Critical Left Ventricular Outflow Tract Obstruction (LVOTO)

Fall Work Weekend
Nov. 2013
Work Weekend Objectives

- General overview of cohort
- Generate feasible research question based on current data – Early question
- Develop long term questions of interest
- Focused chart review and data entry
  - Indication for initial Hybrid procedure
Previous CHSS Work

Discordant managed BVR patients

Survival SAV vs. BAV

Hickey et al., McCrindle et al., Ashburn et al.
Original Cohort Objectives

• Assess emerging management strategies

• Identify risk factors predictive of late outcomes

• Readjust the CHSS calculator
Entry Criteria

• 2005- Present
  – Age < 30 days at admission to CHSS institution
  – AV & VA concordance
  – Ductal dependent systemic circulation
  – Excludes patients with AVSD
## General Profile

<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>% Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enrollment</td>
<td>675</td>
<td>100</td>
</tr>
<tr>
<td>Initial procedure</td>
<td>665</td>
<td>99%</td>
</tr>
<tr>
<td>Total deaths</td>
<td>214</td>
<td>32%</td>
</tr>
<tr>
<td>Deaths after intervention</td>
<td>204</td>
<td>30%</td>
</tr>
<tr>
<td>Deaths prior to intervention</td>
<td>10</td>
<td>1%</td>
</tr>
</tbody>
</table>

435 (65%) have had at least one subsequent procedure
# Morphology Based on Baseline Echo

<table>
<thead>
<tr>
<th>Echocardiographic Abnormality</th>
<th>Yes</th>
<th>% yes</th>
<th>No</th>
<th>%No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoplastic Left Ventricle **</td>
<td>512</td>
<td>78%</td>
<td>101</td>
<td></td>
</tr>
<tr>
<td>Aortic Valve Stenosis</td>
<td>164</td>
<td>25%</td>
<td>404</td>
<td></td>
</tr>
<tr>
<td>Aortic Valve Hypoplasia</td>
<td>66</td>
<td>10%</td>
<td>592</td>
<td></td>
</tr>
<tr>
<td>Aortic Valve Atresia</td>
<td>314</td>
<td>48%</td>
<td>344</td>
<td></td>
</tr>
<tr>
<td>Mitral Valve Stenosis</td>
<td>113</td>
<td>17%</td>
<td>70</td>
<td></td>
</tr>
<tr>
<td>Mitral Valve Hypoplasia</td>
<td>196</td>
<td>30%</td>
<td>70</td>
<td></td>
</tr>
<tr>
<td>Mitral Valve Atresia</td>
<td>222</td>
<td>34%</td>
<td>70</td>
<td></td>
</tr>
<tr>
<td>Aortic Coarctation</td>
<td>182</td>
<td>28%</td>
<td>415</td>
<td></td>
</tr>
<tr>
<td>Hypoplastic Aortic Arch</td>
<td>497</td>
<td>76%</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Interrupted Aortic Arch</td>
<td>15</td>
<td>2%</td>
<td>588</td>
<td></td>
</tr>
</tbody>
</table>

*17 patients w/out baseline echo

**135 (21%) with severe/atretic LV
Index Procedure

N=665

- Ross-Konno N=3
- Aortic Valvotomy N=104
- Yasui N=5
- SVR N=547
- HTX N=5
- SVCPA N=1

Deaths prior to intervention=10
# Surgical Description

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>SANO</td>
<td>Stage 1 Pallation- Norwood-RV-PA shunt</td>
</tr>
<tr>
<td>MBT</td>
<td>Stage 1 Pallation- Norwood-Systemic to PA shunt</td>
</tr>
<tr>
<td>HYBR</td>
<td>Stage 1 Palliation- Hybrid</td>
</tr>
<tr>
<td>HYBNR</td>
<td>Hybrid to Norwood Cross-over</td>
</tr>
<tr>
<td>SVCPA</td>
<td>Stage 2 SVC-PA Anastomosis</td>
</tr>
<tr>
<td>HYBSV</td>
<td>Stage 2-Hybrid Single V</td>
</tr>
<tr>
<td>SVBVCO</td>
<td>Single V to Biventricular Cross Over</td>
</tr>
<tr>
<td>FONTAN</td>
<td>Stage 3 Fontan</td>
</tr>
<tr>
<td>HTX</td>
<td>Heart Transplant</td>
</tr>
<tr>
<td>SVALV</td>
<td>Outflow tract and Arch Repairs</td>
</tr>
<tr>
<td>YASUI</td>
<td>Yasui</td>
</tr>
<tr>
<td>AVR</td>
<td>Aortic Valve replacement</td>
</tr>
</tbody>
</table>
AORTIC VALVOTOMY  
N=104

CATH  
N=73

SANO 2  MBT 3  SVALV 4  HYBR 8  HTX 5  AVR 13  ANFS 34  DIED 4

SVALV 1  MBT 1  HTX 1  AVR 2  ANFS 21  DIED 4

SANO 2  MBT 3  SVALV 4  HYBR 8  HTX 5  AVR 13  ANFS 34  DIED 4

CATH  N=73

SANO 2  MBT 3  SVALV 4  HYBR 8  HTX 5  AVR 13  ANFS 34  DIED 4

SVALV 1  MBT 1  HTX 1  AVR 2  ANFS 21  DIED 4

Dead = 20 (3%)
Norwood
N=440

SANO
N=217

SVCPA 152
YASUI 4
HYBV 1
HTX 3
FONT 1
ANFS 17
DIED 39

SVCPA 143
SVAL 1
HTX 6
FONT 1
ANFS 9
DIED 63

HTX 2
FONT 70
DIED 18
ANFS 52
ANFS 1
DIED 2
ANFS 2
ANFS 2

ANFS 1
DIED 61
ANFS 1
ANFS 1

MBT 223

HTX 3
FONT 1
ANFS 17
DIED 39

HYBV 1
HTX 3
DIED 11
ANFS 52
ANFS 4
DIED 1
ANFS 2
DIED 1

SANO Dead = 57 (8%)
MBT Dead = 84 (12%)
Dead = 41 (6%)
Initial Hybrid Procedure

• N=107
  – 37 = Bilateral PA band alone
  – 67 = Ductal stent + Bilateral PA band
  – 3  = Ductal stent alone

• 5  SVBVC, 2 Yasui

• 26 with subsequent Norwood procedure
  – 10 RV-PA shunts
  – 8 MBT shunts
  – 8 Unknown
OTHERS
N=14

- SVCPA
  - MBT
  - SVCPA
  - ANFS
- AVR
  - DIED
- HTX
  - ANFS
  - DIED
- YASUI
  - ANFS

Dead = 2 (%)
Survival After Initial Procedure

- **N=665**
- **Total Died = 204**
- **2yr=71%**
- **68%**

175 deaths within 2 years of initial procedure
Unadjusted Comparison: Based on Index Procedure

Index Procedure
N=665

Aortic Valvotomy
N=104

Yasui
N=5

SVR
N=547

HTX
N=5

Surgical
N=31

Cath
N=73

MBT
N=223

RV-PA
N=217

Hybrid
N=107
Survival: Aortic Valvotomy vs. SVR

Years after initial procedure

% survival

- Blue line: Aortic valvotomy (N=104)
- Red line: Norwood/Hybrid (N=547)

68% survival for Aortic valvotomy
81% survival for Norwood/Hybrid
Survival after SVR: 3 years

- RV-PA Shunt (N=217) = 76%
- Hybrid procedure (N=107) = 66%
- MBT shunt (N=223) = 63%

Years after initial procedure
Survival after SVR: 6 years

39/42 Hybrid deaths occurred w/in first year

Survival %

Years after initial procedure

= RV-PA Shunt (N=217)

= Hybrid procedure (N=107)

= MBT shunt (N=223)
Moving Forward

- Define succinct research question
  - Baseline data entry is complete (demographic, 1st, 2nd procedures, baseline echo)

- Allocate resources for focused data entry

- Chart Review: Indications for Hybrid
  - Norwood Alternative
  - 1V-2V decision deferral
  - Salvage
  - Pre-transplant palliation
Potential Questions

- What are early risk factors for survival after hybrid?
  - What is driving selection of this population?

- What are the differences among 3 subsets of stage I single ventricular repair?

- Should the aortic stenosis calculator be re-evaluated?
Hazard: Sano, MBT, Hybrid
HAZARD: SVR BVR
Hazard for death after initial procedure

N = 665
Background

• Wide spectrum of morphology

• Evolving surgical techniques
  – New tools at our disposal

• Individualized management strategies not well defined
Gaps in Knowledge

• Optimal management
  – Single ventricular repair
  – Biventricular repair

• Borderline population

• Late outcomes
General Profile

• 675 patients

• 665 index procedures

• Initial procedure
  – 82% represent single ventricular repair
  – 15% represent biventricular repair