Hybrid Procedure for HLHS

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Hypoplastic Left Heart Syndrome

Aorta

Pulmonary Artery
Atrial Morphology

A) Large left atrium, thick prominent septum secondum with thin septum primum adherent
B) Small left atrium with thick, muscular atrial septum
C) Giant left atrium, thin atrial septum with severe mitral regurgitation

Rychik J, et al. JACC 1999
# Atrial Morphology and Pulmonary Vascular Histopathology

<table>
<thead>
<tr>
<th></th>
<th>Type A</th>
<th>Type B</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Arteries</strong></td>
<td>Muscular extension into intra-acinar arteries</td>
<td>Muscular extension into intra-acinar arteries</td>
</tr>
<tr>
<td><strong>Veins</strong></td>
<td>Normal</td>
<td>Thick and dilated with arterialized, &gt;2 elastic laminae</td>
</tr>
<tr>
<td><strong>Lymphatics</strong></td>
<td>Normal or mildly dilated</td>
<td>Severely dilated</td>
</tr>
</tbody>
</table>

Rychik J, et al. JACC 1999
Atrial Morphology

Type A:  
n=12  
Survive: 6

Type B:  
n=4  
Survive: 0

Type C:  
n=2  
Survive: 0

Rychik J, et al. JACC 1999
HLHS with Restrictive Atrial Septum

\[ \text{Control patients} \]

\[ p < 0.001 \]

HLHS with restrictive ASD

At risk:

<table>
<thead>
<tr>
<th>Study</th>
<th>33</th>
<th>12</th>
<th>10</th>
<th>9</th>
<th>5</th>
<th>3</th>
<th>2</th>
<th>2</th>
<th>1</th>
<th>1</th>
<th>1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control</td>
<td>66</td>
<td>45</td>
<td>38</td>
<td>32</td>
<td>23</td>
<td>18</td>
<td>13</td>
<td>11</td>
<td>11</td>
<td>10</td>
<td>7</td>
</tr>
</tbody>
</table>

Stage I Palliation

- Atrial septal stent
- PDA stent
- Pulmonary artery band
RV-PA Shunt to HLHS

The “Sano” modification

- Initially reported ↑ survival from 53% to 89%
- Recent multi-institutional report from Japan
  84% survival after Sano, but 1 year survival was 65% & 2 year 63%
- More recent report 92% survival after Sano, and 5 year survival was 73%
Risk Factor after Repair of HLHS

- Mortality after Stage I Palliation using RV-PA Shunt is 7-8% in our institution and more than 90% of the patients with HLHS reached Stage II BDG
- No risk factor after Stage I (RV-PA Shunt)

- Risk factor in mid-long term result
  Intact atrial septum/ restrictive ASD,
  BW less than 2.5kg
  Associated with Non-Cardiac Anomalies

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Difficulties of catheter intervention for restrictive ASD

- Critical condition
- Thick muscular atrial septum
- Small left atrium
- Limited echo window (space)
- Limited vascular access
- Less efficacy if conventional technique
Recent Treatment Strategy to HLHS

(2005.8~2008.8)

HLHS (71 cases)

- m-Norwood (62 cases)
- Bil PAB (9 cases)

Shock, IAS or rAS
low body weight (+α)

- PAB alone (5 cases)
- Hybrid approach (4 cases)
## Demographic Data

<table>
<thead>
<tr>
<th>Case</th>
<th>BW</th>
<th>Diagnosis</th>
<th>ASD</th>
<th>Fetal Diag.</th>
<th>Time to op. (hrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2700g</td>
<td>HLHS/IAS</td>
<td>-</td>
<td>-</td>
<td>23</td>
</tr>
<tr>
<td>2</td>
<td>2660g</td>
<td>HLHS/IAS</td>
<td>-</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>1650g</td>
<td>HLHS/rAS</td>
<td>2.6m/s (L→R)</td>
<td>-</td>
<td>4 d</td>
</tr>
<tr>
<td>4</td>
<td>2470g</td>
<td>HLHS/IAS</td>
<td>-</td>
<td>+</td>
<td>0</td>
</tr>
</tbody>
</table>
## Hybrid approach: Post Op Course

<table>
<thead>
<tr>
<th>Case</th>
<th>PAB size (mm)</th>
<th>Balloon size (mm)</th>
<th>Post op ASD</th>
<th>re-BAS</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3.5</td>
<td>7</td>
<td>2.9mm, 1.2m/s</td>
<td>× 2</td>
</tr>
<tr>
<td>2</td>
<td>3.0</td>
<td>10</td>
<td>2.3mm, 2.0m/s</td>
<td>stent</td>
</tr>
<tr>
<td>3</td>
<td>2.8</td>
<td>7</td>
<td>3.8mm, 0.9m/s</td>
<td>failure</td>
</tr>
<tr>
<td>4</td>
<td>3.0</td>
<td>7</td>
<td>2.4mm, 1.3m/s</td>
<td>× 1</td>
</tr>
</tbody>
</table>
## Result of Hybrid Approach

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (m)</th>
<th>ASD</th>
<th>TR</th>
<th>Post-op ECMO</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3</td>
<td>3.5mm, 1.5m/s</td>
<td>trivial</td>
<td>+ PH desaturation</td>
<td>HD due to PH</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
<td>8.0mm, 1.3m/s</td>
<td>trivial</td>
<td>-</td>
<td>BDG</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
<td>2.7mm, 1.9m/s</td>
<td>trivial</td>
<td>+ LOS</td>
<td>HD Due to PR</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>2.7mm, 2.0m/s</td>
<td>moderate</td>
<td>-</td>
<td>BDG</td>
</tr>
</tbody>
</table>
Conclusions

- Transcatheter decompression of the LA for patients with HLHS is still a challenging procedure. However, the procedure can be performed safely, reduces the transatrial gradient, and improves oxygenation.

- Catheter intervention can contribute to the survival of this condition compared to conventional emergent Norwood procedure.
Overall survival = 76% (47/62)
Okayama Experience - Patients

Okayama University Hospital

- 62 infants (36 boys & 26 girls)
- Age : 3 – 57 days (median, 9 days)
  >14 days : 11 infants
- Weight : 1.6 – 3.9 kg (median, 2.7 kg)
  7 infants < 2.0 kg
  18 infants < 2.5 kg
- Prematurity <37w : 4 infants

exclude  Bilateral PAB : 7 infants
Radiofrequency-Assisted Atrial Septoplasty

Du Marchie Sarvaas et al. CCI 2002
Radiofrequency perforation and Cutting balloon septoplasty

Hill et al. CCI 2005
Umbilical vein approach

Javois et al. CCI 2005
Transhepatic approach

Pedra et al. CCI 2007
Role of catheter intervention for HLHS

- 33 Patients Identified
  - 3 Surgical Septectomy
    - 2 aborted
      - 1) Perforation
      - 1) Inability to enter LA
  - 30 Catheter-based Approach
    - 23 static balloon
      - 4 Rashkind septostomies
      - 1 intra-atrial stent
        - Trans-septal approach in 16/28 (57%)
    - 6 pts required a Surgical septectomy
    - 22/30 (73%) successful catheter based decompressions

Gossett et al. CCI 2006
Current Surgical Outcome

Stage I Survivors
53/58

- Late Death 2
- Await BDG 3
- Stage II BDG 44
- Await BVR 3
- BVR 1

Stage II BDG 44

- Death 2
- Survivors 42
- Late Death 4
- Await Fontan 13
- Stage III Fontan 25

Overall survival = 76% (44/58)
Role of catheter intervention for HLHS

Gossett et al. CCI 2006
Creation of ASD in Utero

Background

• Historically, surgeons and interventionalists have had a somewhat competitive relationship, especially in adult cardiac disease

• Each have been thinking of how to treat patients by their own speciality

• The management of CHD is evolving due to advances in transcatheter therapies that coincide with surgical strategies to improve outcomes
Hybrid Approach – What is it?

- Collaborative effort between surgeons and interventional cardiologists
- Collaborative effort between physicians and industry
- Sharing of ideas, expertise, equipment, & techniques
- Development of novel treatment strategies
Why?

- Offer best treatment to the patients
- Reduce morbidity & mortality
- Improve quality of life
- Deliver more efficient & cost effective care
Hybrid Cardiac Procedures for CHD

– Group I: intraoperative stents
– Group II: perventricular muscular VSD
– Group III: PA bands & PDA stent
– Group IV: young adults requiring combined interventional & EPS
– Group V: unusual Hybrid procedures
Hypoplastic Left Heart Syndrome

“A Benchmark for the Surgical Treatment of Congenital Heart Disease-
Hybrid Approach to HLHS

**So What’s The Big Deal?**

- **Neurologic & Developmental Morbidity**
  - After Norwood repair: Full Scale IQ
    - Kern, et al: 91
    - Mahle, et al: 86
    - Goldberg, et al: 94
  - Abnormalities of speech & language, oral aversion & poor feeding, poor adaptive behavior, & growth failure
  - Later, there is significant emotional & behavior dysfunction, low self esteem, & psychosocial and physical health issues

*Wernovsky & Newburger, J Peds, Vol 142: Jan, 2003*
Hybrid Approach to HLHS

“Hybrid” Concept Of HLHS Repair

- Less invasive procedures (Bilateral PAB +/- Stent)
- Avoid open heart surgery in Neonate, DHCA
- One comprehensive open heart procedure at an age appropriate for the “big operation”
  - Stage 1 Neo-aortic reconstruction
  - Bidirectional Glenn
Challenge in the management of HLHS
Overall survival rate

One-year survival rate: 80%
Five-year survival rate: 73%

Survival Function

Cum Survival

0.0 0.1 0.2 0.3 0.4 0.5 0.6 0.7 0.8 0.9 1.0
Survival Function
Censored

Survival Function

months f/u till 2007-12
Hybrid Approach

Hybrid Cathe / OR
Stage I HLHS Hybrid Palliation
Self Expandable Zilver stent
8mm x 20mm

It Jumps !!!
Ductal stent overriding the isthmus at 5 months
**Indication of Hybrid Procedure**

- HLHS/IAS, HLHS/rAS
- BW< 2.0-2.5 Kg
- Poor preoperative condition
- Shock
- Severe renal failure, liver failure
- Sepsis
New Hybrid Strategy

New Hybrid Procedure

- Bilateral PAB
- ASD creation

Without using CPB

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Strategy in the management of HLHS - Recent Era

RV-PA Shunt  Bilateral PAB ± Stent
Strategy in the management of HLHS

Recent Era ~ Near Future

*Fetal Intervention*

- To create atrial septal defect
- To decrease PVR
- To stimulate growth of LV and Aorta