Ebstein's and accessory pathways, and how I avoid the AV node

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Research grants: Baylis Medical, Spectrum Dynamics, Catheter Precision
Consultancy: Biosense Webster, Stereotaxis, Medilumics
• Congenital heart disease patients are very different from “normal” patients with regards to:
  – Anatomy of the cardiac chambers (eg. univentricular heart)
  – Location and properties of the conduction system
  – Frequently surgically corrections have altered the substrate
  – Access to target chambers may be difficult
Ablation in ACHD

**Ebstein’s anomaly**

- First described by Wilhelm Ebstein in 1866
  
  
  - characterised by apical displacement of the septal and posterior tricuspid valve leaflets, leading to part of the right ventricle becoming part of the right atrium, with a variable degree of malformation and displacement of the anterior leaflet.

- Epidemiology about 0.5% of cases of CHD

- True prevalence unknown as mild forms often undiagnosed
Ablation in ACHD

Ebstein's anomaly

- The abnormality of the tricuspid valve leads to tricuspid regurgitation. The degree of tricuspid regurgitation is variable, ranging from mild to severe regurgitation.
- Presentation is often between the ages of 10 and 30 years but can present at various stages of life:
  - Fetal life: diagnosed incidentally by echocardiography.
  - Neonatal life and infancy: presents with cyanosis and/or severe heart failure. Symptoms presenting in infancy often improve as the pulmonary vascular resistance decreases.
  - Adult life: fatigue, exertional dyspnoea, cyanosis, tricuspid regurgitation and/or right heart failure and palpitations.
Wilhelm Ebstein first described a patient with cardiac defects typical of Ebstein anomaly in 1866. - German internist, born November 27, 1836, Jauer (Schlesien); died October 22, 1912. Göttingen

In 1927, Alfred Arnstein suggested the name Ebstein's anomaly for these defects
Review of Anatomy

Normal

Ebstein’s Anomaly
Abstract

Objective—To review the anatomical structure of the right atrioventricular junction, including the specialised atrioventricular conduction system, in hearts with Ebstein’s malformation, to identify potential substrates for the abnormalities in conduction.

Methods—Five heart specimens representing the morphological spectrum of Ebstein malformation were examined grossly and histologically.

Results—On the endocardial surface, the atrioventricular junction was marked by a faint line in two hearts, and by a small ridge in the other three. Analysis of the right parietal junction in four hearts revealed only two accessory muscular atrioventricular connections. A plane of fibrofatty tissue separated atrial from ventricular myocardium in the right parietal junction in all hearts. The compact atrioventricular node was closer to the coronary sinus than usual. Accessory nodoventricular connections were present in four hearts, while accessory fasciculo-ventricular connections were found in one. The right bundle branch was hypoplastic or absent in four hearts.

Conclusions—in this small series, the parietal atrioventricular junction was better developed than previously thought. Structural abnormalities of the atrioventricular conduction system, however, were present. These may account for some of the conduction abnormalities frequently observed with the Ebstein malformation.
The body of the atrioventricular (AV) node (within dotted line) is adjacent to the mouth of the coronary sinus in heart 3.

(B) A more inferior section of the same heart shows the inferior nodal extensions (within white dots) at the level of the Thebesian valve.

(C) This section from heart 5 shows the atrioventricular (AV) node at a level close to the mouth of the coronary sinus. The hinge of the septal leaflet is displaced from the atrioventricular junction.

(D) This magnified view shows the extension (white star) of the conduction tissues from the atrioventricular node into the central fibrous body at the transition to the penetrating bundle.

(E) This inferior section also from heart 5 is through the middle of the coronary sinus orifice. It is magnified in (F) to show the continuation of the nodal body at this level.

Ho et al, Heart 2000;83:444–449
12 lead ECG: more than 1 AP?
How to map the AP insertion site I

Cappato et al. Circulation, 1996 Aug 1;94(3):376-83
How to map the AP insertion site II

Cappato et al. Circulation, 1996 Aug 1;94(3):376-83
Epicardial AP ablation RCA guiding catheter

How to map the AP insertion site III

Cappato et al. Circulation, 1996 Aug 1;94(3):376-83
How to map the AP insertion site IV
Review of AP insertion sites

Cappato et al. Circulation, 1996 Aug 1;94(3):376-83
RBH/SE Ebstein experience

Technical recommendations

- CARTO 3
- 3D roadmap (CMR)
- Catheters:
  - SF ST bidirectional
  - His/RV
  - Decapolar in CS
  - (HRA)
- Steerable sheath if necessary
- Assisted sedation
Patient cohort

- Mean age 37±17 yrs
- Mean weight 70.6 ±7.4 kg, mean height 1.68 ±0.17 m
- 5 had a previous surgical repair for Ebstein
- 9 had a previous ablation procedure
- Total of 25 procedures: 7 EP and 18 ablations (7 WPW, 6 AF, 5 AT)
- Remote magnetic navigation used in 8 (32%) procedures
- 22/25 procedures were carried out using a mapping system: 18 EAM (82%), 2 ecVue (9%), 2 HDM (9%)
Procedural Workflow

Non-Invasive ‘Bedside’

Pre-Procedural Planning

252 ELECTRODE VEST

HEART-TORSO GEOMETRY (CT)

SOFTWARE GENERATES 3D ELECTRICAL MAPS

REAL-TIME MAPPING & DIAGNOSIS
Case 1

- 18 y old pt with Ebstein’s anomaly
- previous ablation of a right lateral accessory pathway 2009 now representing with preexcitation and paroxysmal palpitations
Earliest ventricular activation at the postero-lateral tricuspid annulus (yellow)
Spike mapping, low current, no ventricular capture
Spike map

Earliest v activation
Pathway block with ablation
Ventricular activation in SR without preexcitation

Ventricular activation in SR with preexcitation
Example of 3D picture in picture display

13 yrs Ebstein patient  3xTV OP + perc. Melody valve c WPW: Fluoro exposure: 1.1 min
RBH/SE Ebstein experience

Results

- Pre-procedural imaging used in 14/25 (56%): 4 CT and 10 CMR scan
- Acute success 100%
- Median procedural time 165 [120,191] min
- Median DAP 223.4 [122.5, 996.95] mGy*sqcm
- Median FT 3.29 [1.23, 9.35] min

<table>
<thead>
<tr>
<th></th>
<th>All</th>
<th>EP only</th>
<th>WPW</th>
<th>AF</th>
<th>AT</th>
</tr>
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<tbody>
<tr>
<td>Total time (min)</td>
<td>165 [120-191]</td>
<td>85 [66-120]</td>
<td>185 [165.5-190.5]</td>
<td>167 [140-282]</td>
<td>185 [176-219]</td>
</tr>
<tr>
<td>DAP (mGy*sqcm)</td>
<td>223.4 [122.5-996.95]</td>
<td>197 [55.2-382.9]</td>
<td>145.2 [125.3-571.5]</td>
<td>282.5 [16.5-1679.6]</td>
<td>388.3 [137.1-585.9]</td>
</tr>
</tbody>
</table>
Contemporary outcomes Ebstein

### TABLE 3
Patient Characteristics and Procedural Data

<table>
<thead>
<tr>
<th></th>
<th>AP or scAVC n = 25</th>
<th>IART n = 5</th>
<th>CTI-AFL n = 5</th>
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<tbody>
<tr>
<td>Age at first arrhythmia (years)</td>
<td>10 (0, 29)</td>
<td>29 (6, 52)</td>
<td>23 (14, 29)</td>
</tr>
<tr>
<td>Cardioversion before ablation, n (%)</td>
<td>10 (40%)</td>
<td>4 (80%)</td>
<td>3 (60%)</td>
</tr>
<tr>
<td>Age at first ablation (years)</td>
<td>20 (5, 56)</td>
<td>33 (6, 61)</td>
<td>27 (14, 50)</td>
</tr>
<tr>
<td>Number of ablation targets</td>
<td>34</td>
<td>8</td>
<td>5</td>
</tr>
<tr>
<td>Number of interventions</td>
<td>39</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td>First ablation, n (%)</td>
<td>25 (100%)</td>
<td>6 (100%)</td>
<td>5 (100%)</td>
</tr>
<tr>
<td>First redo, n (%)</td>
<td>10 (40%)</td>
<td>–</td>
<td>3 (60%)</td>
</tr>
<tr>
<td>Second redo, n (%)</td>
<td>3 (12%)</td>
<td>–</td>
<td>2 (40%)</td>
</tr>
<tr>
<td>Third redo, n (%)</td>
<td>1 (4%)</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Procedural success</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>First ablation, n (%)</td>
<td>20 (80%)</td>
<td>6 (100%)</td>
<td>4 (80%)</td>
</tr>
<tr>
<td>First redo, n (%)</td>
<td>8 (80%)</td>
<td>–</td>
<td>2 (67%)</td>
</tr>
<tr>
<td>Second redo, n (%)</td>
<td>2 (67%)</td>
<td>–</td>
<td>2 (100%)</td>
</tr>
<tr>
<td>Third redo, n (%)</td>
<td>0 (0%)</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Long-term success, n (%)</td>
<td>16/20 (80%)</td>
<td>4/5 (80%)</td>
<td>5/5 (100%)</td>
</tr>
</tbody>
</table>

Catheter ablation in Ebstein

Summary

• Catheter ablation in Ebstein’s patients is very feasible, but one needs to understand in detail:
  – Cardiac anatomy and degree of atrialized RV
    • Microcatheter in RCA
    • 3D scan (preferable CMR) to mark the tricuspid annulus
  – Delineation of accessory pathway(s!)
    • Overt APs
    • Concealed Aps
  – Pacing manoeuvres to differentiate A from AP and V signals
• There is more arrhythmia to Ebstein than just APs