Electrophysiological Considerations in Congenitally Corrected TGA

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Agenda

1. ECG findings
2. AV node; twin AV nodes
3. Location of the slow pathway
4. Accessory pathways
5. Risk of AV block
6. Cardiac vein anatomy; implications for CRT
ECG Findings in ccTGA
Congenitally Corrected TGA
ECG Findings in ccTGA

- Right-to-left conduction in the interventricular septum
  - Septal Q wave in right precordial leads
  - No septal Q wave in left precordial leads
- Left axis deviation
  - Due to abnormal location of AV node
- High incidence of various degrees of AV block
- High prevalence of left manifest accessory pathway
ECG of ccTGA

- Left axis deviation
- Septal Q in V1
- No septal Q in V5 & V6
AV Node in ccTGA
Twin AV Nodes in ccTGA

- Normal posterior AV node
  - Located at the apex of the triangle of Koch
  - Usually no AV bundle

- Abnormal right anterior AV node
  - Give rise to the long penetrating AV bundle
  - Located antero-superiorly in the area lateral to the pulmonary-mitral continuity
Penetrating AV Bundle in ccTGA

- Penetrating AV bundle from right anterior AV node
  - A superficial course along the anterior aspect of pulmonary outflow tract
  - Descends along the upper interventricular septum
  - In cases of VSD, along anterosuperior margin of VSD

Antero-superior AV bundle
Pulmonic valve
Ventricular septal defect
Penetrating AV Bundle in ccTGA

- Penetrating AV bundle from posterior AV node in some patients
  - Courses to the interventricular septum.
  - Found in patients with the lesser degree of septal malalignment.
  - A sling-like Monckeberg bundle connecting both AV bundles over the anterior margin of VSD.

*Walsh EP, Cecchin F. Circulation 2007;115:534-45*
2 Morphologies of QRS; Twin AV Nodes
2 Morphologies of QRS; Twin AV Nodes
EP Characteristics of Twin AV Nodes

1. Two discrete non-preexcited QRS morphologies
2. Anatomically separate high-frequency His-bundle signals
3. Decremental & adenosine-sensitive antegrade conduction with a normal HV interval
4. Decremental & adenosine-sensitive retrograde conduction
5. A shift to the alternative QRS morphology when the ERP of the primary AV node was reached
Two His signals with normal HV intervals

<table>
<thead>
<tr>
<th>Anterior His</th>
<th>Posterior His</th>
</tr>
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<tbody>
<tr>
<td>III</td>
<td>III</td>
</tr>
<tr>
<td>aVR</td>
<td>aVR</td>
</tr>
<tr>
<td>aVL</td>
<td>aVL</td>
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<tr>
<td>aVF</td>
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<td>V1</td>
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<td>V2</td>
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<td>V6</td>
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<td>ABL d</td>
<td>ABL d</td>
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<tr>
<td>ABL p</td>
<td>ABL p</td>
</tr>
<tr>
<td>RA 1,2</td>
<td>RA 1,2</td>
</tr>
<tr>
<td>RA 3,4</td>
<td>RA 3,4</td>
</tr>
<tr>
<td>RV 1,2</td>
<td>RV 1,2</td>
</tr>
<tr>
<td>RV 3,4</td>
<td>RV 3,4</td>
</tr>
<tr>
<td>200 mm Hg</td>
<td>200 mm Hg</td>
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</tbody>
</table>

42ms

49ms
Multisite A pacing $\rightarrow$ 2 QRS morphologies

Anterior His

Posterior His
Risk of AV Block in ccTGA
AV Block in ccTGA

- High risk of AV block due to long non-branching penetrating AV bundle
- Incidence of spontaneous AV block
  - 5–17% at birth
  - Annually 2%
  - 40% in lifetime
- High risk of iatrogenic AV block
  - During passing the catheter through pulmonic valve in cardiac catheterization
  - During VSD repair
Complete AV Block in ccTGA
DDDR Pacemaker in ccTGA
Slow Pathway in ccTGA
Location of Slow Pathways in ccTGA

- 9 patients with ccTGA & AVNRT
  - Single His bundle in 8
  - Twin His bundles in 1
- Locations of slow pathways
  - 2 at right posterior septum
  - 1 at left posteroseptum
  - 5 at midseptum

How to Map Slow Pathway

157 ms

245 ms
How to Map Slow Pathway
CASE 1; AVNRT in ccTGA

- Patient; 51-year-old male
- C/C; recurrent palpitation
  - Frequency; 1-2 times / week
  - Duration; several hours
- History;
  - At birth, diagnosed with ccTGA with VSD & PS, \{I,D,D\}, functional single ventricle, bilateral SVC
  - At the age of 18 (in 1982), underwent BT shunt
CASE 1; Narrow QRS Tachycardia

HR = 172 /min
CASE 1; After Spontaneous Termination

Referred by: 

Confirmed By: HONG GEU-RU

aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
Decremental concentric VA conduction

St-A = 182 ms → 222 ms
Induction of tachycardia

TCL = 396 ms, VA = 102 ms
V entrainment pacing

VAV response, PPI - TCL = 127 ms, ΔVA = 110 ms
His refractory VPD

Preexcitation index > 100 ms
Slow pathway potential in the right anteroseptum
CASE 1; RFCA for SP in Right Anteroseptum

RAO 45°
CASE 2; AVNRT in ccTGA

- Patient; 45-year-old female
- C/C; recurrent palpitation
  - Frequency; 1-2 times / month
  - Duration; All day long
- History;
  - At the age of 20s years, diagnosed with ccTGA without associated defect, {S,L,L}
CASE 2; Narrow QRS Tachycardia

HR = 131 /min
CASE 2; After Adenosine Injection
Decremental concentric VA conduction
V entrainment pacing; VAV, PPI-TCL=143ms

569 ms

569 ms
Slow pathway potential in left mid-septum
Junctional rhythm during RFCA for SP
CASE 2; RFCA for SP in Left Mid-septum
Accessory Pathways in ccTGA
Accessory Pathways in ccTGA

- Accessory pathways are frequently located in morphological tricuspid annulus
- High prevalence of WPW syndrome & AVRT
Cardiac Vein Anatomy in ccTGA
Cardiac Vein Anatomy in ccTGA

- The coronary sinus and cardiac veins are embryologically originating from the atria.
- The coronary sinus is located between the left atrium and the morphological right (functional left) ventricle.
- CRT implantation is feasible in ccTGA.

*Diller GP, et al. Europace 2006;8:267-72*
Cardiac Venography in ccTGA

ccTGA \{S,L,L\}

ccTGA & PLSVC \{I,D,D\}
CRT Implantation in ccTGA

Rodriguez, et al. PACE 2001;24:235-7
CRT Implantation in ccTGA

Outcomes of CRT in ccTGA

Table 2  Studies evaluating the effect of cardiac resynchronization in patients with congenital heart disease

<table>
<thead>
<tr>
<th>Study</th>
<th>Population studied</th>
<th>Patient no., age range</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Prospective clinical studies</td>
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<tr>
<td>Janousek et al. (^1)</td>
<td>Systemic RV</td>
<td>(n = 8, 7-29) years</td>
<td>Improved systemic right ventricular ejection fraction, augmented interventricular asynchrony, and shortened QRS duration</td>
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<tr>
<td>Rodriguez-Cruz et al. (^36)</td>
<td>Congenitally corrected TGA, pulmonary atresia, and VSD</td>
<td>22 years</td>
<td>Clinical condition, exercise capacity, arterial blood pressure, end-diastolic pressure, and left ventricular contractility improved</td>
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Take-EP Lab Messages

- Patients with ccTGA usually have unique ECG findings.
- Patients with ccTGA usually have twin AV nodes.
- Risk of AV block is high in patients with ccTGA.
- Slow pathway in patients with ccTGA should be meticulous mapped with 3D electroanatomical mapping system.
- CRT implantation is feasible in patients with ccTGA.
Thank you for your attention.