Single Ventricle Heart Disease

Glenn and Fontan Surgeries

Presented by: Leyla Allum, ARNP & Cassie Horner, ARNP

Date: 9/5/19
Disclosure statement

• We do not have any conflict of interest, nor will we be discussing any off-label product use.

• This class has no commercial support or sponsorship, nor is it co-sponsored.
Objectives

• Describe the anatomy and physiology of single ventricle heart disease (focusing on Hypoplastic Left Heart Syndrome)
• Describe the second and third stages of palliation (Glenn & Fontan) after the Norwood procedure
• Distinguish common postoperative challenges for Glenn & Fontan
• Discuss inpatient and outpatient nursing care post Glenn & Fontan, including patient education
• Summarize standards of care for patients post Fontan
• Discuss long term outcomes post Fontan
Variety of CHD that results in single ventricle physiology
- Tricuspid atresia
- Unbalanced atrioventricular canal
- Double inlet left ventricle
- Pulmonary atresia/intact ventricular septum
- Hypoplastic left heart syndrome

Initial palliations and outcomes may differ based on anatomy

Single Ventricle Clinic at SCH est in 2014

Clausen, H. Hypoplastic left heart syndrome. Paediatrics and Child Health, 2015-01-01, Volume 25, Issue 1, Pages 18-22, Copyright © 2014 Elsevier Ltd.
Most common form of SV heart disease is HLHS
- Aortic and mitral atresia: mitral valve, LV, aortic valve and ascending aorta are hypoplastic
- Male predominance (1.5:1)

Birth incidence: 4-8/10,000
- ~1000 children per year born in US with HLHS
- ~1000 children with other forms of SV CHD

Represents 8% of all CHD

Wide variation in prenatal detection rates and termination rates (in WA state approaches 50-60%)
- Prenatal counseling: 60% transplant free survival to 5 years of age

SV heart disease can’t be repaired, but is palliated through staged operations

Twin on the left has HLHS
Clausen, H. Hypoplastic left heart syndrome. Paediatrics and Child Health, 2015-01-01, Volume 25, Issue 1, Pages 18-22, Copyright © 2014 Elsevier Ltd
Various surgical palliations after birth depending on anatomy-about 60% receive a Norwood operation

- Pulmonary trunk is divided from the pulmonary circulation and anastomosed to the ascending aorta, aortic arch is augmented, PDA is ligated, atrial septum is enlarged

- Pulmonary blood flow via a shunt: BT shunt from the innominate artery or proximal right subclavian (pictured) or Sano shunt from the RV

- Goals: Provide unobstructed pulmonary venous return, unobstructed aorta, blood flow to coronary arteries and the right amount of blood flow to the lungs

Clausen, H. Hypoplastic left heart syndrome. Paediatrics and Child Health, 2015-01-01, Volume 25, Issue 1, Pages 18-22, Copyright © 2014 Elsevier Ltd
Stage II: Glenn Procedure

- Occurs at 4-6 months of age
- Pre-Glenn cath to assess PVR
- SVC directly sewn to pulmonary arteries
- Most desaturated blood from brain enters the lungs
- Less volume returning to the ventricle-improves long term systolic function and valve function
- Saturations typically 80-85%
- Improved clinical stability

Clausen, H. Hypoplastic left heart syndrome. Paediatrics and Child Health, 2015-01-01, Volume 25, Issue 1, Pages 18-22, Copyright © 2014 Elsevier Ltd
Pain/headaches from elevated CVP
May be complicated by narrowing at the anastomotic site
Data from NPC-QIC: about 1/3 of patients experience a postoperative complication
  - Unplanned cath/re-operation (15%)
  - Cardiac arrest (3%)
  - Neurologic deficit (seizure) (2%)
  - Feeding/GI issues (15%)
Typical LOS is ~1 week
Very low mortality risk (less than 2%)
Inpatient Nursing Care After Glenn

- Pain management is important
  - Usually managed with Tylenol, ketorolac and narcotics
- Elevated HOB or holding for comfort
- Nutrition continues to be a major focus, but feeding intolerance often improves after Glenn
- Goal sats around 80% but may initially be lower
- General nursing care: infection, CV assessment
• Non local families can usually return home ~1 month post Glenn
• Decreased home monitoring
• Families should expect to see cardiologist regularly in the first few months after operation, visits are then spaced to 6-12 month intervals
• Continued focus on nutrition and development
  – UW cardiac neurodevelopmental clinic at 1 year of age (est 2015)
• Some require return to cardiac cath lab
Stage III: Fontan Procedure

- Occurs between 3-5 years of age depending on cyanosis/activity tolerance
- Pre-Fontan cath to assess PVR
- IVC sewn to pulmonary arteries
- Several technical options but most commonly an extracardiac conduit of synthetic material
- Typically includes a small fenestration to allow a right to left shunt to serve as a “pop off”
- Improves saturations to the 90s (with fenestration >85%)
- Low operative mortality for Fontan <2%
- Typical LOS ~2 weeks (range 1-6 weeks)

Clausen, H. Hypoplastic left heart syndrome. Paediatrics and Child Health, 2015-01-01, Volume 25, Issue 1, Pages 18-22, Copyright © 2014 Elsevier Ltd
Common Postop Issues After Fontan

- Prolonged drainage of pleural chest tubes (low albumin, loss of clotting factors)
- Pleural effusions may recur after CTs are removed
- Chylous drainage (caused by lymphatic leak)
- May need cardiac cath to assess for anatomic or hemodynamic issues if drainage is prolonged

Imazio, M. Brucato, A. & Adler, Y. Is possible to prevent the Post-Pericardiotomy Syndrome? International Journal of Cardiology, 2012-08-09, Volume 159, Issue 1, Pages 1-4, Copyright © 2012 Elsevier Ireland Ltd
Common Postop Issues After Fontan

• Aggressive use of diuretic therapy can lead to electrolyte abnormalities
• Developmental challenges
• Risk for thromboembolism in fenestration
• Risk for sinus node dysfunction
Inpatient Nursing Care After Fontan

- Ambulation and pulmonary toilet
- Developmental support (Child Life, consistent nursing care)
- Difficulty with medication administration
- Chest tube management
- Early postop pain control
- Frequent labs and CXR
- Oxygen and/or Sildenafil to decrease PVR
- Infrequently: NPO/TPN, albumin replacement, IVIG
Some institutions have postop Fontan protocols and a few single site retrospective studies have shown decreased LOS and chest tube duration.

Protocols usually include:
- aggressive diuretic regimen
- fluid restriction to 80% maintenance
- nasal cannula oxygen until chest tube removal
- low-fat diet (30% calories from fat)
- remove CTs at <2 mL/kg in 24 hours
Patient Education after Fontan

• Need for continued diuretic therapy
• Lifelong anti-platelet or anticoagulation therapy
• Risk for recurrent pleural effusions
• Many need to continue low fat diet (6 week from chylous effusion resolution)
• Developmental regression (importance of re-establishing home routines, night terrors not uncommon, difficulty with medical appointments)
Outcomes after Fontan

- Fontan circulation characterized by: low cardiac output & elevated central venous pressure
- Limited ability to increase cardiac output during exercise
- Most children have very little overt limitations in childhood, no heart failure symptoms, majority with normal ventricular function, generally no activity restrictions
- Typically seen by cardiology every 6-12 months
- Patients who have undergone current SV staged palliations are now about 15-20 years old
- Fontans from the 70s: 15-20 year postop survival 60-85%
- Current Fontans (HLHS): 10 year postop survival 72-85%
Long term complications after Fontan

- Neurodevelopmental issues
- Plastic Bronchitis (high lymphatic pressure downstream of Fontan circulation)
- Fontan associated liver disease, Cirrhosis
- Poor exercise intolerance
- Protein losing enteropathy (low albumin leads to progressive volume shifts, ascites)
- Delayed puberty
- Poor growth
- Ventricular failure, valve insufficiency
- Arrhythmias
- Thromboembolism
2018 AHA/ACC guidelines for adults with Fontan palliation

Strong recommendations:

• New atrial tachyarrhythmia requires prompt anticoagulation and EP consultation
• Annual evaluations with echo or cardiac MR
• Cardiac caths should be performed prior to initial Fontan surgery or revision
• New or worsening atrial tach should prompt search for hemodynamic abnormalities via imaging and/or cath
• Anticoagulation with Vitamin K antagonist is recommended for known or suspected thrombus, thromboembolic events or prior atrial arrhythmia without contraindications to anticoagulation
2018 AHA/ACC guidelines for adults with Fontan palliation

Moderate recommendations:

• Encourage a regular exercise program
• Imaging of the liver and laboratory evaluation of liver function for fibrosis, cirrhosis and/or hepatocellular carcinoma are reasonable
• Reasonable to perform liver and renal function testing annually
• Evaluation for transplant is reasonable for signs and symptoms of PLE
• Ablation can be useful in atrial tachyarrhythmia
• Fontan revision surgery including arrhythmia surgery is reasonable for recurrent atrial tachyarrhythmias refractory to medications and ablation
• Pulmonary vasoactive medications can be beneficial to improve exercise capacity
<table>
<thead>
<tr>
<th></th>
<th>1 year post Fontan</th>
<th>4 years post Fontan</th>
<th>7 years post Fontan</th>
<th>10 years post Fontan</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Dexa Scan</td>
<td>Dexa Scan</td>
<td>Dexa Scan</td>
<td>Cath</td>
</tr>
<tr>
<td></td>
<td>Labs</td>
<td>Liver US or Elastograph</td>
<td>Labs</td>
<td>CPET</td>
</tr>
<tr>
<td></td>
<td>Echo</td>
<td>Echo</td>
<td>Echo</td>
<td>Cardiac MRI</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>Exercise</td>
<td>Exercise</td>
<td>Liver MRI or Elastograph</td>
</tr>
<tr>
<td></td>
<td>Nutrition</td>
<td>Nutrition</td>
<td>Nutrition</td>
<td>Labs</td>
</tr>
<tr>
<td></td>
<td>Neuropsych</td>
<td>Neuropsych</td>
<td>Neuropsych</td>
<td>Echo</td>
</tr>
<tr>
<td></td>
<td>Make-a-wish</td>
<td></td>
<td></td>
<td>Exercise</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Holter</td>
<td>Nutrition</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Neuropsych</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>ACHD</td>
</tr>
</tbody>
</table>
Summary

• Only just now seeing long term outcomes from medical and surgical management of HLHS
• Greater emphasis on long term post Fontan monitoring
• Increased focus on developmental implications, social/emotional well being
• Surgical and medical therapies still being developed
Questions?

catherine.horner@seattlechildrens.org

leyla.allum@seattlechildrens.org