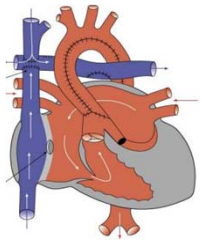
Single Ventricle/Hypoplastic Left Heart Syndrome and Its Variants: Present and Future Medical and Surgical Management



Ram Emani, MD
Boston Children's Hospital

Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

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

Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

2

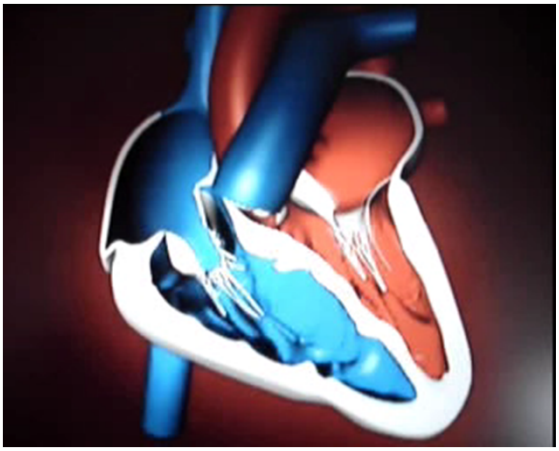
Disclosures

- **Faculty Disclosure**
  As a provider accredited by the ACCME, OptumHealth Education requires the disclosure of any relevant financial relationship a faculty member has with the manufacturer(s) of any product discussed in this educational presentation. The faculty reported the following:
  Dr Emani has nothing to disclose.
  Dr Emani does not intend to discuss unapproved/investigative use of commercial products/devices.
- **Staff Disclosures**
  In compliance with the ACCME's Standards for Commercial Support, employees of OptumHealth Education who have control over content of an activity are required to disclose their relevant financial relationships. No employee has a relevant financial relationship regarding this activity.

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

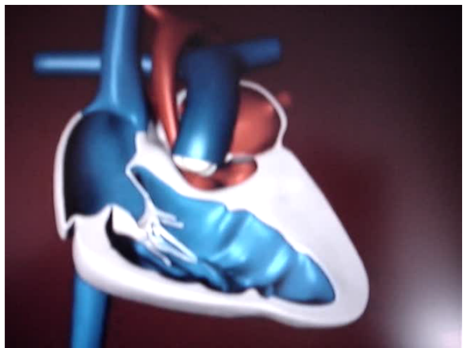


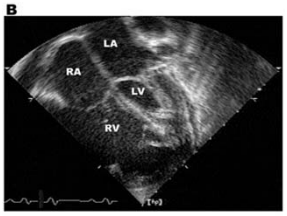
Boston Children's Hospital Heart Center

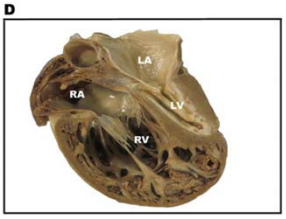
HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

5

Hypoplastic Left Heart Syndrome







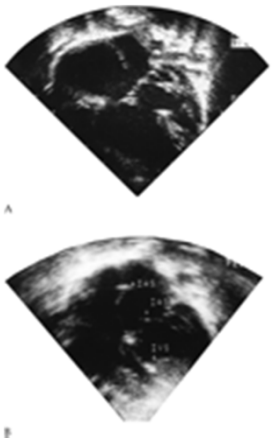
Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

6

Small Left Heart

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



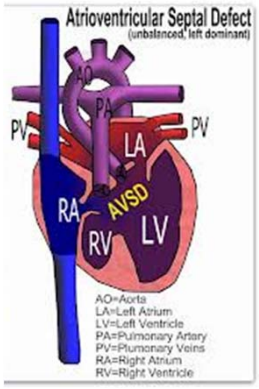
Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

7

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)



Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

8

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

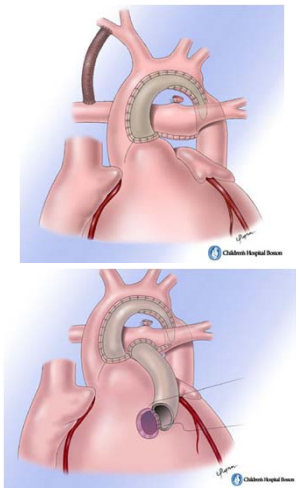
- Small left heart (HLHS)
  - Depends upon PDA for systemic blood flow
  - As Patent Ductus Arteriosus closes, low blood leads to
    - Renal Failure
    - Intestinal Ischemia
    - Acidosis
    - Mortality
- Small right heart (Pulmonary Atresia)
  - Depends upon PDA for pulmonary blood flow
  - As PDA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

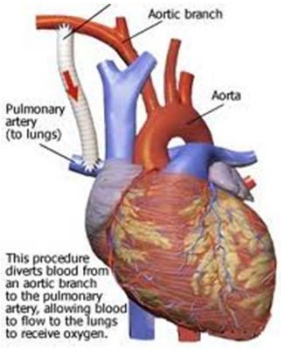
HLHS and small left heart

- 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



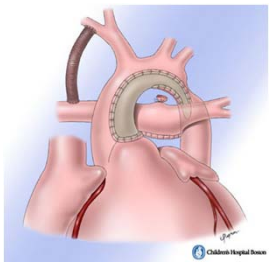
Small Right Heart

- 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%



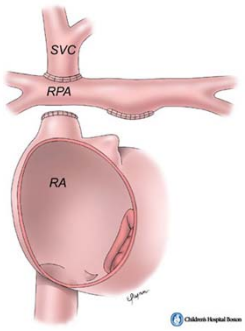
Interstage monitoring

- 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 practioner
  - Early detection and intervention



Second Stage operation

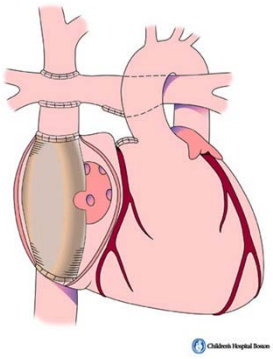
- Bidirectional Glenn or Hemi Fontan
- 4 – 6 months of age
- Partially directs venous return to lungs
- IVC still drains to heart
- Oxygen levels = 80% still
- Less work load on heart
- Heart catheterization prior to procedure to ensure pulmonary arteries unobstructed



Bidirectional Glenn

Final Stage

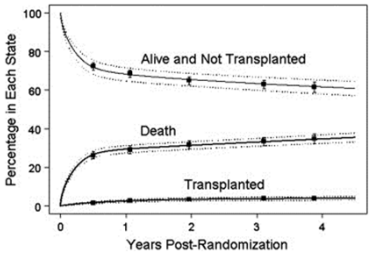
- Fontan Procedure directs IVC blood to pulmonary arteries
- Fenestration to decompress
- Sats 90% until fenestration closed
- Lateral Tunnel vs. Extracardiac
- Pleural effusions postoperatively



Fontan

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

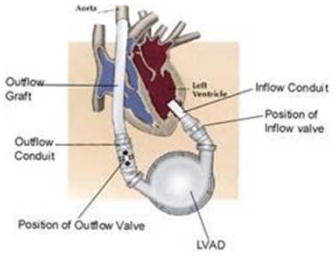


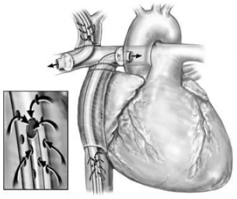
Long term complications

- 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



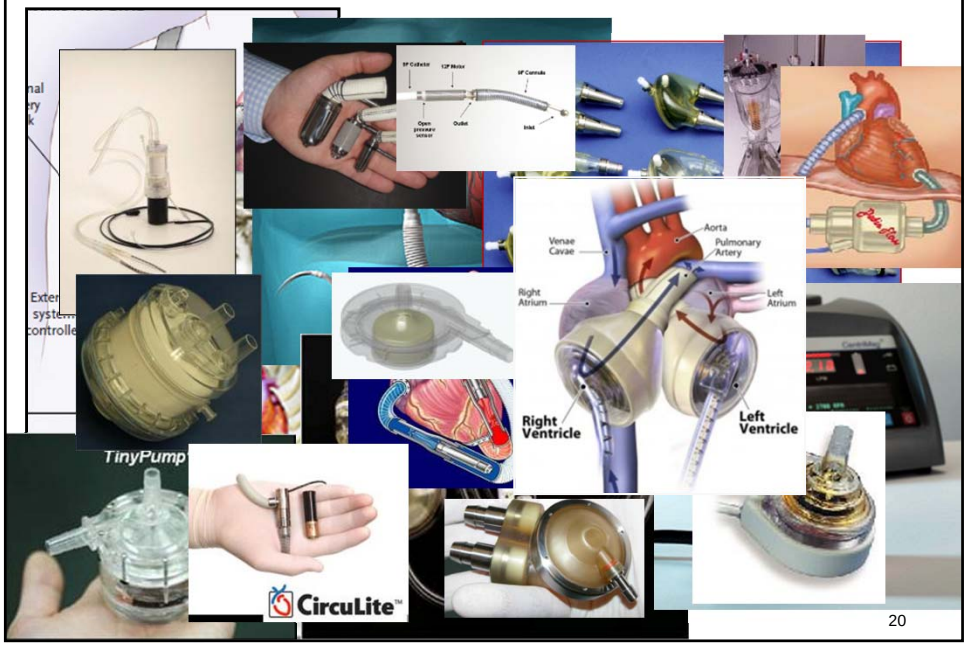


Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

19

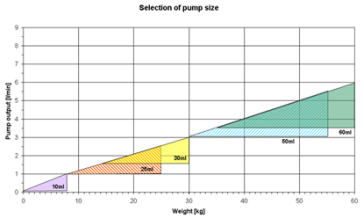
Ventricular Assist Devices



20

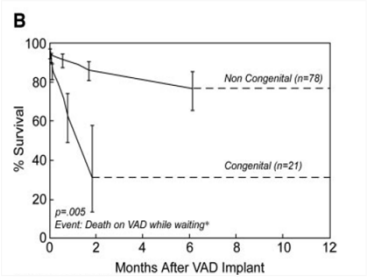
Selection of pump size

- Too small
  - May limit filling
  - Inadequate cardiac output
  - need to run very fast *hemolysis
- Too big
  - Hypertension
  - Need to run slow - thrombus

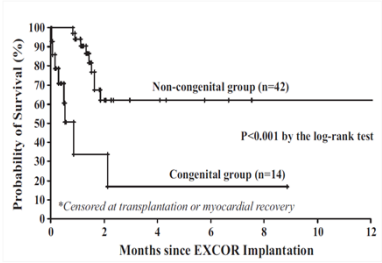


Outcomes

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



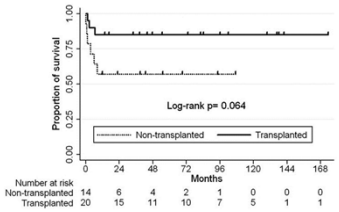
Blume et al, 2006

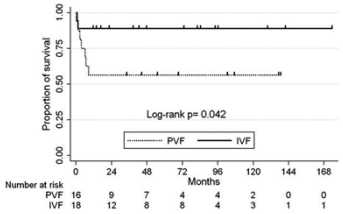


Hertzer et al, 2010

Cardiac Transplantation

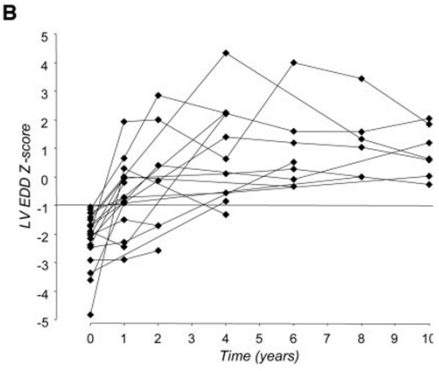
- Risk of allograft right heart failure
- Overall, transplant improves survival
- PRESEVED ventricular function is associated with poor outcome compared to impaired function





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

↓

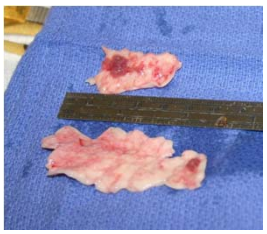
Maneuvers to rehabilitate LV

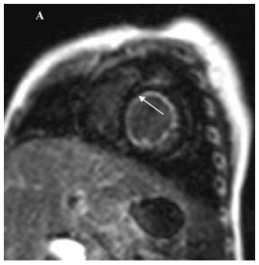
↓

Subsequent biventricular conversion

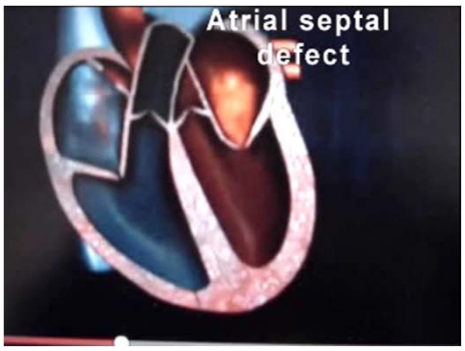
Operative Strategies

- 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-anastamosis





Atrial septal defect restriction

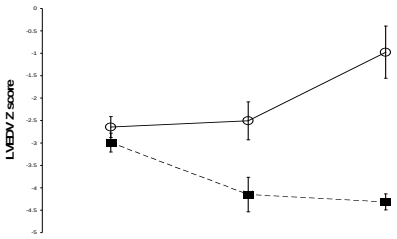


- ASD allows blood to flow to right heart
- Diverts away from left heart
- Restricting size of communication “forces” blood into LV
- Flow = grow
- Promotes growth of LV

Atrial septal defect restriction

- 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



Staged LV Recruitment

Control / Traditional Single v palliation

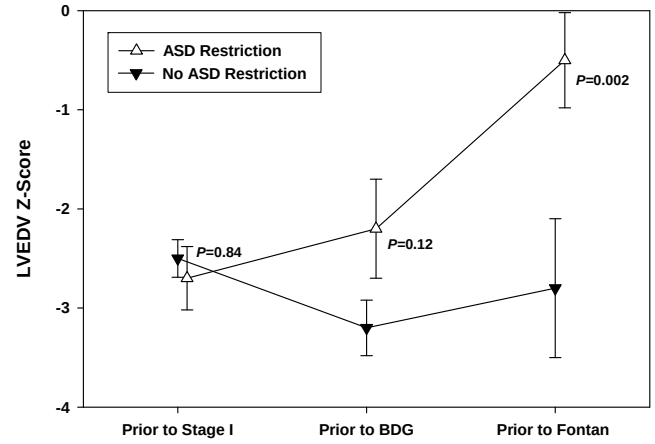
Stage 1 Stage 2 Prior to BiV conversion

Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

29

Effect of ASD restriction on growth



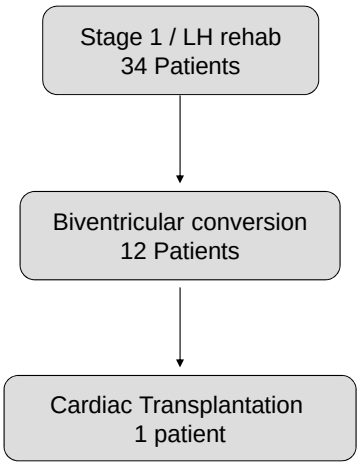
ASD Restriction: N =	17	13	11
No ASD Restriction: N =	13	8	8

Boston Children's Hospital Heart Center

HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

30

LV Recruitment Strategy



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