Ventricular Arrhythmias in CHD patients: How can the Electrophysiologist help?

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Ventricular Arrhythmias in CHD

• Arrhythmias are the main reason for the hospitalization of GUCH patients
• They are an increasingly frequent cause of morbidity and mortality\(^1\).
• The onset of arrhythmias may be a signal of haemodynamic decompensation
• Risk associated with arrhythmias may be amplified in the presence of the often abnormal underlying circulation.

Ventricular Arrhythmias in CHD

• Arrhythmic sudden cardiac death
  – Ventricular fibrillation
  – Polymorphic VT
  – Monomorphomorphic sustained VT
• Symptomatic sustained and non-sustained VT
• Ventricular ectopy
Arrhythmogenic factors in CHD

A Pre-operative

- Sinus Node
  - Abnormal in isomerism
  - Abnormal in left juxtaposition of atrial appendages
- AV Valve Regurgitation
- Atrial Distension
  - Pressure
  - Volume
- AV Node
  - Abnormal location & physiology
  - Twin AV nodes
  - Other conduction system abnormalities
- Great Arteries
  - Regurgitation
- AV Groove
  - Accessory pathways
- Coronary Arteries
  - Anomalous
  - Obstructed
- Ventricular Abnormalities
  - Hypertrophy
  - Dilatation
  - Ischemia
- Cyanosis/hypoxia/acidity

B Post-operative

- Sinus Node
  - Dysfunction
- Coronary Arteries
  - Re-implantation
  - Injury
- Atriotomy
  - Scar → reentrant arrhythmia circuit (IART)
- AV Node
  - Swelling / Injury
  - Complete Heart Block
  - Junctional Ectopic Tachycardia
- Ventricular Abnormalities
  - Ventrilucotomy
  - Dysynchrony
  - Scar → reentrant arrhythmia circuit (VT)

Medications
Electrolyte Disturbances
Repolarization Abnormalities
Systemic Illness
Inflammation
Ventricular Arrhythmias in CHD

<table>
<thead>
<tr>
<th>CHD lesion</th>
<th>Ventricular tachycardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ebstein anomaly</td>
<td>&gt; 2%</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>&gt; 5%</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>10%–15%</td>
</tr>
<tr>
<td>Transposition of the great arteries, atrial switch</td>
<td>7%–9%</td>
</tr>
<tr>
<td>Transposition of the great arteries, arterial switch</td>
<td>1%–2%</td>
</tr>
<tr>
<td>Congenitally corrected transposition of the great arteries</td>
<td>&gt; 2%</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>&gt; 2%</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>&lt; 2%</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>&lt; 2%</td>
</tr>
</tbody>
</table>

- Time dependent incremental risk for VA and SCD
- The five defects with the greatest known risk of late SCD are ToF, TGA, ccTGA, aortic stenosis (AS), and UVHs$^{2,3}$.
- Systemic ventricular dysfunction is a dominant predictor for SCD in CHD patients in general
  - VAs without surgical scar/patch
  - e.g. left heart obstruction
  - d-TGA post atrial switch: 0.5% /yr; polymorphic VT or VF$^4$
- rTOF: 11.9% VT incidence; 8.3% SCD @35yrsfup

1. Van der Bom T, et al, Nat Rev Cardiol 2011;8:50–60
2. Oechslin EN, Am J Cardiol 2000;86:1111 – 1116
Ventricular Arrhythmias in CHD

Therapeutic options:

• Treatment of lesions
• Optimal heart failure management
• ICD implantation
• Anti-arrhythmic drugs
• Anti-arrhythmic catheter or surgical interventions
S/P repair DORV with d-TGA and VSD

- 14 yrs old
- Initial palliation with atrial septostomy and PA banding @ 11 months
- Surgical correction (Rastelli) @ 3 yrs (LAD originating from RCA and crossing infundibulum): Sub-aortic conal resection with creation of an internal hemi-tube associated with a homograft RV to PA
- Homograft stenosis treated by percutaneous dilatation first, then replacement and now restenosis of the replacement
- Recurrent VT since 7yrs of age, treated with Sotalol, Cordarone, beta blockers
Abnormal late potentials
VT Ablation in CHD

- After linear free wall ablation and septal substrate ablation, VT1 was not induced but another, VT2 still with a septal breakthrough: ? Subaortic left sided isthmus
- Original clinical VT not induced.
- 1 month later, homograft replaced and ICD implanted
PACES/HRS consensus: ICD indications in CHD

Class I

• 1. In adults with CHD who are survivors of cardiac arrest due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia after evaluation to define the cause of the event and exclude any completely reversible etiology (LOE: B).

• 2. In adults with CHD and spontaneous sustained ventricular tachycardia who have undergone hemodynamic and electrophysiologic evaluation (LOE: B). Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD therapy in carefully selected patients (LOE: C).

• 3. In adults with CHD and a systemic left ventricular ejection fraction <35%, biventricular physiology, and New York Heart Association (NYHA) class II or III symptoms (LOE: B).

Class IIa

• Reasonable in selected adults with tetralogy of Fallot and multiple risk factors for sudden cardiac death, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration >180 ms, extensive right ventricular scarring, or inducible sustained ventricular tachycardia at electrophysiologic study (LOE: B).

Khairy P et al, Heart Rhythm 2014;11:e102–e165
PACES/HRS consensus: ICD indications in CHD

Class IIb

• 1. May be reasonable in adults with a single or systemic right ventricular ejection fraction <35%, particularly in the presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA functional class II or III symptoms, QRS duration >140 ms, or severe systemic AV valve regurgitation (LOE: C).

• 2. May be considered in adults with CHD and a systemic ventricular ejection fraction <35% in the absence of overt symptoms (NYHA class I) or other known risk factors (LOE: C).

• 3. May be considered in adults with CHD and syncope of unknown origin with hemodynamically significant sustained ventricular tachycardia or fibrillation inducible at electrophysiologic study (Level of evidence: B).

• 4. Maybe considered for nonhospitalized adults with CHD awaiting heart transplantation (LOE: C).

• 5. ICD therapy may be considered for adults with syncope and moderate or complex CHD in whom there is a high clinical suspicion of ventricular arrhythmia and in whom thorough invasive and noninvasive investigations have failed to define a cause (LOE: C)

Khairy P et al, Heart Rhythm 2014;11:e102–e165
Anatomical Isthmuses in rTOF
VT: Anatomical isthmuses

..specific electroanatomical isthmus characteristics, in particular the conduction velocity index through an AI are the key determinants for VT. All VT-related AI had an abnormal, low conduction velocity index of < 0.5 m/s.

Patients without a slow conducting isthmus at baseline or after ablation remained VT free during a follow-up of 262 patient years.


### Table: Characteristics

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>No VT (n = 46)</th>
<th>VT inducible (n = 15)</th>
<th>VT spontaneous and inducible (n = 13)</th>
<th>Overall P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isthmus, n</td>
<td>2.0 (1.0–2.0)</td>
<td>2.0 (2.0–3.0)</td>
<td>2.0 (2.0–3.0)</td>
<td>0.111</td>
</tr>
<tr>
<td>Minimal width (mm)</td>
<td>28 ± 11</td>
<td>22 ± 10</td>
<td>18 ± 5*</td>
<td>0.005</td>
</tr>
<tr>
<td>Maximal length (mm)</td>
<td>16 ± 7</td>
<td>20 ± 7</td>
<td>25 ± 7*</td>
<td>0.001</td>
</tr>
<tr>
<td>EA abnormal isthmus, n pts</td>
<td>5</td>
<td>13*</td>
<td>13*</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Lowest CVi (m/s)</td>
<td>0.78 ± 0.24</td>
<td>0.44 ± 0.44*</td>
<td>0.27 ± 0.09*</td>
<td>&lt;0.001</td>
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</table>
Catheter ablation of VT in CHD

Class I:

• As **adjunctive therapy to ICD** in adults with CHD and **recurrent monomorphic VT, a VT storm, or multiple appropriate shocks** that are not manageable by device reprogramming or drug therapy (LOE: C).

Class IIa:

• Can be considered for symptomatic sustained monomorphic VT in adults with CHD and ICDs as **an alternative to drug therapy** (LOE: B).

Class IIb:

• 1. May be reasonable in adults with postoperative CHD and **nonsustained** or hemodynamically poorly tolerated VT by means of an **empiric anatomic approach** (LOE: C).

• 2. Catheter ablation may be reasonable in adults with CHD and **frequent ventricular ectopy** associated with deteriorating ventricular function (LOE: C).

Khairy P et al, Heart Rhythm 2014;11:e102–e165
Value of EP study in CHD

• Class I: EP testing indicated in adults with unexplained syncope and “high-risk” CHD substrates: TOF, TGA with atrial switch surgery, or significant systemic or single ventricular dysfunction (LOE: C).

• Class IIa: EP testing with programmed stimulation useful in adults with CHD and life-threatening arrhythmias or resuscitated sudden cardiac death when the proximate cause for the event is unknown or there is potential for therapeutic intervention at the time of the electrophysiologic procedure (LOE: B).

Khairy P et al, Heart Rhythm 2014;11:e102–e165
Value of EP evaluation in CHD

Class IIa:

1. Holter monitoring can be beneficial as part of routine follow-up in adults with TGA and atrial switch surgery, Fontan palliation, and in patients with tetralogy of Fallot over 35 years of age (LOE: B).

2. Programmed ventricular stimulation can be useful in risk stratifying adults with tetralogy of Fallot who have additional risk factors for sudden cardiac death, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration >180 ms, and extensive right ventricular scarring (LOE: B).

Class III:

EP study not useful for all TOF or for TGA

<table>
<thead>
<tr>
<th>Tab. 1</th>
<th>Reported risk factors for VT and/or SCD in rTOF</th>
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<tbody>
<tr>
<td>Older age at repair</td>
<td></td>
</tr>
<tr>
<td>Transannular patch</td>
<td></td>
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<tr>
<td>RV parameter (moderate to severe dysfunction, severe dilatation, LGE, akinetic region length, mass/volume ratio ≥0.3 g/ml)</td>
<td></td>
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<tr>
<td>Moderate to severe PVR</td>
<td></td>
</tr>
<tr>
<td>LV parameter (moderate to severe dysfunction, longitudinal strain, end-diastolic pressure)</td>
<td></td>
</tr>
<tr>
<td>History of atrial arrhythmias</td>
<td></td>
</tr>
<tr>
<td>(Pre)Syncope</td>
<td></td>
</tr>
<tr>
<td>QRS duration (≥180 ms), QRS duration increase per year</td>
<td></td>
</tr>
<tr>
<td>Nonsustained VT</td>
<td></td>
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<tr>
<td>Inducible for MVT/PVT at PES</td>
<td></td>
</tr>
</tbody>
</table>

Khairy P et al, Heart Rhythm 2014;11:e102–e165
EP study before adult CHD surgery

Class IIa

- Can be useful in adults with CHD to **identify and map** arrhythmia substrates **that may be addressed surgically** with ablation or incisional lesion sets:
- **History of unexplained syncope or sustained ventricular tachycardia** not attributed to correctable predisposing causes (LOE: B).
- Documented sustained supraventricular tachycardia, excluding atrial fibrillation (LOE: C) Ventricular preexcitation (LOE: B/C).

Khairy P et al, Heart Rhythm 2014;11:e102–e165
Ventricular Arrhythmias in CHD: How can the Electrophysiologist help?

- Diagnosis
- Mechanistic Analysis
- Risk stratification
- Choice of therapeutic strategy
- Implementation of specialised therapy
  - Catheter ablation (substrate mapping and imaging, CF guided ablation)
- Improve prognosis