Hybrid Management of HLHS: Indications, Technique, & Outcomes

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Disclosures

• As a faculty member of SCAI, I have the following disclosures
  – None

• Off label use of FDA approved devices will be discussed
Acknowledgements

• Mark Galantowicz…NCH
• Sharon Cheatham…NCH
• Dietmar Schranz…Giessen
• Carlos Pedra…Sao Paulo
Two Perspectives. Single Focus.
If it’s so good...

- Sano Procedure (RV-PA Conduit)
- Norwood Procedure
- Hybrid Palliation
- Transplantation
- Fetal Intervention
Our Hybrid Concept of HLHS Repair

• Initial palliation with less invasive procedure (Hybrid Stage I) in neonatal period to stabilize to an age and weight appropriate for the “big operation”
  • Control & protect PBF (LPA/RPA bands)
  • Provide reliable systemic cardiac output (PDA stent)
  • Create unobstructed flow from LA (BAS/stent IAS)

• One comprehensive open heart procedure (combine stage I and II)

• Initially transcatheter Fontan completion…now extracardiac pericardial baffle Fontan completion
Good Company…

“The future of treatment for HLHS may eventually involve a single open-heart procedure with initial and final interventions being performed in the catheterization laboratory.”

William I Norwood, MD
Minerva Pediatr, 2004:56:41
Dave’s Top 10 Reasons Why Hybrid Stage I Should be Offered to ALL Patients with HLHS
There has been no significant improvement in over a decade, despite multiple centers, with many bright people trying. Perhaps the anatomy, physiology, and impact of the traditional procedures for HLHS have reached their limit.

It is time to evolve!
As reported by Dr. Ohye at the AHA 2009

- Death/txp at 1 year post-op
  - 36% Norwood
  - 26% Sano

- Serious (non-fatal) complications
  - ~30% both groups
Why Evolve?

• Improve survival – quantity of life
• Improve morbidity – quality of life
• Develop new techniques & treatment strategies to facilitate both
• To better match diagnosis to treatment

“It is not the strongest of the species that survives, nor the most intelligent, but the one most responsive to change”
— Charles Darwin
Evolution of Techniques for Hybrid Stage I

• Phase I
  – All transcatheter: PAFR, PDA stent, BAS

• Phase II
  – PDA stent 1st & BAS, then to O.R. for PA bands
    • Too difficult to access LPA, BAS didn’t last

• Phase III
  – PA bands 1st, then to cath lab for PDA stent & BAS
  – Giessen approach

• Phase IV
  – PA bands, then PDA stent in Hybrid Suite
  – BAS before discharge allowed 2ml balloon
October, 2001
Amplatzer PA Flow Restrictors
Transcatheter Stage I Completed
Hybrid Stage I Palliation

PA Bands

PDA Stent

BAS/Stent
1st LPA/RPA Bands & then PDA Stent
3DRA after Hybrid Stage I
Contraindications To Hybrid Stage I

• Size of patient … NO
• Size of ascending aorta … NO
• Poor RV function/TR … NO
• Eventual biventricular physiology … NO
• Transplant candidate … NO
• Borderline LV … NO
• Retrograde arch obstruction … YES for NCH
  – No for Toronto Sick Kids
Size Doesn’t Really Matter… 1.1 Kg after PA bands & PDA stent (previous aborted fetal intervention)
## Intermediate Results

<table>
<thead>
<tr>
<th></th>
<th>CCH-Hybrid Approach</th>
<th>CHB BTS</th>
<th>CHB RVPA</th>
<th>CHOP BTS</th>
<th>CHOP RVPA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mortality Stage 1</td>
<td>2%</td>
<td>11%</td>
<td>14%</td>
<td>14%</td>
<td>17%</td>
</tr>
<tr>
<td>Mortality Interstage</td>
<td>5%</td>
<td>14%</td>
<td>0%</td>
<td>18%</td>
<td>5%</td>
</tr>
<tr>
<td>Interstage re-intervention</td>
<td>26%</td>
<td>51%</td>
<td>38%</td>
<td>17%</td>
<td>32%</td>
</tr>
<tr>
<td>Rate of weight gain (gm/day)</td>
<td>16 (9-33)</td>
<td>16.5 (10-60)</td>
<td>20.6 (10-40)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate or Severe RV dysfunction pre-2</td>
<td>0%</td>
<td>4%</td>
<td>10%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate or Severe TV regurgitation pre-2</td>
<td>4%</td>
<td>26%</td>
<td>16%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mortality Stage 2</td>
<td>6%</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Overall Survival (“usual risk”)</td>
<td>87% (“92%”)</td>
<td>69%</td>
<td>80%</td>
<td>68% (“86%”)</td>
<td>70% (“86%”)</td>
</tr>
</tbody>
</table>

*Galantowicz et al, Ann Thorac Surg, 2008*
NCH Experience: Overall Mortality

- Hybrid Stage I, Interstage, and Comprehensive Stage II
  - n=36; 4 patients transferred care closer to home
- Excluding those transferred or managed by another institution
  - n=32/131 (24% mortality)
    - Prematurity \( \leq 36 \) weeks gestation 11/32 (34%)
    - Neuro injury 11/32 (34%)
      - 2 of which on ECMO
    - n=1 intact atrial septum
The spirit of a team gets visions to become reality

Ina Michel-Behnke
Interventionalist

Dietmar Schranz
Interventionalist

Jürgen Bauer
HTX / MRI

Matthias Müller
Anaesthesiologist

Hakan Akintürk
Surgeon

Klaus Valeske
Surgeon

Pediatric Heart Center
JUSTUS-LIEBIG-UNIVERSITÄT GIESSEN
Hybrid Stage I

Hybrid stage I: Approach Surgical BPA and Percutaneous DA-stenting

Actuarial survival is 80% after 1 year
78% after 2 years
77% after 14 years

Real: 27/144 after 14 years
81% survival

Actuarial survival curve of all treated by HYBRID SI approach
Hybrid palliation for neonates with hypoplastic left heart syndrome: current strategies and outcomes.
Honjo O, Caldarone CA.
Division of Cardiovascular Surgery, Hospital for Sick Children, Toronto, Ontario, Canada.

“In the last decade the hybrid procedure has emerged as an alternative stage I palliation in neonates with hypoplastic left heart syndrome.”
Interstage Monitoring

• Crucial to overall success
• Home monitoring program
• PE, echo, EKG, O2 sats every 1 - 2 weeks
• Assess antegrade & retrograde flow through PDA stent, PAB flow, and ASD flow
• Beware depressed RV function or increased TR
• Any significant changes, pt goes to the cath lab
  – Need to have a low threshold to recath
  – Most problems can be treated without surgery
Two Challenging Areas

- **Retrograde aortic arch stenosis after the Hybrid Stage I**
- **Pulmonary artery thrombosis after the Comprehensive Stage II**
  - We initiated an anticoagulation protocol post op Comprehensive Stage II...no more cases
Retrograde Aortic Arch Obstruction (RAAO)
The Achilles Heel of the Hybrid Approach
Predictors of Retrograde Aortic Arch Obstruction After Hybrid Palliation of Hypoplastic Left Heart Syndrome

Matthew J. Egan · Sharon L. Hill · Bethany L. Boettner · Ralf J. Holzer · Alistair B. Phillips · Mark Galantowicz · John P. Cheatham · John P. Kovalchin

• Incidence of RAAO underestimated initially
• Currently 29% of patients demonstrate RAAO
• The mean aortic root size smaller in patients who developed RAAO (3.6 vs. 4.4mm)
• Angle between isthmus and PDA larger in those who developed RAAO (86 vs. 63 degree)
• The type of stent (BES vs SES) and the crossing of the RAA were not factors
Retrograde Aortic Arch Stenosis
NCH Experience: Neurodevelopment

• Cerebral blood flow and neurocognitive and developmental outcomes in HLHS infants who undergo Hybrid Stage I procedure is unknown.

• The purpose of this study is to compare neurodevelopmental outcomes and cerebral blood flow in HLHS infants after Hybrid Stage I palliation, with normal, age-matched, controls, as well as norm-referenced control data.

S.L. Cheatham, PhD, ACNP
NRSA F-31 NIH Training Grant
In press Pediatrics
### Results of TIMP

*Significant at .05  
** -0.5 to -1 SD below the norm (low average)  
***-1 to -2 SD below the norm (below average)  
(norm @ 8-9 weeks is 93 ± 18; 16-17 weeks 120 ± 16)

<table>
<thead>
<tr>
<th>Group</th>
<th>Median age</th>
<th>Mean TIMP Score (median, range)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLHS (n=14)</td>
<td>62 days</td>
<td>***63.9 ± 18.1 (61.5, 30-98)</td>
<td>0.002*</td>
</tr>
<tr>
<td>(n=12)</td>
<td>117 days</td>
<td>**108.3 ± 14.9 (111, 83-127)</td>
<td>0.0019*</td>
</tr>
<tr>
<td>Control (n=6)</td>
<td>62</td>
<td>94.5 ± 10 (91, 85-112)</td>
<td></td>
</tr>
<tr>
<td>(n=5)</td>
<td>120 days</td>
<td>133 ± 4 (133, 128-138)</td>
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</table>
Results of Bayley Scales, 3rd edition

<table>
<thead>
<tr>
<th>Group</th>
<th>N Obs</th>
<th>Variable</th>
<th>N</th>
<th>Mean</th>
<th>Median</th>
<th>Minimum</th>
<th>Maximum</th>
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<tbody>
<tr>
<td>HLHS</td>
<td>18</td>
<td>Cognitive</td>
<td>11</td>
<td>90.4</td>
<td>95.0</td>
<td>55.0</td>
<td>110</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Language</td>
<td>11</td>
<td>88.5</td>
<td>89.0</td>
<td>62.0</td>
<td>106</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Motor</td>
<td>11</td>
<td>78.9</td>
<td>74.0</td>
<td>46.0</td>
<td>103</td>
</tr>
<tr>
<td>Control</td>
<td>6</td>
<td>Cognitive</td>
<td>6</td>
<td>99.7</td>
<td>102.5</td>
<td>84</td>
<td>115</td>
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<tr>
<td></td>
<td></td>
<td>Language</td>
<td>6</td>
<td>88.8</td>
<td>94</td>
<td>58</td>
<td>118</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Motor</td>
<td>6</td>
<td>97.8</td>
<td>107</td>
<td>68</td>
<td>110</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Variable</th>
<th>6 month Bayley Cognitive</th>
<th>6 month Bayley Language</th>
<th>6 month Bayley Motor</th>
</tr>
</thead>
<tbody>
<tr>
<td>p-value</td>
<td>0.2883</td>
<td>0.6849</td>
<td>0.0489*</td>
</tr>
</tbody>
</table>

*P value significant at <.05
IS CEREBRAL BLOOD FLOW COMPROMISED IN HLHS FOLLOWING HYBRID STAGE I PALLIATION?
Transcranial Doppler Ultrasound

Blood flow = Velocity $\times \pi \times \text{radius}^2$
Transcranial Doppler of MCA in HLHS
Motor scores are statistically significantly lower in the HLHS group compared to normal controls.

No significant correlation between TCD and NDO measures of cognitive, language, and motor skills.

TCD values are lower at 5-6 months in HLHS patients compared to normal age-matched infants.

HLHS infants who undergo Hybrid Stage I palliation do not appear to have any worse adverse effect on NDO scores compared to previously reported scores after Norwood/Sano Stage I.

Since these abnormalities begin early in life, early intervention is recommended. (Agree with Dr. Newburger)
“LAUNCH Evolution”

- **Lifetime**
- **strAtegies &**
- **oUtcome for**
- **siNgle ventricle**
- **and Complex**
- **Hearts**
Discussion of vision for Single Ventricle Care

- Multiple common themes across groups
- Multiple individual projects within each group
Comprehensive Stage II
The First+Final Surgical Approach

Removal of bilateral PAB+DS
Aortic Arch reconstruction
Atrial Septectomy
Cavo-pulmonary anastomosis

Future:
Remove of PDA stent, if...
Repair of pulmonary arteries
Atrial Septectomy,
Repair of Tricuspid valve, if...
RAoA in beating heart technique
Preparing later TCPC
Stem cell therapy, if...
Hybrid CT Operative Suite For CHD
Exit Angiogram after Comprehensive Stage II
Exit Angiograms We Like To See

PVR & Intrap PA Stents

Comprehensive Stage II
Exit Angiogram We Don’t Like To See But We Fix It!
3DRA versus Conventional Angio
POD #3 after Comprehensive Stage II Repair
Controlling diastolic run-off into PAs
Alternative Surgical Option n=3
Aortic Arch Reconstruction, Remove PDA & RAA Stents, Leave PAB, +/- Atrial Septectomy
Short Valved Homograft from MPA into Aortic Arch
Gerd Hausdorf

Academic achievements:

• Interventional Fontan completion – a hallmark of his innovative thinking
  – “Charite Fontan”

“Hausdorf G; Schneider M; Konertz W. Surgical preconditioning and completion of total cavopulmonary connection by interventional cardiac catheterisation: a new concept. Heart. 1996; 75(4): 403-9”
e-PTFE Covered NuMED CP Stent (Original)
Transcatheter Fontan Completion

23 mo/old with 8Zig-55mm & 8Zig-34mm CCPS on 12mm BiB

Flared to 16mm & 18mm @ IVC
Future Materials

• Biodegradable stent
  – Use platform for DES, PGE1, resveratrol
  – Working with 480 Medical

• Histotripsy
  – Tissue Fractionation US
  – Zhen Xu, PhD
Hybrid Approach to HLHS: Conclusions

• Results of Hybrid therapy are comparable to conventional surgical therapy and results are reproducible, but there is an expected learning curve as with any new procedure.

• Treating retrograde aortic arch obstructions remains a challenge, but stent therapy is the first line of treatment.

• Rule of 6s … If early stent therapy is required in the first 6 weeks, patient will be scheduled for surgical intervention ~ 6 weeks following stent placement (Norwood/Sano, early Comp II, or new alternative surgical technique), irrespective of clinical improvement.

• Neurodevelopmental outcomes are no different from Norwood/Sano & early intervention is indicated in HLHS, regardless of the method of repair.

• Hybrid therapy will find a home in all institutions, regardless of institutional bias…do what you do best!
One Final Reward for a Job Well Done

The Hybrid Martini
You Need Help From Many Friends
Thank You