

Sergey Yalonetsky, MD\textsuperscript{a}, Daniel Tobler, MD\textsuperscript{a}, Matthias Greutmann, MD\textsuperscript{a}, Andrew M. Crean, MD\textsuperscript{a,b}, Bernd J. Wintersperger, MD\textsuperscript{b,c}, Elsie T. Nguyen, MD\textsuperscript{b}, Erwin N. Oechslin, MD\textsuperscript{a}, Candice K. Silversides, MD\textsuperscript{a}, and Rachel M. Wald, MD\textsuperscript{a,d,e,g}

No published studies have evaluated the role of cardiac magnetic resonance (CMR) imaging for the assessment of Ebstein anomaly. Our objective was to evaluate the right heart characteristics in adults with unrepaired Ebstein anomaly using contemporary CMR imaging techniques. Consecutive patients with unrepaired Ebstein anomaly and complete CMR studies from 2004 to 2009 were identified \((n = 32)\). Volumetric measurements were obtained from the short-axis and axial views, including assessment of the functional right ventricular (RV) end-diastolic volume (EDV) and end-systolic volume. The volume of the atrialized portion of the right ventricle in end-diastole was calculated as the difference between the total RVEDV and the functional RVEDV. The reproducibility of the measurements in the axial and short-axis views was determined within and between observers. The median value derived from the short-axis and axial views was 136 ml/m\(^2\) (range 59 to 347) and 136 ml/m\(^2\) (range 63 to 342) for the functional RVEDV, 153 ml/m\(^2\) (range 64 to 441) and 154 ml/m\(^2\) (range 67 to 436) for the total RVEDV, 49\% (range 32\% to 46\%) and 50\% (range 40\% to 64\%) for the functional RV ejection fraction, respectively. The axial measurements demonstrated lower intraobserver and interobserver variability than the short-axis approach for all values, with the exception of the intraobserver functional RVEDV and interobserver total RVEDV for which the limits of agreement and variance were not significantly different between the 2 views. In conclusion, measurements of right heart size and systolic function in patients with Ebstein anomaly can be reliably achieved using CMR imaging. Axial imaging appeared to provide more reproducible data than that obtained from the short-axis views. © 2011 Elsevier Inc. All rights reserved. (Am J Cardiol 2011;107:767–773)
Cardiac MRI

Summary

Ebstein anomaly
- SL & PL to the underlying myocardium
- Downward displacement of the functional annulus
  - > 8 mm/m² BSA
- Atrialized RV dilation
- AL: redundancy, fenestrations, & tethering

Classification
- Echo.
- Carpentier
- Celermajer score (GOSE)
- Anatomic findings during surgery

Dx & Assessment
- mainly echocardiography
- Cardiac MRI