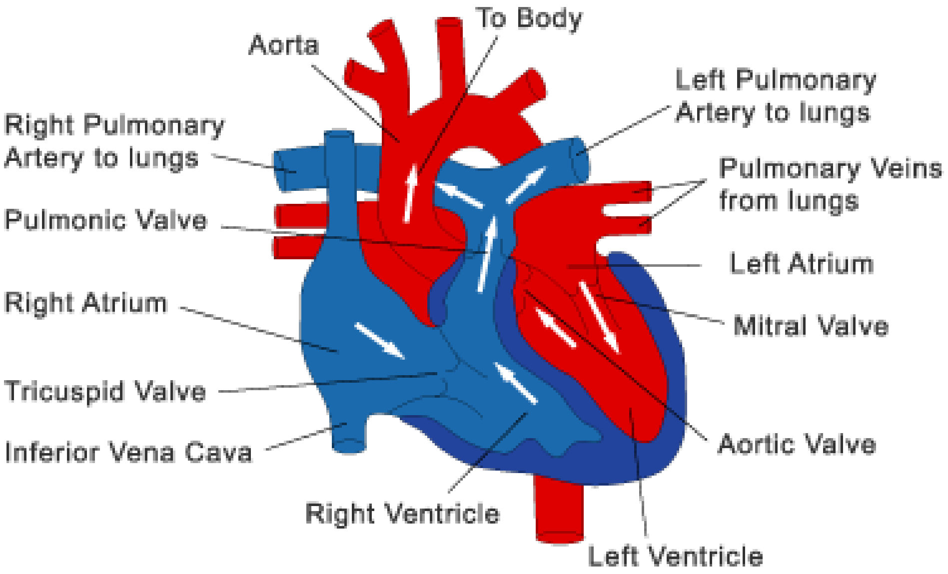
Single Ventricle Cardiac Anomalies

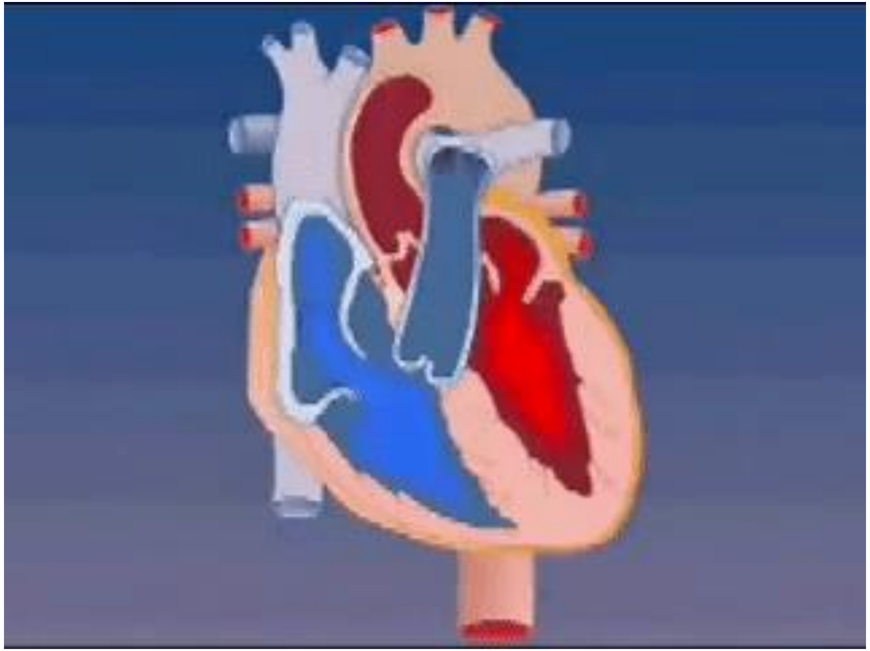
Hypoplastic Left Heart Syndrome & More

Duke Children's

Steve Hepditch, Clinical Educator, Duke Hospital

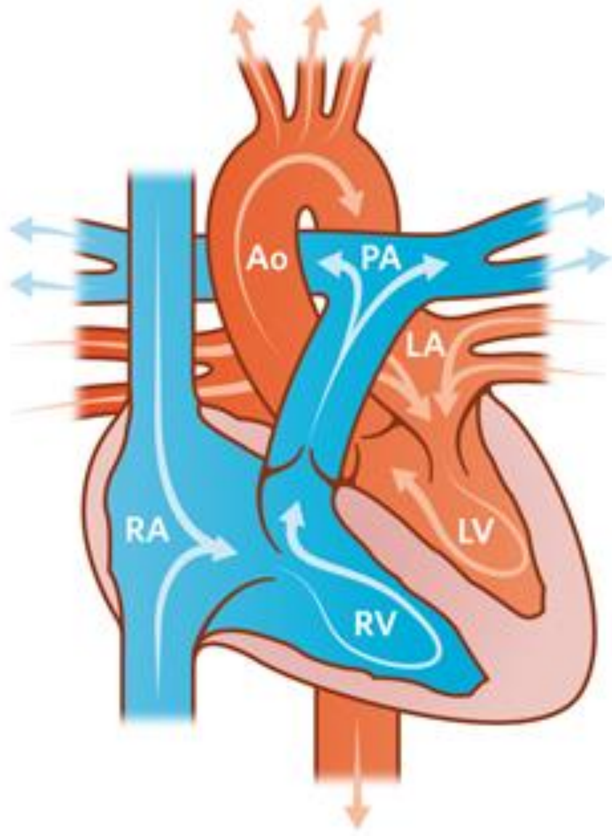
Normal Circulation



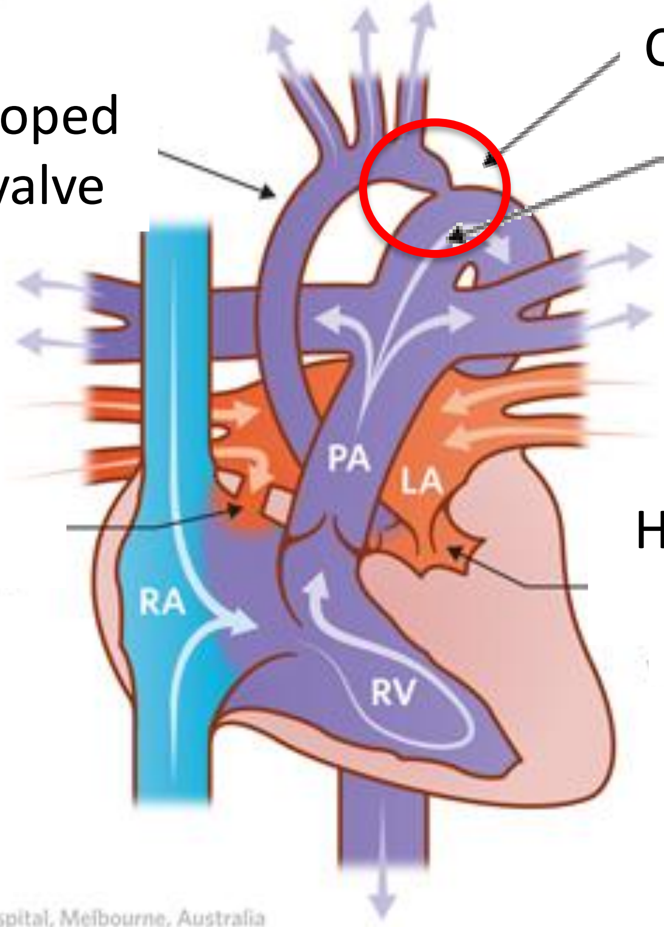


Hypoplastic Left Heart Syndrome (HLHS)

- HLHS is a severe form of congenital heart disease.
  - Without surgical intervention, HLHS is fatal.
- Infants are often diagnosed within 24 to 48 hours of birth
  - Four extremity saturations (pre/post duct differences)
  - Murmur can be heard, cyanosis noted
  - More significant symptoms appear when the PDA begins to close.
- There are three options for treating these children:
  - supportive care until death occurs
  - staged reconstruction of the heart
  - heart transplant

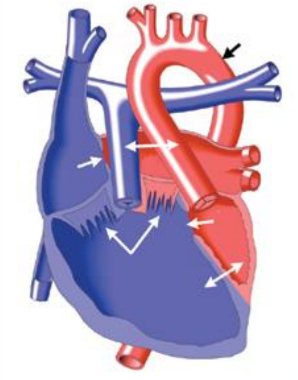


Normal heart and circulation

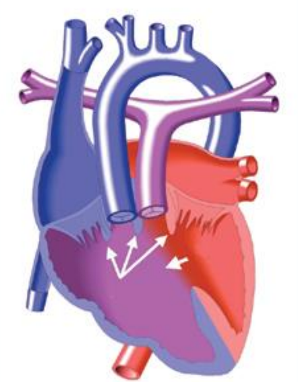


Single Ventricle Lesions

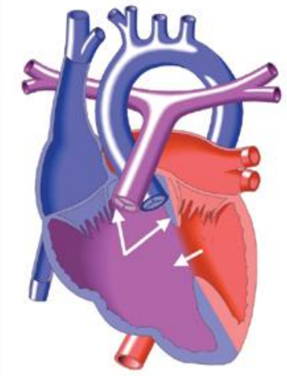
…are not restricted to Hypoplastic Left Heart Syndrome



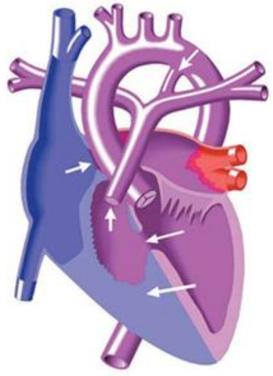
Double Inlet Left Ventricle



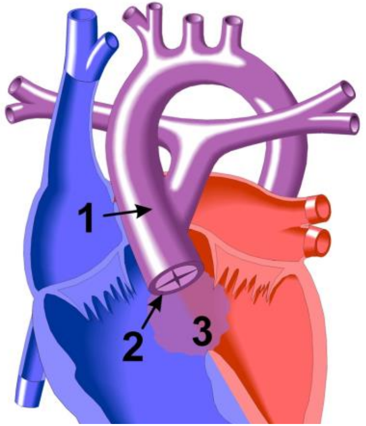
Double Outlet Right Ventricle w/Transposition



Double Outlet Right Ventricle



Tricuspid Atresia



Truncus Arteriosus

Pre-Op Considerations

- Prostaglandins (PGE) maintains ductal patency for adequate systemic blood flow.
  - Concerns for apnea
- Infants with cardiac shock often require intubation, volume & inotropy
- Ensuring adequate systemic perfusion (i.e., balancing Q_P:Q_S) becomes crucial.

Cardiac Output - Q_P:Q_S Ratio

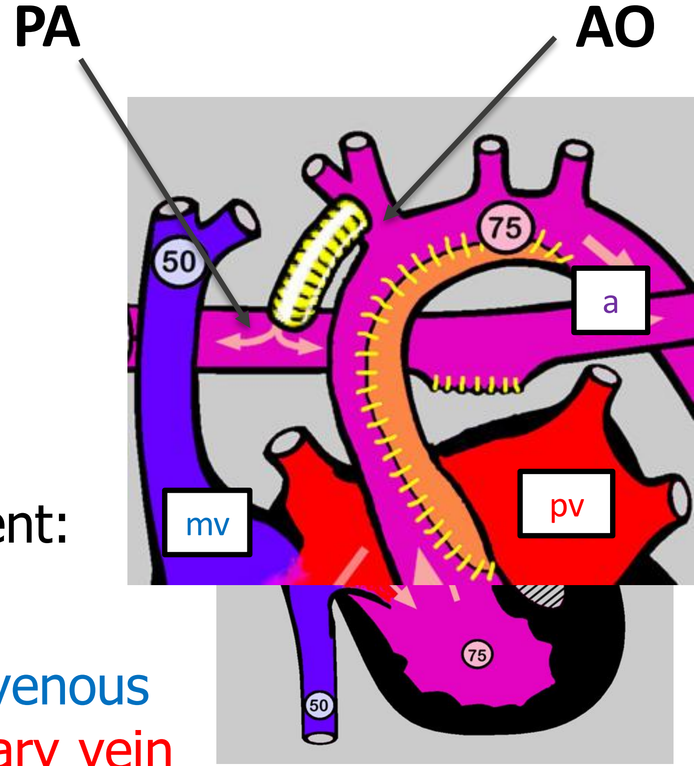
Since Aortic and Pulmonary Blood Flow
both come from the Aorta:
Aortic Sat. = Pulmonary Sat.

$$\frac{Q_P}{Q_S} = \frac{\text{Pulmonary Blood Flow}}{\text{Systemic Blood Flow}}$$

$$\frac{Q_P}{Q_S} = \frac{S_aO_2 \quad S_{mv}O_2}{S_{pv}O2 \quad S_aO_2}$$

In a SV patient:

a= arterial
mv= mixed venous
pv= pulmonary vein
$S_{pa}O_2 = S_aO_2$



$Q_p{:}Q_s$ Ratio

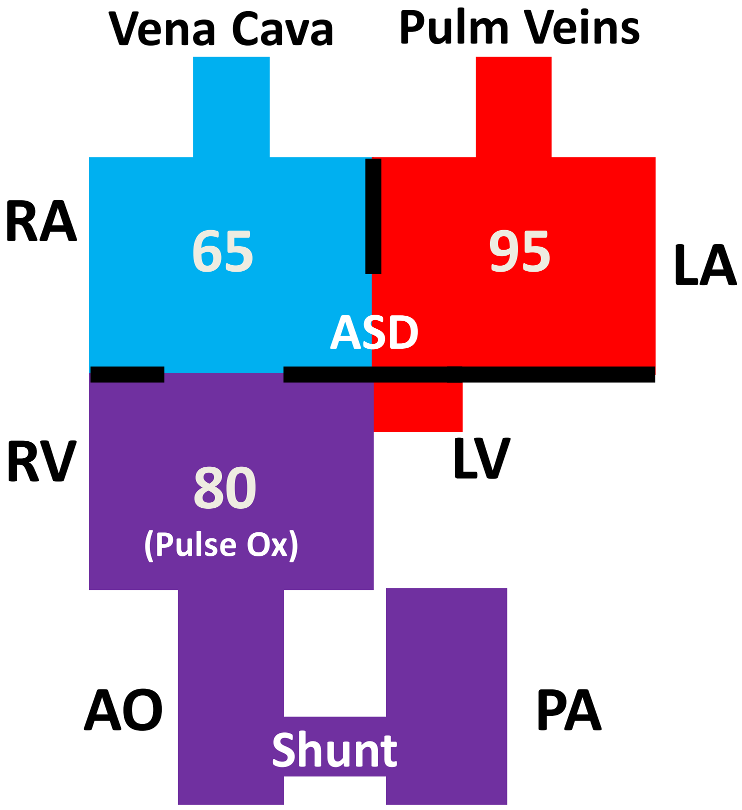
In a SV patient:

$S_{PV}O_2$ = 95-100

Measure:

S_aO_2 & $S_{mv}O_2$

$$Q_p{:}Q_s = \frac{S_aO_2 - S_{mv}O_2}{95 - S_aO_2}$$



$Q_p:Q_s$ Ratio

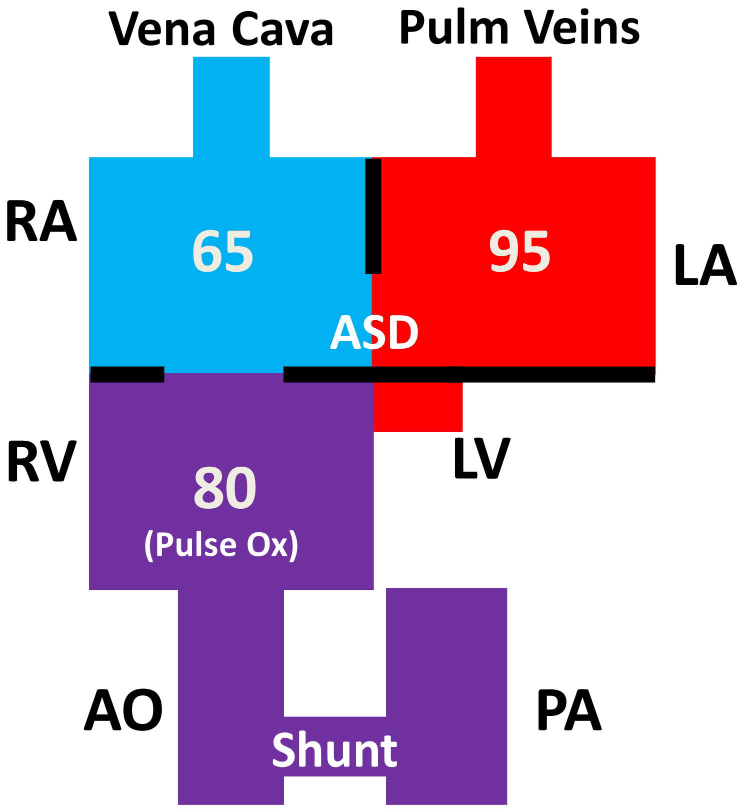
In a SV patient:

$S_{PV}O_2$ = 95-100

Measure:

S_aO_2 & $S_{mv}O_2$

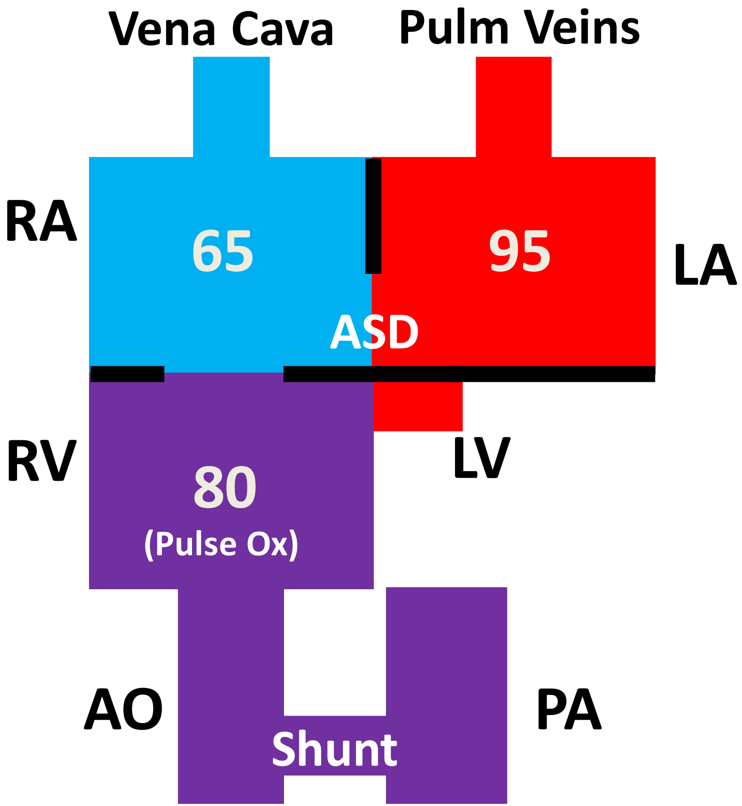
$$Q_p:Q_s = \frac{80 - 65}{95 - 80}$$



Q_p:Q_s Ratio

$$\frac{80 - 65}{95 - 80} = \frac{15}{15} = \frac{1}{1}$$

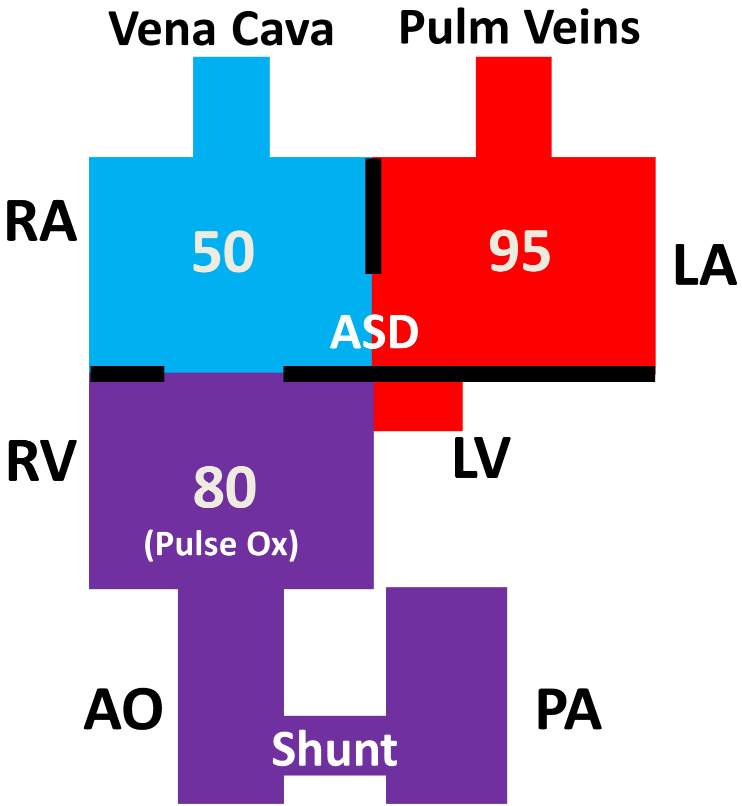
Balanced Pulmonary Blood Flow



Q_p:Q_s Ratio

$$\frac{80 - 50}{95 - 80} = \frac{30}{15} = \frac{2}{1}$$

Excessive Pulmonary Blood Flow



$Q_p:Q_s$ Ratio

$$\frac{80 - 50}{95 - 80} = \frac{30}{15} = \frac{2}{1}$$

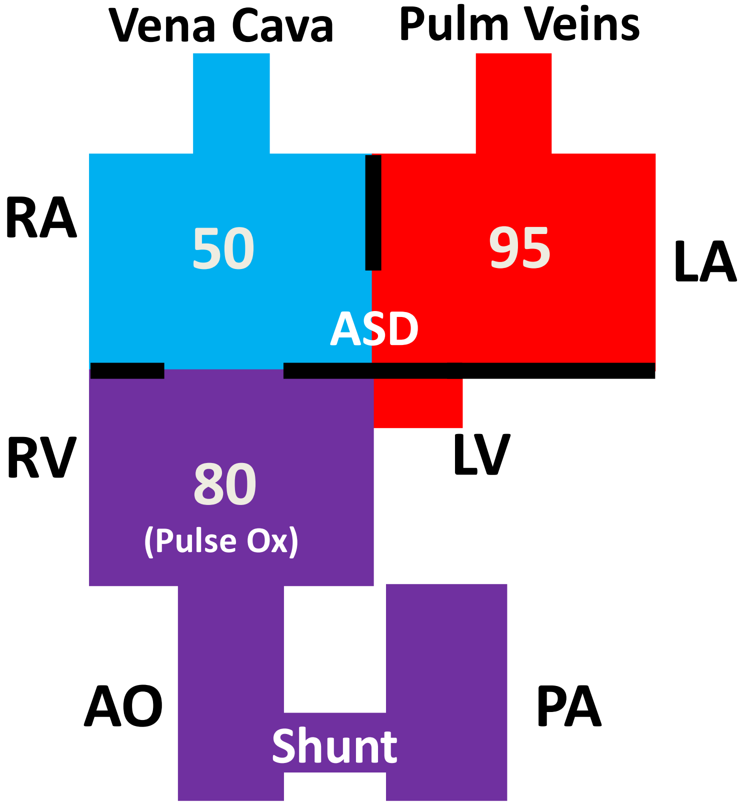
Excessive Pulmonary Blood Flow

Increase PVR

- Keep FiO2 low
- Acidosis

Decrease SVR

- Sedation
- Milrinone



Pulmonary Vascular Resistance

Pulmonary Vasodilators

- Oxygen
- Nitric Oxide
- Alkalosis
- Milirone
- Nipride

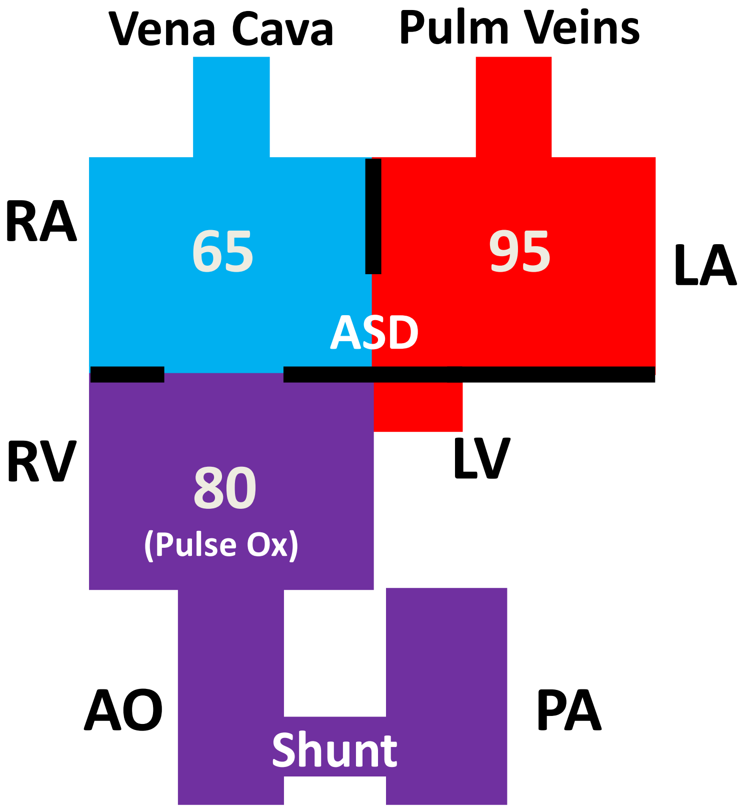
Pulmonary Vasoconstrictors

- Atelectasis
- Hyperinflation
- Acidosis
- Inotropes

Q_p:Q_s Ratio: Shunt Failure

$$\frac{80 - 65}{95 - 80} = \frac{15}{15} = \frac{1}{1}$$

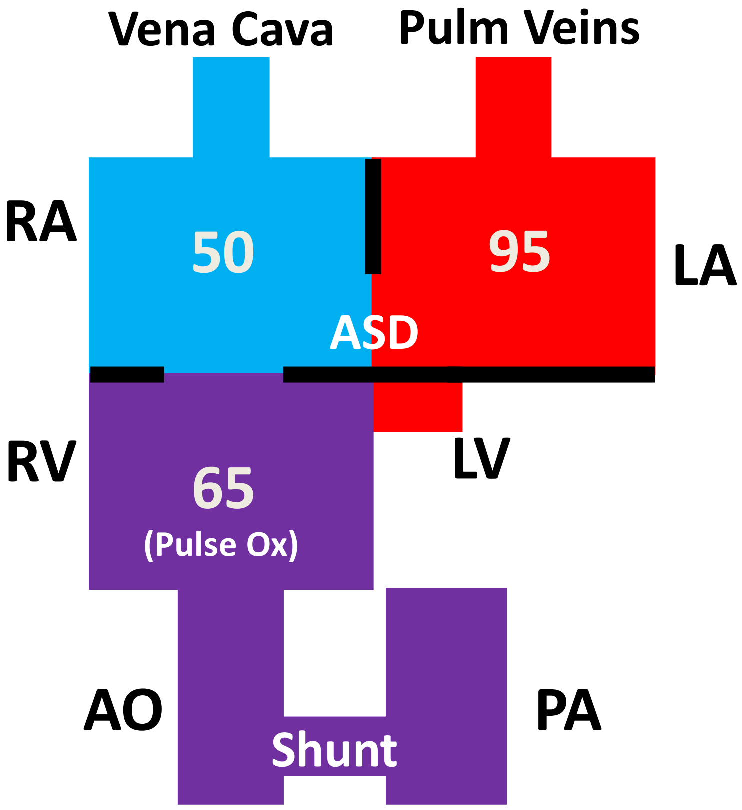
Balanced Pulmonary Blood Flow



Q_p:Q_s Ratio: Shunt Failure

$$\frac{65 - 50}{95 - 65} = \frac{15}{30} = \frac{1}{2}$$

Inadequate Pulmonary Blood Flow



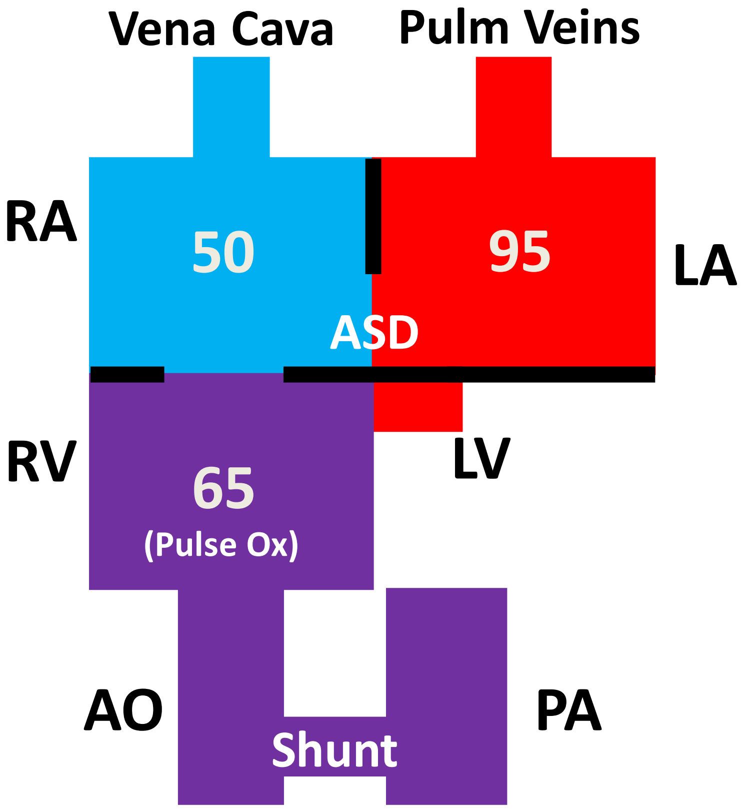
Q_p:Q_s Ratio: Shunt Failure

$$\frac{65 - 50}{95 - 65} = \frac{15}{30} = \frac{1}{2}$$

Inadequate Pulmonary Blood Flow

Interventions: ***Decrease PVR***

- FiO2
- iNO
- Metabolic Alkalosis



Systemic Vascular Resistance

Increases SVR

- Epinephrine, Dopamine, Vasopressin (Exogenous catecholamines)
- Pain / Agitation (Endogenous catecholamines)
- Decreased Body Temp

Decreases SVR

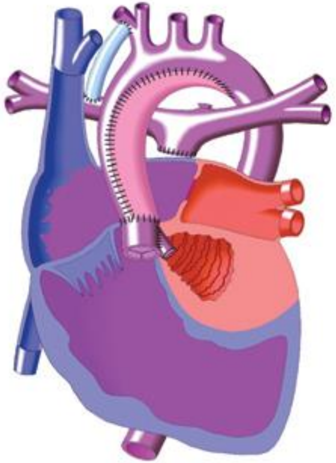
- Versed, Propofol, Ativan, Morphine
- Increased Body Temp
- Milirinone, Nipride, Nicardipine, Esmolol, Dobutamine

Causes of Desaturation

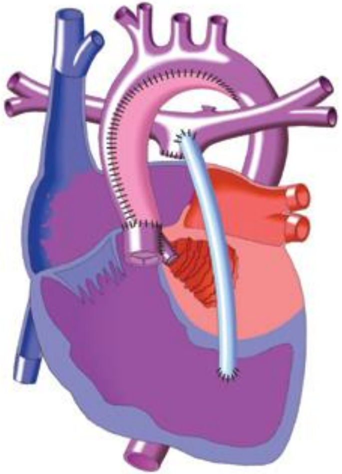
- S_aO_2 is dependent on
  - Mixed Venous Saturation ($S_{mv}O_2$)
    - Low cardiac output syndrome
    - Increased metabolism (agitation, fever)
  - Pulmonary Venous Saturation ($S_{pv}O_2$)
    - A-a gradient, parenchymal issues
  - Alterations in Q_P/Q_S
    - PVR vs. SVR

HLHS Palliation

Stage 1

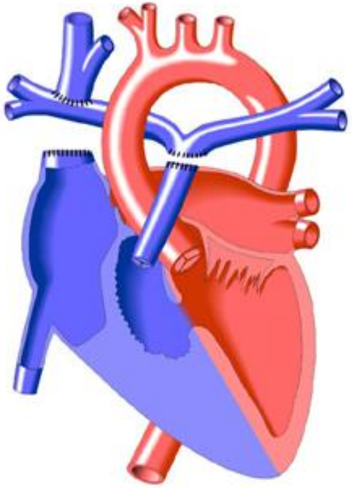


Norwood Procedure



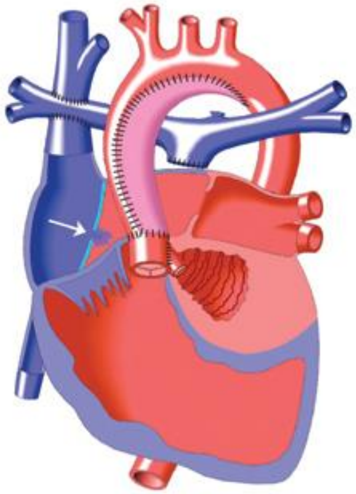
Sano Modification of Norwood Procedure

Stage 2



Glenn Procedure

Stage 3

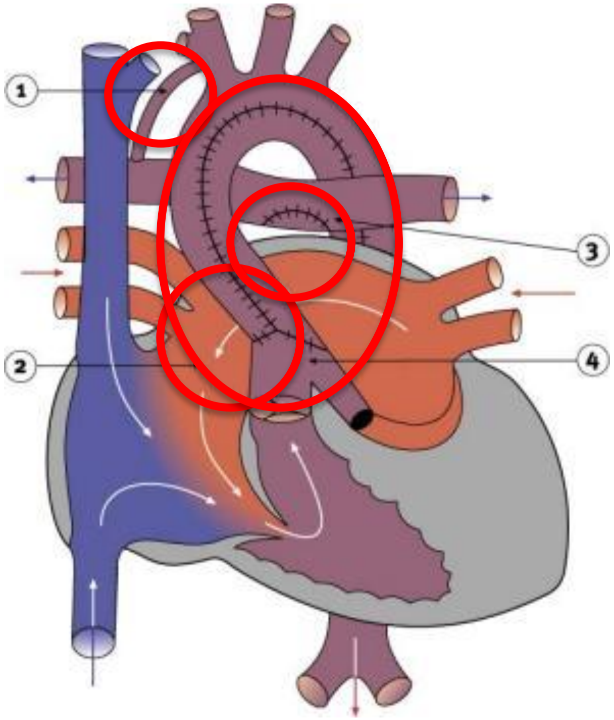


Fontan Procedure

History

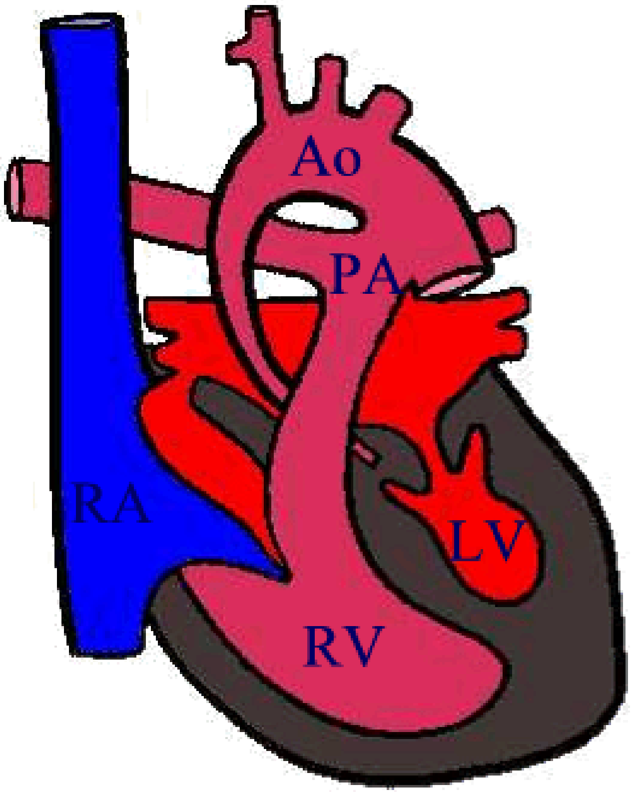
- 1944 Johns Hopkins
  - Alfred Blalock, Helen Taussig, Vivien Thomas
  - "Blue Babies" (TOF, but other shunt-dependent lesions also)
- 1971 – Francois Fontan connected the pulmonary and systemic circulations to one ventricle
  - Modified in 1988 and was the primary surgical approach to hypoplastic palliation in the 1990's
- 2003 – Shunji Sano modifies the palliation to create a RV to PA shunt

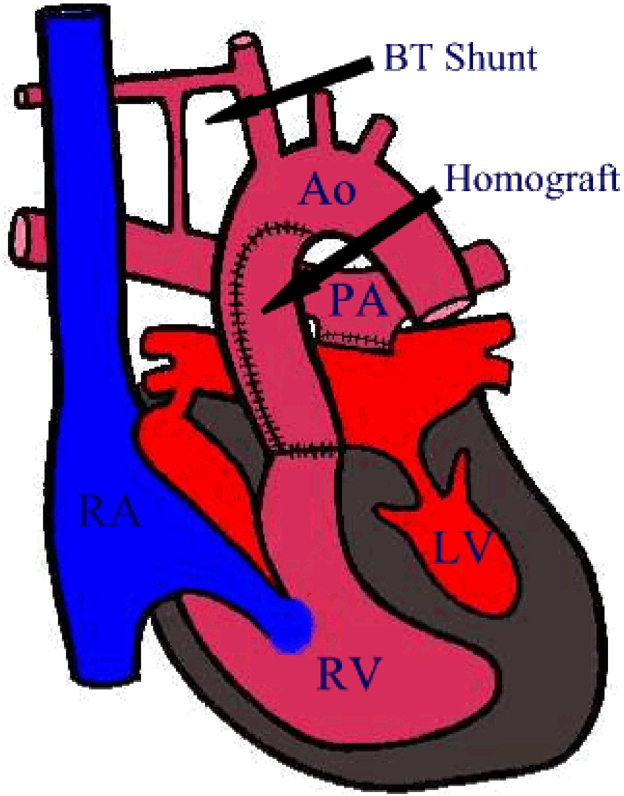
Norwood – Stage I



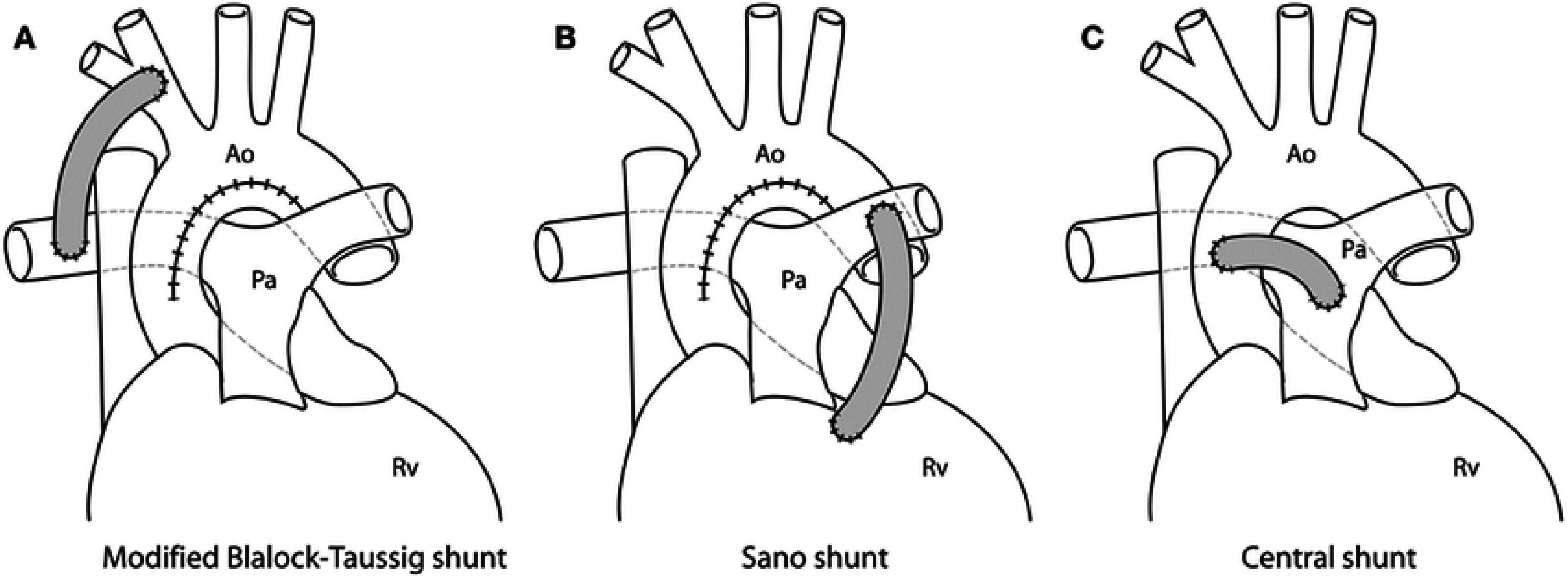
1. Blalock-Taussig shunt (temporary)
2. Atrial septum removed
3. Removal of Ductus Arteriosus
4. Aorta and pulmonary trunk anastomosed together

Norwood – Stage 1: Before and After



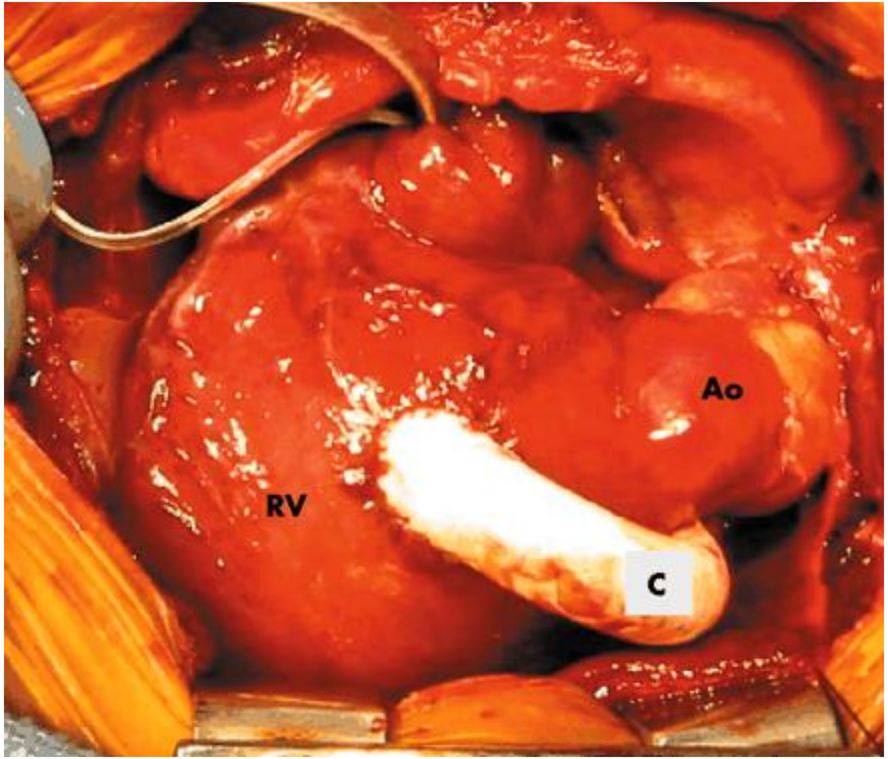


Types of Shunts



Biglino, Frontiers in Pediatrics 1:31 · October 2013

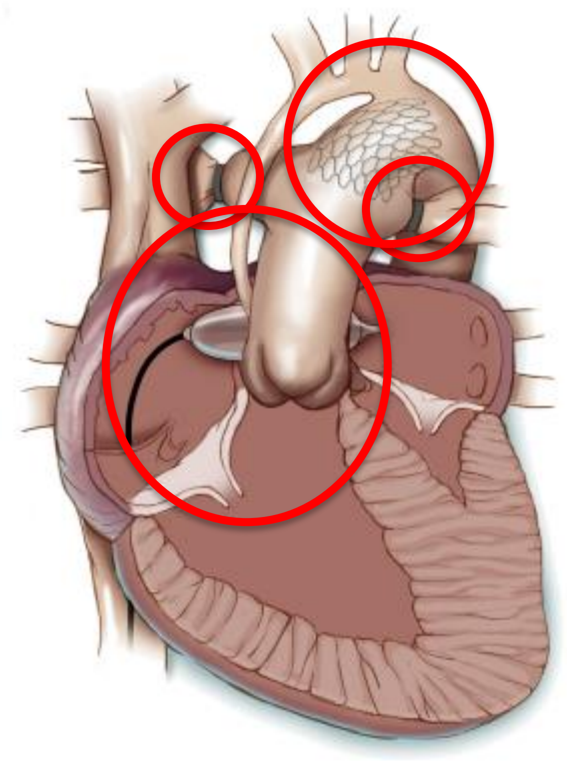
RV-PA Conduit



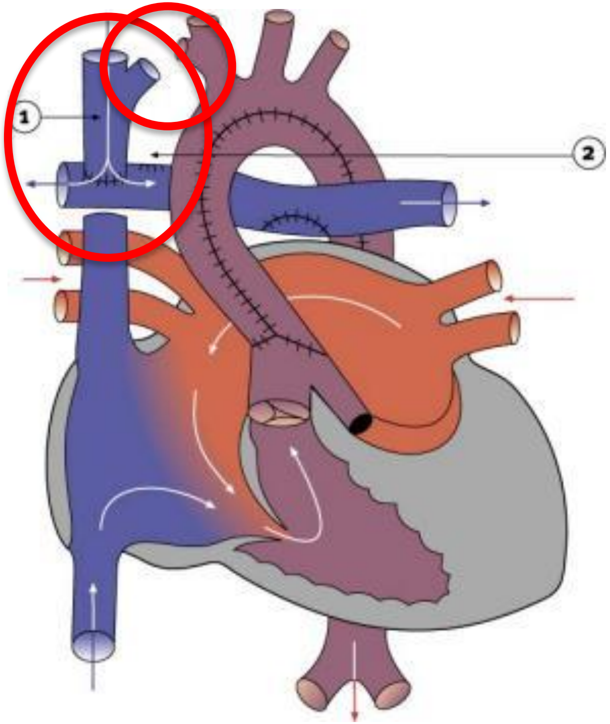
Theilen, ADC Fetal & Neonatal 2005;90:F97-F102

Hybrid Procedure

- PA bands (placed off bypass)
- PDA stent (via cath)
- Balloon atrial septostomy

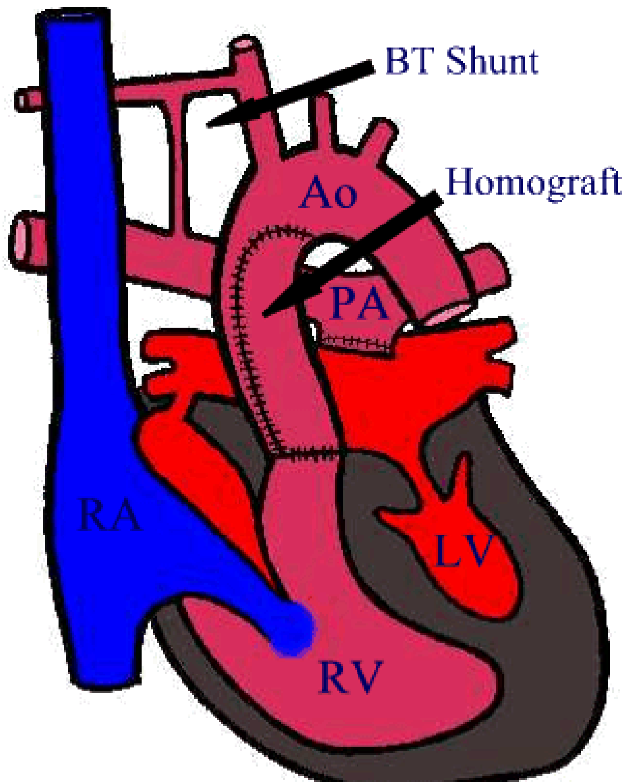


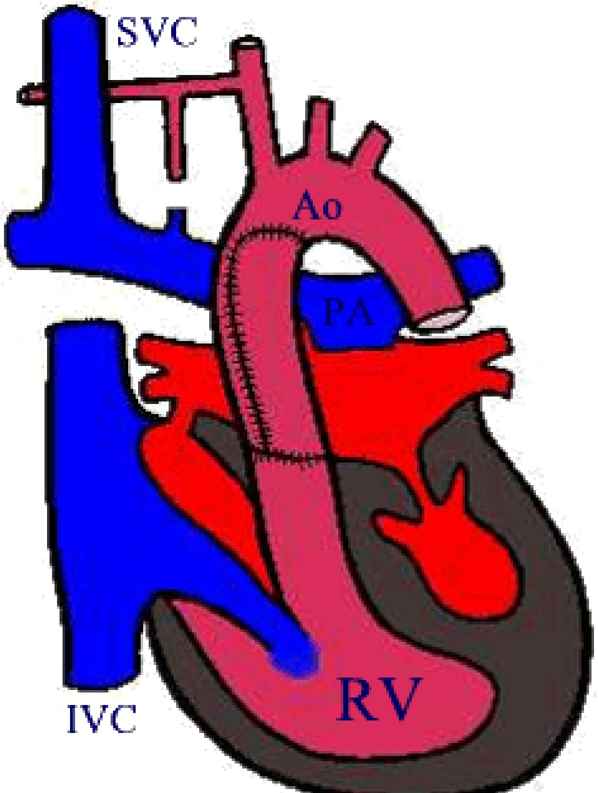
Glenn Procedure – Stage II



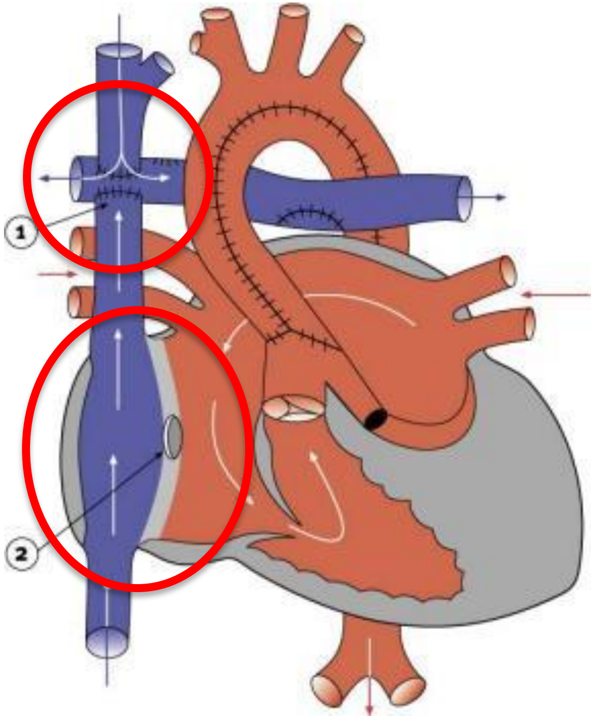
1. Superior vena cava connected to pulmonary artery
2. Takedown of BT shunt

Stage 1 to Stage 2: Glenn Procedure





Fontan – Stage III



1. Conduit joining inferior vena cava and superior vena cava to pulmonary artery
2. IVC connection to RA may have a fenestration added to allow pressure relief for changes in PVR

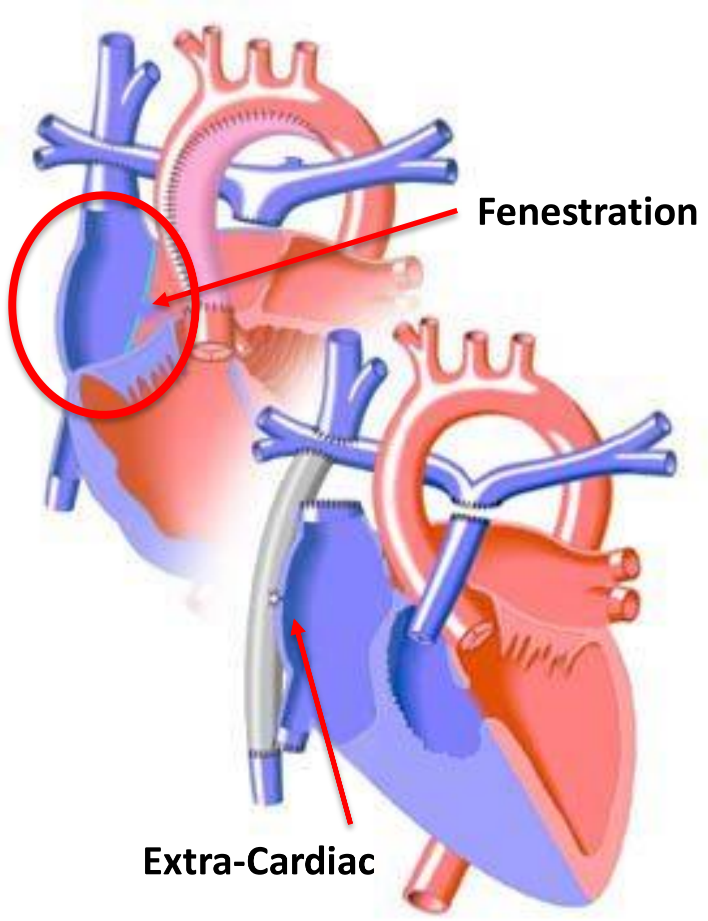
Fontan(s)?

The Fontan procedure has evolved and has 3 interations:

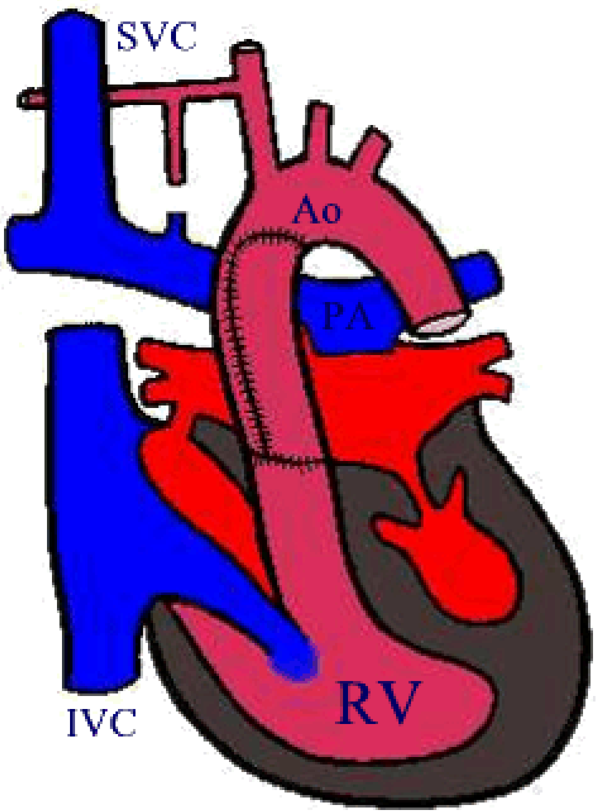
Extra-Cardiac –direct connection of IVC to PA, bypassing the heart

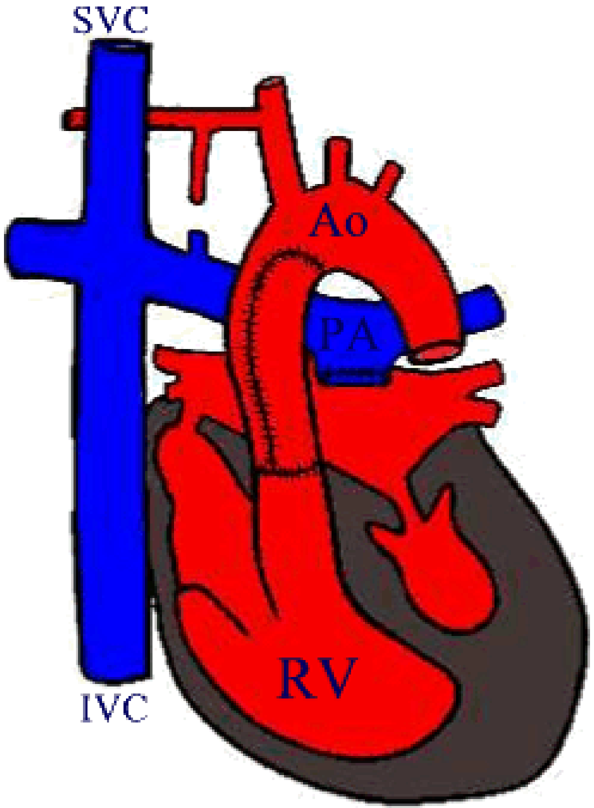
Intra-Cardiac AKA Lateral Tunnel – "baffle" placed to create a "tunnel" through the atrium to the PA

Intra-Cardiac with fenestration – same as above but with a "pressure pop-off" to allow initial high RA pressures (created by resistance to added flow to PA) to be relieved into the ventricle



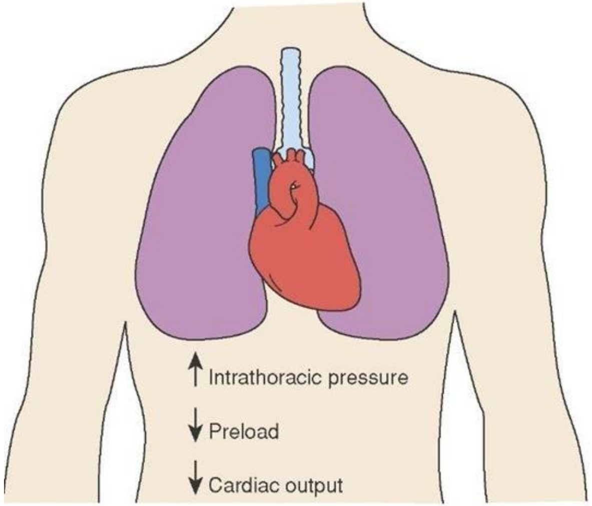
Stage 3: Glenn to Fontan





Complications of HLHS Palliation

- Higher incidence after stage I vs. II
- Addition of PVR to SVC drainage elevates CVP
- Anything that increases the resistance to the passive venous return to the RA will have
  - Pulmonary Hypertension
  - PA hypoplasia
  - Tricuspid or RV insufficiency
  - Restrictive ASD



Complications of HLHS Palliation

- Protein Losing Enteropathy (PLE)
  - Excessive loss of proteins in the intestinal mucosa
    - Ascites, effusions, edema
- Plastic Bronchitis
  - Aerosolized Urokinase? TPA?

Respiratory Considerations

- Mechanical ventilation raises intrathoracic pressure, potentially compromising passive venous return to the heart
  - Early extubation should always be the goal
  - Keep PEEP and MAP at minimum levels required to maintain ABG goals
    - SaO2 70's-80's
  - iNO or Sildenafil to mitigate any pulmonary hypertension
  - *Any* parenchymal disease can cause major issues
  - Watch for diaphragmatic concerns caused by intra-operative phrenic nerve injury
  - Effusions common re: disruption of lymphatic drainage & post op inflammation

Why Not Transplant?

- Donor availability
  - 70-100 neonatal hearts in North America annually
  - Wait list of several months with high mortality
  - Stage I palliation with 6-25% mortality
    - Better than waitlist mortality, especially for HLHS patients
  - Perioperative mortality for transplant similar or worse than Norwood numbers (~20%)
- ABO transplant to widen donor pool?
  - Long term mortality better in transplant patients (survival 20+yrs)

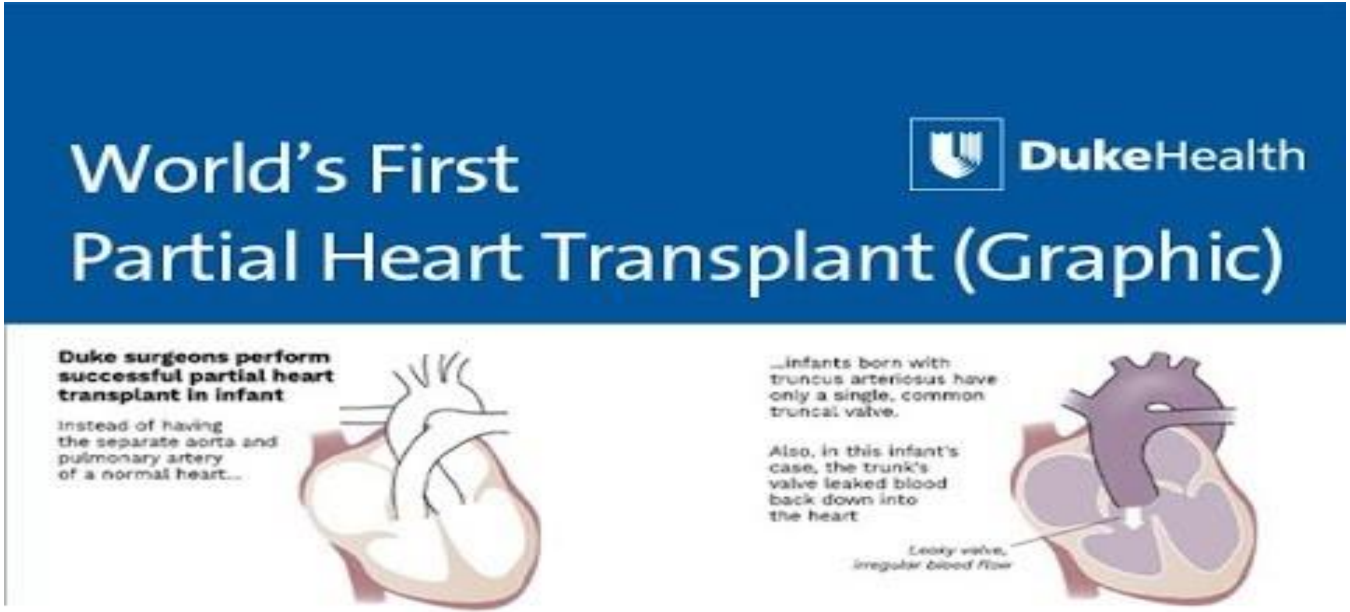
World's First Heart Transplant, Cultured Thymus Implantation Performed at Duke

Milestone Could Eliminate Need for Anti-Rejection Medications

By Morgan deBlecourt
March 09, 2022

Share:

Easton Sinnamon smiles alongside his sister, Ivy, at his one-year birthday celebration. Credit: Sinnamon Family.


World's First
Partial Heart Transplant (Graphic)
DukeHealth
Duke surgeons perform successful partial heart transplant in infant
Instead of having the separate aorta and pulmonary artery of a normal heart...
...infants born with truncus arteriosus have only a single, common truncal valve.
Also, in this infant's case, the trunk's valve leaked blood back down into the heart
Leaky valve, irregular blood flow

Thank You For Your Time!

Stephen.Hepditch@duke.edu