THE EVOLUTION OF SURGICAL THERAPY FOR HYPOPLASTIC LEFT HEART SYNDROME: Current Techniques, Issues, Future Directions

THOMAS L. SPRAY, M.D.
Chief, Cardiothoracic Surgery
Alice Langdon Warner Endowed Chair
The Children’s Hospital Of Philadelphia
Professor of Surgery
The University of Pennsylvania

Definition

• Atresia or hypoplasia of multiple “left-sided” structures of the heart
  – mitral valve
  – left ventricle
  – aorta/ aortic arch
• Variants include other univentricular hearts with subaortic obstruction: typically with arch hypoplasia and coarctation
Epidemiology of HLHS

- CHD in 3 per 1,000 livebirths
- HLHS in ~1 per 5,000 livebirths
- HLHS ranks 5th-11th in frequency of CHD in multiple studies

Epidemiology of HLHS (cont’d)

- Consistent male preponderance
  - BWIS reported 58.6% males
- Equal distribution among races
- Non-cardiac anomalies occur in 11-37% patients with HLHS in clinical and autopsy studies.
Epidemiology of HLHS
BWIS 1997 (N = 4,390 cases)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No. (Rank)</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>1,411 (1)</td>
<td>32.1</td>
</tr>
<tr>
<td>PS</td>
<td>395 (2)</td>
<td>9.0</td>
</tr>
<tr>
<td>ASD (2&quot;)</td>
<td>340 (3)</td>
<td>7.7</td>
</tr>
<tr>
<td>AVC</td>
<td>326 (4)</td>
<td>7.4</td>
</tr>
<tr>
<td>TOF</td>
<td>297 (5)</td>
<td>6.8</td>
</tr>
<tr>
<td>TGA</td>
<td>208 (6)</td>
<td>4.7</td>
</tr>
<tr>
<td>Coarctation</td>
<td>203 (7)</td>
<td>4.6</td>
</tr>
<tr>
<td><strong>HLHS</strong></td>
<td><strong>167 (8)</strong></td>
<td><strong>3.8</strong></td>
</tr>
<tr>
<td>AS</td>
<td>128 (9)</td>
<td>2.9</td>
</tr>
<tr>
<td>Bicuspid AV</td>
<td>84 (13)</td>
<td>1.9</td>
</tr>
</tbody>
</table>

Single Ventricle - Prevalence/1000 Live Births

**Congenital Heart Defects**
- All Congenital Heart Defects 8.0
- Ventricular Septal Defects 1.3
- Tetralogy of Fallot 0.3
- **Hypoplastic Left Heart Syndrome** 0.2
- Tricuspid Atresia 0.07

**Other Anomalies**
- Down’s Syndrome 1.5
- Pyloric Stenosis 1.5
- Congenital Diaphragmatic Hernia 0.3
- Esophageal Atresia 0.2
- Hirschsprung’s Disease 0.2
Pathophysiology

- Complete mixing of systemic and pulmonary venous return
- Atrial septum and ductus arteriosus must be patent

Physiology of HLHS
Natural History

**HLHS**

**DEVELOPMENT OF FIRST STAGE PALLIATION (RECONSTRUCTION)**
**HLHS**

**PRINCIPLES OF PALLIATION**


---

**HLHS**

**PRINCIPLES OF PALLIATION**

“...we consider palliation in selected cases of hypoplasia of the left side of the heart to be indicated, for the current rapid advances in cardiac therapy hold the promise of the development of a “curative” procedure in the near future.”

HLHS

LITWIN PROCEDURE FOR IAA (1972)


HLHS

MOHRI (1979)
(4 cases, no survivors)

From: Mohri et al: JTCVS 1979;78:223-28
From: Doty et al JTCVS 1977;74:624-30

Early Procedures

Norwood et al
JTCVS 82:511, 1981

HLHS
BEHRENDT/ROCCHINI 1981

Early Procedures

Norwood et al
JTCVS 82:511, 1981

Stage 1 Procedure

Jacobs Mastery CT
Surgery 1998
Famous Baby With HLHS

October, 1984

Babies of the Century | FIGHTING TO SAVE THE YOUNGEST LIVES

Baby Fae

In his quest to find a cure for the thousands of infants born with incurable heart disease in North America each year, pediatric heart surgeon Leonard L. Bailey performed radical surgery on a baby he named Fae. In October 1984, he replaced the infant’s failing heart with that of a baboon’s at Loma Linda University Medical Center in California. Although baby Fae died 21 days later of organ failure, doctors discovered that she hadn’t rejected the heart as many had expected she would. That gave hope to the doctors and inspired continued research to find a cure for neonatal heart disease. The attention the media showered on baby Fae also helped increase awareness of the need for organ donation and better medical procedures for infants in general.

WHERE IS SHE NOW?

Baby Fae is gone, but the lessons she taught doctors live on. Her medical team purified what they learned from the procedure into more successful infant transplants later on. A year after Fae died, they successfully performed a human-to-human heart transplant on a 4-day-old infant; two years later, they operated on a 3-hour-old infant. Since then, the survival rate for infant heart transplants has been 80 percent, with nearly all the survivors still doing well.
Hypoplastic Left Heart Syndrome

STAGED RECONSTRUCTION

VS

TRANSPLANTATION

Hypoplastic Left Heart Syndrome

STAGED RECONSTRUCTION

AND PRIMARY

TRANSPLANTATION ARE

EFFECTIVE PALLIATIVE

INTERVENTIONS
Transplantation as Primary Treatment for HLHS

Razzouk et al.

Transplantation for HLHS
Survival for Transplantation and Staged Surgery

From: Jenkins et al, JACC 36:1178-1185, 2000
Hypoplastic Left Heart Syndrome

Survival (Low Risk Institutions)


Cardiac Tx vs. Staged Reconstruction
Transplantation for HLHS

Marginal survival benefit of transplantation exists largely because strategy used in only small proportion of patients at risk. Extension to larger proportion would decrease benefit by increasing mortality while waiting, and posttransplant mortality from PVD.

Transplantation for HLHS

The fundamental problem is that the supply of human donor hearts will never meet the demand.
Transplantation for HLHS

Is it justifiable to promote a strategy, even with marginal survival benefit, that cannot be applied to all patients at risk or even a small fraction of patients at risk?

Hypoplastic Left Heart Syndrome

STAGED RECONSTRUCTION DOES NOT PRECLUDE LATER FURTHER PALLIATION BY TRANSPLANTATION
RESULTS WITH THE NORWOOD OPERATION AT THE CHILDRENS HOSPITAL OF PHILADELPHIA

Stage I Norwood at The Children’s Hospital of Philadelphia
1984-2002 (n = 1061 Average 56/year*)

Currently stable at 50-60/year since 2002
Operative Mortality

- Weight > 2.5 kg and No Associated Anomaly: 12% Dead, 88% Alive
- Weight < 2.5 kg or an Associated Anomaly: 37% Dead, 63% Alive

Hospital mortality has been 8% for patients with no associated risk factors who underwent the Norwood procedure after 1/1/2000.

HLHS: Historical Cohort vs. Current Pts.

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stage I</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mortality</td>
<td>~40%</td>
<td>~10%</td>
</tr>
<tr>
<td>Core Cooling</td>
<td>11 ± 4 min</td>
<td>16 ± 2 min</td>
</tr>
<tr>
<td>DHCA</td>
<td>57 ± 13 min</td>
<td>37 ± 8 min</td>
</tr>
<tr>
<td>CPB</td>
<td>99 ± 27 min</td>
<td>82 ± 16 min</td>
</tr>
<tr>
<td><strong>Intermediate Staging</strong></td>
<td>Rare</td>
<td>~100%</td>
</tr>
<tr>
<td>Hemi Fontan/BDG Mortality</td>
<td>15%</td>
<td>1%</td>
</tr>
<tr>
<td><strong>Fontan</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mortality</td>
<td>~25%</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>Length of Stay</td>
<td>~1 month</td>
<td>~1 week</td>
</tr>
<tr>
<td>Effusions (&gt;14d)</td>
<td>common</td>
<td>rare</td>
</tr>
</tbody>
</table>
HLHS: Evolution in Management

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I Norwood</td>
<td>Superior CPC - “Stage II”</td>
<td>BDG/Hemi-Fontan</td>
<td>Hybrid RV-PA Shunt</td>
</tr>
<tr>
<td></td>
<td>Hepatic Vein Exclusion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fetal Dx</td>
<td>Improvements in CPB (MUF, ↓DHCA, ↑core cooling, etc.)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Post-Op Care</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

HLHS- Cumulative Survival - Staged Reconstruction

\[ N = 1431 \text{ Stage I Procedures} \]

- 2001-2002 “Standard” Risk Patients = 7.3%
- 2001-2002 All Patients = 15.2%
- 1995-98
- 1998-99
- 1992-94
- 1989-91
- 1984-88

Natural History
HLHS: Evolution at CHOP


Stage I Reconstruction: early approach
Focus: survival
Mortality > 50%
Increase PVR (iCO₂)
Dopamine, epinephrine
Paralysis
Avoid oxygen
Delayed Sternal Closure

Stage I Reconstruction: current approach
Focus: neurodevelopmental outcome
Mortality < 10%
Decrease SVR
Milrinone
No paralysis
Oxygen as needed
Chest closure
Early extubation

THE CHILDREN'S HOSPITAL OF PHILADELPHIA
RECENT TRENDS IN HLHS SURGERY

- Post-op ventricular support (NOMO)
- Fontan modifications (extracardiac, fenestration, off-pump, etc)
- Avoidance of DHCA
- Coarctation excision/native tissue reconstruction, aortic reimplantation, many technical surgical modifications, perioperative strategies and monitoring
- RV to PA shunt (Sano)
- Strategies for intact atrial septum
- Hybrid approach (PA banding/PDA stent)
- Out-of-hospital monitoring programs

CONTEMPORARY USE OF THE MBTS IN STAGE I NORWOOD

- Postop management straightforward – “overcirculation” a myth related to inadequate systemic output by increased systemic resistance (Milrinone)
- Tailoring of patch and experience in shunt tailoring decreases early distortion/intervention
- Lower diastolic pressure, but higher systolic pressure – unknown effect on coronary perfusion (resolves in days)
- Better PA growth; PA reconstruction easier at Stage II
- ? Benefit of anticoagulation on shunt patency
RV-PA SHUNT -CONCERNS

- Early stenosis common at RV origin – larger shunt size requires larger ventriculotomy
- PA growth not consistent – branch stenosis common
- Dissection more difficult at Stage II
- Early postop stability similar to MBTS
- ? Better tolerance of increased PVR
- RV incision may be associated with increased fibrosis and abnormalities of RV function
  - ? long-term effect

ISSUES

- Despite Markedly Improved early Survival With Norwood, Mortality Still High (Up to 15-20%) In Some Subgroups
- Groups With Higher Risk (LBW, Genetic Syndromes, Anatomic Features Such As AA/MS, Coronary Fistulae, RV Dysfunction, TR) Varied And Hard To Define
  - Mechanism Of Increased Risk
- Non-Cardiac Morbidity/Mortality Still A Problem (Sepsis, Necrotizing Enterocolitis, Neuro Issues)
- Impact Of Newer Monitoring, Followup Approaches On Morbidity/Mortality Still Not Clear
- Ability To Target Surgical Approach To Risk Factor Analysis A Goal For Improvement In Outcomes
SVR TRIAL: Comparison of BT Shunt and RVPA Shunt

From: Ohye, RG et al. NEJM 2010;362:1980-92
SVR TRIAL: Comparison of BT Shunt and RVPA Shunt

From: Ohye, RG et al. NEJM 2010;362:1980-92

From: Tweddell, JS, et al. JTCVS 2012;1-8
SVR: SURVIVAL BY PRESENCE OF OBSTRUCTED PV

From: Tweddell, JS, et al. JTCVS 2012;1-8

---

SVR: IMPACT OF GENETIC SYNDROME

From: Tweddell, JS, et al. JTCVS 2012;1-8
SVR: AS/MS LOW-RISK IMPACT OF SHUNT TYPE, SES, AO DIAMETER

From: Tweddell, JS, et al. JTCVS 2012;1-8

SVR: RISK FACTORS FOR PERINATAL DEATH

From: Atz, AM, et al. JTCVS 2010;140;1245-50
SVR TRIAL RESULTS

• ECMO or CPR - Outcomes significantly worse if ECMO or CPR required …no difference in shunt type on outcome

• RV Function – RV function, cardiac and valvar dimensions and function and neoaortic flow patterns are similar for both shunt types

• Risk Factors for Hospital Mortality and Morbidity – Shunt type did not affect hospital Mortality or Morbidity. Patient factors, pre-op condition and lower center volume were risk factors

• Neurodevelopmental Outcome – Patient factors and morbidity were risk factors. Shunt type, perfusion strategy, HCT, pH strategy and anatomy not risk factors

SVR RISK FACTORS FOR MORTALITY

• Independent Risk Factors For Mortality Include: Lower SES, Genetic Syndrome, Lower GA, and Shunt Type.


• For 48%, Shunt Type Had No Impact
SVR INTERSTAGE MORTALITY

- Overall ISM 12% - 6% RV/PA; 18% MBTS
- ISM Independently Associated With Gestational Age <37 wks., Hispanic Ethnicity, AA/MA, Increased Number Of Postop Complications, Poverty Level, And Use Of MBTS In Patients With No Or Mild AVVR
- Majority Of Deaths Occur In Hospital At Median of 1.6 mo., and are due to cardiac causes

The existing data suggest that there are clinical situations in which short and intermediate term mortality after stage 1 reconstruction is:

better with the RV-PA shunt,
better with the MBTS,
and, importantly, not different between shunt types.

There is insufficient evidence to determine which shunt type is associated with better long-term transplant-free survival.

In order to optimize survival and functional status, it is likely we will need to individualize patient management, rather than simply always choosing one shunt type.
HLHS with intact atrial septum

Before stent

Highly Restrictive Atrial Septum
“There is no branch of the modern scientific care of sick children that has not been carefully considered in the formulation of the future plans, and indeed it may here be suggested that this institution might well consider the possibility of a maternity department as a necessary part of the hospital. Among the class from which we draw our patients prenatal work is of vital importance, and so closely allied with pediatrics that the line of demarcation between them is exceedingly narrow.”

Edward S. Sayres, President, Board of Managers, 1914

New Facilities: Offering Comprehensive Services Through A Special Delivery Unit at CHOP

• 8 bed delivery unit, specifically designed for delivery of newborns with congenital anomalies
• Obstetricians and pediatric subspecialists all in one site offering state-of-the-art care to mothers and infants
• Unique opportunity for patient care across traditional disciplines
• Unique opportunity for research
Hospital Expansion – West Tower

2008

OR, Cath Labs
SDU
CICU
Highly Restrictive Atrial Septum

After stent

“IMPACT” Procedure = IMmediate Postpartum Access to Cardiac Therapy

<table>
<thead>
<tr>
<th>Indication for IMPACT Procedure</th>
<th>N</th>
<th>Alive</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLHS with intact atrial septum</td>
<td>10</td>
<td>6</td>
</tr>
<tr>
<td>Single ventricle, heterotaxy syndrome with CHB</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Maternal lupus mediated CHB</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>HLHS with total anomalous pulmonary veins</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>HLHS with severe ventricular dysfunction</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Giant PAVM</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>TOTAL</td>
<td>16</td>
<td>9 (56%)</td>
</tr>
</tbody>
</table>
NORWOOD STAGE I: What I Think Is Important

- **AA/MA** Coronary Fistulae: Not Important
- **AA/MS** Hypertensive LV: Not Important
- **AA/MS** Coronary Fistulae: Not Important
- **AS/MA**: Not Important
- **AS/MS** Lower Risk?: Not Important
- **PV Obstruction** Important
- **Severe AVVR** Important
- **Preop Shock** ? Important
- **VLBW (<2 Kg.)** Important
- **Older Age At Surgery** Important
- **Associated Congenital Anomalies** Important
**NORWOOD STAGE I**

**BYPASS STRATEGY**

- DHCA
- Antegrade Perfusion
- Alpha vs. pH Stat

**NORWOOD STAGE I**

**TECHNICAL ASPECTS**

- Small Shunt
- Coarctectomy
- Patch Aortic Arch
- Reimplantation Ao to PA
- Tailoring Aortic Patch
- Cut-Back PA For Ao Anastomosis
- MBTS
- RV-PA Shunt

In High-Volume Centers ?AA
### NORWOOD STAGE I

#### PERIOPERATIVE STRATEGIES
- Phenoxybenzamine
- Aprotinin
- Hct >25%
- Open Sternum
- NOMO
- Milrinone
- NIRS Monitoring

- Not important
- Not Important
- ? Important
- Not Important
- Not Important
- Important
- ? Not Important

#### POSTOPERATIVE STRATEGIES
- Paralysis/Sedation
- Low FiO2
- CO2 Administration
- Lactate Monitoring
- SVO2 Monitoring
- Peritoneal Dialysis
- Heparin

- Not Important
- Not Important
- Not Important
- Important (Trend)
- Not Important
- Not Important
- Not Important
WHAT I THINK IS IMPORTANT

- PV Obstruction
- Severe AVVR
- Preop Shock (+/-)
- VLBW
- Older Age At Surgery
- Associated Congenital Anomalies
- Coarctectomy (Selected)
- Tailoring of Aortic Patch
- Milrinone

A Contemporary Comparison of the Impact of Shunt Type in Hypoplastic Left Heart Syndrome on the Hemodynamics and Outcome at the Stage 2 Reconstruction

JA Ballweg MD, TE Dominguez MD, C Ravishankar MD, J Kreutzer MD, BS Marino MD, GL Bird MD, PJ Gruber MD PhD, G Wernovsky MD, JW Gaynor MD, SC Nicolson MD, TL Spray MD, S Tabbutt MD PhD

The Children’s Hospital of Philadelphia
and the University of Pennsylvania School of Medicine
Prior to Stage 2 Reconstruction (S2R)

<table>
<thead>
<tr>
<th></th>
<th>mBTS (n = 81)</th>
<th>RV-PA (n = 46)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Body surface area (m²)</td>
<td>0.31 (.23-.46)</td>
<td>0.3 (.24-.44)</td>
<td>0.87</td>
</tr>
<tr>
<td>Medications prior to S2R</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Digoxin</td>
<td>33 (41%)</td>
<td>13 (28%)</td>
<td>0.15</td>
</tr>
<tr>
<td>ACE-I</td>
<td>35 (43%)</td>
<td>15 (33%)</td>
<td>0.14</td>
</tr>
<tr>
<td>Diuretic</td>
<td>59 (73%)</td>
<td>30 (65%)</td>
<td>0.19</td>
</tr>
<tr>
<td>Anticoagulant</td>
<td>74 (91%)</td>
<td>40 (87%)</td>
<td>0.32</td>
</tr>
<tr>
<td>NG/GT feedings</td>
<td>23 (28%)</td>
<td>11 (24%)</td>
<td>0.37</td>
</tr>
<tr>
<td>Hemoglobin (gm/dL)</td>
<td>14.8 (12-19)</td>
<td>15.7 (13-21)</td>
<td>0.01</td>
</tr>
<tr>
<td>Home oxygen prior to S2R</td>
<td>5 (6%)</td>
<td>9 (20%)</td>
<td>0.03</td>
</tr>
<tr>
<td>Age at S2R (days)</td>
<td>176 (80-318)</td>
<td>153 (108-340)</td>
<td>0.03</td>
</tr>
</tbody>
</table>

Pre Stage 2 Reconstruction
ECHO: *Qualitative Ventricular Systolic Function*

- **Percent**
  - **bts**: 12 % (none: 20%, mild: 6%, moderate: 2%, severe: 4%)
  - **rv-pa**: 31 % (none: 6%, mild: 9%, moderate: 15%, severe: 22%)

*p = 0.02*
## Pre Stage 2 Reconstruction Catheterization

<table>
<thead>
<tr>
<th></th>
<th>mBTS (n=78)</th>
<th>RV-PA (n=46)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Common atrial pressure</td>
<td>8 (3-15)</td>
<td>8 (4-14)</td>
<td>0.79</td>
</tr>
<tr>
<td>Pulmonary artery pressure (or PVWp)</td>
<td>15 (8-22)</td>
<td>14 (10-25)</td>
<td>0.34</td>
</tr>
<tr>
<td>SVC saturation (%)</td>
<td>50 (30-66)</td>
<td>47 (26-63)</td>
<td>0.13</td>
</tr>
<tr>
<td>Aortic SBP (mm Hg)</td>
<td>91 (68-135)</td>
<td>92 (56-132)</td>
<td>0.77</td>
</tr>
<tr>
<td>Aortic DBP (mm Hg)</td>
<td>40 (20-57)</td>
<td>48 (29-60)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Coronary perfusion pressure</td>
<td>31 (12-47)</td>
<td>39 (21-53)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Aortic saturation (%)</td>
<td>77 (57-89)</td>
<td>74 (58-85)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Qp:Qs</td>
<td>1.6 (0.47-4.6)</td>
<td>1.3 (1.19-3.6)</td>
<td>0.03</td>
</tr>
</tbody>
</table>

## Stage 2 Reconstruction Operative Data

<table>
<thead>
<tr>
<th></th>
<th>mBTS (n = 73)</th>
<th>RV-PA (n = 46)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>CPB (min)</td>
<td>60 (13-184)</td>
<td>66 (16-152)</td>
<td>0.29</td>
</tr>
<tr>
<td>Aortic cross-clamp (min)</td>
<td>24 (5-61)</td>
<td>30 (16-64)</td>
<td>0.09</td>
</tr>
<tr>
<td>DHCA (min)</td>
<td>24 (9-61)</td>
<td>28 (16-60)</td>
<td>0.15</td>
</tr>
<tr>
<td>Additional procedures</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bilateral BDG</td>
<td>13</td>
<td>7</td>
<td>0.92</td>
</tr>
<tr>
<td>Tricuspid Valvuloplasty</td>
<td>4</td>
<td>6</td>
<td>0.27</td>
</tr>
</tbody>
</table>
### Stage 2 Reconstruction Hospital Course

<table>
<thead>
<tr>
<th></th>
<th>mBTS (n = 73)</th>
<th>RV-PA (n = 46)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total days ventilated</td>
<td>0 (0-79)</td>
<td>0 (0-8)</td>
<td>0.71</td>
</tr>
<tr>
<td>Hospital length of stay</td>
<td>5 (2-100)</td>
<td>6 (2-60)</td>
<td>0.09</td>
</tr>
<tr>
<td>Chest tube placement*</td>
<td>18% (11/60)</td>
<td>20% (8/40)</td>
<td>0.99</td>
</tr>
<tr>
<td>Surgical survival</td>
<td>99% (72/73)</td>
<td>98% (45/46)</td>
<td>0.69</td>
</tr>
</tbody>
</table>

*including hospital readmissions for pleural effusions within 30 days of surgery*

### Center Fontan Volume

(Average/year = 43)
Fontan Type

ERA 1
n=227

ERA 2
n=195

ERA 3
n=349


Early Death
Takedown

p=<0.001

Mortality 1/92-12/09
n=771
SUMMARY

- Expeditious, technically accurate operation key to success
- Most “advances” are center-specific, and individually have had only minor, if any, demonstrable effect on outcome
- Most problems still shunt-related: Thrombosis with MBTS, Stenosis with RV-PA shunt
- Major risk factors common in all studies are non-surgical: VLBW, Shock, AVVR, Older age, Associated Genetic Anomalies, ?AA

Norwood Stage I
Summary-HLHS After 30 Years

• Short and mid-term *Survival* for children with HLHS-especially low-risk subgroup-now approaches that of other complex CHD

• Management strategies must now focus on minimizing long-term *Morbidity*
  – CNS
  – Arrhythmia
  – PLE
  – Ventricular Function

Summary

• Non-CPB initial palliation (PA banding and ductal stenting) may delay CPB surgery to later age when CNS morbidity less, and may be most applicable to the high-risk subgroups, but Comprehensive Stage II difficult and no improvement in survival

• Hybrid procedures may be useful as staging to Stage I Norwood or bridge to transplant
**Summary**

- Critical evaluation of modifications to the staged reconstruction or transplant pathways for HLHS will require multi-institutional, prospective studies to confirm the theoretical benefits on late morbidity and mortality

- Selective use of various modifications (RV-PA shunt, hybrid procedures, antegrade cerebral perfusion, etc.) based on individual age, anatomy, genetic makeup best chance of reducing residual mortality and morbidity

**Summary**

- Decreasing the residual mortality in HLHS will be exponentially more difficult in the future

- Most risk factors are unknown…only about 20% of variability in outcomes accounted for by definable risk factors

- As for other complex CHD, biggest problem may now be what to do with long-term survivors …are we creating a large population of poor Fontan patients with few therapeutic options?