Children with Single Ventricle Physiology: The Possibilities

William I. Douglas, M.D.

Pediatric Cardiovascular Surgery
Children’s Memorial Hermann Hospital
The University of Texas Health Science Center at Houston
(UTHealth) Medical School
Single Ventricle Physiology

Definition

• A cardiac defect in which there is only one functioning ventricle

• The single ventricle may be a morphological right or left ventricle, or indeterminate

• May be due to underdeveloped chamber, valve, or outflow tract
  – There may be two good-sized ventricles where the inflow and/or outflow tracts cannot be separated
Single Ventricle Physiology

Diagnoses

- Tricuspid atresia
- Pulmonary atresia with intact septum
- HLHS
- Unbalanced AVSD
- D-TGA w/ HV
- L-TGA w/ HV

- Heterotaxy
- Ebstein’s anomaly
- Double inlet LV
- DORV
- AVC with TGA
Normal Circulation

Qp

Qs

Lungs

Right

Left

Body
Single Ventricle Physiology

Possibilities at Birth

Principles of SVP

– Pulmonary Resistance < Systemic Resistance
– Ideal Qp:Qs ≈ 1
  • Pulmonary stenosis necessary to achieve ideal Qp:Qs
– There is no “balanced” systemic obstruction
– The pulmonary outflow and systemic outflow cannot both be obstructed
Single Ventricle Physiology
Possibilities at Birth

Principles of SVP

– Pulmonary Resistance < Systemic Resistance
– Ideal Qp:Qs ≈ 1
  • Pulmonary stenosis necessary to achieve ideal Qp:Qs
– There is no “balanced” systemic obstruction
– The pulmonary outflow and systemic outflow cannot both be obstructed
Single Ventricle Physiology

Possibilities at Birth

Principles of SVP

- Pulmonary Resistance < Systemic Resistance
- Ideal $Q_p:Q_s \approx 1$
  - Pulmonary stenosis necessary to achieve ideal $Q_p:Q_s$
- There is no “balanced” systemic obstruction
- The pulmonary outflow and systemic outflow cannot both be obstructed
Single Ventricle Physiology

Possibilities at Birth

Principles of SVP

- Pulmonary Resistance < Systemic Resistance
- Ideal $Q_p:Q_s \approx 1$
  - Pulmonary stenosis necessary to achieve ideal $Q_p:Q_s$
- There is no “balanced” systemic obstruction
- The pulmonary outflow and systemic outflow cannot both be obstructed
Single Ventricle Physiology
Possibilities at Birth

Principles of SVP

– Pulmonary Resistance < Systemic Resistance
– Ideal Qp:Qs ≈ 1
  • Pulmonary stenosis necessary to achieve ideal Qp:Qs
– There is no “balanced” systemic obstruction
– The pulmonary outflow and systemic outflow cannot both be obstructed
Single Ventricle Physiology
Possibilities at Birth

Principles of SVP

- Pulmonary Resistance < Systemic Resistance
- Ideal Qp:Qs ≈ 1
  - Pulmonary stenosis necessary to achieve ideal Qp:Qs
- There is no “balanced” systemic obstruction
- The pulmonary outflow and systemic outflow cannot both be obstructed
Single Ventricle Physiology

Possibilities at Birth

• Unobstructed systemic blood flow
  – Appropriately balanced Pulmonary stenosis
    • No Surgery
    – Insufficient PBF (Severe PS or atresia)
      • BT Shunt
    – Excessive PBF (unobstructed PA)
      • PA Band

• Obstructed systemic blood flow
  – Complex Neonatal Surgery (Norwood)
Single Ventricle Physiology
Possibilities at Birth

• Unobstructed systemic blood flow
• Appropriately balanced Pulmonary stenosis
  – No Surgery
Single Ventricle Physiology

Possibilities at Birth

• Unobstructed systemic blood flow
• Unobstructed pulmonary blood flow
  – Excessive PBF
Single Ventricle Physiology

Possibilities at Birth

• Unobstructed systemic blood flow
• Excessive PBF (unobstructed PA)
  – PA Band
Single Ventricle Physiology

Possibilities at Birth

- **Unobstructed systemic blood flow**
- **Insufficient PBF (Severe PS or atresia)**
Single Ventricle Physiology

Possibilities at Birth

• Unobstructed systemic blood flow
• Insufficient PBF (Severe PS or atresia)
  • Systemic-Pulmonary Artery Shunt Shunt

\[ Q_p \quad \text{and} \quad Q_s \]

Lungs

Single ventricle heart

Body
Single Ventricle Physiology

Possibilities at Birth

- Obstructed systemic blood flow
- Implies unobstructed flow to the lungs

Make $Q_s$ arrow smaller
Single Ventricle Physiology

Possibilities at Birth

- Obstructed systemic blood flow
- Complex Neonatal Surgery (DKS, Norwood)
Single Ventricle Physiology

Possibilities at Birth

- Obstructed systemic blood flow
- Complex Neonatal Surgery (DKS, Norwood)
Surgical Treatment

- Single ventricle physiology
- Unobstructed flow to the body
- Moderately obstructed blood flow to the body
- **No surgery as a newborn**
Bored Surgeon
Surgical Treatment

- Single ventricle physiology
- Unobstructed flow to the body
- Severely obstructed blood flow to the lungs
- Systemic-pulmonary artery shunt (BT shunt)
  - Usually performed without cardiopulmonary bypass

- **Operative mortality: ≈5%**
Blalock-Taussig Shunt

TRICUSPID ATRESIA
Modified Blalock-Taussig Shunt
Surgical Treatment

- Single ventricle physiology
- Unobstructed flow to the body
- Unobstructed flow to the lungs
- Surgery: Pulmonary artery band (PA Band)
  - Almost always without cardiopulmonary bypass
- Operative Mortality: \( \approx 5\% \)
Pulmonary Artery Band
Surgical Treatment

• Obstructed blood flow to the body
  – Blood flow to the lungs always unobstructed

• These are the complex procedures

• Re-route the pulmonary outflow to the aorta
  – Add back pulmonary blood flow (e.g., BT shunt)
  – Always require CPB; often circulatory arrest

• Norwood or Damus-Kaye-Stansel

• Operative mortality: ≈15%
Norwood Procedure
Single Ventricle Physiology

WHAT HAPPENS NEXT
Single Ventricle Physiology

SPA Shunt: Disadvantages

- Arterial blood not fully saturated
- Difficult to precisely control amount of pulmonary blood flow
- Inherent volume overload
- *Doomed for failure*
Single Ventricle Physiology

Solution: Fontan Procedure

- Total cavopulmonary anastomosis
- Allows the pulmonary and systemic circuits to be in series
- Patients are fully oxygenated
- Volume overload on the ventricle is removed
Superior Cavopulmonary Anastomosis
Staged Fontan
Single Ventricle Physiology

Staged Approach

• Stage I: neonatal physiology
  – Small percentage won’t need surgery
  – BT Shunt or PA Band if pulm blood flow << or >>
    • ≈5% operative risk
  – Norwood needed if aortic obstruction present
    • ≈15% risk procedures

• Stage II: Glenn (sup. cavopulmonary shunt)
  – Six months of age

• Stage III: total cavopulmonary anastomosis (Fontan)
  – 2-3 years of age
Children with Single Ventricle Physiology

OUTCOMES
Outcomes in Pediatric Cardiac Surgery

• Risk adjustment methods
• Basic outcome statistics
Risk Adjustment Methods

• Mortality to *hospital discharge* is the current metric for judging program outcomes
  – 30 day mortality not appropriate
  – 30 day mortality not benchmarked
• Raw mortality not sufficient as programs have different profiles of case complexity
Risk Adjustment Methods

• RACHS-1 is first method created
  – All subsequent methods relatively similar

• 79 identified procedures were divided into 5 groups (by experts)
  – Originally 6 groups (one group too small)
# RACHS-1

<table>
<thead>
<tr>
<th>Level 1 (Simple)</th>
<th>Level 2 (Straightforward)</th>
<th>Level 3 (Moderately Complex)</th>
<th>Level 4 (Complex)</th>
<th>Level 5-6</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA Ligation</td>
<td>VSD Closure</td>
<td>Complete AVC</td>
<td>ASO + VSD</td>
<td>Norwood</td>
</tr>
<tr>
<td>ASD closure</td>
<td>TOF Repair</td>
<td>Fontan</td>
<td>Truncus</td>
<td>Damus-Kaye-Stansel</td>
</tr>
<tr>
<td>CoA Repair (&gt;30 days)</td>
<td>Partial AV Canal</td>
<td>Arterial Switch</td>
<td>TAPVR (&lt;30 days)</td>
<td>Truncus-IAA (Level 5)</td>
</tr>
<tr>
<td>PAPVR</td>
<td>Glenn</td>
<td>Valve Replacements</td>
<td>Interrupted Aortic Arch</td>
<td></td>
</tr>
<tr>
<td>TAPVR (&gt;30 days)</td>
<td>BT Shunts and PA Bands</td>
<td>Hypoplastic Arch Repair</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Other Risk Adjustment Methods

• Aristotle
  – 130 operations each with its own score
  – Grouped into 4 Levels

• STS-EACTS
  – Very similar to RACHS-1, except data has redefined which operations are grouped

• State of Texas (AHRQ)
  – Privileged methodology
  – 6 risk levels (like RACHS-1) with some non-cardiac co-morbidities
Important Insight

- All primarily use PROCEDURE to stratify risk
- Few use any patient related variables
- None use any cardiac co-morbidities:
  - A 6-hour old child with a single ventricle and highly obstructed TAPVR undergoes emergent TAPVR repair
  - A 2 week-old with unobstructed TAPVR undergoes semi-elective repair
  - THEY CARRY THE SAME RISK PROFILE; THEY HAD THE SAME PROCEDURE
- STS Data is Voluntarily Submitted
Bottom Line

- Data is useful for screening programs which are outliers
- Wide variability in accuracy
- Not good for distinguishing between good programs
## Outcomes: Sources

<table>
<thead>
<tr>
<th>Source</th>
<th>Society of Thoracic Surgeons</th>
<th>STS-EACTS</th>
<th>Single Ventricle Trial</th>
</tr>
</thead>
<tbody>
<tr>
<td>Source</td>
<td>Comprehensive US Database</td>
<td>Combined US-European Database</td>
<td>Prospective trial with focused population (HLHS)</td>
</tr>
<tr>
<td>First Author</td>
<td>NA</td>
<td>O’Brien</td>
<td>Ohye</td>
</tr>
<tr>
<td>Note</td>
<td>• Complete and well analyzed</td>
<td>• Varied centers, so difficult to interpret results</td>
<td>• Prospectively collected data on a uniform lesion</td>
</tr>
<tr>
<td></td>
<td>• Not readily available</td>
<td>• Fairly complete list of mortality (by lesion) published</td>
<td>• Very well analyzed</td>
</tr>
<tr>
<td></td>
<td>• Data is voluntarily submitted</td>
<td></td>
<td>• Describes natural history of HLHS</td>
</tr>
<tr>
<td></td>
<td>• How most programs are judged</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
STS 2010

• 29,000 operations submitted
• 24,000 analyzed in 20,000 patients
• Discharge mortality: 3.2%
• Discharge mortality by age group:
  – Neonates (0-30 days): 9.1%
  – Infants (1 month – 1 year): 2.8%
  – Children (1-18 years): 1.1%
  – Adults (>18 years): 2.7%
STS 2010
Mortality for Key Procedures

• Norwood: 15%
  – Not just Classic HLHS
• TAPVR: 9.6%
  – Obstructed and unobstructed
• Interrupted Aortic Arch: 5.6%
• Systemic-Pulmonary Artery Shunt: 4.7%
• Arterial Switch: 2.0%
• Arterial Switch + VSD: 4.0%
• Arterial Switch + Arch Repair: 11.5%
STS 2010
Mortality for Key Procedures

• Complete AV Canal: 2.9%
• Tetralogy of Fallot: 0.9% (no RV-PA conduit)
• PA Band: 6.7%
• Ross: 1.1%
• Ross-Konno: 13.6%
Combined European + STS

- Overall: 4.3% (2002-2007)
- Coarctation repair: 2.5%
- Arterial switch: 4.8%
  - ASO + aortic arch repair: 11%
- Aortic arch repair: 7.9%
- TOF repair: 2%
- Complete AV Canal: 4.6%
- Truncus: 14.3%
- Norwood: 23.7%
Single Ventricle Trial (HLHS)

- Death or Transplantation (12 months): 31.3%
- Time to initial extubation: 5.25 days
- Average ICU stay: 14 days
- Incidence of ECMO: 10%
- Incidence of CPR: 16%
- Excludes patients with any major extracardiac abnormality affecting the likelihood of survival (?)
  - Cohort did include patients <2.5 kg
Summary

• Newborns with single ventricle physiology can be grouped into 4 treatment categories
• Simplifies initial explanation to families
• Pediatric cardiac surgery still carries a significant operative mortality
  – Neonatal pediatric cardiac surgery exceptionally high risk
  – Mortality for neonatal cardiac surgery: 9-10%