Single Ventricle Anatomy, Physiology, and Nomenclature
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Disclosures

- Medronic Inc., Data Safety and Monitoring Board, Harmony Transcatheter Pulmonary Valve
- Chair, STS Congenital Heart Surgery Database Taskforce
Single Ventricle Physiology

Diagram showing the differences between normal heart anatomy and single ventricle physiology. The diagram highlights the following structures:

- **AO**: Aorta
- **PA**: Pulmonary Artery
- **LA**: Left Atrium
- **LV**: Left Ventricle
- **RA**: Right Atrium
- **RV**: Right Ventricle
- **SV**: Single Ventricle

The left side of the diagram represents normal heart anatomy, while the right side illustrates single ventricle physiology.

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# Types of Single Ventricle

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>STS Diagnosis Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single ventricle, Double inlet single LV</td>
<td>0790</td>
</tr>
<tr>
<td>Single ventricle, Double inlet single RV</td>
<td>0800</td>
</tr>
<tr>
<td>Single ventricle, Mitral atresia (usually single RV)</td>
<td>0810</td>
</tr>
<tr>
<td>Single ventricle, Tricuspid atresia</td>
<td>0820</td>
</tr>
<tr>
<td>• Normally related great arteries</td>
<td></td>
</tr>
<tr>
<td>• Transposed great arteries</td>
<td></td>
</tr>
<tr>
<td>Single ventricle, Unbalanced AV canal</td>
<td>0830</td>
</tr>
<tr>
<td>Single ventricle, Heterotaxia syndrome</td>
<td>0840</td>
</tr>
<tr>
<td>Single ventricle, Other</td>
<td>0850</td>
</tr>
<tr>
<td>Single ventricle + TAPVC</td>
<td>0851</td>
</tr>
<tr>
<td>HLHS</td>
<td>0730</td>
</tr>
</tbody>
</table>
Single Ventricle: Natural History

Mortality: (all forms of untreated single ventricle) is 90% by 1 year of age

Other morbidities:
- Cyanosis- Reduced exercise capacity and polycythemia
- Stroke- “Paradoxical” embolus, in situ thrombosis
- Brain abscess
Double Inlet Single Ventricle
Mitral Atresia

Restrictive left atrial outflow

Ao
PDA
PA
LA
RA
rPFO
RV

Mitral atresia
Aortic atresia+
intact VS
Tricuspid Atresia with Normally Related Great Arteries
Tricuspid Atresia with Transposition of the Great Arteries
Single Ventricle with Heterotaxy
Normal Heart

Hypoplastic Left Heart Syndrome (AA/MA)
Single Ventricle Physiology

Normal

Single Ventricle
Single Ventricle: Physiologic Problems

• Cyanosis (reduced oxygen levels in arterial blood)
  • Pulmonary venous blood (red) mixes with Systemic venous blood (blue) and “mixed” (desaturated) blood is pumped to both lungs and body

• Congestive heart failure (excess pulmonary blood flow, potentially at expense of systemic blood flow)

• Pulmonary vascular obstructive disease
  • Excess pulmonary blood flow
  • Elevated pulmonary artery pressures
Single Ventricle- Surgical Treatment

• Ultimate goal is to separate systemic venous return (blue, deoxygenated blood) from pulmonary venous return (red, fully oxygenated blood) so that only blue blood goes to the lungs and only red blood goes to the body.

• Ultimate operation to achieve this separation is the Fontan procedure
  • Systemic venous (blue) blood passes through the lungs without a ventricle to overcome pulmonary vascular resistance
  • Requires low resistance for “passive” blood flow through the lungs
Single Ventricle- Initial Surgical Treatment

• Staged management required (elevated pulmonary vascular resistance in first months of life)

• **Goals**
  • Provide sufficient blood flow to lungs to allow viable oxygenation
  • Limit pulmonary blood flow and pressures to:
    • Prevent heart failure and inadequate blood flow to body
    • Prevent development of elevated resistance in pulmonary circulation
    • Repair obstructions to systemic circulation
Single Ventricle-Initial Surgical Treatment

• Increase pulmonary blood flow:
  • Systemic to Pulmonary artery shunts
    • Modified Blalock-Taussig shunt. 1590
    • Central shunt (from aorta) 1600
    • Central shunt (Mee shunt) 3130
    • Potts-Smith shunt 3230
    • Shunt-other 1610

• Limit pulmonary blood flow
  • Pulmonary Artery Band 1640
Modified Blalock Taussig Shunt
STS Procedure Code 1590

This procedure diverts blood from an aortic branch to the pulmonary artery, allowing blood to flow to the lungs to receive oxygen.
Pulmonary Artery Band
STS Procedure code 1640
HLHS: The Norwood Procedure

- Aorta
- Ligated PDA
- PA
- BT-shunt
- Neo-aorta
HLHS: Norwood Operation (STS procedure code 0870)
Blalock-Taussig shunt  Sano shunt
Single Ventricle 2\textsuperscript{nd} Stage Management

• Goals
  • Reduce volume load on single ventricle
  • Maintain “adequate” oxygenation

• Operations
  • Bidirectional Cavopulmonary Shunt (Bidirectional Glenn)
  • Ligate or maintain existing systemic to pulmonary artery shunt
Bidirectional Cavopulmonary Shunt
(Bidirectional Glenn)
STS Procedure codes 1670, 1690, 2130, 2330
Bidirectional Cavopulmonary Shunt (Hemi-Fontan)
STS procedure code 1700
Single Ventricle Management-Third Stage-Fontan

• **Goals of the operation**
  • Complete separation of the systemic venous blood (blue, deoxygenated) from the pulmonary venous blood (red, oxygenated)
  • Normal (or near normal) oxygen levels in blood reaching the body
    • Improved exercise tolerance
    • Reduce risks of right to left shunt (stroke, brain abscess)

• **Risks**
  • Short-term: Pleural effusion
  • Longer term: Protein losing enteropathy, liver injury/cirrhosis, arrhythmias
Surgical repair of tricuspid atresia has been carried out in three patients; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, 'ventrilocized', to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cavo-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.

Only palliative operations (systemic vein to pulmonary artery anastomosis; systemic artery to pulmonary artery anastomosis) have been performed in tricuspid atresia. Although these procedures are valuable, they result in only a partial clinical improvement, because they do not suppress the mixture of venous and oxygenated blood.

We have initiated a corrective procedure for tricuspid atresia, which completely suppresses blood mixing. The entire vena caval return undergoes arterIALIZATION in the lungs and only oxygenated blood comes back to the left heart. This procedure is not an anatomical correction, which would require the creation of a right ventricle, but a procedure of physiological pulmonary blood flow restoration, with suppression of right and

![Diagram](image.png)

**FIG. 1.** Case 2. Tricuspid atresia type II B. Drawing illustrates steps in surgical repair: (1) end-to-side anastomosis of distal end of right pulmonary artery to superior vena cava; (2) end-to-end anastomosis of right atrial appendage to proximal end of right pulmonary artery by means of an aortic valve homograft; (3) closure of atrial septal defect; (4) insertion of a pulmonary valve homograft into inferior vena cava; and (5) ligation of main pulmonary artery.
Fontan Operations
Extracardiac conduit: cavo-pulmonary connection
# Single Ventricle Management - Third Stage

<table>
<thead>
<tr>
<th>Fontan operations</th>
<th>STS procedure codes</th>
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<tbody>
<tr>
<td>Atrio pulmonary connection</td>
<td>0950</td>
</tr>
<tr>
<td>Atrio-ventricular connection (Bjork)</td>
<td>0960</td>
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<tr>
<td>Total cavopulmonary connection TCPC</td>
<td></td>
</tr>
<tr>
<td>Lateral tunnel, fenestrated</td>
<td>0970</td>
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<tr>
<td>Lateral tunnel, non-fenestrated</td>
<td>0980</td>
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<tr>
<td>External conduit, Fenestrated</td>
<td>1000</td>
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<tr>
<td>External conduit, Non-fenestrated</td>
<td>1010</td>
</tr>
<tr>
<td>Intra/extracardiac, Fenestrated</td>
<td>2780</td>
</tr>
<tr>
<td>Intra/extracardiac, Non-fenestrated</td>
<td>2790</td>
</tr>
<tr>
<td>External conduit, hepatic veins to PA, fenestrated</td>
<td>3310</td>
</tr>
<tr>
<td>External conduit, hepatic veins to PA, non-fenestrated</td>
<td>3320</td>
</tr>
<tr>
<td>Fontan revision or conversion</td>
<td>1025</td>
</tr>
<tr>
<td>Fontan + atrioventriculoplasty</td>
<td>2340</td>
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</table>
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Questions?