Single Ventricle/Hypoplastic Left Heart Syndrome and Its Variants: Present and Future Medical and Surgical Management

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Learning Objectives

At the end of this educational activity, participants should be able to

- Identify various forms of single ventricle anatomy
- Outline current medical and surgical management patterns and considerations in single ventricle patients to identify long-term limitations and complications
- List negative prognostic risk factors
- Recognize new treatment strategies and the prospects for the future
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Outline

• What is Hypoplastic Left Heart Syndrome?
• Other Single Ventricle Variants
• Current management strategy
• Outcomes
• Ventricular support
• Transplantation
• Strategy to rehabilitate left heart
Cardiac Anatomy 101

Hypoplastic Left Heart Syndrome

[Images of cardiac anatomy and ultrasound]
Small Left Heart

- HLHS
- Critical Aortic stenosis
- Congenital mitral stenosis
- Severely unbalanced AV Canal defect (dominant RV)
- Often have hypoplastic aorta and coarctation

Small Right Heart

- Tricuspid atresia (dominant LV)
- Pulmonary atresia with intact ventricular septum, RV dependant coronary circulation, and hypoplastic right ventricle (dominant LV)
- Severely unbalanced AV Canal defect (dominant LV)
Other Variants

- Double inlet ventricle (dominant LV or RV)
- Straddling AV valve
- L-loop transposition of the great arteries with pulmonary atresia and univentricular hypoplasia
- Double outlet right ventricle with mitral atresia.

Natural History

- Small left heart (HLHS)
  - Depends upon PDA for systemic blood flow
  - As Patent Ductus Arteriosus closes, low blood leads to
    - Renal Failure
    - Intestinal Ischemia
    - Acidosis
    - Mortality
- Small right heart (Pulmonary Atresia)
  - Depends upon PDA for pulmonary blood flow
  - As PDA closes, hypoxemia
- Prostaglandins to reopen PDA
Single Ventricle Palliation

- Goal is to eventually use single ventricle (right or left) to pump to body
- Allow passive drainage of systemic venous blood into pulmonary artery to provide oxygenation
- Resistance of flow through lungs determines timing of surgical approach
  - Very high after birth = Need high pressure to pump blood through lungs

HLHS and small left heart

- Neonate - Stage 1 (Norwood)
- Create unobstructed outflow from ventricle to aorta (through pulmonary valve)
- Augment the aorta
- Ensure coronary blood flow (aortopulmonary connection)
- Create source of pulmonary blood flow (arterial pressure)
  - Innominate artery
  - Ventricle
**Small Right Heart**

- Neonate - Blalock Taussig shunt to provide pulmonary blood flow
- Do not need aortopulmonary connection
- Shunt thrombosis is major risk
- Balanced circulation important to prevent over circulation or cyanosis
- Goal pulse oximetry sats 80%

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**Interstage monitoring**

- Risk of mortality (10%) in HLHS
- Risk of hypoxia with small right heart
- Home monitoring program improves outcomes
  - Home pulse oximetry
  - Daily weights
  - Regular contact with nurse practitioner
  - Early detection and intervention
Second Stage operation

- Bidirectional Glenn or Hemi Fontan
- 4 – 6 months of age
- Partially directs venous return to lungs
- IVC still drains to heart
- Oxygen levels = 80% still
- Less work load on heart
- Heart catheterization prior to procedure to ensure pulmonary arteries unobstructed

Final Stage

- Fontan Procedure directs IVC blood to pulmonary arteries
- Fenestration to decompress
- Sats 90% until fenestration closed
- Lateral Tunnel vs. Extracardiac
- Pleural effusions postoperatively
Single ventricle palliation - outcomes

- 10-20% mortality following stage 1 (Norwood)
- 50-70% 10 year survival
- 5% require cardiac transplantation
- Role for medical management
  - ACE inhibitors

Long term complications

- Protein losing enteropathy (PLE)
- Plastic bronchitis
- Arrhythmias
- Tricuspid / Mitral regurgitation
- Thrombosis in baffles
- Progressive ventricular dysfunction
- Cirrhosis
Ventricular Assist Device for Single V

- Single ventricle support
- Right heart support is technically difficult
- Currently as Bridge to Transplantation
- Successful in anecdotal cases

Ventricular Assist Devices
Selection of pump size

• Too small
  – May limit filling
  – Inadequate cardiac output
  – need to run very fast  *hemolysis

• Too big
  – Hypertension
  – Need to run slow - thrombus

Outcomes

• Patients with congenital heart disease (CHD) are known to have worse outcomes on VAD support than patients without CHD

Blume et al, 2006
Hertzer et al, 2010
Cardiac Transplantation

- Risk of allograft right heart failure
- Overall, transplant improves survival
- PRESEVED ventricular function is associated with poor outcome compared to impaired function

Growth Potential

- Observation – Children have enormous healing and growth capabilities
- Left Ventricle can grow too
- How to stimulate growth?

McElhenney et al. 2005 (CHB)
Staged LV recruitment

Initial Single Ventricle Palliation

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Maneuvers to rehabilitate LV

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Subsequent biventricular conversion

Operative Strategies

• Flow = Grow
• LV Rehabilitation
  – EFE resection
  – Accessory pulmonary blood flow
  – Restriction of ASD
  – Aortic valve repair
  – Mitral valve repair
• Biventricular conversion procedure
  – Ross procedure
  – Direct re-anastomosis
Atrial septal defect restriction

- ASD allows blood to flow to right heart
- Diverts away from left heart
- Restricting size of communication “forces” blood into LV
- Flow = grow
- Promotes growth of LV

Downside – If pressure builds up in left atrium, lungs can get damaged
- Particularly problematic in single ventricle patients
- Traditionally with single ventricle – HERESY to restrict atrial septum
- If left heart does not grow, then risk of lung injury
Left Heart size by stage

Effect of ASD restriction on growth
LV Recruitment Strategy

- Stage 1 / LH rehab
  - 34 Patients
- Biventricular conversion
  - 12 Patients
- Cardiac Transplantation
  - 1 patient

Future Directions

- Improvement in interstage care of single V patient
- Improve devices for Failing Fontan
- Refine indications for transplantation
- Increased Role for biventricular conversion
- Effect of LV recruitment upon single ventricle function
- Multi Site pacing for dyssynchrony
- Management of the Adult with single ventricle
Thank You!

Questions?

Please direct questions regarding the activity to OptumHealth Education at moreinfo@optumhealtheducation.com