Arrhythmias in Adult patients with Congenital Heart Disease

Waleed Al-Maneeea
Ped cardiology - EP
Case:

- 18 years old young man

- Tricuspid atresia post modified Fontan

- Had repeated narrow QRS tachycardia since age 15 y not controlled with meds
What is the most likely mechanism of SVT in this patient?
With medications - bradycardia
Options:
- Pacemaker implantation + meds
- Surgical maze (high risk, SN dysfunction)
What type of pacemaker?

Best is pacemaker with antitachycardia pacing features
AAI pacing
pacing by the pacemaker
Recurrence of IART
Atrial Re-entry tachy
SCAR MAPPING

ABLATION LINES
Topics

- Clinical implication of conduction system for rhythm disturbances
- Substrates for rhythm disturbances
- Factors influencing rhythm disturbances
- Stepwise approach to evaluate the risk of rhythm disturbances
- Current management strategies
Conduction System

Normal Situs solitus

Abnormal location
( L-TGA, AVSD, SV )
Bilateral (Rt isomerism)

Absent (Lt Isomerism)
Factors Influencing Rhythm Disturbance in ACHD

- Hemodynamic condition
  - Valvular function
  - Ventricular function
- Cyanosis
- Aging
Effects of Aging on Cardiac Structure and Function

- Thickening and stiffness of the myocardium
- Atrial diameter and thickness (aged 60 to 69 years)
- Dilatation of pulmonary veins

Huonker et al, 2002
Gardin et al, 1979
Nieman et al, 2002
Najjar et al, 2005
Pan et al, 2008
Tsao et al, 2005
Effects of Aging on Atrium & Pulmonary Veins

Pan NH et al. Chest 2008;133:190-6
**Specific Arrhythmias & Associated Defects in ACHD**

**Tachycardia**

- **IART**
  - s/p Mustard; s/p Senning; s/p Fontan
- **Atrial fibrillation**
  - MV disease; aortic stenosis; unrepaired SV
- **Accessory pathway**
  - Ebstein anomaly; L-TGA
- **Twin AV node**
  - Heterotaxy syndrome
- **Ventricular tachycardia**
  - TOF; congenital AS
Specific Arrhythmias & Associated Defects in ACHD

Bradycardia

- **Congenital sinus node dysfunction**
  - Heterotaxy syndrome
- **Acquired sinus node dysfunction**
  - s/p Mustard; s/p Senning; s/p Fontan
- **Congenital AV block**
  - L-TGA; atrioventricular septal defect
- **Acquired AV block**
  - VSD closure; Subaortic stenosis; AV valve replacement
## Postoperative CHD and the Incidence of Late Arrhythmias

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Incidence of Arrhythmia</th>
<th>Incidence of Reoperation</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD / PAPVR</td>
<td>5-15% SVT</td>
<td>1 – 9%</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>40-80% SVT</td>
<td>&gt; 25%</td>
</tr>
<tr>
<td>Fontan repair</td>
<td>30-60% SVT</td>
<td>&gt; 25%</td>
</tr>
<tr>
<td>TGA s/p atrial switch</td>
<td>30-50% SVT</td>
<td>27%</td>
</tr>
<tr>
<td>TOF</td>
<td>7% VT, 35% SVT</td>
<td>8-14%</td>
</tr>
<tr>
<td>TV repair</td>
<td>35% SVT</td>
<td>&gt; 35%</td>
</tr>
<tr>
<td>VSD</td>
<td>&lt; 2% SVT, VT</td>
<td>&lt; 5%</td>
</tr>
</tbody>
</table>

Khairy et al, 2006
Monro et al, 2003
Overgaard et al, 1999
Nollert et al, 1997
Management of Emergencies in ACHD

Kaemmerer et al. Am J Cardiol 2008;101:521-525
Survival Curves for Patients who underwent Closure of ASD at the Mayo Clinic

Murphy et al. 1990
Symptomatic Atrial Arrhythmias and Transcatheter Device Closure of ASD in Adults

Silversides et al. Heart 2004; 90:1198-8
Risk Factors for Sudden Cardiac Death in Patients with Postoperative TOF

- Older age at initial repair
- Moderate or severe pulmonary regurgitation
- History of sustained ventricular dysfunction
- QRS duration of 180 ms or more
- Rapid increase in QRS duration

Gatzoulis 2000; Lancet 356:975-981
Gatzoulis 1995; Circulation 92:231-237
Therrien 2003; Lancet 362:1305-1313
Karamlou 2006; Ann Thorac Surg 81:1786-1793
Harrison 2001; AM J Card 87:584-588
Friedli 1999; Pediatr Cardiol 20:326-330
## Estimates of the Incidence of Sudden Death After TOF Surgery

<table>
<thead>
<tr>
<th>Author</th>
<th>Findings</th>
<th>Incidence per decade, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Murphy et al.</td>
<td>6% of 163 cases followed for 30 years</td>
<td>2.0</td>
</tr>
<tr>
<td>Nollert et al.</td>
<td>3% of 490 cases f/u for 25 years</td>
<td>1.2</td>
</tr>
<tr>
<td>Silka et al.</td>
<td>2 death per 1000 patient-years</td>
<td>2.0</td>
</tr>
<tr>
<td>Norgaard et al.</td>
<td>5.6% of 125 cases f/u for 25 years</td>
<td>2.2</td>
</tr>
<tr>
<td>Gatzoulis et al.</td>
<td>6% of 793 cases f/u for 21 years</td>
<td>3.0</td>
</tr>
</tbody>
</table>
Risk Factors for Arrhythmias in Patients Who Underwent the Fontan Operation

- Preoperative AV valve regurgitation
- Older age at operation
- Poor preoperative functional status
- Previous atrial septectomy
- Preoperative atrial tachyarrhythmias
- Pulmonary artery reconstruction
- Atriopulmonary anastomosis
- Early postoperative atrial tachyarrhythmia
- Postoperative sinus node dysfunction
- Length of follow-up

Gelatt et al. J Am Coll Cardiol 1994;24:1735-1741
Kaplan-Meier Curves of Freedom from Atrial Flutter as Function of Type of Fontan Operation

Fishberger et al. 1996
Stamm et al. 2001
1. **Pacemaker component**
   - *SA node*
     - Congenital – atrial isomerism
     - Acquired – surgical damage

2. **Slow impulse conduction component**
   - *Atrium conduction delay*
     - Congenital / acquired
Simple Stepwise Approach to Rhythm Disturbance in ACHD

3. **Slow impulse conduction component**
   - **AV node**
     - Congenital – atrial isomerism
   - **Accessory pathway**
     - L-TGA, Ebstein anomaly
   - Dual (twin) AV node

4. **Rapid impulse conduction component**
   - **His-Purkinje system damage**
     - Congenital / acquired
5. **Risk stratification**

- Atrial arrhythmia
  - Fontan
- Ventricular arrhythmia
  - S/P ventriculotomy
- Risk of SCD?
- Hemodynamic condition?
- Surgical correction?
Management of Rhythm Disturbance in ACHD

**Tachycardia**
- Most drugs – ineffective
  - Amiodarone (less negative inotropic effect)
- Low acute success
- High recurrence rate
  - Outcome is improving
- ICD – VT, SCD
- PPM – chronotropic incompetence
  - ATP Rx
- Surgery + concomitant arrhythmia surgery

**Bradycardia**
- Not application

**Medication**

**R F C A**

**Device**

**Surgery**

**PPM**
- Technical consideration
  - Epicardial/Endocardial
  - Thoracoscopic Pericardial
  - Dual chamber/ Single chamber
  - Biventricular (CRT)

**EPS**
Evolution of RF Ablation Techniques for Intraatrial Reentrant Tachycardia (IART)

<table>
<thead>
<tr>
<th>Dates</th>
<th>Technique</th>
<th>Acute Success</th>
<th>Recurrence Rates</th>
</tr>
</thead>
<tbody>
<tr>
<td>92 – 95</td>
<td>Entrainment/ 4-mm tip</td>
<td>60-75%</td>
<td>&gt; 50%</td>
</tr>
<tr>
<td>95 – 98</td>
<td>“Cooled” tip</td>
<td>80-90%</td>
<td>30 – 50%</td>
</tr>
<tr>
<td>99 – 00</td>
<td>3D mapping</td>
<td>80-90%</td>
<td>30 – 50%</td>
</tr>
<tr>
<td>00 – 05</td>
<td>Demonstrate block</td>
<td>90-100%</td>
<td>25- 30%</td>
</tr>
<tr>
<td>05 -</td>
<td>Advanced map, cooled tip, cryo</td>
<td>90-100%</td>
<td>?? ??</td>
</tr>
</tbody>
</table>

Love et al, 2001
Walsh et al, 2007

Saul JP PACE 2008;31-51-12
# Clinical Series of Surgical “Fontan Conversions”

<table>
<thead>
<tr>
<th>Author</th>
<th>No. of Patients</th>
<th>Freedom from postoperative AA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kim et al.</td>
<td>16</td>
<td>88%</td>
</tr>
<tr>
<td>Sheikh et al.</td>
<td>15</td>
<td>73%</td>
</tr>
<tr>
<td>Weinstein et al.</td>
<td>10</td>
<td>89%</td>
</tr>
<tr>
<td>Mavroudis et al.</td>
<td>111</td>
<td>82%</td>
</tr>
<tr>
<td>Morales et al.</td>
<td>35</td>
<td>89%</td>
</tr>
</tbody>
</table>

Morales et al, Annals of Thoracic Surgery, 2005; 80(4), 1445-1452*
Atrial Arrhythmia Surgery in Congenital Heart Disease
Catheter Ablation of VT after repair of Congenital Heart Disease

Zeppenfeld et al. 2007
EPS/RFCA approach for VT after repair of TOF

Adult
QRS > 180 ms
Severe dilated RV
Ventricular scar (+)

EPS?

High LVEDP
Nonsustained VT
Prior to surgery for PVR

EPS / RFCA

Sinus rhythm
Geometry scar

VT induction (+)

Stable VT

Unstable VT

Activation mapping
Entrainment

Pace mapping
Voltage mapping

3D mapping

Successful RFCA ? ICD ? Surgery ?
Summary

- Minimize potential substrates
- Adequately assess the rhythm disturbances
- Choose optimal management
  - RFCA
  - Medications
  - Concomitant arrhythmia surgery