Le Cardiopatie Congenite nell’Adolescente e nell’Adulato

LE ARITMIE

Berardo Sarubbi

U.O.C. di Cardiologia
U.O.S. Cardiopatie Congenite dell’Adulto
Seconda Università degli Studi di Napoli - A.O. Monaldi
Cause di Morte nei GUCH

- Sudden 26%
- Non-cardiac 17%
- Perioperative 18%
- Other CVS 18%
- CHF 21%

Oechsling et al Am J Cardiol 2000
Late Death in Repaired Tetralogy

793 adult pts (1985-95)
33 pts died (4.2% mortality)

Gatzoulis et al. Lancet 2000
The principal reasons for 373 medical admissions to the Royal Brompton Hospital GUCH unit in 1997. Abbreviations: assess, assessment; BE, bacterial endocarditis; med, medical; MRI, magnetic resonance imaging; TOE, transoesophageal echocardiography. From JS, GUCH Unit database.
Ricoveri “GUCH” Anno 2009

Percentuale di Fallot ricoverati per aritmie

A.O. Monaldi
Napoli
**EVENTI CLINICI MAGGIORI DOPO INTERVENTO:** disfunzione ventricolare, aritmie, reintervento

<table>
<thead>
<tr>
<th>Evento</th>
<th>Percentuale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Comunicazione interatriale</td>
<td></td>
</tr>
<tr>
<td>Stenosi polmonare</td>
<td>5%</td>
</tr>
<tr>
<td>Drenaggio venoso polmonare</td>
<td></td>
</tr>
<tr>
<td>Canale parziale</td>
<td>10-15%</td>
</tr>
<tr>
<td>Canale Completo</td>
<td>50%</td>
</tr>
<tr>
<td>Valvulotomia aortica</td>
<td></td>
</tr>
<tr>
<td>Mustard</td>
<td>100%</td>
</tr>
<tr>
<td>Senning</td>
<td></td>
</tr>
<tr>
<td>Fontan</td>
<td></td>
</tr>
<tr>
<td>Fallot</td>
<td></td>
</tr>
</tbody>
</table>
Modifiche della composizione della popolazione GUCH

2010

2020
“Pediatric congenital cardiac becomes a postoperative adult: the changing population of congenital heart disease”
Perloff JK. Circulation 1973; 47:606-619

...it is simple a matter of time before a population of adult with congenital heart disease would emerge.
## Table 3  Healthcare professionals involved in caring for ACHD patients in Europe

<table>
<thead>
<tr>
<th></th>
<th>Number of centres that employ this professional n (%)</th>
<th>Number of professionals in the centre(^a) median (Q1–Q3)</th>
<th>Hours per week spent in ACHD(^a, b) median (Q1–Q3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Specialist centers</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ACHD cardiologist with paediatric background</td>
<td>19 (40%)</td>
<td>2 (1–2)</td>
<td>7.5 (3–15)</td>
</tr>
<tr>
<td>ACHD cardiologist with adult cardiology background</td>
<td>33 (90%)</td>
<td>2 (1–3)</td>
<td>14 (5.8–28.25)</td>
</tr>
<tr>
<td>Paediatric cardiologist</td>
<td>38 (79%)</td>
<td>4 (2.75–7)</td>
<td>3.9 (0.8–11.25)</td>
</tr>
<tr>
<td>Congenital heart surgeon</td>
<td>31 (85%)</td>
<td>2 (2–3)</td>
<td>5 (2–9.5)</td>
</tr>
<tr>
<td><strong>Electrophysiologist</strong></td>
<td><strong>42 (87%)</strong></td>
<td><strong>2 (1–3)</strong></td>
<td><strong>1 (1–4.75)</strong></td>
</tr>
<tr>
<td>Nurse specialist</td>
<td>20 (42%)</td>
<td>2 (1–2)</td>
<td>20 (6.7–35)</td>
</tr>
<tr>
<td>Non-specialist centres</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ACHD cardiologist with paediatric background</td>
<td>7 (26%)</td>
<td>2 (1–4.25)</td>
<td>3 (1–5)</td>
</tr>
<tr>
<td>ACHD cardiologist with adult cardiology background</td>
<td>18 (78%)</td>
<td>1 (1–4)</td>
<td>4 (1.25–9.25)</td>
</tr>
<tr>
<td>Paediatric cardiologist</td>
<td>14 (70%)</td>
<td>2 (1–3)</td>
<td>6 (2.25–24.5)</td>
</tr>
<tr>
<td>Congenital heart surgeon</td>
<td>7 (30%)</td>
<td>2 (2–2)</td>
<td>5 (1.75–23.75)</td>
</tr>
<tr>
<td>Electrophysiologist</td>
<td>10 (43%)</td>
<td>2 (1–3.25)</td>
<td>3.75 (3–8.5)</td>
</tr>
<tr>
<td>Nurse specialist</td>
<td>5 (22%)</td>
<td>1 (1–4.5)</td>
<td>16 (3–17.5)</td>
</tr>
</tbody>
</table>
Cardiopatici Congeniti Adulti
Stratificazione del Rischio Aritmico

✓ Storia Clinica
✓ Parametri ECG Standard
✓ SAECG/LP
✓ SEE
✓ VD/VS Emodinamica, Volume, Funzione
✓ Caratterizzazione “tissutale”
“The essence of wisdom is the ability to make the right decision on the basis of inadequate evidence”

Alan Gregg
Arrhythmias in GUCH
RISK STRATIFICATION

Storia Clinica

✓ Precedenti Interventi CCH
✓ Precedenti Interventi Palliativi
✓ Età all’intervento
✓ Tecnica/Approccio Chirurgico
✓ Durata Follow-up
**TOF: Rischio Aritmico**

Incidenza di SD da 0.5 a 5.5%

TV da rientro

“Scar related”

- Ventricolotomia
- Patch
- Interventricolare
- Patch RVOT
Tachicardie in CHD - Forme incisionali

- **Fontan**: 60%
- **Mustard/Senning**: 25%
- **TOF**: 25%
- **CAV**: 15%
- **DIA**: 10%

**Fontan operation** (cavopulmonary connection): Electroanatomic map of an IART circuit involving the anterolateral surface of the right atrium.

Predictors of Sudden Cardiac Death After Mustard or Senning Repair for Transposition of the Great Arteries

Janneke A. E. Kammeraad, MD,* Carolien H. M. van Deurzen, MD,* Narayanswami Sreeram, MD,* Margreet Th. E. Bink-Boelkens, MD,† Jaap Ottenkamp, MD,‡ Willem A. Helbing, MD,§ Jan Lam, MD,‖ Martha A. Sobotka-Plojhar, MD,¶ Otto Daniels, MD,# Seshadri Balaji, MD**

Utrecht, Groningen, Leiden, Rotterdam, Amsterdam, and Nijmegen, the Netherlands; and Portland, Oregon

✓ Presence of symptoms of Arrhythmia or Heart Failure
✓ History of documented AFL/AF

The best predictors of SCD
Cardiopatici Congeniti Adulti
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✓ Caratterizzazione “tissutale”
<table>
<thead>
<tr>
<th></th>
<th>sVT</th>
<th>A. Flutter</th>
<th>SD</th>
<th>No Arrhy.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>QRS</strong></td>
<td>198.9</td>
<td>177.8</td>
<td>193.8</td>
<td>142.5</td>
</tr>
<tr>
<td><em>p</em> value</td>
<td><em>&lt;.0001</em></td>
<td><em>&lt;.0001</em></td>
<td><em>=.01</em></td>
<td></td>
</tr>
<tr>
<td><strong>CTR</strong></td>
<td>0.67</td>
<td>0.64</td>
<td>0.63</td>
<td>0.53</td>
</tr>
<tr>
<td><em>p</em> value</td>
<td><em>&lt;.01</em></td>
<td><em>&lt;.002</em></td>
<td><em>&lt;.04</em></td>
<td></td>
</tr>
</tbody>
</table>

SD not related to width of QRS


✓ Measurement of QRS is difficult
✓ Can be operator dependent
✓ Can be influenced by the presence of conduction abnormalities which reduce its accuracy and reproducibility.
Gatzoulis et al. Lancet 2000
Arrhythmogenic substrate in young patients with repaired tetralogy of Fallot: Role of an abnormal ventricular repolarization

Berardo Sarubbi¹,*, Giuseppe Pacileo³, Valentino Duceschi², Maria Giovanna Russo³, Carola Iacono³, Carlo Pisacane¹, Aldo Iacono¹, Raffaele Calabro³

¹Secondo Università degli Studi di Napoli, Divisione di Cardiologia Pediatrica-Azienda Ospedaliera V. Monaldi, Napoli, Italy
²Istituto Medico-Chirurgico di Cardiologia-Cattedra di Cardiologia, Napoli, Italy

Received 18 June 1999; received in revised form 25 August 1999; accepted 2 September 1999
Cardiopatici Congeniti Adulti
Stratificazione del Rischio Aritmico

- ✔ Storia Clinica
- ✔ Parametri ECG Standard
- ✔ SAECG/LP
- ✔ SEE
- ✔ VD/VS Emodinamica, Volume, Funzione
- ✔ Caratterizzazione “tissutale”
Signal Average ECG

High accuracy of Signal Average ECG to predict severe VA

Pts operated on for TOF:

<table>
<thead>
<tr>
<th></th>
<th>CONTR OLS</th>
<th>ALL PTS</th>
<th>PTS WITH MINOR ARRHYTHMIA</th>
<th>PTS WITH SEVERE ARRHYTHMIA</th>
</tr>
</thead>
<tbody>
<tr>
<td>QRS 40 (ms)</td>
<td>125 ± 4 *</td>
<td>162 ± 29</td>
<td>156 ± 29 #</td>
<td>181.5 ± 19.6</td>
</tr>
<tr>
<td>LAS 40 (ms)</td>
<td>33.6 ± 13.4</td>
<td>32 ± 22</td>
<td>28.5 ± 19.8 §</td>
<td>45.1 ± 26.7 §</td>
</tr>
<tr>
<td>RMS 40 (μV)</td>
<td>26 ± 8</td>
<td>41 ± 32</td>
<td>45.3 ± 34.6</td>
<td>26 ± 16</td>
</tr>
</tbody>
</table>

*p<0.001 vs pts with minor and severe arrhythmias.
#< 0.01 vs pts with severe arrhythmias

* Time domain

* Frequency domain
**Comparison Between QRS Duration at Standard ECG and Signal-Averaging ECG for Arrhythmic Risk Stratification After Surgical Repair of Tetralogy of Fallot**

GIULIA RUSSO, M.D., ANTONIO FRANCO FOLINO, M.D., Ph.D., ELISA MAZZOTTI, M.D., LUCA REBELLATO, M.D., and LUCIANO DALIENTO, M.D., F.A.C.C., F.E.S.C.

From the Department of Cardiology, University of Padua, Padua, Italy

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**Table**: Comparison of QRS duration at standard ECG and signal-averaging ECG for VT and non-VT patients.

<table>
<thead>
<tr>
<th></th>
<th>No VT</th>
<th>VT</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients (no.)</td>
<td>54</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>QRS duration at standard ECG</td>
<td>152 ± 16</td>
<td>159 ± 20</td>
<td>0.15</td>
</tr>
<tr>
<td>Filter 25–250 Hz</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>fQRS (ms)</td>
<td>175 ± 19</td>
<td>188 ± 18</td>
<td>0.02</td>
</tr>
<tr>
<td>HFLA (ms)</td>
<td>24 ± 15</td>
<td>29 ± 13</td>
<td>0.28</td>
</tr>
<tr>
<td>RMS (μV)</td>
<td>51 ± 33</td>
<td>40 ± 21</td>
<td>0.24</td>
</tr>
<tr>
<td>Filter 40–250 Hz</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>fQRS (ms)</td>
<td>165 ± 17</td>
<td>179 ± 18</td>
<td>0.01</td>
</tr>
<tr>
<td>HFLA (ms)</td>
<td>37 ± 23</td>
<td>47 ± 30</td>
<td>0.19</td>
</tr>
<tr>
<td>RMS (μV)</td>
<td>30 ± 20</td>
<td>26 ± 14</td>
<td>0.5</td>
</tr>
</tbody>
</table>

fQRS = duration of filtered QRS; HFLA = duration of high-frequency low-amplitude signal < 40 μV; QRS = duration of unfiltered QRS; RMS = the root mean square of the mean voltage in the terminal portion (last 40 ms) of the filtered QRS.

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**Table**: Comparison of LVEDV and LVEF between VT and non-VT patients.

<table>
<thead>
<tr>
<th></th>
<th>No VT</th>
<th>VT</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVEDV (mL/m²)</td>
<td>61.9 ± 15</td>
<td>84.4 ± 44</td>
<td>0.003</td>
</tr>
<tr>
<td>LVEF (%)</td>
<td>61.7 ± 6</td>
<td>54.8 ± 10</td>
<td>0.005</td>
</tr>
<tr>
<td>RVEDV (mL/m²)</td>
<td>110.2 ± 31</td>
<td>129.2 ± 36</td>
<td>0.07</td>
</tr>
<tr>
<td>RVEF (%)</td>
<td>53 ± 8</td>
<td>48 ± 9</td>
<td>0.07</td>
</tr>
</tbody>
</table>

LVEDV = left ventricular end-diastolic volume; LVEF = left ventricular ejection fraction; RVEDV = right ventricular end-diastolic volume; RVEF = right ventricular ejection fraction.

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**Figure 1**: Correlation between right ventricular end-diastolic volume (RVEDV) and filtered QRS (fQRS) at 25–250 Hz (panel A) and 40–250 Hz (panel B).
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✓ Caratterizzazione “tissutale”
EPS inducible sustained VT $\iff$ VT or SCD

Khairy et al, Circulation 2004
7% of pts with neg. VSTIM studies died during follow-up

37% of pts with documented sustained VT/VF had no inducible ventricular arrhythmia with VSTIM

- Very low positive predictive value (20%) of VSTIM to predict SCD
- Proarrhythmia of antiarrhythmic drugs
- Management of pts with spontaneous VT and non inducible arrhythmias

Cardiopatici Congeniti Adulti
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✓ SAECG/LP
✓ SEE
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✓ Caratterizzazione “tissutale”
1 normal; 2 mild depression; 3 moderate depression; 4 severe depression.

Gelatt M J et al. JACC, Jan 1997: 29 (1); 194-201
Mechanoelectrical Interaction in Tetralogy of Fallot

QRS Prolongation Relates to Right Ventricular Size and Predicts Malignant Ventricular Arrhythmias and Sudden Death

Michael A. Gatzoulis, MD; Jan A. Till, MD;
Jane Somerville, MD, FRCP; Andrew N. Redington, MD, FRCP

Chronic right ventricular volume overload after tetralogy of Fallot repair is related to diastolic function and correlates with QRS prolongation. The risk of symptomatic arrhythmia is high when marked right ventricular enlargement and QRS prolongation develop.

follow-up, 235 patients who were operated on by one surgeon and who were prospectively studied with a 12-lead ECG, chest radiography, and two-dimensional and Doppler echocardiography. Nine patients (mean follow-up, 17 years) from the total group of 178 were identified as having had sustained ventricular tachycardia (8 with near-miss sudden death), and their ECGs, Holter monitor readings, electrophysiological studies, and chest radiographs were reviewed. The case notes of an additional 4 patients with postoperative sudden cardiac death also were available for review. QRS duration in the 41 patients in whom mechanoelectrical interaction was sought ranged between 90 and 200 milliseconds and correlated with cardiothoracic ratio (CTR) on chest radiography ($r=64$, $P<0.01$) and with right ventricular size on echocardiography ($r=43$, $P<0.02$). Twenty of the 41 patients had restrictive right ventricular Doppler physiology (reduced ventric.

$P<0.01$, respectively, compared with patients without life-threatening arrhythmias). All patients with documented sustained ventricular tachycardia and the 4 patients with sudden death had a QRS duration of $>180$ milliseconds (100% sensitivity).

Conclusions: Chronic right ventricular volume overload after tetralogy of Fallot repair is related to diastolic function and correlates with QRS prolongation. The risk of symptomatic arrhythmia is high when marked right ventricular enlargement and QRS prolongation develop. A QRS duration on the resting ECG of $>180$ milliseconds is the most sensitive predictor of life-threatening ventricular arrhythmias yet described. (Circulation. 1995;92:231-237.)

Key Words • tetralogy of Fallot • arrhythmia • death, sudden • ventricles

The combination of QRS \( \geq 180\text{ms} \) and significant LV systolic dysfunction has a positive predictive value for SCD of 66% and negative predictive value of 93%.
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MYOCARDAL FIBROSIS AND LIFE THREATENING VENTRICULAR ARRHYTHMIAS
VT ablated at site RVOT scar

RVOT scar
Come prevenire e/o trattare le aritmie nei GUCH?
Abbiamo veramente necessità di tanti fattori di rischio?
Malignant arrhythmias occur even in patients with:

- no residual lesion
- no QRS prolongation
- no ventricular dysfunction

The recognition of those who would benefit from an ICD remains a clinical challenge.
Outcome of implantable cardioverter defibrillators in adults with congenital heart disease: a multi-centre study

Sing-Chien Yap¹, Jolien W. Roos-Hesselin², Elke S. Hoendermis², Werner Budts³, Hubert W. Vliegen⁴, Barbara J.M. Mulder⁵,⁶, Arie P.J. van Dijk⁷, Martin J. Schalij⁴, and Willem Drenthen²

¹Department of Cardiology, Thoraxcentre, Erasmus MC, Dr Molewaterplein 40, 3015 GD Rotterdam, The Netherlands; ²Department of Cardiology, University Medical Centre Groningen, University of Groningen, Groningen, The Netherlands; ³Department of Cardiology, University Hospitals Leuven, Leuven, Belgium; ⁴Department of Cardiology, Leiden University Medical Centre, Leiden, The Netherlands; ⁵Department of Cardiology, Academic Medical Centre, Amsterdam, The Netherlands; ⁶University Medical Centre Utrecht, Utrecht, The Netherlands; and ⁷Department of Cardiology,
The finding that the diagnosis of TOF was associated with less appropriate shocks might imply that the abundance of risk factors described for this subgroup has decreased the threshold to consider ICD therapy in this group (more TOF patients had an ICD as primary prevention...)

"...Se si guarda troppo fisso una stella, si perde di vista il firmamento..."
Issues for the use of AICD in ACHD

✓ Indications
 ✓ Inappropriate shocks and lead failure
 ✓ Unique anatomical situations in CHD
 ✓ Technical difficulties
CHD patients are not mentioned as a different group and it is assumed that general guidelines are applicable to these patients as there are not yet clear indications for AID therapy in this group.
High Incidence of Appropriate and Inappropriate ICD Therapies in Children and Adolescents with Implantable Cardioverter Defibrillator

THOMAS KORTE, HARALD KÖDITZ,* MICHAEL NIEHAUS, THOMAS PAUL,† and JÜRGEN TEBBENJOHANNS

From the *Department of Cardiology and Pediatric Cardiology, Medical School Hannover, Hannover, Germany, and †the Department of Pediatric Cardiology, Medical University of South Carolina, Charleston, SC

✓ 20 pts aged 16±6yrs
✓ 11 CHD
✓ 6 Epicardial; 14 transvenous
✓ Therapy-rate 2.8 per patient-years of F-U
✓ 53% appropriate; 47% inappropriate
✓ 1.5 appropriate per patient-year of FU
✓ 1.3 inappropriate per patient-year of FU

PACE 2004; 27:924–932
Implications of Implantable Cardioverter Defibrillator Therapy in Congenital Heart Disease and Pediatrics

MARK E. ALEXANDER, M.D., FRANK CECCHIN, M.D., EDWARD P. WALSH, M.D., JOHN K. TRIEDMAN, M.D., LAURA M. BEVILACQUA, M.D., and CHARLES I. BERUL, M.D.

From the Arrhythmia Service, Department of Cardiology, Children’s Hospital, Boston, Massachusetts, USA; and the Department of Pediatrics, Harvard Medical School, Boston, Massachusetts, USA

### Predictors of Lead Failure

<table>
<thead>
<tr>
<th>Predictor</th>
<th>Odds Ratio (95% CL)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Univariate Analysis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Increase in body surface area</td>
<td>329.67 (8.61–12627.50)</td>
<td>0.002</td>
</tr>
<tr>
<td>Smallest patients</td>
<td>4.45 (1.37–14.46)</td>
<td>0.013</td>
</tr>
<tr>
<td>Subcutaneous array</td>
<td>4.04 (0.81–20.20)</td>
<td>0.089</td>
</tr>
<tr>
<td>Single-chamber device</td>
<td>4.02 (0.85–19.05)</td>
<td>0.079</td>
</tr>
<tr>
<td>Youngest patients</td>
<td>2.9 (0.95–8.87)</td>
<td>0.062</td>
</tr>
<tr>
<td>Change in height</td>
<td>1.09 (1.02–1.16)</td>
<td>0.007</td>
</tr>
<tr>
<td>Change in weight</td>
<td>1.06 (1.01–1.11)</td>
<td>0.028</td>
</tr>
<tr>
<td>Weight</td>
<td>0.99 (0.97–1.02)</td>
<td>0.62</td>
</tr>
<tr>
<td>Height</td>
<td>0.98 (0.95–1.01)</td>
<td>0.27</td>
</tr>
<tr>
<td>Age at implant</td>
<td>0.94 (0.88–1.02)</td>
<td>0.12</td>
</tr>
<tr>
<td>Year of implant</td>
<td>0.93 (0.80–1.09)</td>
<td>0.39</td>
</tr>
<tr>
<td>Generator manufacturer</td>
<td>0.76 (0.26–2.20)</td>
<td>0.60</td>
</tr>
<tr>
<td>Body surface area</td>
<td>0.55 (0.12–2.46)</td>
<td>0.44</td>
</tr>
<tr>
<td>Lead manufacturer</td>
<td>0.42 (0.12–1.48)</td>
<td>0.18</td>
</tr>
<tr>
<td>Subclavicular location</td>
<td>0.41 (0.13–1.27)</td>
<td>0.12</td>
</tr>
<tr>
<td>Multivariate Analysis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Increase in body surface area</td>
<td>73 (3.5–1529)</td>
<td>0.006</td>
</tr>
</tbody>
</table>

### Chronic Complications

<table>
<thead>
<tr>
<th>Complication Type</th>
<th>Patients [n (%)]</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lead failure</td>
<td>16 (21%)</td>
</tr>
<tr>
<td>Inappropriate shocks (no lead failure)</td>
<td>14 (18%)</td>
</tr>
<tr>
<td>ICD storm</td>
<td>3 (7%)</td>
</tr>
<tr>
<td>Elevated pacing thresholds</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Diaphragmatic oversensing</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>L SVC occlusion</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Overall complications</td>
<td>38</td>
</tr>
<tr>
<td>Total no. patients affected*</td>
<td>29 (38%)</td>
</tr>
</tbody>
</table>
Epicardial lead malfunction is common on long-term follow-up. Some leads have a failure of 28% at 4yrs.
A Multicenter Experience with Novel Implantable Cardioverter Defibrillator Configurations in the Pediatric and Congenital Heart Disease Population

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✓ Subcutaneous array placed around the thorax
✓ Transvenous design ICD lead placed on the epicardium
✓ Transvenous design ICD lead placed in a subcutaneous position

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High Incidence of Appropriate and Inappropriate ICD Therapies in Children and Adolescents with Implantable Cardioverter Defibrillator

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[Diagram showing freedom of ICD therapies over months after implantation, with oversensing, VT, ST, AF/AFL, and VF as categories.]
Take-home message

- Le tachicardie sono la causa principale di morbilità e mortalità nella popolazione GUCH
- Sono la principale causa di ospedalizzazione
- La terapia farmacologica spesso è poco efficace
- Esistono difficoltà anatomiche alla terapia non-farmacologica
- Le aritmie possono essere correlate alle specifiche CHD, al peggioramento emodinamico, alla correzione chirurgica
- E’ necessario che i cardiologi che lavorano nelle GUCH Unit abbiano un’adeguata esperienza aritmologica (diagnosi, terapia farmacologica, terapia non-farmacologica, problematiche correlate alle cardiopatie congenite)
Aritmie e CHD: quali aritmie in quali CHD

**TABLE 1. Specific Arrhythmias and Associated Defects in Adults With Congenital Heart Disease**

<table>
<thead>
<tr>
<th>Arrhythmias</th>
<th>Associated Defects</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Tachycardias</strong></td>
<td></td>
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<tr>
<td>Accessory pathways</td>
<td>Ebstein’s anomaly; L-TGA</td>
</tr>
<tr>
<td>Twin AV nodes</td>
<td>Heterotaxy syndrome</td>
</tr>
<tr>
<td>Intra-atrial reentrant tachycardia (atrial flutter)</td>
<td>Postoperative Mustard; postoperative Senning; postoperative Fontan; others</td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>Mitral valve disease; aortic stenosis; un repaired single ventricle</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>Tetralogy of Fallot; congenital aortic stenosis; others</td>
</tr>
<tr>
<td><strong>Bradycardias</strong></td>
<td></td>
</tr>
<tr>
<td>Congenital sinus node dysfunction</td>
<td>Heterotaxy syndrome</td>
</tr>
<tr>
<td>Acquired sinus node dysfunction</td>
<td>Postoperative Mustard; postoperative Senning; postoperative Fontan; postoperative Glenn; others</td>
</tr>
<tr>
<td>Congenital AV block</td>
<td>Endocardial cushion defects; L-TGA</td>
</tr>
<tr>
<td>Acquired AV block</td>
<td>VSD closure; subaortic stenosis relief; AV valve replacement</td>
</tr>
</tbody>
</table>

VSD indicates ventricular septal defect.

Bradicardie in CHD: Fontan

- BAV III: 10%
- Disfunzione SA: 50%

Fontan Procedure
First stage: Bi-directional Glenn:
A graft re-routes blood flow to bypass the right atrium. Blood flows from the superior vena cava directly to the pulmonary artery and then to the lungs to pick up oxygen.

Second stage: Fontan:
A graft and an internal baffle (wall) re-route blood flow from the inferior vena cava to the pulmonary artery.

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Meccanismi causa di aritmia in CHD

- tipici di alcune cardiopatie congenite
- conseguenza dello stress emodinamico o ipossico
- secondari a sequele chirurgiche
Bradicardie in CHD: DIA

Disfunzione del nodo del seno per incannulazione vena cava superiore.
Bradicardie in CHD: DIA

P.M. anni 19.
Difetto interatriale tipo seno venoso s/p correzione radicale.

Studio elettrofisiologico endocavitario. Disfunzione del nodo del seno. TRNS: tempo di recupero del nodo del seno

4700 msec
Istologia: frammentazione del tessuto di conduzione in pz con BAV post operatorio (chiusura DIV con patch).

Bradicardie in CHD: DIV

NAV e fascio di His in rapporto a patch posto a chiusura di DIV perimembranoso sottoaortico.

Istologia: frammentazione del tessuto di conduzione in pz con BAV post operatorio (chiusura DIV con patch).
V.A. anni 16.
Difetto interventricolare s/p correzione radicale.

Studio elettrofisiologico endocavitario: Blocco Atrio-ventricolare sottohissiano.
Bрадидардие in CHD: Mustard

• Disfunzione del nodo seno-atriale
• Distruzione chirurgica di tessuto atriale
• Aree di ritardata attivazione atriale
• Dispersione della refrattarietà atriale

TGA s/p Mustard operation
Bradicardie in CHD: Mustard

- BAV III
- Blocco SA