Unusual & Unexpected Location of Accessory Pathways

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Agenda

1. Unusual accessory pathways in usual anatomy
   1) Multiple accessory pathways
   2) Lown-Ganong-Levine syndrome
   3) Epicardial accessory pathway

2. Unusual accessory pathways in unusual anatomy
   1) Accessory pathway in CS diverticulum
   2) Accessory pathway in PLSVC
   3) Accessory pathway in Ebstein anomaly
Multiple Accessory Pathways
Multiple Accessory Pathways

- **ECG**
  - Atypical or unusual pattern of preexcitation
  - ≥ 2 delta wave morphologies
  - ≥ 2 P wave morphologies during orthodromic AVRT

- **EP study**
  - Antegrade-retrograde mismatch (> 1cm)
  - ≥ 2 atrial exit sites during orthodromic AVRT
  - AP-to-AP AVRT; preexcited tachycardia using 2^{nd} AP retrogradely
CASE 1: 32/M, Recurrent palpitation
CASE 1: Induction of AP-to-AP AVRT; antegrade right AP & retrograde left AP AV (with hidden H)

The earliest A
The earliest V
The earliest V

Onset of QRS & the earliest V
CASE 1: RFCA for right manifest AP & left concealed AP
Antidromic AVRT vs AP-to-AP AVRT
## Preexcited Tachycardias

<table>
<thead>
<tr>
<th></th>
<th>Antidromic AVRT</th>
<th>AP-to-AP AVRT</th>
<th>Preexcited AVNRT</th>
<th>Preexcited AT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preexcitation</td>
<td>Maximal</td>
<td>Maximal</td>
<td>Various</td>
<td>Various</td>
</tr>
<tr>
<td>HV interval</td>
<td>–</td>
<td>–</td>
<td>+ or –</td>
<td>+ or –</td>
</tr>
<tr>
<td>AV relationship</td>
<td>1:1</td>
<td>1:1</td>
<td>Usually 1:1</td>
<td>Various</td>
</tr>
<tr>
<td>Atrial activation</td>
<td>Concentric</td>
<td>Ec- or concentric</td>
<td>Concentric</td>
<td>Various</td>
</tr>
<tr>
<td>His conduction</td>
<td>Retrograde</td>
<td>Ante- or retro-</td>
<td>Antegrade</td>
<td>Antegrade</td>
</tr>
<tr>
<td>Adenosine</td>
<td>Terminated</td>
<td>Not terminated</td>
<td>Terminated</td>
<td>Various</td>
</tr>
<tr>
<td>V entrainment</td>
<td>VAV</td>
<td>VAV</td>
<td>VAV</td>
<td>VAAV</td>
</tr>
<tr>
<td>$\Delta HA$</td>
<td>$H_{tachy} = H_{V \text{pacing}}$</td>
<td>$H_{tachy} = H_{V \text{pacing}}$</td>
<td>$H_{tachy} &lt; H_{V \text{pacing}}$</td>
<td></td>
</tr>
<tr>
<td>Reset by early As</td>
<td>Constant VA</td>
<td>Constant VA</td>
<td>Constant VA</td>
<td>Inconstant VA</td>
</tr>
</tbody>
</table>
Lown-Ganong-Levine Syndrome
CASE 2: 29/F, Recurrent palpitation
CASE 2: 29/F, Recurrent palpitation
CASE 2: Sinus rhythm & A pacing → short AH interval
Lown-Ganong-Levine Syndrome

- **Theories**
  - 1\textsuperscript{st}: Intranodal AP
  - 2\textsuperscript{nd}: Atriohisian AP (Brechenmacher’s fiber)
  - 3\textsuperscript{rd}: Atrionodal AP (James’ fiber)
  - 4\textsuperscript{th}: Enhanced AV nodal conduction

- **Treatment**
  - Catheter ablation for AP
  - Beta-blocker, verapamil or diltiazem, class III AADs
  - AV node ablation & pacemaker
Accessory Pathways & Variants

Normal

WPW (Kent fiber)

LGL (James fiber)

Mahaim (Mahaim fiber)

Normal PR
No delta

Short PR
Delta (+)

Short PR
No delta

Normal PR
Delta (+)
Epicardial Accessory Pathways
Epicardial Accessory Pathways

- AP in the middle cardiac vein
- AP in the coronary sinus diverticulum
- AP in the vein (or ligament) of Marshall
- AP in the appendage
CASE 3: 67/F, Recurrent palpitation
CASE 3: 67/F, Recurrent palpitation
CASE 3: 67/F, Cardiac venography
CASE 3: Antegrade mapping & diverticulography
CASE 3: Retrograde mapping & diverticulography
Accessory Pathways in Posteroseptal Region

From Wood MA. Catheter Ablation of Cardiac Arrhythmias. 2015

Coronary sinus is wrapped by myocardial coat!
## ECG Findings of Epicardial APs

<table>
<thead>
<tr>
<th>Finding</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>PPV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steep negative delta in II</td>
<td>87%</td>
<td>79%</td>
<td>50%</td>
</tr>
<tr>
<td>Steep positive delta in aVR</td>
<td>61%</td>
<td>98%</td>
<td>88%</td>
</tr>
<tr>
<td>Deep S in V6</td>
<td>70%</td>
<td>87%</td>
<td>57%</td>
</tr>
<tr>
<td>CS diverticulum</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>
Accessory Pathways in Patients with Persistent Left SVC
CASE 4: 50/F, Palpitation, secundum ASD
CASE 4: PLSVC & great cardiac vein
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CASE 4: The CS catheter should be in the GCV!
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Accessory Pathways in Patients with Ebstein Anomaly
CASE 5: 35/M, WPW syndrome & Ebstein anomaly
CASE 5: 35/M, Ebstein anomaly (Carpentier C)
CASE 5: 35/M, Right ventriculography

Atrialized RV
CASE 5: Initially, normal sinus rhythm
CASE 5: Atrialized RV signals at proximal aRV
CASE 5: Atrialized RV signals at distal aRV
CASE 5: Atrialized RV signal during V pacing at proximal aRV
CASE 5: Atrialized RV signals changed during RFCA
Activation Sequence in Atrialized RV

Sinus rhythm before RFCA

V pacing before RFCA

After RFCA
Location of APs in Ebstein Anomaly


Accessory Pathways in Ebstein Anomaly

**Downward displacement** of the tricuspid septal leaflet

**Discontinuity** of the central fibrous body and septal AV ring with direct muscular

Creating a potential substrate for accessory AV connections and preexcitation
Tricuspid Annuli in Ebstein Anomaly

- True annulus
- Functional annulus
- Electrophysiological annulus (= RCA line)

Atrialized RV

T, E, F
Take-EP Lab Messages

- **ECG** is important for prediction of unusual accessory pathways.
- **Cardiac venography** should be performed in patients with posteroseptal accessory pathway.
- **The CS catheter** should be placed in the great cardiac vein in patients with PLSVC.
- **Right coronary angiography** should be performed for identifying the electrophysiological annulus in patients with Ebstein anomaly.
Thank you for your attention.