A Contemporary Assessment of Sudden Cardiac Arrest in Patients with Congenital Heart Disease

Michael J Silka MD
Childrens Hospital Los Angeles
University of Southern California

Sudden Cardiac Arrest – Congenital Heart Disease
An Evolving Medical Problem

- Improved Outcomes and Survival of CHD Patients
  - More Complex Forms of CHD
  - Longer Term Follow-up – New Risk Factors
  - Increased Awareness of the Problem
Sudden Death in Post-op CHD patients (1957-70):

One Year Mortality: Non-Paced Surgical AVB

![Bar chart showing one year mortality for various diagnoses including VSD, TOF, AVC, and L-TGA](chart.jpg)

HRS 2006: 3:601-4

Sudden Death in Post-op CHD patients (1970-79):

Surgically Induced Right Bundle-Branch Block with Left Anterior Hemiblock
An Ominous Sign in Postoperative Tetralogy of Fallot

By Grace S. Wolff MD, Thomas W Rowland MD and R. Curtis Ellison MD

Circulation 1972; 46: 587-96

![ECG image](ecg.jpg)
Sudden Death in Post-op CHD patients (1979-90):  

**Status of the Adult and Adolescent After Repair of Tetralogy of Fallot**

Garson A Jr, Nihill MR, McNamara DG, Cooley DA

*Circulation* 1979; 59:1232-40

---

**Status of the Adult and Adolescent after Repair of TOF**

![Pie chart showing PVC-186 vs PVC+ and SCD 8 vs RVSP > 70, RVEDP > 8 Alive 13]

*Circulation* 1979; 59:1232-40
Incidence of SCD after Surgery for Tetralogy of Fallot

![Graph showing the incidence of SCD over years follow-up.](image)

Meta-Analysis of Factors Associated with Sudden Death After Repair of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Factor</th>
<th>Sudden Death</th>
<th>Alive</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>n=57</td>
<td>n=4570</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular arrhythmia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ECG</td>
<td>61%</td>
<td>8%</td>
<td>0.001</td>
</tr>
<tr>
<td>Exercise</td>
<td>50%</td>
<td>18%</td>
<td>0.001</td>
</tr>
<tr>
<td>Holter</td>
<td>93%</td>
<td>46%</td>
<td>0.001</td>
</tr>
<tr>
<td>EPS</td>
<td>38%</td>
<td>17%</td>
<td>0.01</td>
</tr>
<tr>
<td>Abnormal RVSP</td>
<td>77%</td>
<td>20%</td>
<td>0.001</td>
</tr>
<tr>
<td>Abnormal RVEDP</td>
<td>85%</td>
<td>20%</td>
<td>0.001</td>
</tr>
<tr>
<td>Ventricular arrhythmia and RVSP/EDP</td>
<td>80%</td>
<td>8%</td>
<td>0.0001</td>
</tr>
</tbody>
</table>

Cardiol Young 1991 1:177-181
Incidence of Sudden Death After Tetralogy of Fallot Surgery

<table>
<thead>
<tr>
<th>Study</th>
<th>Findings</th>
<th>Incidence per Decade, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Murphy</td>
<td>6% of 163 cases followed up for 30 yrs</td>
<td>2.0</td>
</tr>
<tr>
<td>Nollert</td>
<td>3% of 490 cases followed up for 25 yrs</td>
<td>1.2</td>
</tr>
<tr>
<td>Silka</td>
<td>2 Deaths per 1000 pt-yrs 445 cases</td>
<td>2.0</td>
</tr>
<tr>
<td>Norgaard</td>
<td>5.6% of 125 cases followed up for 25 yrs</td>
<td>2.2</td>
</tr>
<tr>
<td>Gatzoulis</td>
<td>6% of 793 cases followed up for 21 yrs</td>
<td>3.0</td>
</tr>
</tbody>
</table>

Mechanelectrical Interaction in Tetralogy of Fallot: QRS Prolongation Relates to RV Size and Predicts Malignant Ventricular Arrhythmias and Sudden Death

Circulation 1995; 92:231-7
Risk Factors for Arrhythmia and Sudden Cardiac Death Late After Repair of Tetralogy of Fallot: a Multicentre Study

Lancet 2000: 356:975-81

Relationship of Inducible VT and Subsequent Events in Post-Op TOF

Primary Prevention ICDs in TOF: Annualized Rates of Appropriate Shocks for Various Clinical Parameters

Circulation 2008; 117:363-70

Tetralogy Of Fallot: Current Risk Factors for SCA

- Multiple Potential Risk Factors
  Low Event Rate (~2% / decade) → Minimal PPV of Any One Factor

- Primary Risk Factors:
  - QRS duration >180 ms
  - RV volume overload ≥ moderate PI
  - Inducible Sustained-VT
  - LV dysfunction (EF <40% or EDP >12 mm Hg)

- ICD shock ≠ Sudden Cardiac Death
  7% ICD shock / year ≠ 2% SCD / decade
Sudden Cardiac Arrest – Congenital Heart Disease

An Evolving Medical Problem

Is Sudden Cardiac Death in Postoperative Tetralogy of Fallot a Valid Model for the Study of SCD in Other Forms of Congenital Heart Disease?

Bi-ventricular Heart (TOF)
Systemic Right Ventricle (d-TGA or I-TGA) ?
Uni-ventricular Heart (Fontan) ???

SCD in Patients with a Systemic Right Ventricle

D-TGA s/p Mustard / Senning
Discordant AV / VA Connection
Progression of Ventricular Dysfunction: L-TGA

![Graph showing progression of ventricular dysfunction in L-TGA patients]

Group I (n=121)
Group II (n=47)
P = 0.081

JACC 2000: 36:255-61

Incidence of SCD / Decade post Mustard or Senning for d-TGA

<table>
<thead>
<tr>
<th>Author</th>
<th>Date</th>
<th># patients</th>
<th>Follow-up years</th>
<th>#SCD (total)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Flynn</td>
<td>1983</td>
<td>372</td>
<td>1674</td>
<td>9</td>
</tr>
<tr>
<td>Turina</td>
<td>1988</td>
<td>220</td>
<td>2266</td>
<td>8</td>
</tr>
<tr>
<td>Gewillig</td>
<td>1991</td>
<td>226</td>
<td>2644</td>
<td>37</td>
</tr>
<tr>
<td>Gelatt</td>
<td>1997</td>
<td>478</td>
<td>5544</td>
<td>31</td>
</tr>
<tr>
<td>Silka</td>
<td>1998</td>
<td>172</td>
<td>1413</td>
<td>7</td>
</tr>
<tr>
<td>Moons</td>
<td>2004</td>
<td>257</td>
<td>4369</td>
<td>10</td>
</tr>
<tr>
<td>Dos</td>
<td>2005</td>
<td>137</td>
<td>767</td>
<td>3</td>
</tr>
</tbody>
</table>

TOTAL 1,862 18,677 105 (5.6%)
### Predictors of SCD after Atrial Switch (d-TGA)

<table>
<thead>
<tr>
<th>Significant</th>
<th>P Value</th>
<th>Odds Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart failure</td>
<td>0.001</td>
<td>4.44</td>
</tr>
<tr>
<td>IART / A fib</td>
<td>0.003</td>
<td>21.6</td>
</tr>
<tr>
<td>CHF + syncope</td>
<td>&lt;0.0005</td>
<td>6.45</td>
</tr>
<tr>
<td>Cardiomegaly (CXR)</td>
<td>0.53</td>
<td>2.2</td>
</tr>
<tr>
<td>QRS / QT duration</td>
<td>0.1 - 0.6</td>
<td>0.6</td>
</tr>
<tr>
<td>Non-sinus rhythm</td>
<td>0.79</td>
<td></td>
</tr>
</tbody>
</table>

*JACC 2004; 44:1095-102*

### D-TGA s/p Atrial Repair: Mechanisms / Causes of SCD

IART with a 1:1 Ventricular Response
D-TGA s/p Atrial Repair: Mechanisms / Causes of SCD
Polymorphic VT as a Primary Arrhythmia

D-TGA s/p Atrial Repair: Incidence of Appropriate ICD shocks and Survival in Primary and Secondary Prevention
D-Transposition of the Great Arteries: Risk Factors for SCA

- Event Rate: 5-6% / decade

- Primary Risk Factors: (?)
  - IART - rapid ventricular response
  - RV dysfunction → Polymorphic VT
  - EPS / NOT PREDICTIVE

- ICD shock ≠ Sudden Cardiac Death
  6% / year ICD shock ≠ 5-6% / decade SCD

SCD in the Univentricular Heart

Variation in the Univentricular Heart

- HLHS (Left Heart)
- HRH (TA / PA)
- DILV / Heterotaxy
**SCD in the Univentricular Heart**

Variation in the Fontan Palliations

Atrio-Pulmonary  Lateral Tunnel  Extra-Cardiac

**Freedom from Death or Transplantation Based on Type of Fontan**

![Graph showing freedom from death or transplantation based on type of Fontan. The graph includes three types: RA-RV connection, Total cavopulmonary connection, and RA-PA connection. The x-axis represents time from Fontan surgery in years, ranging from 0 to 25. The y-axis represents freedom from death or transplantation in percentage. Logrank P=0.0018.](Circulation 2008;117:85-92)
Cumulative Hazard by Mode of Death Post-Fontan

Event Rate ~ 5 % / decade: Diverse Causes of SCD

Primary Risk Factors: ( ? )
- IART
- RV (systemic) dysfunction
- EPS / NOT PREDICTIVE

ICD shock vs Sudden Cardiac Death
NO DATA to DATE
Sudden Cardiac Arrest – Congenital Heart Disease

An Evolving Medical Problem

Demographics of Patients with Congenital Heart Disease 2011


JACC 2010: 56:1149-57
Causes of Mortality in Cyanotic and Non-Cyanotic CHD

SCD Risk in Patients with Congenital Heart Disease

**Risk of SCD: CHD Patients vs Adult Population**

- Total CHD population < 25 yrs: 1 / 1,000 pt-yrs
- Male > 35 yr: 1 - 5 / 1,000 pt-yrs
- Tetralogy of Fallot: 20 / 1,000 pt-yrs
- D-TGA / Single Ventricle: 50 / 1,000 pt-yrs
- Prior coronary event: 50 / 1,000 pt-yrs
- EF < 30% or CHF: 150 / 1,000 pt-yrs
- Prior SCD: 200 / 1,000 pt-yrs
- Arrhythmias, low EF: 300 / 1,000 pt-yrs
RISK stratification for SCD following Surgery for CHD

- Critical issue for congenital heart disease patients / physicians
- We can define defects – but not individuals - at risk of SCD
- Primary prevention ICDs - when / are they indicated in CHD?
- Adult CHD patients - will risk factors be additive or exponential?