

## Hypoplastic Left Heart Syndrome Guideline

What the Nurse Caring for the Patient with CHD Needs to Know

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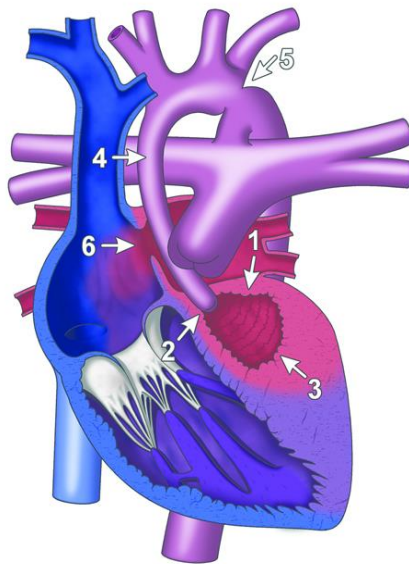
University of Michigan, CS Mott Children's Hospital

### Embryology

- Formation of Atrioventricular (AV) cardiac valves
  - Days 34 to 36
  - Formed from endocardial cushions
- Formation of the ventricles: Days 22-35
- Spectrum of underdevelopment of left sided heart structures

### Anatomy

- Hypoplasia or agenesis of the tricuspid valve (TV) or mitral valve (MV)  
(As indicated by #1 in Illustration)
- Hypoplasia or agenesis of the aortic valve (AV) or pulmonary valve (PV)  
(As indicated by #2 in Illustration)
- Hypoplasia of the left ventricle (LV)  
(As indicated by #3 in Illustration)
- Hypoplasia of the ascending aorta with/without coarctation or interrupted aortic arch  
(As indicated by numbers 4 & 5 in Illustration)
- Possible thick or sclerotic endocardium
- Atrial septal defect  
(As indicated by #6 in Illustration)
- Patent ductus arteriosus (PDA)



Hypoplastic Left Heart Syndrome

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## **Physiology**

- Hypoplastic left heart syndrome (HLHS)
  - Blood enters the left atrium (LA)
    - Cannot exit due to hypoplasia/agenesis of the MV
    - Crosses the atrial defect into the right atrium (RA)
  - Blood then crosses the tricuspid valve (TV) and enters the right ventricle (RV)
  - Blood enters the pulmonary artery (PA) through the pulmonary valve (PV)
  - Blood then enters the lungs through the PA and shunts right to left through the PDA to the systemic circulation
  - Balance of pulmonary blood flow
    - Dependent on the respective pulmonary and systemic resistance
    - Size of the PDA
  - Coronary blood flow provided by retrograde filling of the aorta through the PDA
- Hypoplastic left heart syndrome with restrictive ASD
  - Blood enters the left atrium
    - Cannot exit due to hypoplasia/agenesis of MV
    - Attempts to cross the atrial defect into RA
    - Due to the atrial constriction
      - Blood then is unable to exit LA
      - Backs up into pulmonary veins and pulmonary vasculature.
  - Results in pulmonary edema, pulmonary hypertension, desaturation and low cardiac output.

## **Interventions**

### **Medical**

- Prostaglandin (PGE<sub>1</sub>) to maintain ductal patency and systemic perfusion
  - Infant with postnatal diagnosis
    - Becomes symptomatic as ductus closes and systemic perfusion increasingly compromised
    - Early symptoms
      - Irritable
      - Not easily consoled
      - Feeding difficulties
      - Tachypnea
      - Color changes
        - Pale
        - Dusky
    - Late symptoms
      - Lethargy
      - Increasing respiratory distress
      - Cyanosis
    - Leads to profound low cardiac output and pending cardiovascular collapse

- Presents to PCP or emergency department in cardiac failure/shock
- Management
  - Initiate PGE<sub>1</sub> at 0.1mcg/kg/minute at time of presentation of postnatal diagnosis, even if diagnosis uncertain
  - Must also rule out other etiologies of cardiovascular compromise, specifically sepsis
    - Initiate course of antibiotics until cultures negative
    - Investigation warranted even when diagnosis confirmed
- At birth with prenatal diagnosis
  - Initiated at initial resuscitation generally at lower dose typically 0.05mcg/kg/minute
  - Reduce PGE dose
    - Once ductal patency established
    - To minimum required to maintain ductal patency
    - To minimize side effects
- Manage side effects of PGE<sub>1</sub>
  - Observation for PGE side effects including apnea, fever, seizures
  - Initiation of theophylline derivative for prevention of PGE associated apnea
- Additional neonatal non-interventional management (See Pediatric Neonatal Guidelines for Neonatal Care)
  - Indicated by specific patient status and degree of low cardiac output
    - Intubation and vasoactive support
    - Use of oxygen, nitrogen and afterload reduction
      - To balance pulmonary and systemic blood flow
      - Treat hypoxemia preoperatively
  - Continuous evaluation of end organ function as reflection of adequacy of cardiac output
    - Renal function and use of diuretic as indicated to maintain euvolemic state
    - Cranial ultrasound and observation for altered neurologic status especially for infants requiring postnatal resuscitation
    - Observation for bowel ischemia, hematochezia or pneumatosis on x-ray, due to preoperative shock or low diastolic pressure and hypoxemia
  - Nutritional support initiated at 24 hours of age
    - Either enteral or parenteral
    - Depends on institutional preference

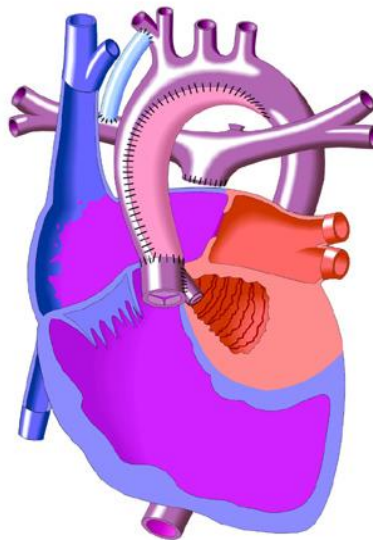
### **Interventional Cardiac Catheterization**

- Neonatal intervention [See Pediatric Neonatal Guidelines on Neonatal Care and Venous-Arterial Extracorporeal Membrane Oxygenation (VA ECMO)]
  - Balloon atrial septostomy (BAS)
    - Open atrial defect
    - Decompress left atrium and pulmonary veins
    - Indicated for pre- and postnatal diagnosis of restrictive ASD
  - Restrictive ASD - Emergent procedures

- Indicated for severe low cardiac output and hypoxemia at birth
- ECMO as exit procedure for cath lab interventions
- Initiation of ECMO in delivery room
- Emergent hybrid procedure
- Fetal interventions
  - Prenatal intervention
    - Balloon atrial septostomy - attempts to relieve restrictive atrial septum
    - Aortic balloon dilation - optimize growth of LV and aortic valve (AV)
      - Used in situations with marginal hypoplasia of LV and AV
      - May prevent single ventricle reconstruction
  - Generally 24 weeks gestation

### **Surgical interventions**

- Stage I reconstruction: Norwood/Sano or Norwood/systemic to pulmonary shunt
  - Aortic arch reconstruction for repair of arch obstruction and establishment of systemic outflow and unobstructed coronary blood flow
  - Creation of nonrestrictive ASD
  - Creation of controlled source of pulmonary blood flow
    - Classic Blalock-Taussig shunt: end to side anastomosis subclavian to right or left branch pulmonary artery shunt (rarely performed)
    - Modified Blalock-Taussig shunt: Gortex<sup>®</sup> interposition graft between subclavian or innominate artery and right or left branch pulmonary artery (See illustration below)



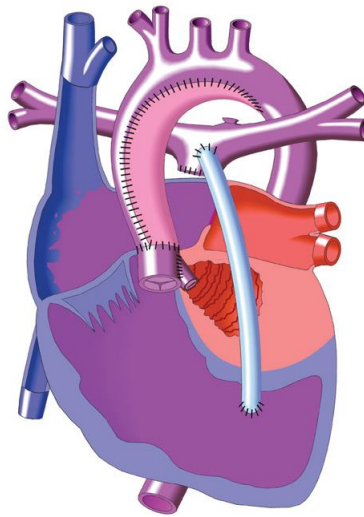
Norwood with systemic - pulmonary shunt

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- Central shunt: Gortex<sup>®</sup> interposition graft between aorta and main pulmonary artery

- Sano: right ventricular to pulmonary artery non-valved Gortex® tube  
(See illustration below)

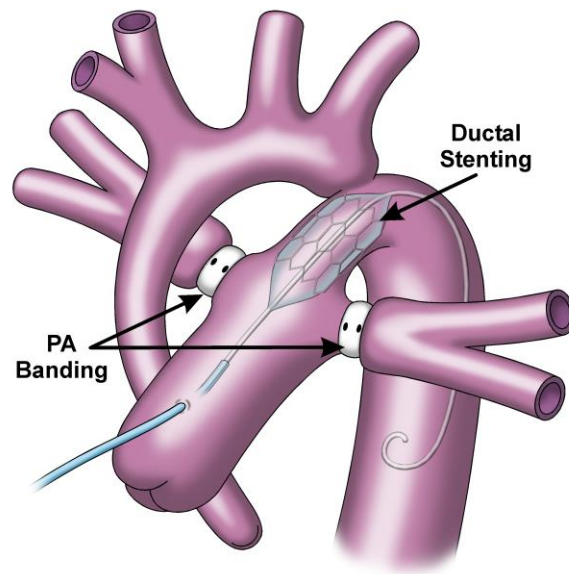


**Sano Modification of  
Norwood Procedure**

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- Complications of Norwood
  - Pulmonary overcirculation and systemic undercirculation resulting in systemic hypoperfusion and low cardiac output
  - Pulmonary undercirculation resulting in hypoxemia and good systemic perfusion
  - Residual arch obstruction
  - Restrictive ASD
  - Later complications:
    - Thromboembolic events of the systemic to pulmonary shunt or Sano
    - Congestive heart failure (CHF)
    - Poor growth of central pulmonary arteries
- Stage I reconstruction: Hybrid
  - PDA stent (See Illustration below)
    - Assure secure source of systemic and coronary artery blood flow
  - Pulmonary artery bands (See Illustration below)
    - Regulate pulmonary blood flow to prevent pulmonary overcirculation
    - Balance of pulmonary and systemic blood flow
  - Atrial septostomy/septectomy
    - Unobstructed outflow of blood from lungs, pulmonary veins and left atrium
    - Unobstructed mixing of oxygenated and unoxygenated blood at atrial level



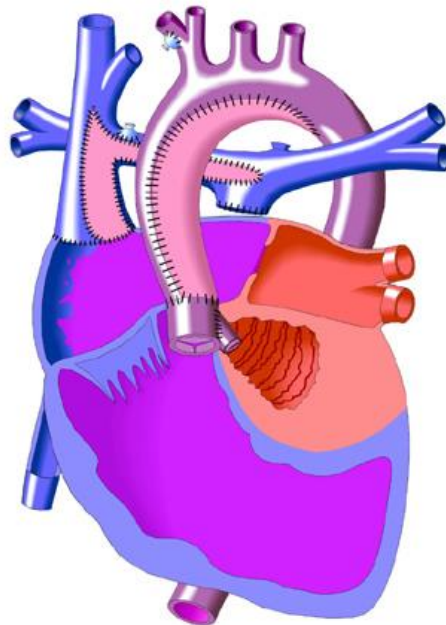
Hybrid procedure

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- Complications of Hybrid
  - Perforation
  - Embolism
  - Congestive heart failure
  - Restriction of ASD
- Discharge after Stage I reconstruction (See Pediatric Neonatal Guidelines on Interstage Monitoring for Infants with Hypoplastic Left Heart Syndrome)
  - Includes both Norwood Procedures with BT shunt and Sano Shunt and Hybrid Procedure
  - Infants at high risk for:
    - Growth failure
    - Developmental delays
    - Hypoxia
    - Interstage morbidity and mortality
  - Discharge planning extremely important
    - Direct communication with community practitioners
      - Scheduled follow up with both community practitioner and cardiologist/cardiologist nurse practitioner/interstage coordinator
      - Ongoing monitoring and communication
    - Parent education
      - Use “teach back” methods
      - Include:

- Signs and symptoms of respiratory distress and increasing cardiovascular compromise
- Medication administration and monitoring
- Nutrition with supplements to formula/breast milk and caloric and weight gain monitoring
- Developmental assessment and interventions
- Structured program available for interstage monitoring (See Pediatric Neonatal Guidelines for Interstage Monitoring for Hypoplastic Left Heart Syndrome)
- Stage II reconstruction: Hemi-Fontan or Bidirectional Glenn
  - Connection of superior vena cava (SVC) to pulmonary arteries
  - Provide controlled pulmonary blood flow based on pulmonary resistance and ventricular function/end diastolic pressure
    - Increases SVC pressure
    - Decreases volume load to ventricle and increases diastolic blood pressure and coronary perfusion
  - Ligation of Sano or systemic to pulmonary shunt
  - Complications of Hemi-Fontan and Bidirectional Glenn
    - SVC syndrome
    - Pleural effusion
    - Hypoxemia



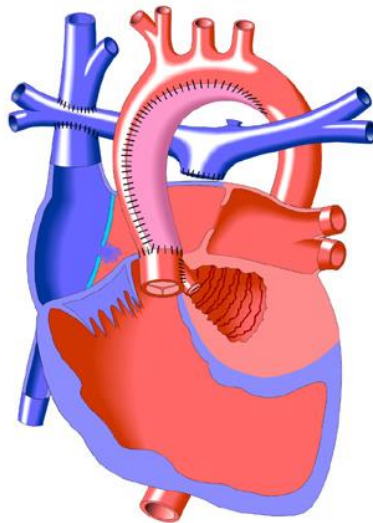
Glenn Shunt

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- Stage III reconstruction: Modified Fontan
  - Physiologic correction for single ventricle lesion

- Pulmonary blood flow achieved through SVC/inferior vena cava (IVC) /PA to LA pressure gradient (transpulmonary gradient)
- Despite surgical technique achieve systemic flow (IVC/SVC) directly into PA's bypassing ventricular contribution
- Fenestration utilized to assist hemodynamic adjustment to acutely elevated venous pressures
- Surgical options for Fontan operation
  - Lateral tunnel: Gortex<sup>®</sup> graft placed inside RA to direct IVC flow through RA/SVC junction and into main pulmonary artery (MPA)
  - Extracardiac: Gortex<sup>®</sup> or Dacron circumferential tube graft from IVC to MPA
  - Direct RA to PA anastomosis: connection of right atrial appendage to PA (not preformed currently)



Lateral Tunnel Fontan

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- Long term complications with interventions: Fontan (See Pediatric Neonatal Guidelines for Postoperative Management, Open Sternotomy – Delayed Chest Closure, Hemodynamic Monitoring, Neonatal Care, Infection Prevention, Nutrition, ECMO)
  - Arrhythmia: ablation, pacemaker, ICD, medications, conversion to lateral tunnel or extracardiac Fontan connection with plication of RA ( See to Adult and Pediatric Neonatal Problem Guidelines on Arrhythmia Management for further discussion and management)
  - Ventricular dysfunction: rhythm and transplant (See Adult Problem Guidelines on Systemic Ventricular Failure for further discussion and management.)



- Atrioventricular valve regurgitation (AVVR): Valve repair/replacement
- Fontan pathway obstruction: reoperation for relief of conduit stenosis
- Protein Losing Enteropathy (PLE): ( See Pediatric Neonatal Guidelines on Nutrition for further discussion and management of PLE)
  - Loss of protein into abdomen
  - Diarrhea
  - Edema
  - Etiology/definitive treatment unknown
  - Treatment may include conversion Fontan, creation of ASD, or transplant
- Plastic bronchitis: casts that occlude bronchus, no treatment
- Thromboembolic events: anticoagulation varies from center to center but minimally lifelong aspirin (See Adult and Pediatric Neonatal Guidelines for Anticoagulation Management)

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