

# Hypoplastic Left Heart Syndrome

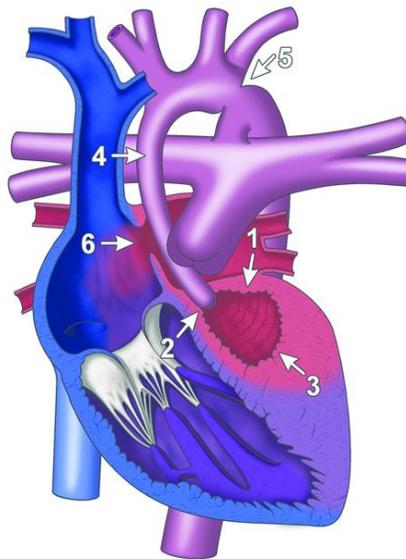
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## I. Embryology

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

## II. Anatomy

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



Hypoplastic Left Heart Syndrome

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### III. Physiology

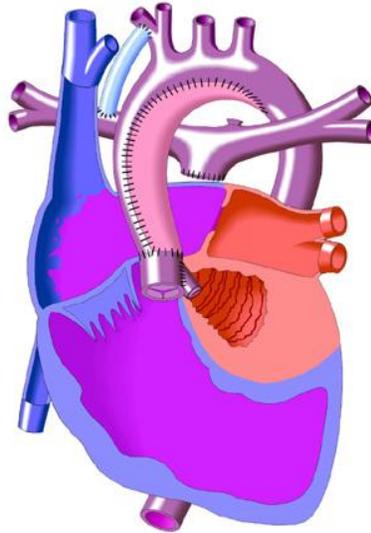
- A. Hypoplastic left heart syndrome (HLHS)
  1. Blood enters the left atrium and cannot exit due to hypoplasia/agenesis of the MV and crosses the atrial defect into the right atrium.
  2. Blood then crosses the tricuspid valve and enters the right ventricle. Blood enters the pulmonary artery through the pulmonary valve.
  3. Blood then enters the lungs through the pulmonary artery and shunts right to left through the PDA to the systemic circulation.
  4. The balance of pulmonary blood flow is dependent on the respective pulmonary and systemic resistance and the size of the PDA.
  5. Coronary blood flow is provided by retrograde filling of the aorta through the PDA.
- B. Hypoplastic left heart syndrome with restrictive ASD
  1. Blood enters the left atrium and cannot exit due to hypoplasia/agenesis of the MV and attempts to cross the atrial defect into the right atrium. Due to the atrial constriction blood then is unable to exit the left atrium and backs up into the pulmonary veins and lungs.
  2. This results in pulmonary edema, pulmonary hypertension, desaturation and low cardiac output.

### IV. Medical interventions

- A. Prostaglandin to maintain ductal patency and systemic perfusion
- B. Balloon atrial septostomy to open atrial defect and decompress left atrium and pulmonary veins

### V. Surgical interventions

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

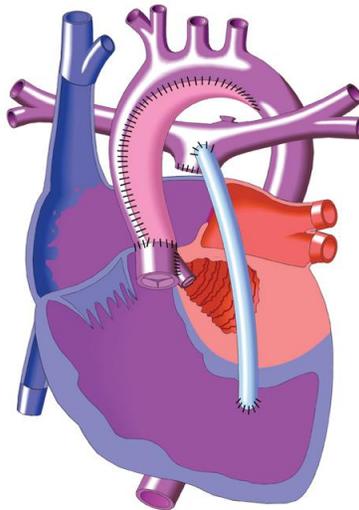


Norwood with systemic - pulmonary shunt

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- c. Central shunt: Gortex interposition graft between aorta and main pulmonary artery
- d. Sano: right ventricular to pulmonary artery non-valved Gortex tube  
(Refer to illustration below)



**Sano Modification of  
Norwood Procedure**

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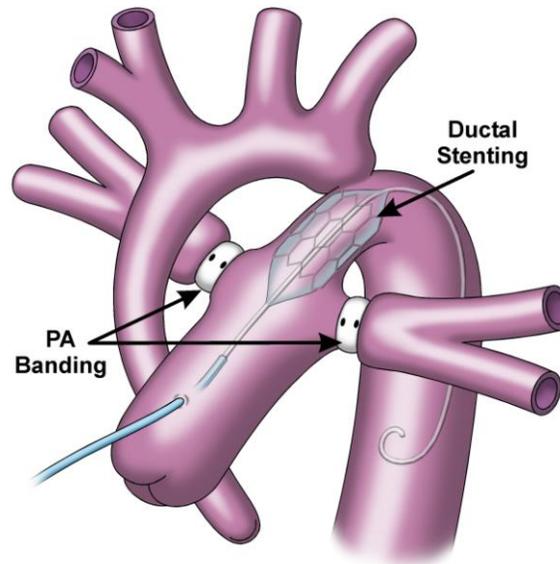
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- 3. Complications of Norwood
  - a. Pulmonary overcirculation and systemic undercirculation resulting in systemic hypoperfusion and low cardiac output

- b. Pulmonary undercirculation resulting in hypoxemia and good systemic perfusion
- c. Residual arch obstruction
- d. Restrictive ASD
- e. Later complications: thromboembolic events of the systemic to pulmonary shunt or Sano, congestive heart failure (CHF), poor growth of central pulmonary arteries

**B. Stage I reconstruction: Hybrid**

1. PDA stent (See Illustration below)
  - a. Assure secure source of systemic and coronary artery blood flow
2. Pulmonary artery bands (See Illustration below)
  - a. Regulate pulmonary blood flow to prevent pulmonary overcirculation
  - b. Balance of pulmonary and systemic blood flow
3. Atrial septostomy/septectomy
  - a. Unobstructed outflow of blood from lungs, pulmonary veins and left atrium
  - b. Unobstructed mixing of oxygenated and unoxygenated blood at atrial level



Hybrid procedure

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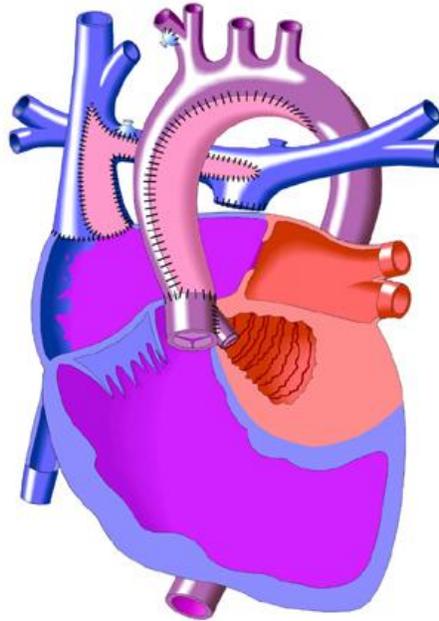
**4. Complications of Hybrid**

- a. Perforation
- b. Embolism
- c. Congestive heart failure
- d. Restriction of ASD

**C. Stage II reconstruction: Hemifontan or Bidirectional Glenn**

1. Connection of superior vena cava (SVC) to pulmonary arteries
2. Provide controlled pulmonary blood flow based on pulmonary resistance and ventricular function/end diastolic pressure

- a. Increases SVC pressure
  - b. Decreases volume load to ventricle and increases diastolic blood pressure and coronary perfusion
3. Ligation of Sano or systemic to pulmonary shunt
4. Complications of Hemifontan and Bidirectional Glenn
- a. SVC syndrome
  - b. Pleural effusion
  - c. Hypoxemia

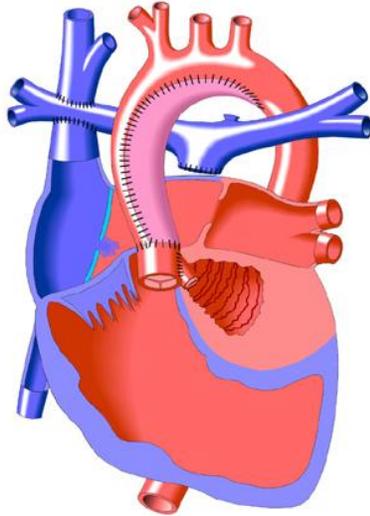


Glenn Shunt

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- D. Stage III reconstruction: Modified Fontan
- 1. Physiologic correction for single ventricle lesion
  - 2. Pulmonary blood flow achieved through SVC/inferior vena cava (IVC) /PA to left atrium (LA) pressure gradient (transpulmonary gradient)
  - 3. Despite surgical technique achieve systemic flow (IVC/SVC) directly into PA's bypassing ventricular contribution
  - 4. Fenestration utilized to assist hemodynamic adjustment to acutely elevated venous pressures
  - 5. Surgical options for Fontan operation
    - a. Lateral tunnel: Gortex graft placed inside RA to direct IVC flow through RA/SVC junction and into main pulmonary artery (MPA)
    - b. Extracardiac: Gortex or Dacron circumferential tube graft from IVC to MPA
    - c. Direct RA to PA anastomosis: connection of right atrial appendage to PA (not preformed currently)



#### Lateral Tunnel Fontan

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6. Long term complications with interventions: Fontan
  - a. Arrhythmia: ablation, pacemaker, ICD, medications, conversion to lateral tunnel or extracardiac Fontan connection with plication of RA ( Refer to Problem Section on Arrhythmias for further discussion and management)
  - b. Ventricular dysfunction: rhythm and transplant (Refer to Problem Section on Systemic Ventricular Failure for further discussion and management.)
  - c. Atrioventricular valve regurgitation (AVVR): Valve repair/replacement
  - d. Fontan pathway obstruction: reoperation for relief of conduit stenosis
  - e. Protein Losing Enteropathy (PLE):
    - (1) Loss of protein into abdomen
    - (2) Diarrhea
    - (3) Edema
    - (4) Etiology/definitive treatment unknown
    - (5) Treatment may include conversion Fontan, creation of ASD, or transplant
  - f. Plastic bronchitis: casts that occlude bronchus, no treatment
  - g. Thromboembolic events: anticoagulation varies from center to center but minimally lifelong aspirin

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