Alder Hey Children's NHS
NHS Foundation Trust

Physiological effects of Congenital Heart Disease

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Inspired by Children

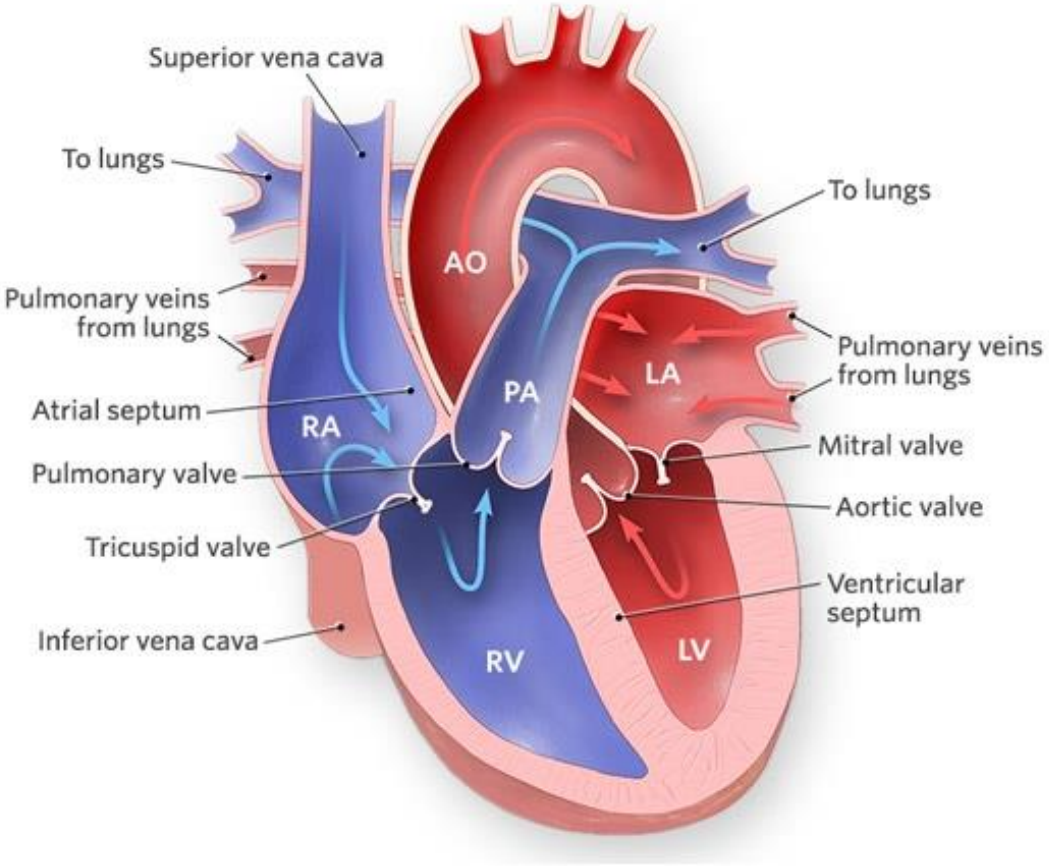
Physiological effects of CHD

There are 3 basic effects on the body

- Increased pulmonary blood flow (PBF)
- Decreased pulmonary blood flow
- Decreased systemic blood flow

Remember that blood will always flow through the path of least resistance

Normal Heart

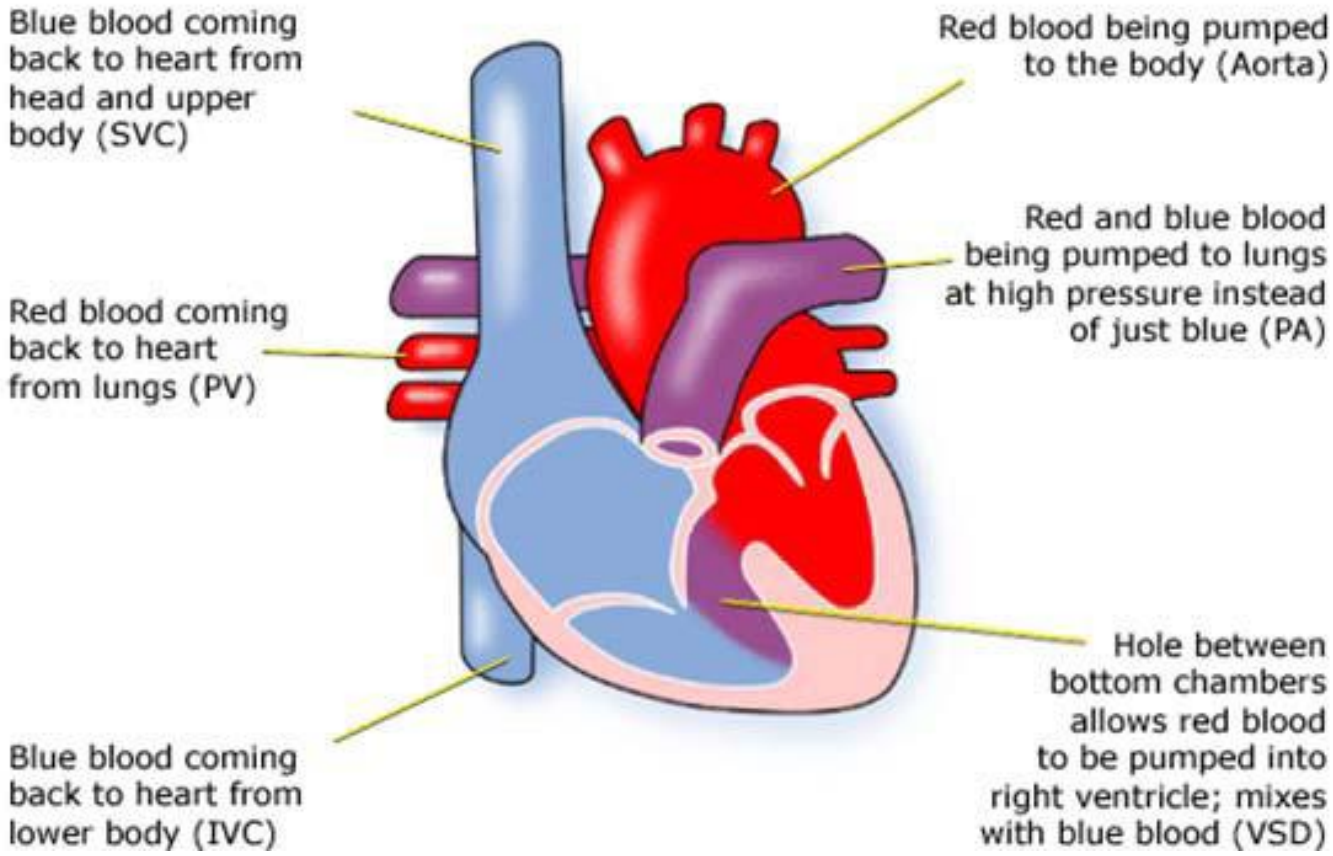


Increased pulmonary blood flow

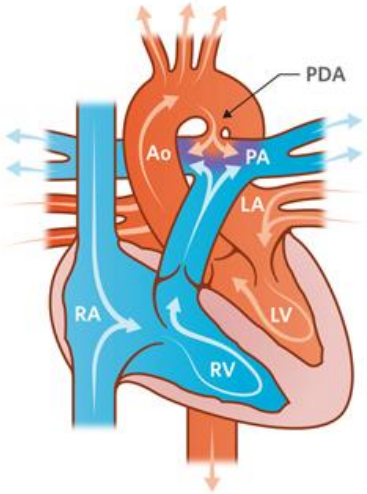
- Left side of heart is at higher pressure than the right side
- Increase in blood in the right side of heart leads to increased pulmonary blood flow

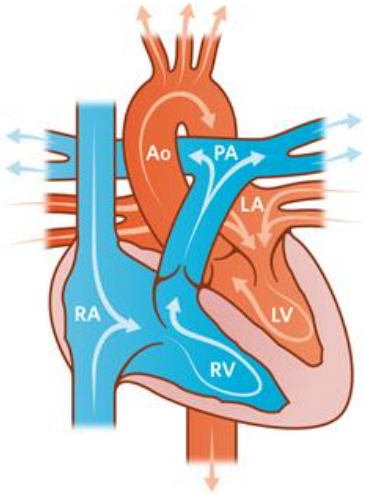
- Ventricular Septal Defect (VSD)
- Atrial Septal Defect (ASD)
- Patent ductus arteriosus (PDA)
- Complete or partial Atrioventricular Defect (CAVSD, PAVSD)
- Total or Partial Anomalous Pulmonary Venous Drainage (TAPVD, PAPVD)
- Truncus Arteriosus
- Transposition of the Great Arteries (TGA)

VSD (Ventricular Septal Defect)

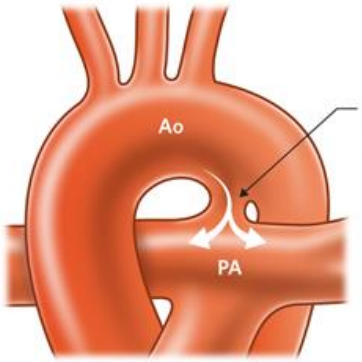


Patent ductus arteriosus (PDA)



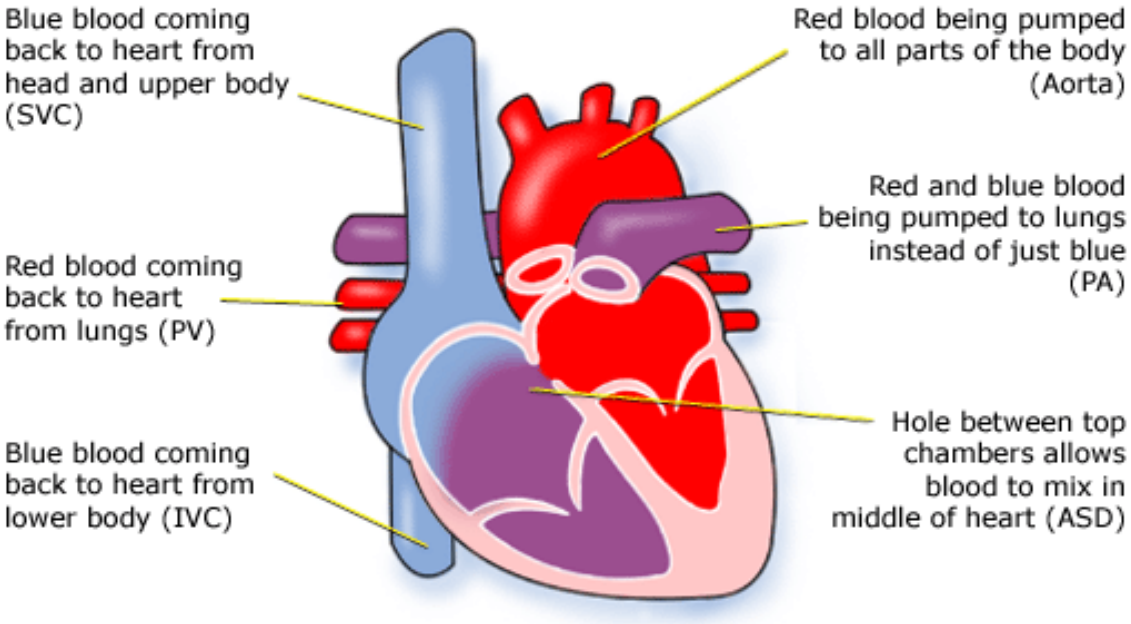


Normal heart and circulation

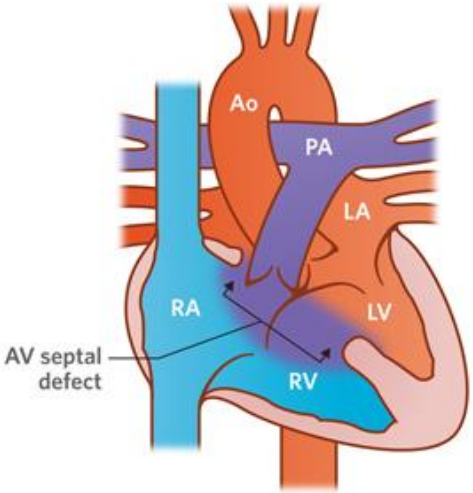


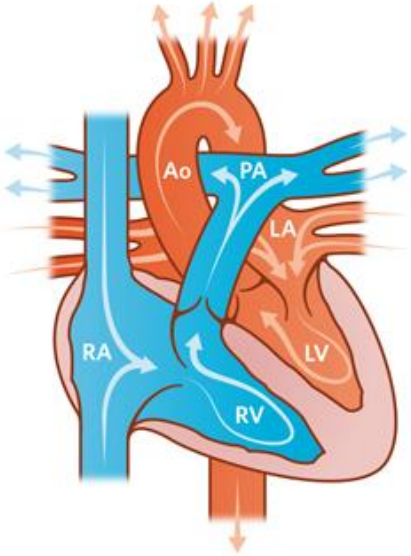
Blood flows from aorta through patent ductus into the pulmonary artery

ASD (Atrial Septal Defect)

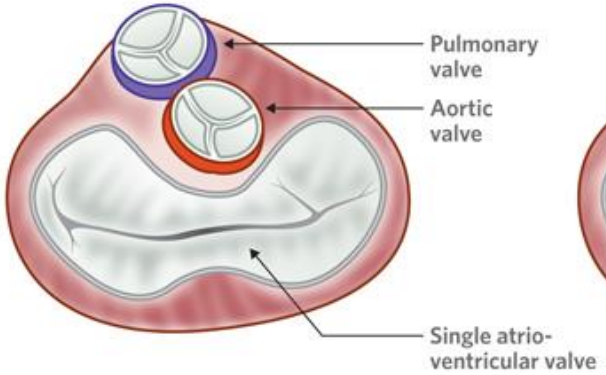


Atrioventricular septal defect

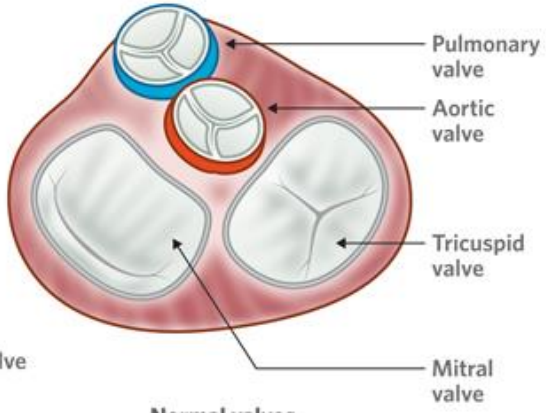




Normal heart and circulation

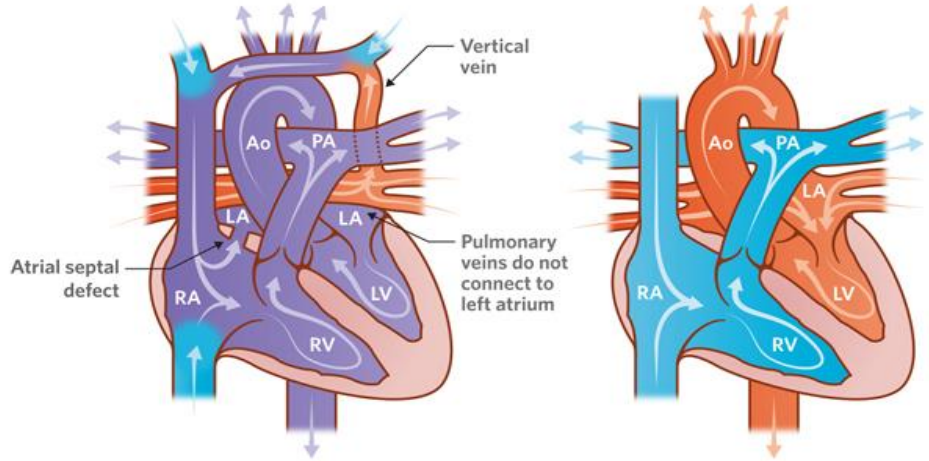


'Shared' atrioventricular valve

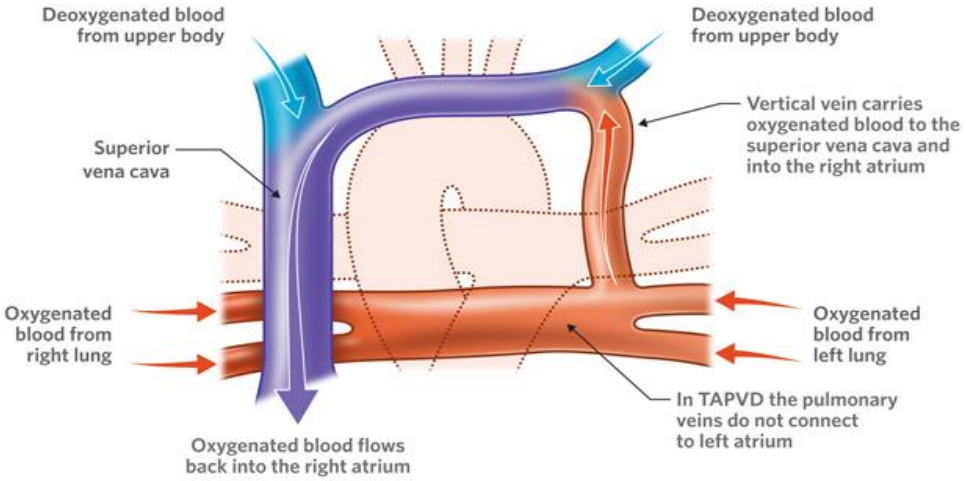


Normal valves

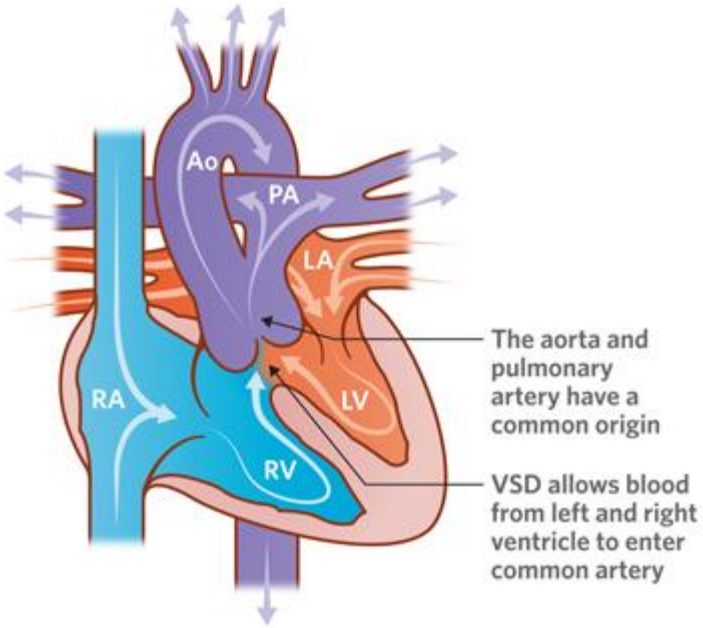
Total anomalous pulmonary venous drainage (TAPVD)

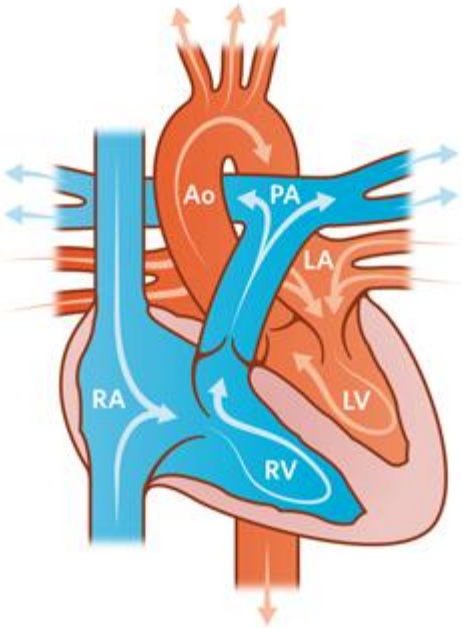


Normal heart and circulation



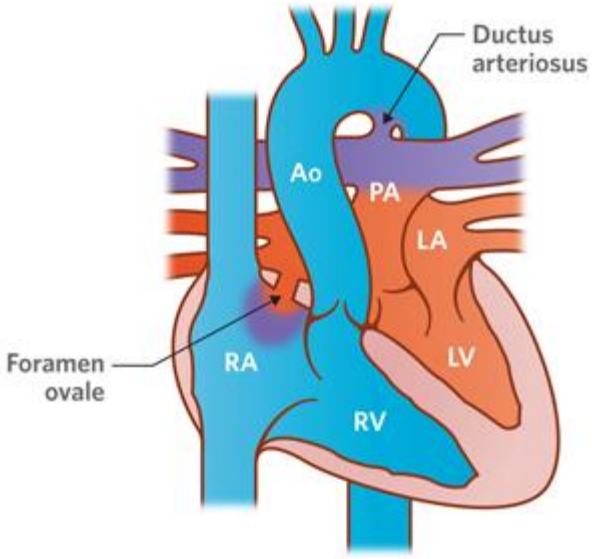
Truncus arteriosus

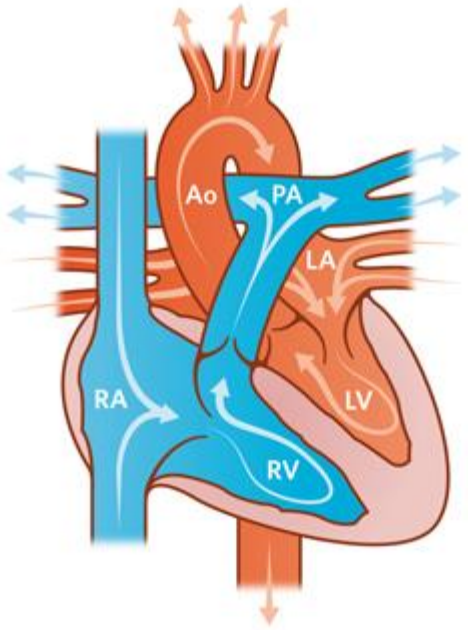




Normal heart and circulation

Transposition of the great arteries





Normal heart and circulation

Congestive heart failure (CHF)

- Cardiomegaly
- Tachycardia
- Hypotension
- Tachypnoea
- Pulmonary oedema
- Pleural effusions
- Poor urine output
- Retention of fluid and sodium
- Failure to thrive (FTT)

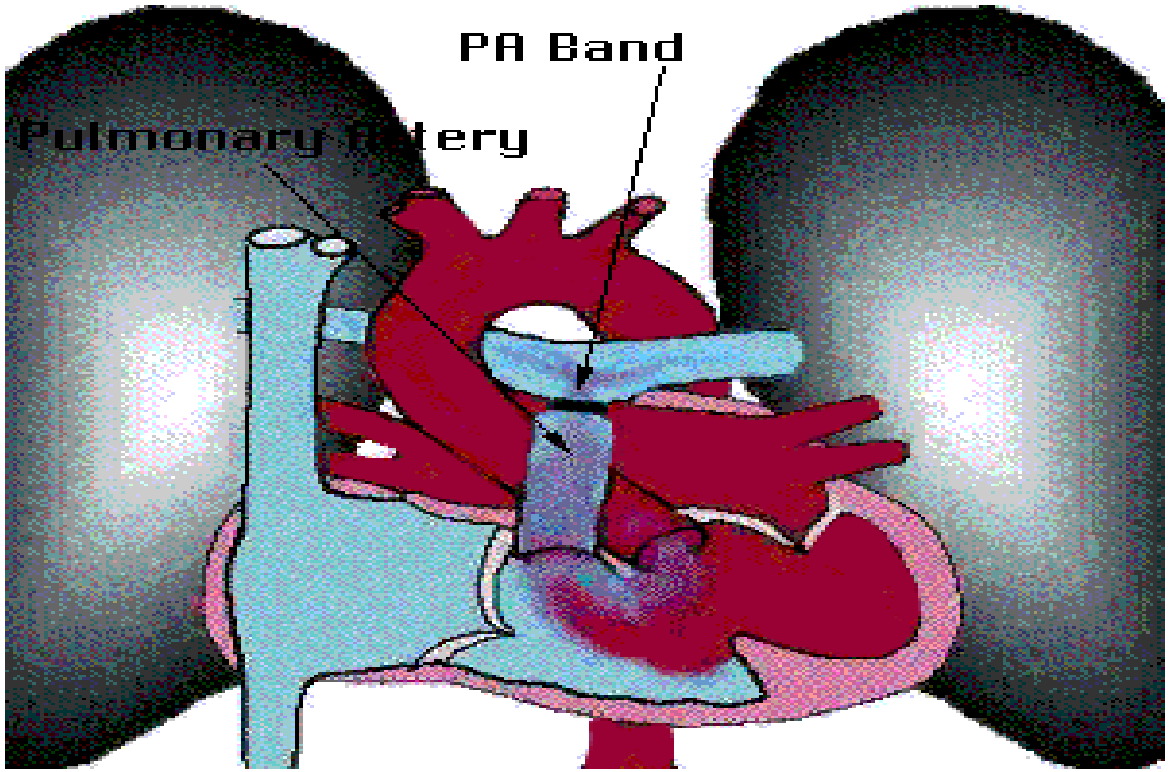
Potential consequences of ↑PBF

- Excessive pulmonary blood flow ultimately leads to Pulmonary Hypertension (PHT)
- If PHT, then reversal of L-R shunt can occur producing the cyanotic child – Eisenmenger syndrome

What are we going to do?

- Repair the defect
- Palliate the defect – Pulmonary Artery Band
- Medical management of symptoms – diuretics, ACE inhibitors, nutritional support
- Respiratory support
- ?Palivizumab if under 2 years
- CCNT support if has NG/PEG feeds, saturation monitoring not usually required

PA Band

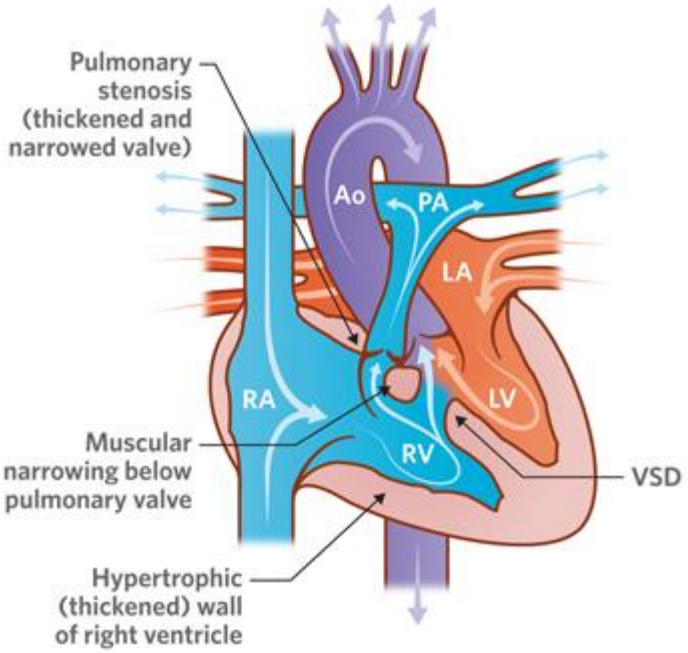


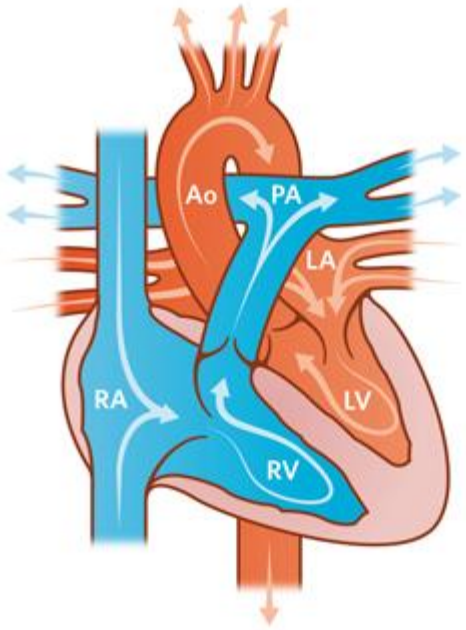
Decreased pulmonary blood flow

Defects which obstruct blood flow to the lungs

- Pulmonary Stenosis
- Pulmonary Atresia
- Tetralogy of Fallot
- Tricuspid Atresia

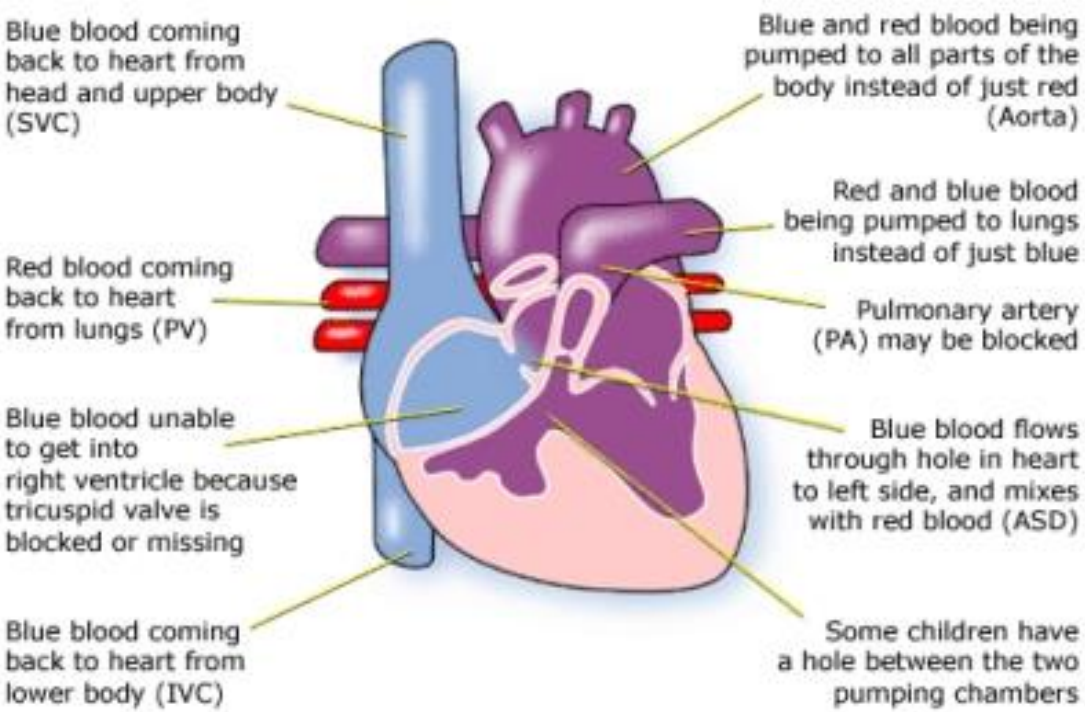
Tetralogy of Fallot



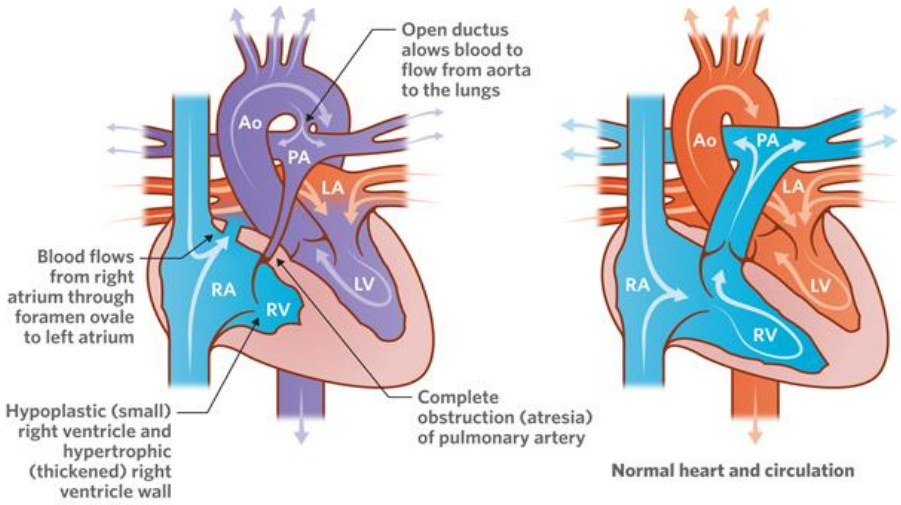


Normal heart and circulation

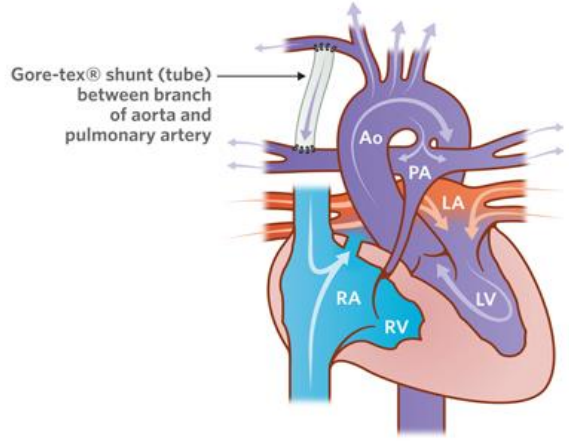
Tricuspid Atresia



Pulmonary atresia with intact ventricular septum

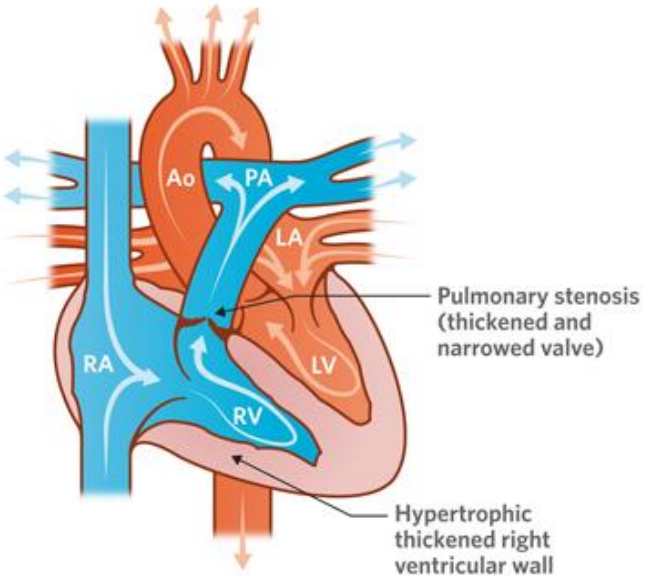


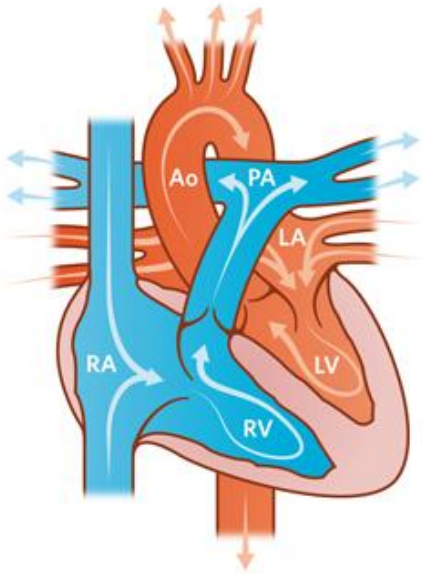
Normal heart and circulation



Shunt operation

Pulmonary stenosis (PS)





Normal heart and circulation

Pulmonary stenosis

Healthy pulmonary valve

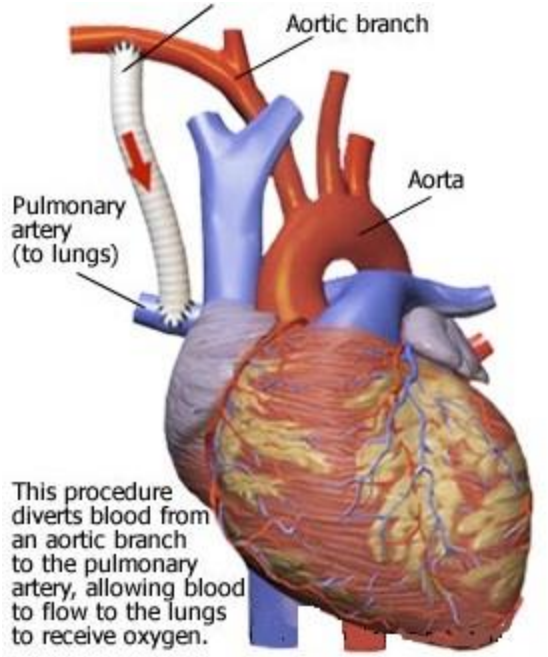
Potential consequences of ⬇ PBF

- Cyanosis
- Hypercyanotic spells in Fallot's babies(TET spells)

What are we going to do?

- Maintain PDA – Prostin, PDA stent
- Cardiac Catheter intervention – PDA Stent, Ballooning of pulmonary valve, Radiofrequency (RF) perforation of pulmonary valve
- Surgical intervention –Pulmonary Valvotomy, Modified Blalock-Taussig Shunt (MBTS), Right Ventricle to Pulmonary Artery (RV-PA) shunt (Sano)
- Community saturation monitoring/ Single Ventricle Home Monitoring Programme & open access.
- ?Palivizumab if under 2 years old

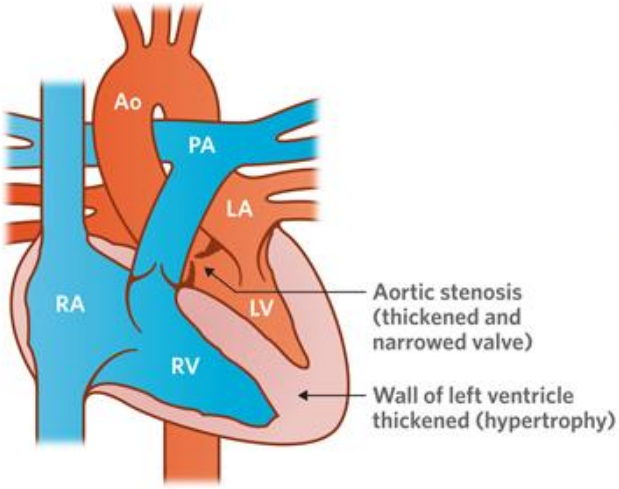
BT shunt

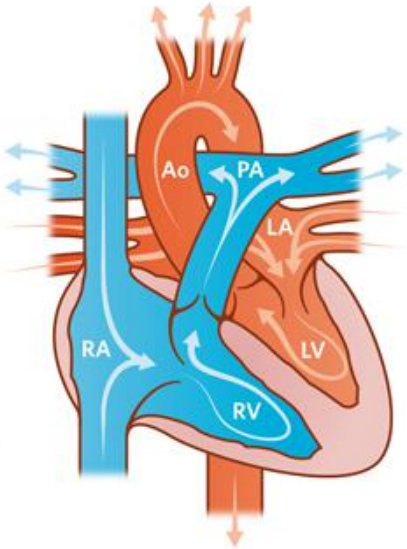

Aortic branch
Aorta
Pulmonary artery (to lungs)
This procedure diverts blood from an aortic branch to the pulmonary artery, allowing blood to flow to the lungs to receive oxygen.

Reduced systemic blood flow

- Aortic Stenosis
- Coarctation of the Aorta
- Interrupted Aortic Arch
- Hypoplastic Left Heart Syndrome

Aortic stenosis (AS)



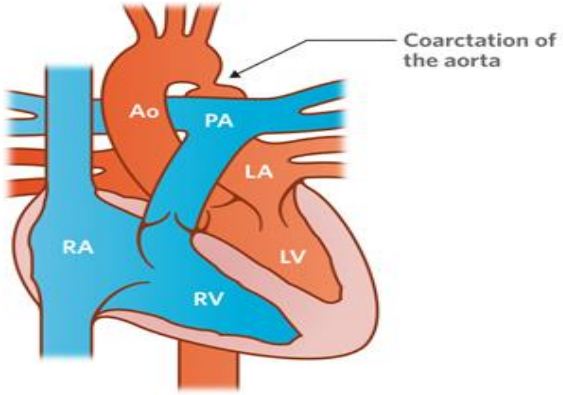


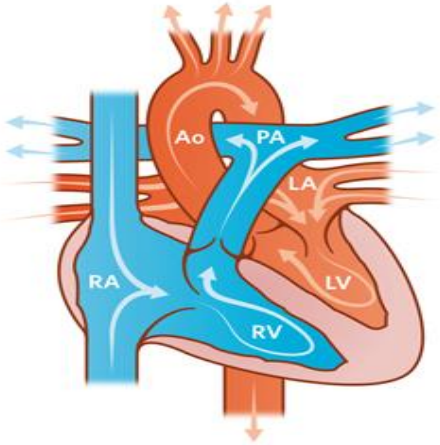
Normal heart and circulation

Aortic stenosis

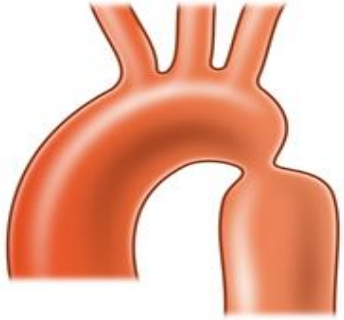
Healthy aortic valve

Coarctation of the aorta





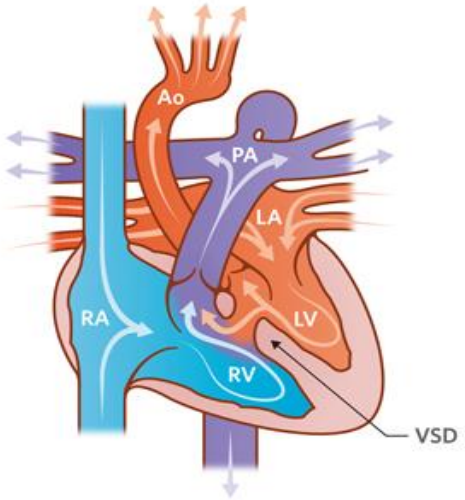
Normal heart and circulation

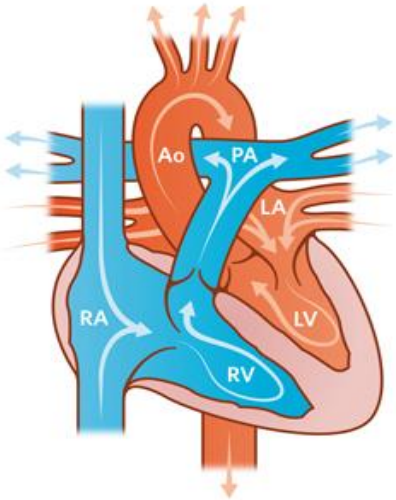


Coarctation of the aorta

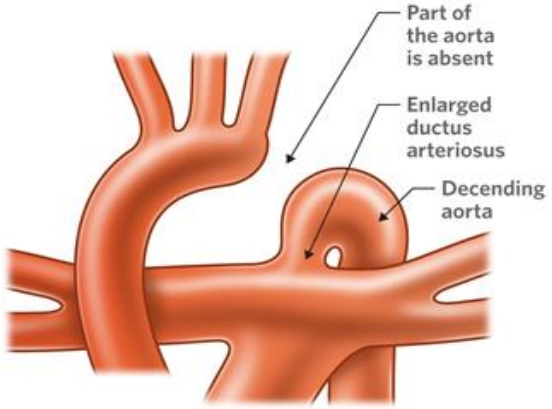
Normal aorta

Interrupted aortic arch

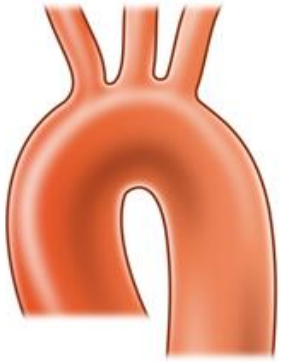




Normal heart and circulation

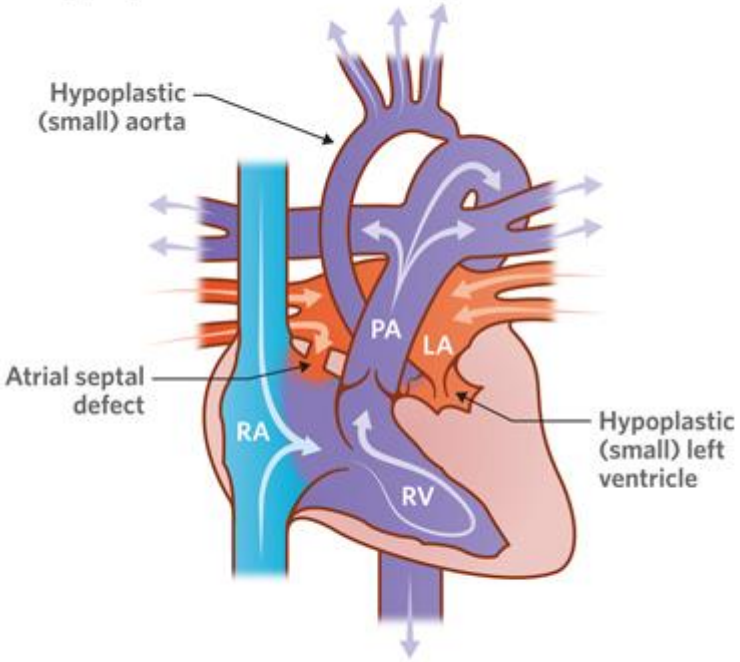


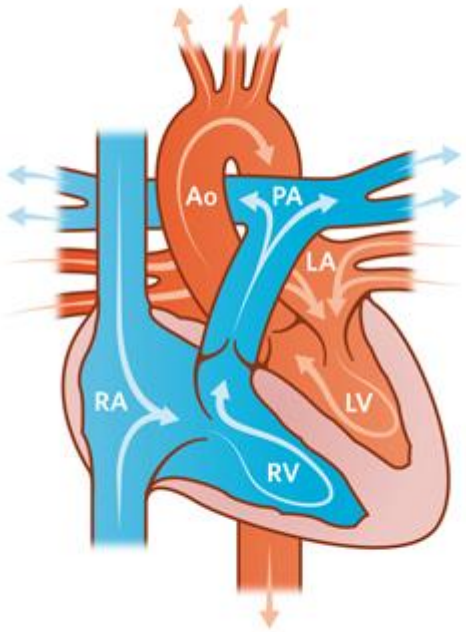
Interrupted aortic arch



Normal aorta

Hypoplastic left heart syndrome





Normal heart and circulation

Potential consequences of ↓ systemic blood flow

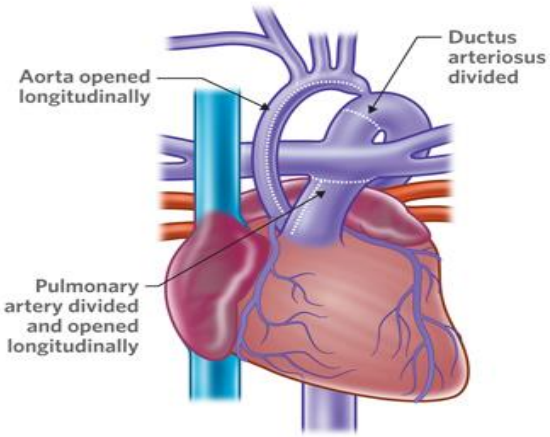
Low cardiac output

- Diminished pulses
- Poor colour
- Poor capillary refill time
- Decreased urine output
- Necrotising enterocolitis (NEC)

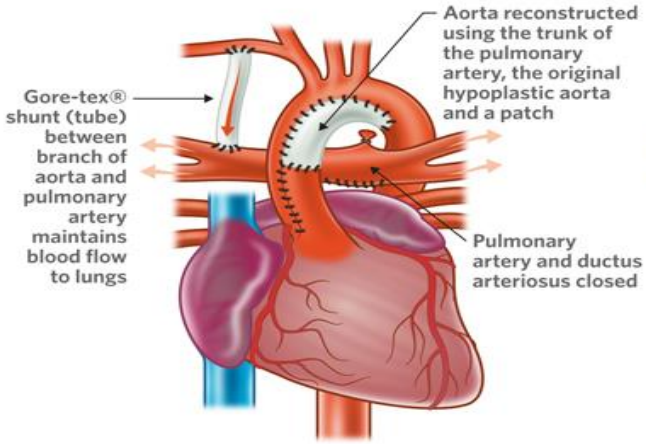
What are we going to do?

- Cardiac catheter- ballooning of Aortic Valve (AS)
- Surgical repair – Coarctation of Aorta (CoAo), Interrupted Aortic Arch (IAA), Aortic Valvotomy for AS
- Palliative surgery – Damus-Kaye Stansel Anastomosis or Norwood Procedure
- Single Ventricle Home Monitoring Programme if palliative surgery. Open access at DGH. "Fragile" babies.
- ? Palivizumab if under 2 years old

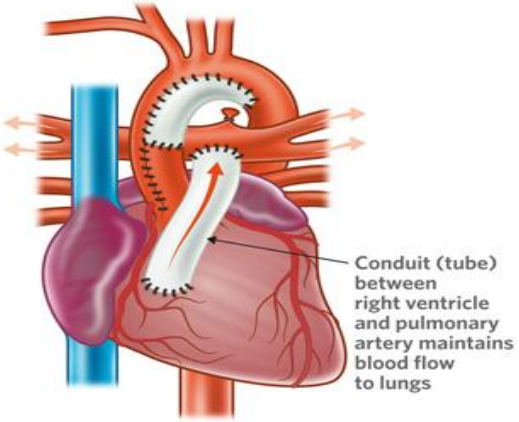
Norwood operation



Pre-operative anatomy

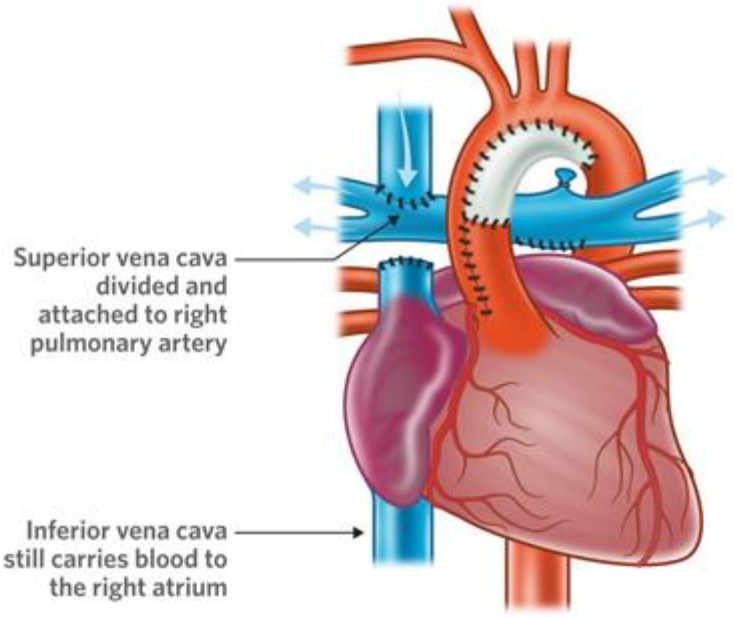


Norwood operation

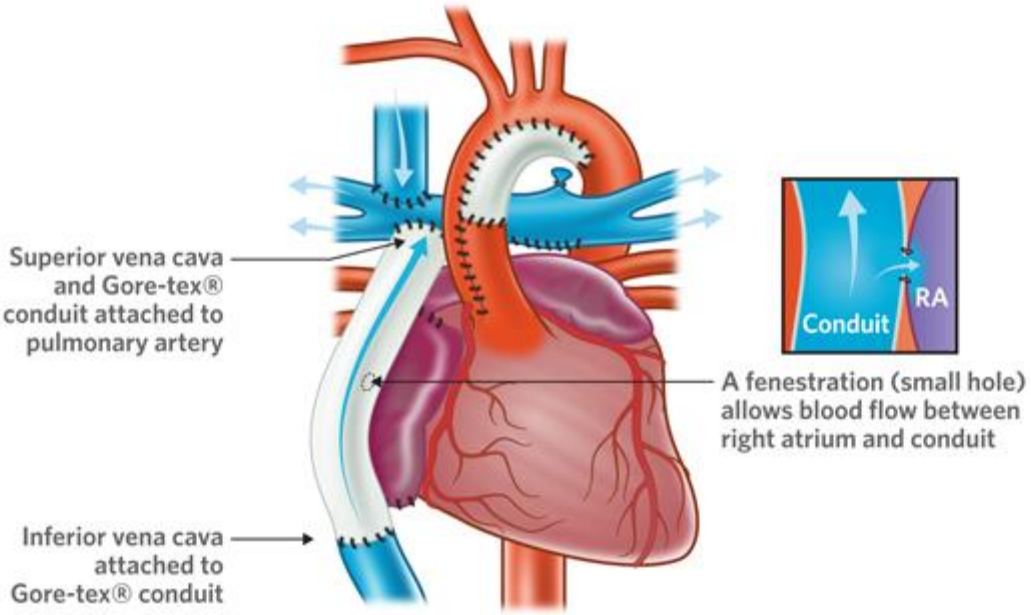


Norwood operation (Sano)

Bidirectional cavo-pulmonary connection (BCPC)



Fontan operation



Fontan operation with extracardiac conduit


We're Done.
Questions?
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