Hybrid for HLHS - Objectives

• Background – Recognized problems and theoretical solutions
• Technique – Tips and Tricks of the Trade
• Indications and outcomes – Do we really know?
• Starting a New Cath Service – Advice on how to achieve success (or cushion failure)
• Early career advice
Hybrid for HLHS - Objectives

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• Indications and outcomes – Do we really know?
• Starting a New Cath Service – Advice on how to achieve success (or cushion failure)
Hypoplastic Left Heart Syndrome (HLHS)

Norwood Procedure for HLHS

1. No permanent source of Aortic blood-flow

2. Restrictive atrial septum = obstructed pulmonary flow

3. Unrestricted Pulmonary blood-flow = Aortic “steal”

With new thinking, more is possible.
Norwood Procedure for HLHS

Restrictive atrial septum = obstructed pulmonary flow

1. No permanent source of aortic blood-flow
2. Unrestricted pulmonary blood-flow = aortic "steal"
3. Restrictive atrial septum = obstructed pulmonary flow

With new thinking, more is possible.
Surgical Approaches

• Norwood
• Norwood – Sano modification
• Modifications of modifications
• Transplant
• When there are multiple modifications, it’s for a reason
Operative mortality as a function of institutional volume from 1998 to 2007 for stage 1 palliation (S1P) of hypoplastic left heart syndrome.

Three-stage Palliation of Hypoplastic Left Heart Syndrome in the University HealthSystem Consortium
Kimberly E. McHugh, MD,*
Congenit Heart Dis. 2010;5:8–15
HLHS - Neurodevelopmental

• HLHS-Norwood Neurodevelopmental Outcome
  – Traditional Norwood - > 5% onset of new seizures
  – HLHS has lower than normal IQ, increased ADHD, and increased psychiatric diagnoses
  – Why?
HLHS - Neurodevelopmental

- Reasons for Neurodevelopmental Outcome
  - Multifactorial
  - Age at time of cardiopulmonary bypass
Cardiopulmonary bypass

Neonates vs. Infants

• Periventricular Leukomalacia (PVL) is one of the hallmarks of brain injury
• There is an age-dependent window of vulnerability to injury of developing white matter during myelination.
Cardiopulmonary bypass
Neonates vs. Infants

- Infant brain – “relatively resistant to injury from conditions such as hypoxia-ischemia”
- Neonate brain - “differentiating oligodendrocytes are exquisitely sensitive to a variety of insults, including hypoxia, ischemia, reactive oxygen species, and inflammatory mediators”

“Periventricular leukomalacia is common after neonatal cardiac surgery.”
Cardiopulmonary bypass
Neonates vs. Infants

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“Periventricular leukomalacia is common after neonatal cardiac surgery.”
Cardiopulmonary bypass
Neonates vs. Infants

- MRI performed on neonates and older infants after cardiopulmonary bypass
- Neonates - PVL was seen in 54% post-op neonates
- Older Infants - PVL was only seen in 4% post-op infants.
- Thus, younger age at time of operation is a potential predictor of poor neurodevelopmental outcome

“Periventricular leukomalacia is common after neonatal cardiac surgery.”
Can CPB and circulatory arrest be avoided?
Hypoplastic Left Heart Syndrome (HLHS)

No permanent source of Aortic blood-flow

1

Restrictive atrial septum = obstructed pulmonary flow

3

Unrestricted Pulmonary blood-flow = Aortic “steal”

2

Some success

With new thinking, more is possible.
Amplatzer PA Flow Restrictor
Evolution of Technique

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

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

• Phase II
  – PDA stent 1st & BAS, then to O.R. for PA bands
    • Difficult to access LPA for band

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

• Phase IV
  – PA bands, then PDA stent…the beginning of Hybrid approach
    • Started in conventional O.R. then migrated to Hybrid Suites
  – BAS before discharge
Hypoplastic Left Heart Syndrome (HLHS)

1. No permanent source of Aortic blood-flow
2. Unrestricted Pulmonary blood-flow = Aortic “steal”
3. Restrictive atrial septum = obstructed pulmonary flow
PDA stent for aortic flow

PA bands regulate pulmonary flow

BAS for unrestricted atrial septum (later)
Intraoperative PDA Stent & RPA/ LPA Bands In HLHS Phase IV in the O.R.
PDA Stent

• PDA stenting for Hybrid HLHS:
  – What type of stent?
PDA Stent Types

– Balloon expandable
  • Pro: Can overcome PDA narrowing
  • Con: Obstructs systemic output during inflation
  • Caution: surgical access point into MPA needs to be far enough from stent that proximal balloon is in the vessel

– Self expandable
  • Pro: Can be slowly deployed without obstructing ductal dependent systemic flow
  • Con: relatively little outward radial force
  • Caution: can “jump” at end of exposure
Pre-mounted Balloon Expandable Stents

Palmaz Genesis
(Closed Cell)

Cook Formula 418
(Open Cell)

With new thinking, more is possible.
Self-Expandable Stents

With new thinking, more is possible.
Protégé – eV3

Different lengths:
- 20 mm
- Overlap multiple stents if need more length

Different diameters:
- 6-10 mm

Different delivery system lengths:
- 80 cm

(Diameters up to 14mm)

With new thinking, more is possible.
PDA Stent

• PDA stenting for Hybrid HLHS:
  – What type of stent?
  – Technique
    • Access
    • Preparation
    • Landmarks
With new thinking, more is possible.
MA – PDA lateral
MA – PDA lateral

CATH 4
Ex: R200705011215542
Ped 30 frs Low Contrast
C: UNKNOWN
Se: 7/10
Im: 7/199 (Fr: 1/199)

University of Minnesota Medical Center Fairview
AMERMOCHE MOHAMED
2007 Apr 25 M 0051023143
Acc: 7539222
2007 May 01
Img Tm: 12:47:58:000
MA – PDA lateral
MA – PDA lateral
• PGE shut off in lab – Transferred to PICU
• Extubated and off inotropes < 24 hours
• Echo next day showed question of obstruction of proximal PDA
• 3 days later, there was significant obstruction - PGE reinitiated & obstruction resolved
• Returned to the lab 6 days after Hybrid
With new thinking, more is possible.
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
- Retrograde aortic arch flow obstruction…Debatable
Interstage Monitoring

- **Crucial** to overall success
- PE, echo, O2 sats every 1 - 2 weeks
- Personal preference – single individual follows all hybrid patients in specific clinic
- Assess antegrade & retrograde flow through PDA stent, PAB flow, and ASD flow
- Beware depressed RV function or increased TR
  - Early sign of a problem
- Any significant changes, pt goes to the cath lab
  - Need to have a low threshold to recath
  - Most problems can be treated without surgery
Post Mortem Exam: After Hybrid Stage I Palliation For HLHS

Stenosis Of Transverse Aortic Arch Ostium

LPA Band

RPA Band

PDA Stent

Atretic Asc Ao

LPA Band
Retro-Aortic Arch Obstruction

- Warning signs:
  - Poor feeding, diaphoresis, fussiness
  - Increased or new TR
  - RV dysfunction
- Pre-mounted balloon-expandable
- Retro-aortic (and coronary) flow is occluded during inflation
Another Solution

“Reverse” R MBT shunt:

Chris Caldarone, TSKH
Who gets offered a Hybrid?
CONGENITAL CARDIOVASCULAR INTERVENTIONAL STUDY CONSORTIUM (CCISC)

- Pediatric/SHD Interventionalists
- 50+ centers in US and International
- Multicenter Studies
- Email server – Study updates, clinical cases, discussion topics
Types of Pediatric Hybrid

- PA band/PDA stent
- Perventricular VSD
- Stenting
- Other

- Cut-down access
- PA/IVS
- Balloon angioplasty
- Perventricular valves
- Perivalvar leak
- Bidirectional Glenn
- Fetal
Why aren’t more doing it?

With new thinking, more is possible.
Follow-up Question

• What is your institutional approach to offering PA bands/PDA stent for HLHS?
  – Hybrid is an equal option to Norwood
  – Hybrid is preferred option to Norwood
  – Hybrid is offered to sub-optimal Norwood candidates
Institutional Approach to Offering Hybrid Palliation for HLHS

- Equal option to surgery
- Hybrid is preferred option
- Hybrid is only offered for "sub-optimal" surgical candidates
Development of a new service/procedure

• Identified a problem
  – Outcomes/Survival
    - Neurodevelopmental

• Consider new approaches
  – Discussion about Hybrid Approach
  – ISHAC 2006 – Nationwide Children’s Hospital
U of Minnesota Amplatz Children’s Experience

• 2006 – ISHAC meeting at Ohio State
• Discussion with entire pediatric cardiac group
• Consensus – Hybrid approach would be offered to all patients with HLHS and “appropriate anatomy” (i.e. no significant risk of retrograde Ao arch obstruction)
• Continue to offer until 10 HLHS patients have undergone approach – then review
FIRST HYBRID HLHS STAGE I

- May 1, 2007
U of Minnesota Amplatz Children’s Experience

• After 12 patients underwent hybrid branch PA banding and PDA stent (only 9 with HLHS or variant anatomy)

• Analyzed in 3 groups:
  – Group A – HLHS
  – Group B – HLHS, but “bad Norwood candidates”
  – Group C – other anatomy; BDG not intended next surgical goal
Patients

• 12 Hybrid PA band/PDA stent (5/08 – 7/10)
  – Group A: HLHS (n=7)
  – Group B: HLHS “bad Norwood” (n=2)
  – Group C: non-HLHS; not intended for BDG as next operation (n=3)
Patients

• Group B: HLHS “bad Norwood candidates” (n=2)
• Originally discussed plan was to not offer hybrid to these patients
  – Patient LC – Mitral atresia/VSD/ Hypoplastic LV
    • No prenatal Dx; born outside institution
    • Retrograde arch obstruction
    • Severe RV dysfunction and TR
    • Heart Center consensus – too high risk for Norwood
  – Patient EK – HLHS/retrograde arch obstruction
    • No prenatal Dx; born outside institution
    • Severe RV dysfunction
    • Lactate = 15; Seizures on arrival with abnormal EEG
    • Heart Center consensus – too high risk for Norwood
Results – Stage I Hybrid PA band/PDA stent

• Survival to Hospital Discharge
  – 100% (all Groups)
  – 81-93 % 30 day survival for Norwood
Results – Hybrid PA band/PDA stent

• Open Chest
  – 0 % Hybrids (all Groups)
  – (74-76% in Norwood BT vs Sano NEJM paper)

• ECMO
  – 0 % Hybrids (all Groups)
  – (8-12% in Norwood BT vs Sano NEJM paper)
Results – Hybrid PA band/PDA stent

• Time to extubation
Hybrid Stage I - Time to Extubation

Norwood BT vs Sano (NEJM)
Median = 7 days

HLHS
Range = POD 0-4
Mean = 1.1
Results – Hybrid PA band/PDA stent

• Time to discontinuation of inotropes
Hybrid Stage I - Time on Inotropes

- HLHS
  - Range = POD 0-4
  - Mean = 0.75

Norwood BT vs. Sano
(NEJM)
N/A

All Groups
A
B
C
Results – Hybrid PA band/PDA stent

• Time to discharge from PICU
Hybrid Stage I - Time in PICU

Nnorwood BT vs Sano (NEJM)
Median = 14 days

HLHS
Range = POD 3-11
Mean = 6.9
Results – Hybrid PA band/PDA stent

- Time to discharge from hospital
Hybrid Stage I - Day of Discharge

Norwood BT vs Sano (NEJM)
Median = 24 days

HLHS
Range = POD 4-32
Mean = 16.1
Results – Interstage

• Interstage Deaths – 0 patients (4-15% in Norwood)
• Additional unplanned interventions between Stage I discharge and Stage II – 5 interventions/4 patients (42 procedures/100 pts)
  – BAS = 3
  – Retrograde Ao stent (group B)
  – PDA stent
  – mBT shunt = 56 procedures/100 patients
  – Sano = 96 procedures/100 pt
Hybrid Stages I vs. Norwood

• Is it fair to compare Stage I Hybrid to Norwood?

• Are we just “shifting the risk”?
Combined Stage I & II

Norwood BT vs Sano (NEJM)
Median = 32 days

Norwood BT vs Sano (NEJM)
Median = 9 days

Hybrid
Mean = 24 days

Hybrid
Mean = 4 days

Extubate POD
Inotropes POD
Discharge Day

With new thinking, more is possible.
Toronto study

• Retrospectively compared Hybrid to Norwood experience from 2004-2007
• 58 patients underwent palliation
  – Norwood (BT shunt); n = 39
  – Hybrid; n = 19

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Opinion

• I am not convinced that the Hybrid approach to HLHS is the best option

• The hybrid may be a good option for HLHS centers if applied correctly and if the proper infrastructure and team are assembled

• The hybrid is **very forgiving**
  – Most problems are tolerated well and are corrected without repeat CPB in cath lab
  – Interstage mortality is exceedingly rare (none at U of M) and ventricular function is preserved (in worst cases, ventricular function improves!)
Opinion

• There is a terrible deficit of data, particularly in regards to applying this approach to normal-risk patients.

• Is it time for a multi-center study?
Advice on Starting New Procedure

• Clearly identify what the problem is that you are trying to address
• Keep an open mind about alternative procedures and opposing viewpoints
• Learn how to perform, learn from other’s early mistakes, build a collaborative relationship with more experienced
• If at all possible, work to establish a consensus agreement to try new procedure, but include a minimum number to perform and collect data to compare to alternative treatments
Advice on Starting New Procedure

• Initial poor outcomes do not mean that it is not worth pursuing/modifying (initial good outcomes do not mean that you’ve found a better way)
• Failure of new procedure does not mean that you have failed
• Go out of your way to acknowledge and thank those involved with success (cath team, surgical team, cardiology and surgical colleagues, anesthesia, PICU, administration) – (1) it’s the right thing to do (2) you WILL have other new ideas that you want to try – the more that others feel that they are a part of something successful, the more that they will want to be a part of future successes (and more understanding of failures)
Advice on Starting Your Career

• You’re smart, you’re well trained, you’re eager
  – Go slow
  – Better to be 99-then-1 than it is to be 1-then-99
• Be in the upper-right corner
<table>
<thead>
<tr>
<th>Quality</th>
<th>Maintenance</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>High</td>
<td>High</td>
<td>Can be successful… but don’t always last</td>
</tr>
<tr>
<td>Low</td>
<td>Low</td>
<td>YOU!!!</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Low</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Slip under the radar?</td>
</tr>
</tbody>
</table>
Advice on Starting Your Career

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  - Go slow
  - Better to be 99-then-1 than 1-then-99
- Be in the upper-right corner
- Be a part of a team
- Track what you’re doing, identify the positives, promote what you and your team have accomplished
Advice on Starting Your Career

• Get involved
Pediatric/Congenital Interventional Cardiology Early-Career Society (PICES)

- Taskforce of Congenital Heart Council of SCAI
- International membership – Fellows committed to interventional catheterization and early career interventionalists (~10 years post-training)
Pediatric/Congenital Interventional Cardiology Early-Career Society (PICES)

- Networking
- Multi-center research projects
- Promote involvement in committees
- Education
  - Breakout sessions at PICS and SCAI
Welcome, colleagues, to the greatest job in the world!

We look forward to collaborating with you for the rest of our careers

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University of Minnesota Amplatz Children’s Hospital

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