Ablation of Atrial Arrhythmias in the Fontan patient

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No disclosures
Factors Predisposing to Arrhythmias in Adults with CHD

PACES/HRS Expert Consensus Statement on Management of Arrhythmias in Adult Congenital Heart Disease

Heart Rhythm 2014

Pre-operative

- Sinus Node
  - Abnormal in isomerism
  - Abnormal in left juxtaposition of atrial appendages

- Great Arteries
  - Regurgitation

- AV Groove
  - Accessory pathways

- Coronary Arteries
  - Anomalous
  - Obstructed

- Atrial Distension
  - Pressure
  - Volume

- AV Valve Regurgitation

- AV Node
  - Abnormal location & physiology
  - Twin AV nodes
  - Other conduction system abnormalities

- Cyanosis/hypoxia/acidosis

Post-operative

- Sinus Node
  - Dysfunction

- Coronary Arteries
  - Re-implantation
  - Injury

- Atriotomy
  - Scar – reentrant arrhythmia circuit (JAT)

- AV Node
  - Swelling / Injury
    - Complete Heart Block
    - Junctional Ectopic Tachycardia

- Patch
  - AV block

- Great Arteries
  - Regurgitation

- Coronary Arteries
  - Re-implantation
  - Injury

- AV Node
  - Swelling / Injury
    - Complete Heart Block
    - Junctional Ectopic Tachycardia

- Ventricular Abnormalities
  - Ventrilubotomy
  - Dyssynchrony
  - Scar – reentrant arrhythmia circuit (VT)

- Medications
- Electrolyte Disturbances
- Repolarization Abnormalities
- Systemic Illness
- Inflammation
Factors Predisposing to Atrial Arrhythmias in Congenital Heart Disease

1. Scarring due to multiple surgical suture lines
2. High wall stress due to pressure and volume overload
3. Chamber enlargement
4. Abnormal anatomy associated with the underlying congenital lesion
5. Changes in atrial refractoriness associated with sinus node dysfunction and bradycardia

- Multiple tachycardia morphologies frequently occur
- Up to 20% risk of SCD in Atrial Flutter in CHD
Cumulative incidence of atrial arrhythmia in ACHD 50% by age 65

Bouchardy et al Circ 2009
Surgical repair of tricuspid atresia

F. Fontan and E. Baudet

Centre de Cardiologie, Université de Bordeaux II, Hôpital du Tondu, Bordeaux, France

Surgical repair of tricuspid atresia has been carried out in three patients; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, ‘ventricillized’, to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.
Carins, Iyenegar, Nisbet et al, AATS, 2015
Why is this important?

- **Intra-atrial reentrant tachycardia** (atypical atrial flutter / incisional tachycardia) commonest mechanism

- **Atrial rates slower** than typical flutter (150-250 bpm) but can **conduct 1:1** in context of young healthy AV node so risk of syncope, heart failure or cardiac arrest

- **Thromboembolic complications** may occur – may require TOE before cardioversion if the duration of the tachycardia is unknown or >48 hours
Atrial arrhythmia in ACHD

Increased morbidity

Increased mortality

Bouchardy et al Circ 20
Rhythm control in adults with CHD and IART or atrial fibrillation

- Identify and treat precipitating factors
- Consider catheter ablation

Simple CHD

- Systemic ventricular hypertrophy or
- Systemic or subpulmonary ventricular dysfunction?

  No
  - Floccainide*
  - Propafenone*
  - Sotalol†

  - Amiodarone‡
  - Dofetilide‖

Moderate CHD

- Systemic or subpulmonary ventricular dysfunction?

  No
  - Amiodarone‡
  - Dofetilide"†

  - Sotalol†

Complex CHD

- Systemic or subpulmonary ventricular dysfunction?

  Yes

  - Amiodarone‡
  - Dofetilide‖
28 year old s/p Fontan repair. Atrial flutter (rx Sotalol) and Rec syncope
## Atrial Tach in CHD: Acute ablation success rates

<table>
<thead>
<tr>
<th>Study</th>
<th>N</th>
<th>Age (yrs)</th>
<th>Success rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hebe, 2000</td>
<td>69</td>
<td>25 ± 18</td>
<td>90%</td>
</tr>
<tr>
<td>Triedman, 2002</td>
<td>177</td>
<td>25 ± 12</td>
<td>79%</td>
</tr>
<tr>
<td>Blaufox, 2002</td>
<td>31</td>
<td>18 ± 5</td>
<td>96%</td>
</tr>
<tr>
<td>Kannankeril, 2003</td>
<td>47</td>
<td>28 ± 13</td>
<td>87%</td>
</tr>
<tr>
<td>Tanner, 2004</td>
<td>36</td>
<td>Median 46</td>
<td>94%</td>
</tr>
<tr>
<td>Lukac, 2005</td>
<td>83</td>
<td>Median 47</td>
<td>88%</td>
</tr>
<tr>
<td>Seiler, 2007</td>
<td>40</td>
<td>52 ± 12</td>
<td>88%</td>
</tr>
<tr>
<td>Yap, 2010</td>
<td>118</td>
<td>40 ± 13</td>
<td>69%</td>
</tr>
<tr>
<td>De Groot, 2010</td>
<td>53</td>
<td>38 ± 15</td>
<td>65%</td>
</tr>
<tr>
<td>Drago, 2011</td>
<td>31</td>
<td>26 ± 17</td>
<td>87%</td>
</tr>
<tr>
<td><strong>SUMMARY</strong></td>
<td>685</td>
<td></td>
<td><strong>81%</strong> [95% CI 79%-84%]</td>
</tr>
</tbody>
</table>
Ablation of arrhythmias in Fontan patients
“set yourself up for success!”

• Preparation is key
• Understand the anatomy
• Review the imaging
• Find the surgical operation notes (if possible)
• Consider access issues in advance and be prepared to use TOE or ICE to facilitate
IART in Fontan Approach to mapping and ablation

- **P wave morphology**
  - Limited utility for accurate localization
- **Standard Multipolar Activation Mapping**
  - Wavefront direction
  - Detect changes in activation pattern
- **Entrainment Mapping**
  - Post-pacing interval - yes
  - Endocardial concealment/fusion – not very helpful
  - Surface concealment/fusion – not very helpful
- **3-D Mapping System** – high density mapping
Examples
Atriopulmonary Fontan

- Summary of features
36 yr old male

- Double inlet left ventricle, ASD and pulmonary atresia
- BT shunt at birth
- Atrio-pulmonary Fontan repair age 4
- Recurrent tachycardia since age 24
- Several DCRs
- Sotalol and flecainide
- Palpitations and exertional breathlessness
Fontan AT CL 270 msec

Septal view

Inferior view
Fontan AT
CL 270 msec

Fontan AT
CL 320 msec

Septal view
Fontan AT CL 320 msec

Rt lateral view

Septal view
22 year old lady

- Tricuspid atresia, hypoplastic RV, VSD, pulmonary stenosis
- BT shunt at birth
- Atrio-pulmonary Fontan age 3
- Symptomatic bradycardia requiring epicardial AAI PPM 2006
- Troublesome palpitations, documented atrial flutter since 2007 (age 17), syncope, “seizures”
- Tilt table test – no syncope or HR/BP change
- EEG - normal
- Implantable loop recorder
CL 480 ms

CL 560 ms
3D Activation map – superior view

Oversewn SVC

AP connection

AP connection

Oversewn SVC
Pacing confirms complete linear block post-ablation
Lateral Tunnel Fontan

B. Lateral tunnel (intra-atrial baffle)

Anastomosis of enlarged cardiac end of SVC to RPA

Placement of baffle inside right atrium, forming a channel with a decreased diameter
Split potentials
TCPC – total cavo-pulmonary connection

“extra cardiac” Fontan
# AT: Focal vs Macro vs Small circuit

<table>
<thead>
<tr>
<th></th>
<th>Focal</th>
<th>Macro</th>
<th>Small circuit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Most common SHD</td>
<td>None</td>
<td>SHD/CHD/AF RFA</td>
<td>AF RFA, SHD/CHD</td>
</tr>
<tr>
<td>ECG isoelectric interval</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Record from &gt;80% of CL</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Entrain with PPI=TCL</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Entrain PPI=TCL, 2 sites&gt;2 cm apart</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Activation pattern</td>
<td>Radial</td>
<td>Loop</td>
<td>Small loop</td>
</tr>
<tr>
<td>Ablation approach</td>
<td>Focal</td>
<td>Linear or Focal</td>
<td>Linear or Focal</td>
</tr>
</tbody>
</table>
Multiple unstable right atrial circuits: Extensive substrate
- Consider substrate ablation

RA Rt Lat Activation

RA Rt Lat Voltage

RA Posterior Voltage
IART in ACHD

- Huge Chamber Enlargement
- Extensive scarring
- Multiple Tachycardias
  - Low Flow (esp Fontan)

“Scar” Map

 Identify Channels

Empiric “Bridging” Lesions
Cooled Tip Catheter
**RMH Fontan 3D Mapping Procedures**

Nisbet et al, APHRS 2016

<table>
<thead>
<tr>
<th><strong>16 RFA procedures in 13 patients</strong></th>
<th><strong>Mean (SD)</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at RFA (years)</td>
<td>26 (6)</td>
</tr>
<tr>
<td>Mean time since Fontan (years)</td>
<td>19 (5)</td>
</tr>
<tr>
<td>Number of IART circuits mapped</td>
<td>2 (1.7)</td>
</tr>
<tr>
<td>Fluoroscopy duration (mins)</td>
<td>25 (12)</td>
</tr>
<tr>
<td>Total RFA duration (mins)</td>
<td>19 (14)</td>
</tr>
<tr>
<td>Acute procedural success</td>
<td>75%</td>
</tr>
<tr>
<td>Complications</td>
<td>0</td>
</tr>
<tr>
<td>Mean follow up duration</td>
<td>65 mths</td>
</tr>
<tr>
<td>Late recurrence</td>
<td>9pts (56.3%)</td>
</tr>
</tbody>
</table>

3 underwent successful repeat procedure
### Table 1. IART Clinical Severity Score

<table>
<thead>
<tr>
<th>Documented IART</th>
<th>Frequency of Cardioversion (Prior 3 Months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>Nonsustained IART only</td>
<td>One cardioversion</td>
</tr>
<tr>
<td>History of sustained IART</td>
<td>AAdIT cardioversions</td>
</tr>
<tr>
<td>Incessant IART</td>
<td>Two or more cardioversions</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>IART Severity</th>
<th>Antiarrhythmic Medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>No symptoms</td>
<td>None or digoxin only</td>
</tr>
<tr>
<td>Palpitations only</td>
<td>Class II or class IV</td>
</tr>
<tr>
<td>Syncope/CHF/thrombosis</td>
<td>Class I or class III</td>
</tr>
<tr>
<td>Cardiac arrest</td>
<td>Amiodarone toxicity</td>
</tr>
</tbody>
</table>

Score of 0 to 12 points is calculated as the sum of the highest score achieved in each of the four categories. Thrombosis determined by either echocardiographic evidence of intracardiac thrombosis or clinical evidence of thromboembolic event. AAAdIT cardioversions are defined as one or more automatic or manual cardioversions performed using an implanted atrial pacemaker and not requiring any additional intervention. Amiodarone toxicity includes documented abnormalities of thyroid, hepatic or pulmonary function attributed by clinician to amiodarone administration, whether or not necessitating discontinuation of medication.

CHF = congestive heart failure, determined by review of clinical records and/or hemodynamic measurement; IART = intra-atrial re-entrant tachycardia.

Triedman et al. JACC 2002
IART severity assessment

- Mean reduced from 7 to 4 post ablation (p=0.016)
- 1 case who had an increase in score post RFA - Ultimately referred for extra-cardiac Fontan conversion
- Mean number of DCRs per year reduced from 5 to 1 post RFA.
- 14/16 continued on antiarrhythmic drugs post RFA
Conclusions

• Arrhythmias in the Fontan patient are often complex and multiple
• Careful mapping and entrainment is necessary for a successful outcome
• Consider the possibility of haemodynamic instability
• Consider your strategy in case of complete intra-atrial block during RFA
• Long term “cure” rates are low but significant palliation of symptoms is achievable