EP In Adults with Congenital Heart Disease

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Cohen Children’s Medical Center
EP in ACHD

Table 8.1. Substrates associated with a relatively high prevalence of congenital and postoperative sinus node dysfunction

Congenital sinus node dysfunction
Left-sided juxtaposition of the atrial appendages
Left atrial isomerism (polysplenia, heterotaxy syndrome)
Postoperative sinus node dysfunction
Mustard baffle
Senning baffle
Hemi-Fontan or Fontan surgery; atriopulmonary and total cavopulmonary connections
Glenn shunt
Sinus venosus atrial septal defect
Ebstein anomaly
Arterial switch operation for transposition of the great arteries
(chronotropic incompetence)

Tetralogy of Fallot

Khairy P, Van Hare GF, Balaji S et al. 2014; Heart Rhythm:11:e102-165
Sinus Node Dysfunction in the Post-op Tetralogy of Fallot
SAN Dysfunction in the Post-op Tetralogy of Fallot

Study designed to assess HR response to exercise in ACHD

Evaluate HR response and exercise capacity and answer the question: is chronotropic incompetence (CI) a prognostic factor in adults with repaired/palliated CHD?

Retrospective study 1999-2005; standard Bruce protocol

Measured ventilation oxygen uptake, CO$_2$ production, HR, BP response. \[ \text{Chronotropic Index} = \frac{\text{Peak HR} - \text{Rest HR}}{220 - \text{age} - \text{Rest HR}} \]

(Normal = 0.8 – 1.3)

SAN Dysfunction in the Post-op Tetralogy of Fallot

Results of the study – CI in 62% of patients
Fontan 84%, Eisenmengers 90%
ToF and VSD – 52% (lowest of the CHD patients)

Largest group was ToF – 228 patients

HR reserve in ToF pts was highest of all others – 80+/- 26;
peak HR 162 +/- 26; 98% at rest in NSR
There were 5 post-op ToF patients who died out of a total of 38 deaths in 727 patients.

Three top groups were: “Complex” anatomy – 10; Fontan – 7; and ToF – 5

HR reserve (10 bpm) predicted mortality in patients after repaired ToF (HR=0.66/10 bpm; 95% CI 0.48 to 0.91, p<0.05)
# SAN Dysfunction in Post-op CHD

## Table 2

Distribution of Parameters of Chronotropic Incompetence, Peak Oxygen Consumption, Presence of Sinus Rhythm, and Use of Antiarrhythmic Medication by Underlying Anatomy

<table>
<thead>
<tr>
<th></th>
<th>Low CI</th>
<th>HRR (beats/min)</th>
<th>Peak Pulse (beats/min)</th>
<th>Peak ( \text{VO}_2 ) (ml/kg/min)</th>
<th>Sinus Rhythm</th>
<th>AAD Treatment</th>
<th>Deceased During FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD (n = 42)</td>
<td>60%</td>
<td>69 ± 30</td>
<td>154 ± 34</td>
<td>21.6 ± 7.9</td>
<td>89%</td>
<td>32%</td>
<td>1</td>
</tr>
<tr>
<td>ccTGA (n = 25)</td>
<td>68%</td>
<td>70 ± 33</td>
<td>151 ± 38</td>
<td>21.8 ± 9.6</td>
<td>95%</td>
<td>43%</td>
<td>2</td>
</tr>
<tr>
<td>CoA (n = 23)</td>
<td>59%</td>
<td>73 ± 24</td>
<td>156 ± 27</td>
<td>28.9 ± 7.9</td>
<td>95%</td>
<td>30%</td>
<td>1</td>
</tr>
<tr>
<td>Complex (n = 75)</td>
<td>81%</td>
<td>56 ± 28</td>
<td>138 ± 32</td>
<td>20.2 ± 7.7</td>
<td>88%</td>
<td>35%</td>
<td>10</td>
</tr>
<tr>
<td>Ebstein (n = 32)</td>
<td>53%</td>
<td>77 ± 28</td>
<td>158 ± 31</td>
<td>21.5 ± 5.2</td>
<td>90%</td>
<td>35%</td>
<td>3</td>
</tr>
<tr>
<td>Eisenmenger (n = 53)</td>
<td>90%</td>
<td>51 ± 23</td>
<td>136 ± 24</td>
<td>12.8 ± 5.7</td>
<td>100%</td>
<td>40%</td>
<td>1</td>
</tr>
<tr>
<td>Fontan (n = 58)</td>
<td>84%</td>
<td>59 ± 27</td>
<td>140 ± 33</td>
<td>20.9 ± 6.1</td>
<td>96%</td>
<td>53%</td>
<td>7</td>
</tr>
<tr>
<td>Mustard (n = 56)</td>
<td>58%</td>
<td>77 ± 26</td>
<td>160 ± 28</td>
<td>25.8 ± 6.9</td>
<td>94%</td>
<td>29%</td>
<td>1</td>
</tr>
<tr>
<td><strong>TOF (n = 228)</strong></td>
<td>52%</td>
<td>80 ± 26</td>
<td>162 ± 26</td>
<td>25.7 ± 8.4</td>
<td>98%</td>
<td>19%</td>
<td>5</td>
</tr>
<tr>
<td>Valvar (n = 78)</td>
<td>47%</td>
<td>76 ± 31</td>
<td>159 ± 29</td>
<td>26.9 ± 12.8</td>
<td>97%</td>
<td>17%</td>
<td>2</td>
</tr>
<tr>
<td>VSD (n = 25)</td>
<td>52%</td>
<td>70 ± 27</td>
<td>157 ± 26</td>
<td>22.2 ± 7.1</td>
<td>100%</td>
<td>13%</td>
<td>0</td>
</tr>
</tbody>
</table>

Plus-minus values are mean ± standard deviation.
Heart Rate Reserve and Mortality

## HRR and mortality in ACHD

**Table 4. Multivariate Predictors of Mortality**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Hazard Ratio*</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart rate reserve (10 beats/min)</td>
<td>0.86 (0.74–0.99)</td>
<td>0.04</td>
</tr>
<tr>
<td>Antiarrhythmic therapy (incl. digoxin)</td>
<td>3.7 (1.7–8.1)</td>
<td>0.0008</td>
</tr>
<tr>
<td>NYHA functional class</td>
<td>2.0 (1.2–3.4)</td>
<td>0.007</td>
</tr>
<tr>
<td>Peak VO₂ (ml/kg/min)</td>
<td>—</td>
<td>NS</td>
</tr>
</tbody>
</table>

| Heart rate reserve (10 beats/min)      | 0.83 (0.72–0.96)| 0.01    |
| Amiodarone therapy                     | 4.7 (2.4–9.5)   | <0.0001 |
| NYHA functional class                  | 2.1 (1.3–3.5)   | 0.002   |
| Peak VO₂ (ml/kg/min)                   | —              | NS      |

*Hazard ratios for heart rate reserve and peak oxygen consumption are per 10 beats/min and 1 ml/kg/min, respectively. Abbreviations as in Table 3.

Studied CI in adolescents and adults with CHD after cardiac surgery

Premise: heart failure would be more apparent with exercise as measured by reduced capacity and decrease peak VO$_2$

361 ACHD pts aged 14-50 yrs; ToF 1 of 7 groups
94 pts, age at op 7.3 +/- 0.6
30/94 (32%) had CI

Sinus Node Dysfunction in the Post-op CHD Patient

Using bicycle ergometry measured O₂ uptake throughout and at RER >= 1.1; VO₂_AT and VO₂ at highest value within 30 sec of terminating exercise. ToF was shown to be a moderate risk along with valve disease (single V, TGA and CoA were higher risks)

Fig. 1. The probability of chronotropic incompetence over age and type of heart defect. SV, single ventricle; TGA, transposition of the great arteries; CoA, coarctation of the aorta; TOF, tetralogy of Fallot; VD, aortic and pulmonary valve disease: LRS, left-to-right-shunt.
Atrial Tachyarrhythmias in Fontan Patients

Intra-atrial reentry tachycardia (IART)
Atrial fibrillation (Afib)
IART + Afib
SVT due to a macro-reentry circuit (AP mediated, AVNRT)

Study * of 166 adult patients with long-term follow-up
42% had arrhythmias (incidence 2.23 per 100 pt-yrs)
M/F was 49%
Mean age of arrhythmia group was older - 34.3 +/- 10.5 yrs (non-arr group 25.3 +/- 5.6)
Time from surgery 14.2 +/- 6.4, median 14, IQR 11-19)

Figure 1. Kaplan–Meier curve showing the arrhythmia-free survival for the total Fontan population.

Predictors of Arrhythmias in the Fontan

Predictors of Arrhythmias in the Fontan

Figure 1 Hazard of IART Following the Fontan Operation, With 95% Confidence Bands.
Date of diagnosis was missing for 1 patient, thus the number at risk initially is 519. IART intra-atrial re-entrant tachycardia.

Figure 2 Freedom From IART Following the Fontan Operation by Type of Fontan Procedure
Values are adjusted for predominant rhythm and CHQ physical summary score. Number of patients at risk in each surgical group is shown in the adjoining panel. IART = intra-atrial re-entrant tachycardia.

# Catheter Ablation of Atrial Tachyarrhythmias

Table 7.1. Acute success rates for catheter ablation of atrial tachyarrhythmias in CHD

<table>
<thead>
<tr>
<th>First Author</th>
<th>Year</th>
<th>N</th>
<th>Mean age (years)</th>
<th>Acute success</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hebe</td>
<td>2000</td>
<td>69</td>
<td>25 +/- 18</td>
<td>90%</td>
</tr>
<tr>
<td>Triedman</td>
<td>2002</td>
<td>177</td>
<td>25 +/- 12</td>
<td>79%</td>
</tr>
<tr>
<td>Blaufox</td>
<td>2002</td>
<td>31</td>
<td>18 +/- 5</td>
<td>96%</td>
</tr>
<tr>
<td>Kannankeril</td>
<td>2003</td>
<td>47</td>
<td>28 +/- 13</td>
<td>87%</td>
</tr>
<tr>
<td>Tanner</td>
<td>2004</td>
<td>36</td>
<td>Median 46 (9 to 67)</td>
<td>94%</td>
</tr>
<tr>
<td>Lukac</td>
<td>2005</td>
<td>83</td>
<td>Median 47 (9 to 73)</td>
<td>88%</td>
</tr>
<tr>
<td>Seiler</td>
<td>2007</td>
<td>40</td>
<td>52 +/- 12 years</td>
<td>88%</td>
</tr>
<tr>
<td>Yap</td>
<td>2010</td>
<td>118</td>
<td>40 +/- 13 years</td>
<td>69%</td>
</tr>
<tr>
<td>de Groot</td>
<td>2010</td>
<td>53</td>
<td>38 +/- 15 years</td>
<td>65%</td>
</tr>
<tr>
<td>Drago</td>
<td>2011</td>
<td>31</td>
<td>26 +/- 17 years</td>
<td>87%</td>
</tr>
</tbody>
</table>

Summary 

<table>
<thead>
<tr>
<th>N</th>
<th>Acute success</th>
</tr>
</thead>
<tbody>
<tr>
<td>685</td>
<td>81% [95% CI (79%-84%)]</td>
</tr>
</tbody>
</table>

Khairy P, Van Hare GF, Balaji S et al. 2014; Heart Rhythm:11:e102-165
## EP in ACHD

**Table 8.2. Congenital heart disease substrates associated with a relatively high prevalence of congenital and postoperative AV block**

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital AV block</td>
</tr>
<tr>
<td>Congenitally corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Atrioventricular septal defect (endocardial cushion defect)</td>
</tr>
<tr>
<td>L-Looped single ventricles</td>
</tr>
<tr>
<td>Anomalous left coronary artery arising from the pulmonary artery (ALCAPA)</td>
</tr>
<tr>
<td>Postoperative AV block</td>
</tr>
<tr>
<td>Cardiac surgery in patients with displaced AV conduction systems (congenitally corrected transposition of the great arteries, atrioventricular septal defect)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
</tr>
<tr>
<td>Valve surgery, especially mitral valve and multivalve surgery involving the tricuspid valve</td>
</tr>
<tr>
<td>Left ventricular outflow surgery, subaortic stenosis</td>
</tr>
</tbody>
</table>

Khairy P, Van Hare GF, Balaji S et al. 2014; Heart Rhythm:11:e102-165
## Causes of Death and Sudden Death following Repair of CHD

### Table 9.1. Causes of sudden cardiac death following surgical repair of CHD

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>No. events</th>
<th>Arrhythmic</th>
<th>Embolic</th>
<th>MI/CHF</th>
<th>Aneurysm</th>
</tr>
</thead>
<tbody>
<tr>
<td>Silka et al 1998</td>
<td></td>
<td>41</td>
<td>30 (73.2%)</td>
<td>5 (12.2%)</td>
<td>4 (9.8%)</td>
<td>2 (4.9%)</td>
</tr>
<tr>
<td>Nieminen et al 2007</td>
<td></td>
<td>88</td>
<td>73 (83.0%)</td>
<td>5 (17.9%)</td>
<td>5 (17.9%)</td>
<td>5 (17.9%)</td>
</tr>
<tr>
<td>Koyak et al 2012</td>
<td></td>
<td>213</td>
<td>171 (80.3%)</td>
<td>8 (37.6%)</td>
<td>5 (2.3%)</td>
<td>19 (8.9%)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td></td>
<td><strong>342</strong></td>
<td><strong>274 (80.1%)</strong></td>
<td><strong>18 (5.3%)</strong></td>
<td><strong>14 (4.1%)</strong></td>
<td><strong>26 (7.6%)</strong></td>
</tr>
</tbody>
</table>

CHD = congenital heart disease; CHF = congestive heart failure; MI = myocardial infarction.

### Table 9.2. Causes of death following surgical repair of CHD

<table>
<thead>
<tr>
<th>Authors</th>
<th>Years</th>
<th>Patients</th>
<th>Deaths</th>
<th>SCD</th>
<th>CHF</th>
<th>Other CV</th>
<th>Noncardiac</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oeschlin et al 1981-1996</td>
<td></td>
<td>2609</td>
<td>197</td>
<td>26%</td>
<td>21%</td>
<td>34%</td>
<td>18%</td>
</tr>
<tr>
<td>Silka et al 1958-1996</td>
<td></td>
<td>3589</td>
<td>176</td>
<td>23%</td>
<td>13%</td>
<td>35%</td>
<td>12%</td>
</tr>
<tr>
<td>Nieminen et al 1953-1998</td>
<td></td>
<td>5919</td>
<td>582</td>
<td>15%</td>
<td>27%</td>
<td>31%</td>
<td>8%</td>
</tr>
<tr>
<td>Verheugt et al 2001-2009</td>
<td></td>
<td>6933</td>
<td>197</td>
<td>19%</td>
<td>26%</td>
<td>32%</td>
<td>23%</td>
</tr>
<tr>
<td>Zomer et al 2001-2010</td>
<td></td>
<td>8595</td>
<td>231</td>
<td>22%</td>
<td>26%</td>
<td>29%</td>
<td>24%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td></td>
<td><strong>27,645</strong></td>
<td><strong>1,383</strong></td>
<td><strong>19%</strong></td>
<td><strong>24%</strong></td>
<td><strong>36%</strong></td>
<td><strong>15%</strong></td>
</tr>
</tbody>
</table>

CHD = congenital heart disease; CHF = congestive heart failure; Noncardiac = noncardiac cause of death; Other CV = other cardiovascular cause of death; SCD = sudden cardiac death.
Table 11.3. Prophylactic arrhythmia surgery in adults with CHD

<table>
<thead>
<tr>
<th>Congenital heart substrate</th>
<th>Arrhythmia</th>
<th>Technique</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan revision or conversion</td>
<td>IART, atrial fibrillation</td>
<td>Modified right atrial Maze +/- left atrial Cox Maze III</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>Accessory connection</td>
<td>Dissection and division or ablation</td>
</tr>
<tr>
<td></td>
<td>IART</td>
<td>Modified right atrial Maze</td>
</tr>
<tr>
<td></td>
<td>Atrial fibrillation</td>
<td>Left atrial Cox Maze III with right-sided lesion set +/- left atrial</td>
</tr>
<tr>
<td></td>
<td></td>
<td>appendectomy or oversew orifice</td>
</tr>
<tr>
<td>Right heart conduit revisions, tricuspid valve repair or</td>
<td>IART</td>
<td>Cavotricuspid isthmus ablation or modified right atrial Maze</td>
</tr>
<tr>
<td>replacement, congenital lesions with atrial dilation</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Atrial fibrillation</td>
<td>Left atrial Cox Maze III with cavotricuspid isthmus ablation, left atrial</td>
</tr>
<tr>
<td></td>
<td></td>
<td>appendectomy or oversew orifice</td>
</tr>
<tr>
<td>Left-sided valve repair/replacement</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Atrial fibrillation</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect closure</td>
<td>IART</td>
<td>Cavotricuspid isthmus ablation, (modified) right atrial Maze</td>
</tr>
<tr>
<td></td>
<td>Atrial fibrillation</td>
<td>Left atrial Cox Maze III with cavotricuspid isthmus ablation left atrial</td>
</tr>
<tr>
<td></td>
<td></td>
<td>appendectomy or oversew orifice</td>
</tr>
</tbody>
</table>

Khairy P, Van Hare GF, Balaji S et al. 2014; Heart Rhythm:11:e102-165
EP of the Post-op Mustard procedure for d-TGA

Surgical Anatomy

EP Anatomy

Figure 1 Electroanatomical map of the atria after the Mustard operation (Carto® combined to RMN) merged with CT-based anatomy. The patient has ongoing peri-tricuspid IART. The green dots show the sites with positive entrainment, the dark red dots are the ablation sites. IVC, inferior vena cava; LAA, left atrial appendage; SVC, superior vena cava; TV, tricuspid valve.


Fig. 5: An illustration of the steps of the Mustard operation. After the right atriotomy (top left), the atrial septum is largely removed, and the coronary sinus may be incised or otherwise redirected; followed by placement of a "pant-legs" shaped baffle (right) which directs the caval returns to the mitral valve and allowing the pulmonary venous returns to enter the tricuspid valve. The upper-most suture line is most likely to damage the sinus node or its blood supply, and the lower/anterior suture line may damage the AV node. Last, the right atriotomy is closed (lower).

Figure 2. Electroanatomic maps of the right atrium demonstrating atrial macroreentry that developed in an adult with tetralogy of Fallot many years after surgical repair. Surgery included a right lateral atriotomy incision (hatched area). A: Typical counterclo...

Edward P. Walsh

**Sudden death in adult congenital heart disease: Risk stratification in 2014**


http://dx.doi.org/10.1016/j.hrthm.2014.07.021
Figure 1. Pathologic specimen from a young patient with unrepaired tetralogy of Fallot demonstrating the intrinsic anatomic features that can contribute to macroreentrant ventricular tachycardia. A portion of the anterior right ventricle (RV) has been removed ...

Edward P. Walsh

Sudden death in adult congenital heart disease: Risk stratification in 2014


http://dx.doi.org/10.1016/j.hrthm.2014.07.021
Basic Issues In EP for ACHD

For palliated/operated patients:

• A concerted effort to obtain old records. Patients frequently do not have old records at home and may be confused as to their diagnosis and what surgeries they have had performed.

• Prior to any interventional procedure, it is almost imperative that you obtain the surgical report from previous operations. This greatly assists in planning what type of procedure is to be done and expectations for success. Especially true for the single ventricle patients.

• Many patients will benefit from advanced imaging techniques such as CTA and/or MRI/MRA. Images can be imported into electroanatomic mapping systems; 3-D models can be printed.

• Patients undergoing implantation of CIEDs can have their venous anatomy determined accurately and other procedures, such as stenting open an SVC/atrial junction can be performed in conjunction with CIED implant.
Procedural workflow

Gijsbert F.L. Kapel et al. Circ Arrhythm Electrophysiol.
2015;8:102-109
Schematic overview and prevalence of the 6 identified anatomic isthmuses.

Occurrence of sustained monomorphic ventricular tachycardia (SMVT) during long-term follow-up.

Gijsbert F.L. Kapel et al. Circ Arrhythm Electrophysiol. 2015;8:102-109
Basic Issues In EP for ACHD

For un-operated CHD patients:

- Includes patients with CCAVB, CCTGA, VSDs, ASDs, aortopathies, valve stenoses, coronary artery anomalies, etcetera.

- Age appropriate evaluation of coronary arterial patency, including coronary angiography if appropriate (. 40 yrs of age), hemodynamic assessment in the cath lab by a qualified congenital interventional cath physician.

For all ACHD patients scheduled for an intervention:

A qualified cardiologist to care for patients with ACHD and

- Pre-interventional consultation with (preferably) CV anesthesiologist.
- Pre-interventional consultation with a CHD surgeon.
- Full pre-brief with cath lab personnel and assurance that adult resuscitation equipment is available and personnel educated on use (ACLS qualified).
Thank You
EXTRA SLIDES
Thromboprophylaxis in adults with CHD and IART or atrial fibrillation

Acute cardioversion

Duration unknown or ≥48 hours?

Yes

Moderate/complex CHD

Anticoagulation ≥3 weeks or TEE*

Cardioversion

No

Simple CHD

Long-term thromboprophylaxis

Moderate/complex CHD

VKA

Prosthetic valve or significant valve disease?

Yes

Simple CHD

CHA₂DS₂-VASc score

0

No thromboprophylaxis

1

• No thromboprophylaxis
• ASA

≥2

• VKA
• NOAC

*Patients with Fontan palliation are at particularly high risk of thromboembolic complications such that TEE may be prudent prior to cardioversion even if therapeutic anticoagulation is received for ≥3 weeks

TEE denotes transesophageal echocardiography; CHD, congenital heart disease; VKA, vitamin K antagonist; NOAC, newer oral anticoagulant; CHA₂DS₂-VASc: Congestive heart failure; Hypertension; Age (≥75 years, 2 points; 65-74 years, 1 point); Diabetes; Stroke, transient ischemic attack, or thromboembolism (2 points); Vascular disease; Sex category (female); ASA, aspirin

Khairy P, Van Hare GF, Balaji S et al. 2014; Heart Rhythm:11:e102-165
CRT Implantation and Complex Anatomy

A
B
C

MORPHOLOGIC LV

SGW
GW
MORPHOLOGIC RV

A
B

ANT
RV
LAT
MCV
CS
RV
LV

A
B
C
D

S1
S2
GDE
RV
ANT
LAT
RA
S1
S2
GDE
RV ICD
LV

Niazi, I et al. PACE 2014;37:1181-1188
Simple colour map representation of average activation patterns across the RV in both ToF and congenital PS, with and without TAP repair.

Carla M. Plymen et al. Europace 2015;17:274-280
Graph showing absolute change (±SD) in QRS duration with various pacing modalities.