Care of the Child with Congenital Heart Disease

presentation by

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Learning Outcomes

Following this session, you will be able to:

- Name some common facts about CHD
- Describe anatomy and physiology of foetal circulation and foetal shunts
- Describe changes in circulation at birth
- Label a diagram of the key structures of the normal heart and blood vessels
- Name some common facts about CHD
- Discuss duct dependant CHD lesions
- Have a brief understanding of single ventricle pathway and stages of operations
- Discuss Assessment and management of a child with a cardiac defect
- Have a greater understanding of the collaborative role of specialist nurse and community teams in caring for patients with CHD

Incidences of CHD – some facts

- CHD is the most common congenital anomaly
- Each day, around 13 babies in the UK are diagnosed with congenital heart disease
- **CHD** happens in about 8 babies out of every 1,000 (0.8%)
- Before the BHF was founded in 1961, the majority of babies born with congenital heart disease died before their first birthday. Now, thanks to research, research has helped to turn this around, with 8 out of 10 babies surviving to adulthood. (source – British Heart Foundation)

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CHD Statistics and Research

- One in 125 babies is born with Congenital Heart Disease (CHD)
- More than 6,000 babies are born with major CHD each year in the UK
- In around 33% of cases, the CHD is detected prenatally
- Around 1,000 newborns leave UK hospitals each year with undetected CHD
- CHD is the most common congenital anomaly
- Up to 15% of CHD may remain undiagnosed at death
- More than 300 infants die every year in the UK from CHD
- CHD accounts for up to 12 % of all infant deaths – 1 in 10
- When detected, we know that many heart conditions can be treated – surgery survival rates are very high and 80% of babies born with a congenital heart defect survive past 16 years of age. The NHS has some of the world's leading heart surgeons and doctors, and units around the UK are achieving remarkable things every day.
- (information source:- Tiny Tickers website CHD Statistics and Research - Tiny Tickers)

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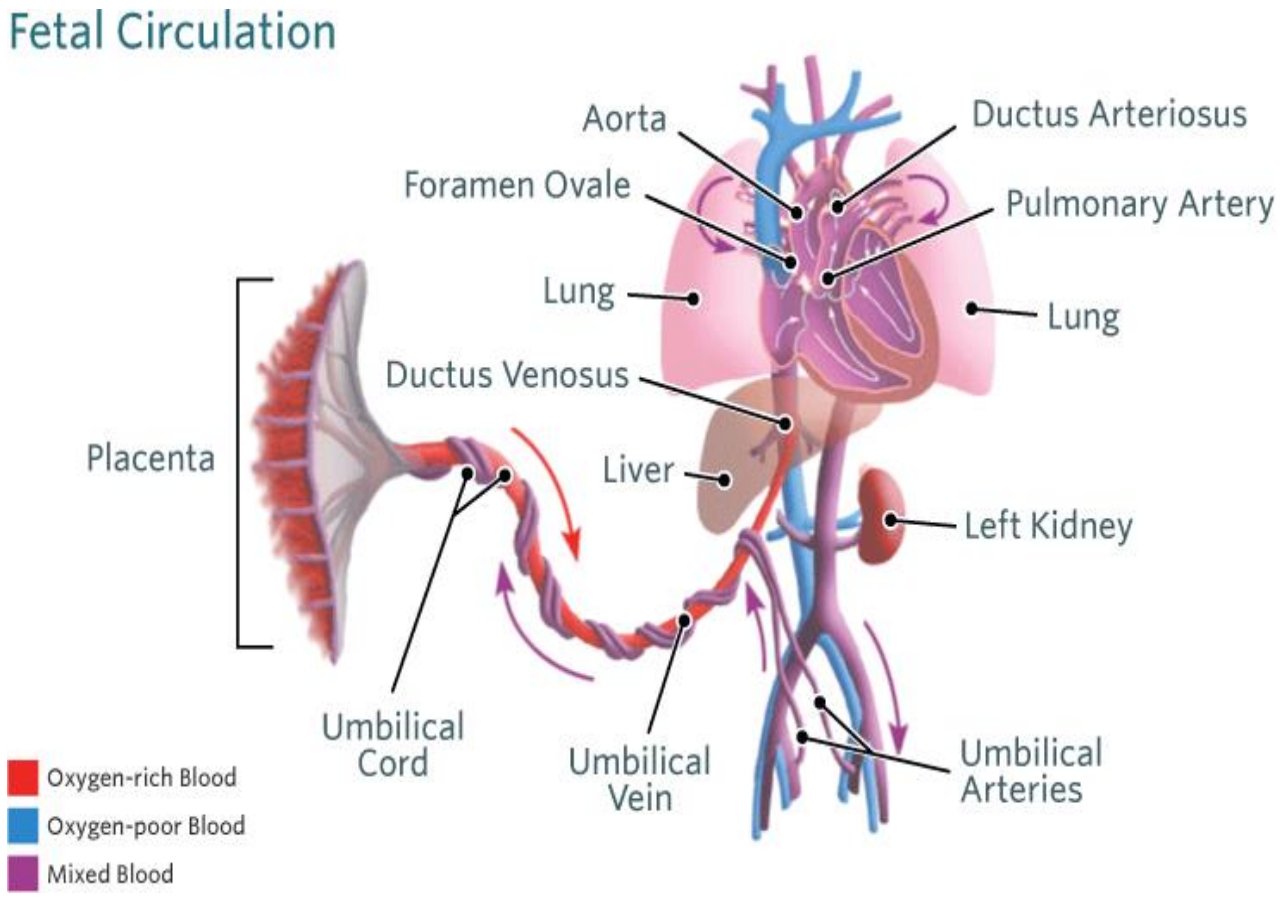
Foetal Circulation



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How does the foetal circulatory system work?

During pregnancy, the foetal circulatory system works differently than after birth:

- The fetus is connected by the umbilical cord to the placenta, the organ that develops and implants in the mother's uterus during pregnancy.
- Through the blood vessels in the umbilical cord, the fetus receives all the necessary nutrition, oxygen, and life support from the mother through the placenta.
- Waste products and carbon dioxide from the fetus are sent back through the umbilical cord and placenta to the mother's circulation to be eliminated.

Foetal Shunts

- 1. Ductus Venosus
- 2. Foramen Ovale
- 3. Ductus Arteriorus

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1st Shunt - Ductus Venosus

- From the placenta the blood travels via the umbilical vein toward the liver.
- At the liver it meets the **1st foetal shunt- the Ductus Venosus.** Some blood will be sent to the liver but the majority of the blood bypasses the liver and continues on to the Inferior Vena Cava

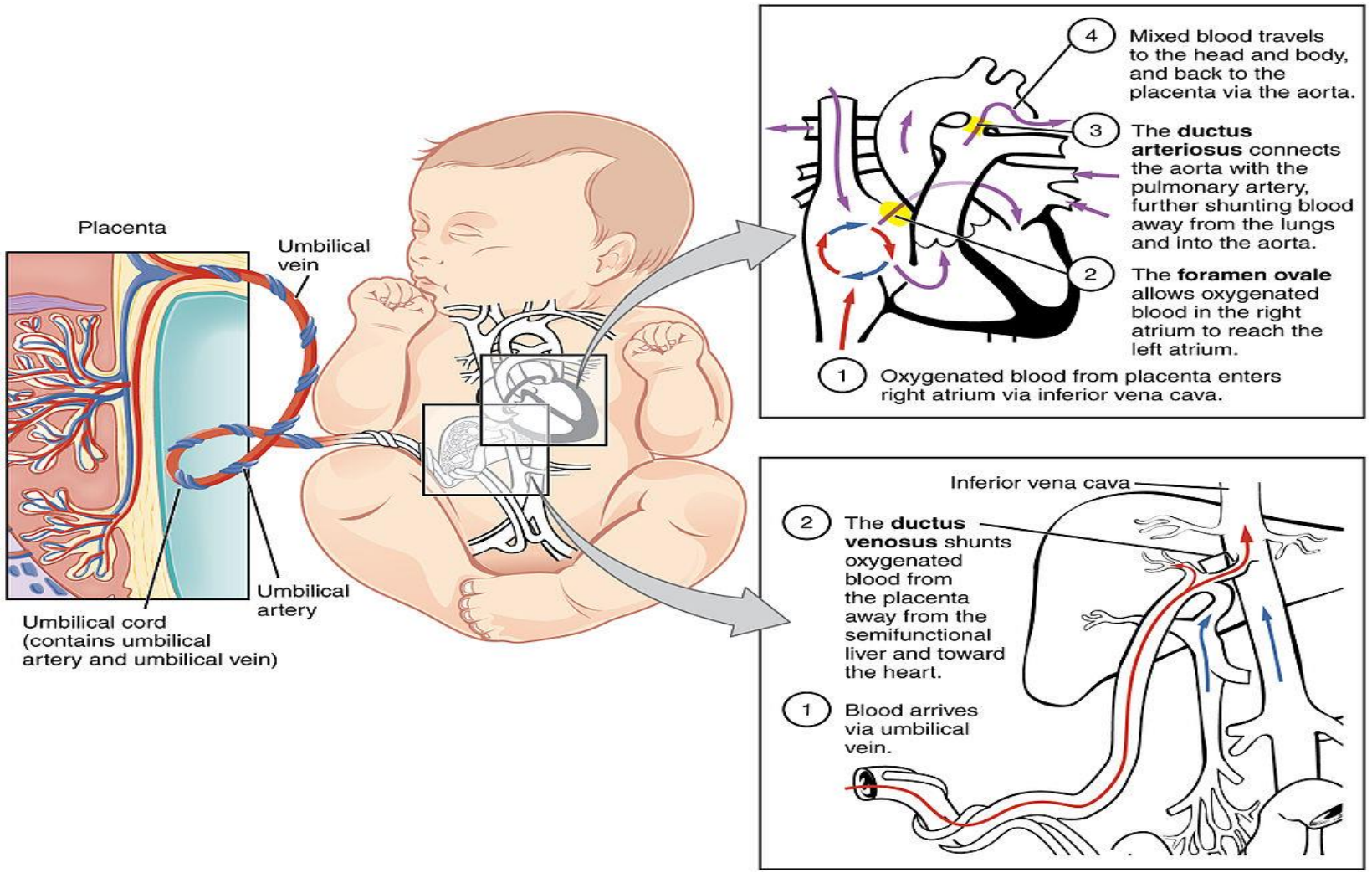
2nd Shunt - Foramen Ovale

The **2nd shunt** is an atrial communication called the **Fore man Ovale**.

- Blood flows from right to left at atrial level through this communication as in the foetal heart the right sided pressures are higher than the left.
- This is due to the foetal lungs being filled with fluid creating a high pulmonary vascular resistance (PVR) and therefore higher right sided pressures.
- This blood then continues down the descending aorta to the rest of the circulatory system and organs.

3rd Shunt - Ductus Arteriosus

- The patent ductus arteriosus (PDA) provides a shunt between the pulmonary artery and aorta. The PDA is kept **patent** in utero by Prostaglandin produced by the mother.
- The blood, which has travelled through the PDA, then joins the blood in the descending aorta. This blood is therefore also delivered to the tissues and organs.
- The de-oxygenated blood filled with fluid waste products returns to the placenta via the umbilical artery to be removed by the mother.



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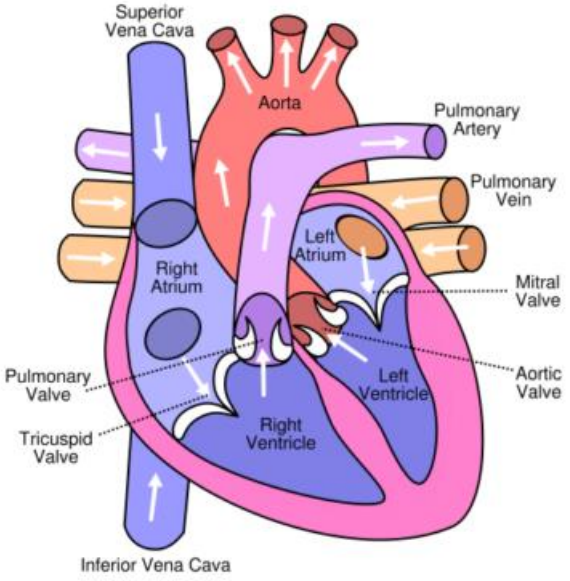
Changes to Circulation at Birth

- At birth, the umbilical cord is clamped and the baby no longer receives oxygen and nutrients from the mother.
- With the first breaths of life, the lungs begin to expand. As the lungs expand, the alveoli in the lungs are cleared of fluid. An increase in the baby's blood pressure and a significant reduction in the pulmonary pressures reduces the need for the **ductus arteriosus to shunt blood.** These changes promote the closure of the shunt. These changes increase the pressure in the left atrium of the heart, which decrease the pressure in the right atrium. The shift in pressure stimulates the **foramen ovale to close.**
- The **closure of the ductus arteriosus and foramen ovale** completes the transition of fetal circulation to newborn circulation.

Cardiac Anatomy

The right side of the heart gets blood from the body and pumps it to the lungs, while the left side does the opposite by getting blood from the lungs and pumping it to the rest of the body. The heart is divided into four chambers:

- The left atrium collects blood coming from the lungs;
- The left ventricle pumps blood to the body;
- The right atrium collects blood from the body;
- The right ventricle pumps blood to the lungs.



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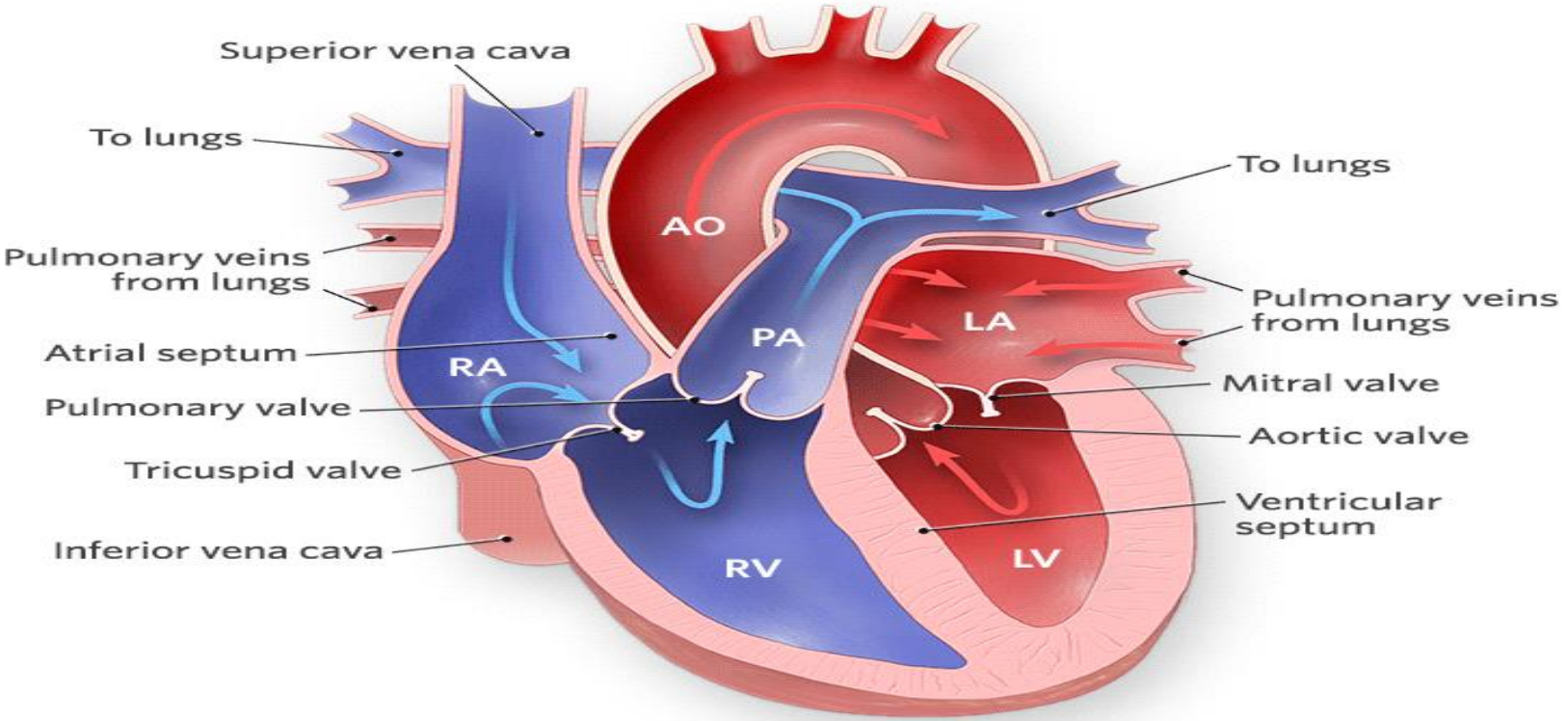
Normal Heart



Oxygen-rich blood
Oxygen-poor blood

AO: Aorta
LA: Left atrium
RA: Right atrium
PA: Pulmonary artery
LV: Left ventricle
RV: Right ventricle

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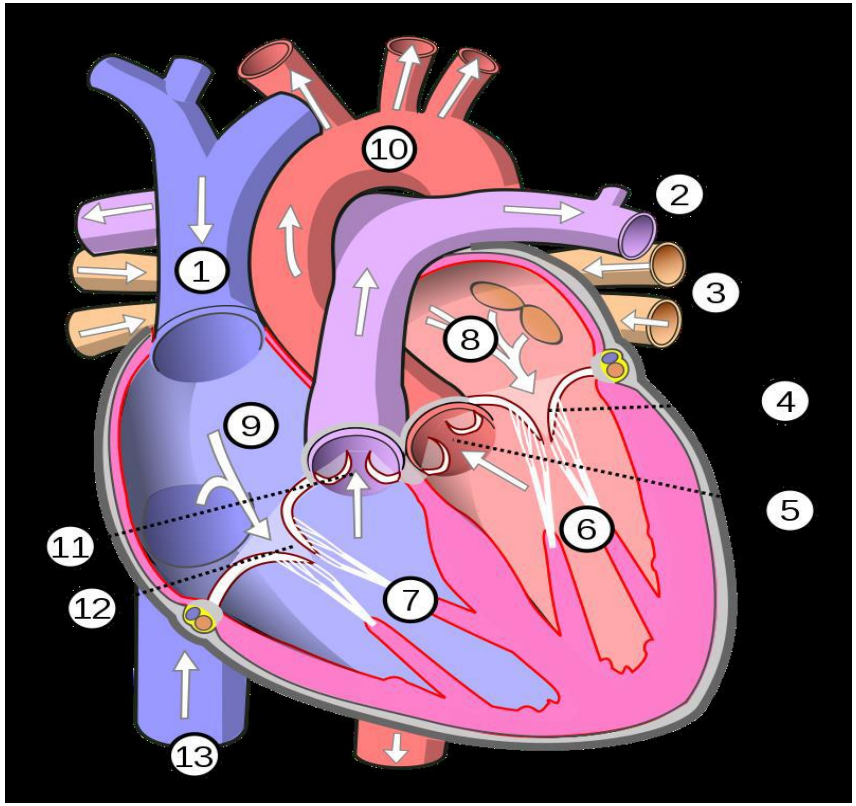
Structure of the Heart



Label this diagram of the heart and draw the direction of the blood flow. Identify whether the blood is oxygenated or deoxygenated

Aorta
Pulmonary Artery
Superior Vena Cava
Right Atrium
Right Ventricle
Left Atrium
Left Ventricle
Aortic Valve
Pulmonary Valve
Mitral Valve
Tricuspid Valve
Inferior Vena Cava
Pulmonary veins

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Blood Pressure

- Evaluate the carotid, brachial, femoral and pedal pulses to detect differences between sides and upper and lower extremities. If pulses are unequal, obtain four extremity blood pressures. A marked difference may be caused by coarctation of the aorta.
- Cuff size is critical. A cuff that is too narrow gives falsely high readings and too large a cuff may yield low readings.
- Comparison of upper and lower limb blood pressure (BP) is recommended by standard paediatric cardiology textbooks to aid the clinical diagnosis of coarctation of the aorta (CoA). A BP 20 mm Hg higher in the arms than in the legs in neonates with CoA or interrupted aortic arch is widely reported, although there is concern about the possibility of false negative testing. Suspicion of a cardiac defect will require ultrasound to exclude.

Assessment of child with CHD

Assessment of cardiac child can be is similar to that of any sick child but need to look for signs for any potential deterioration

- Changes in colour
- Change in saturations (higher or lower)
- Change in respiratory rate
- Change in murmur
- Weak pulses
- Irritability/difficult to console
- Increased sleepiness
- Sweating, especially when feeding
- Poor weight gain

Duct Dependant CHD

DUCT DEPENDANT PULMONARY CIRCULATION (right heart obstruction)	DUCT DEPENDANT SYSTEMIC CIRCULATION (left heart obstruction)
Pulmonary Atresia with intact ventricular septum	Interrupted aortic arch
Pulmonary Atresia with VSD	Critical Aortic stenosis
Pulmonary Atresia with single ventricle	Hypoplastic Left Heart syndrome
Critical pulmonary stenosis	Critical coarctation of aorta
Rare TOF with critical RVOT	
Severe Ebstein anomaly	
<u>Admixture lesions</u> Transposition of great arteries with intact ventricular septum	

Single Ventricle Defects – What are they?

Rare disorders affecting one lower chamber of the heart. The chamber may be smaller, underdeveloped, or missing a valve.

- **Hypoplastic Left Heart Syndrome (HLHS)** - An underdeveloped left side of the heart. The aorta and left ventricle are too small and the holes in the artery and septum did not properly mature and close.
- **Pulmonary Atresia/Intact Ventricular Septum** - The pulmonary valve does not exist, and the only blood receiving oxygen is the blood that is diverted to the lungs through openings that normally close during development.
- **Tricuspid Atresia** - There is no tricuspid valve in the heart so blood cannot flow from the body into the heart in the normal way. The blood is not being properly refilled with oxygen so it does not complete the normal cycle of body–heart–lungs–heart–body.

Single Ventricle Pathway

- This is a series of operations to create a connection between the veins returning low-oxygen blood to the heart and the pulmonary artery. The goal is to allow the right ventricle to pump only oxygenated blood to the body and to prevent or reduce cyanosis (lower than normal blood oxygen levels). The are usually three staged operations to achieve this during the first few years of life.
- Many patients go onto to require a heart transplant if they are suitable, including many patients with HLHS. Although it can provide the infant with a heart that has normal structure, the infant will require life-long medications to prevent rejection. Many other transplant-related problems can develop following transplant and the patient will require lifelong medications and care.

What is a BT shunt?

- A Blalock-Taussig (BT) shunt is a small tube that connects the arterial circulation to the pulmonary circulation in order to get more blood to the lungs. This is the first in a series on operations to correct complex congenital heart defects. It is only a temporary fix, as the tube is only a certain size, but it allows the baby to grow and prepare for his or her next operation. It is not a long-term solution. A growing child will “outgrow” the shunt, eventually needing more blood flow to the lungs to meet the body’s oxygen needs.
- BT shunts can be used to treat conditions such as pulmonary atresia, pulmonary stenosis, Tetralogy of Fallot, hypoplastic left heart syndrome and tricuspid atresia.

BT Shunt – the operation

- The Blalock-Taussig (BT) shunt mimics the role of the ductus arteriosus, meaning it allows blood to flow from a major artery through a connection to the pulmonary artery. Not only does this allow more blood to be oxygenated by the lungs, it also encourages the pulmonary arteries to grow, making the next surgery easier.
- A BT shunt is tiny, measuring less than 5 millimeters (0.20 inches) in diameter. A surgeon attaches the two ends of the shunt to a major blood vessel, such as the subclavian artery, and to the pulmonary artery. The high-pressure arterial system will force blood through the BT shunt to the lungs to pick up more oxygen. The shunt helps to return healthy color to a child whose skin, lips or fingernails may have appeared blue (cyanosis) because of low oxygen levels in the blood. But even with a BT shunt, children likely will not have normal oxygen saturation levels and can appear blue, or cyanotic. The child's oxygen saturation levels after a BT shunt will depend on the type of heart defect that is being repaired.

Stenting of PDA

- Stenting of the patent ductus arteriosus (PDA) is **the primary treatment for patients with duct-dependent pulmonary circulation**. It serves as a temporary bridge for later surgical repair [1]. It is considered a better alternative to conventional shunt surgery, Why are PDA stents done?
- Stenting of the PDA is a better alternative to modified BT shunt as it **eliminates problems associated with thoracotomy in neonatal period and long-term problems of scarring**, which may cause major difficulty in future definitive surgery.
- PDA stenting is an effective method of palliation for patients with duct-dependent pulmonary circulation. It has low morbidity and mortality rates. Stenting difficult ducts have become more feasible with evolving materials and techniques.

Hypoplastic Left Heart Syndrome

What is it?

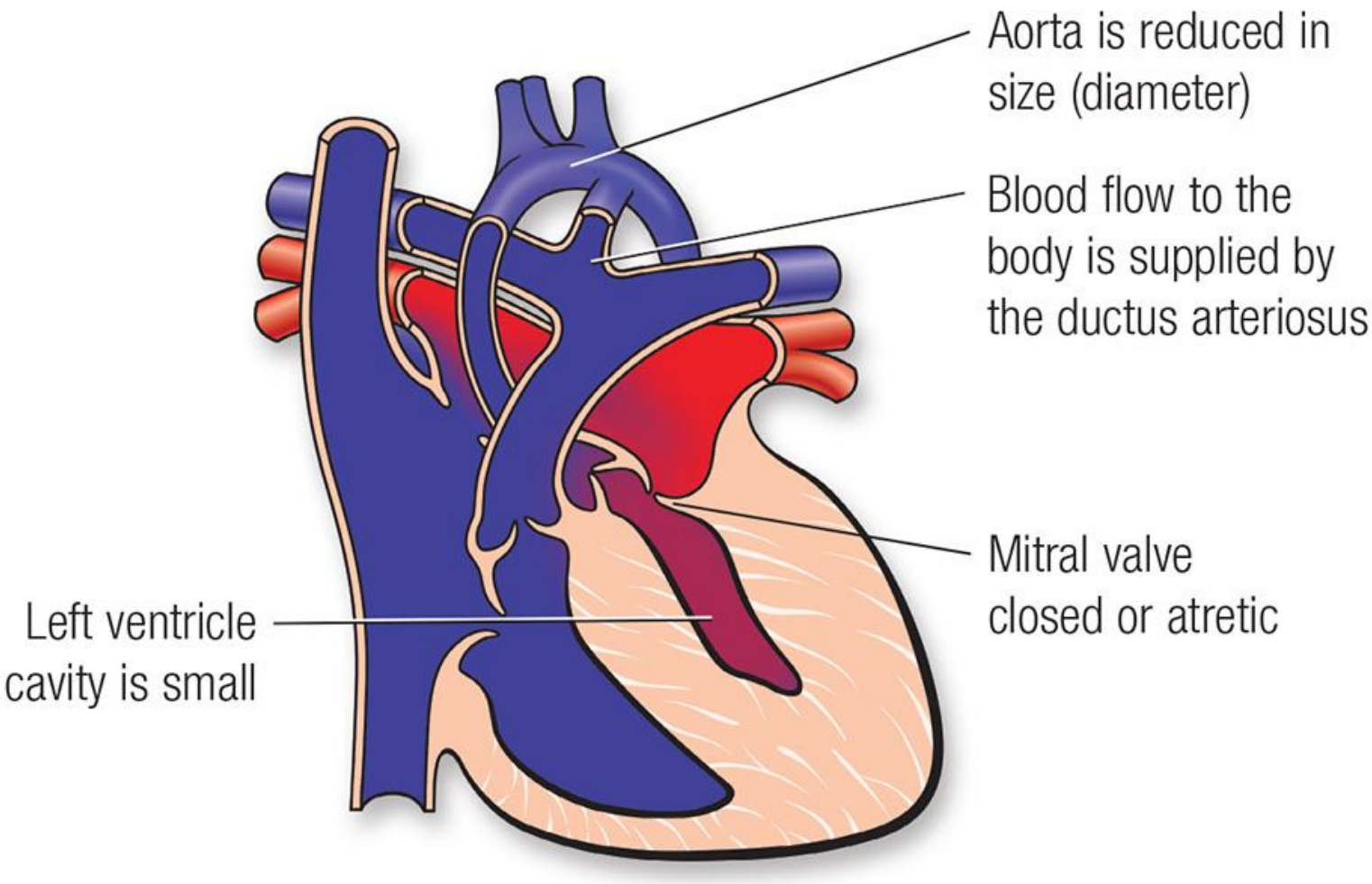
- In hypoplastic left heart syndrome (HLHS), the heart's left side — including the aorta, aortic valve, left ventricle and mitral valve — is underdeveloped.

What causes it?

- In most children, the cause isn't known. Some children can have other heart defects along with HLHS.

How does it affect the heart?

- In HLHS, blood returning from the lungs must flow through an opening in the wall between the atria (atrial septal defect). The right ventricle pumps the blood into the pulmonary artery and blood reaches the aorta through a patent ductus arteriosus (see diagram).



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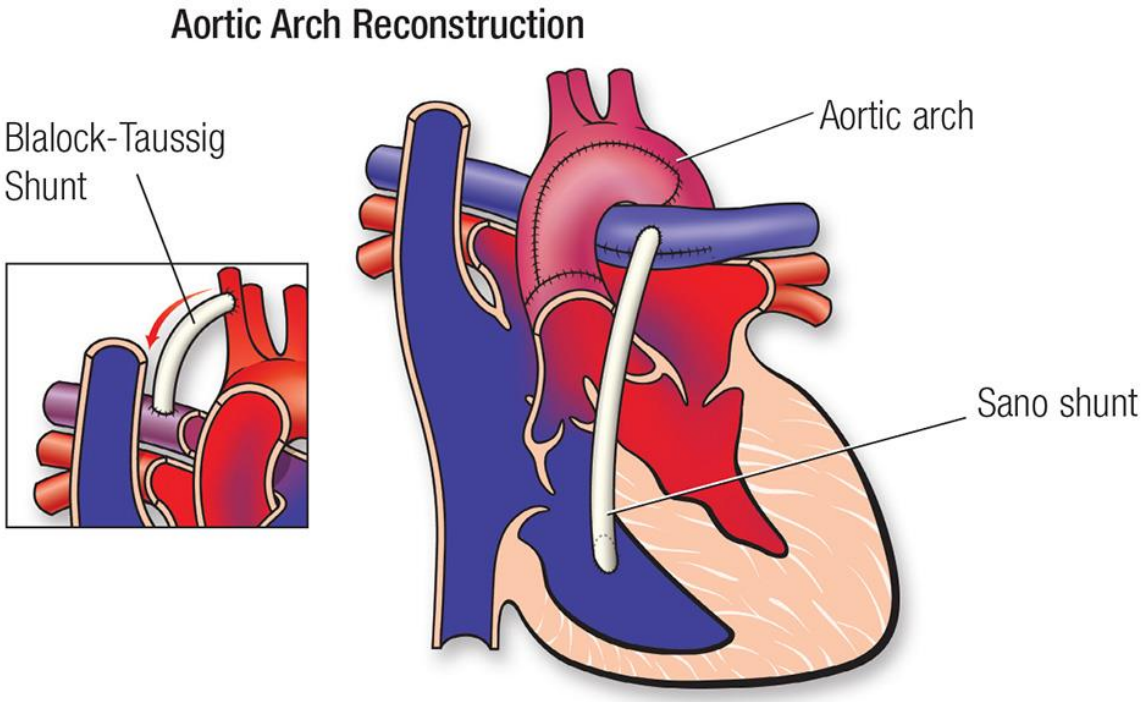
1st stage – Norwood Procedure



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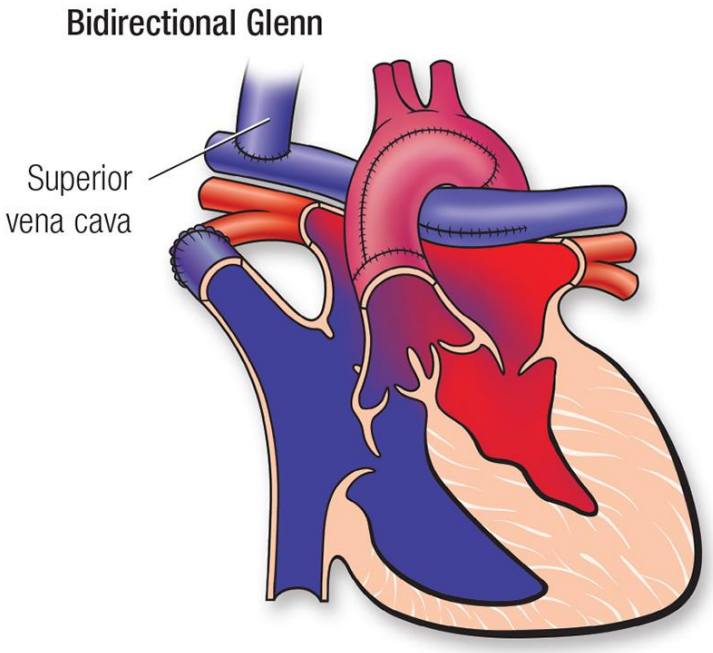
2nd Stage – Bi-directional/Glenn shunt



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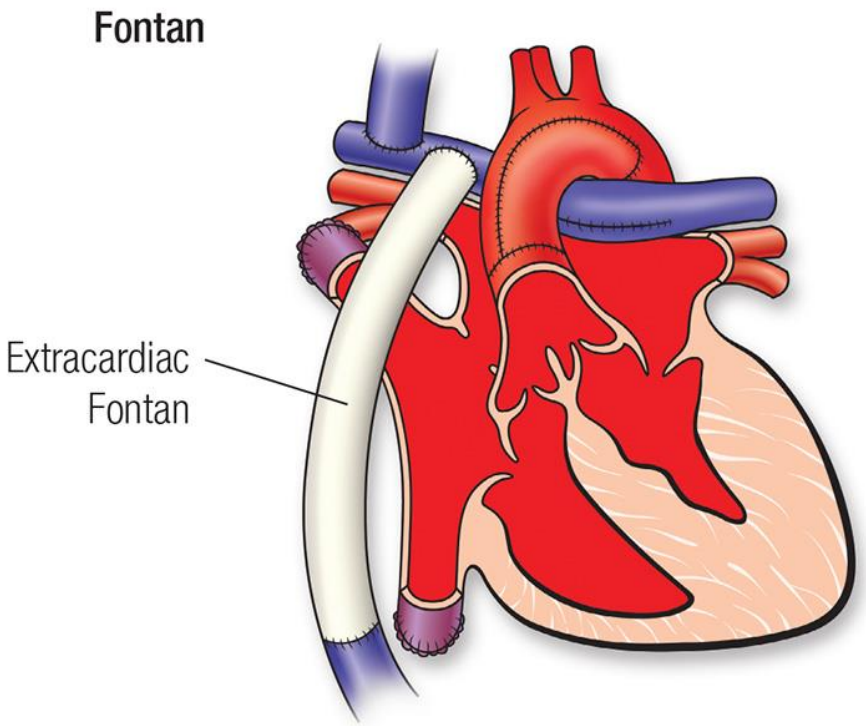
3rd Stage – Fontan Procedure



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Norwood Procedure

- The Norwood procedure is the first of three surgeries required to treat hypoplastic left heart syndrome (HLHS). Because the left side of the heart can't be fixed, the series of HLHS surgeries rebuilds other parts of the heart to redirect the blood flow.
- Hypoplastic left heart syndrome once was considered inoperable. Advances such as the three procedures currently used to treat the condition give more babies the chance to live full lives.
- This first step in the series of surgeries is performed in the baby's first or second week of life.

Goals of Norwood Procedure

- Build a new aorta.
- Direct blood from the right ventricle through the new aorta and on to the rest of the body.
- Direct the right ventricle to pump blood to the lungs until the next surgery.

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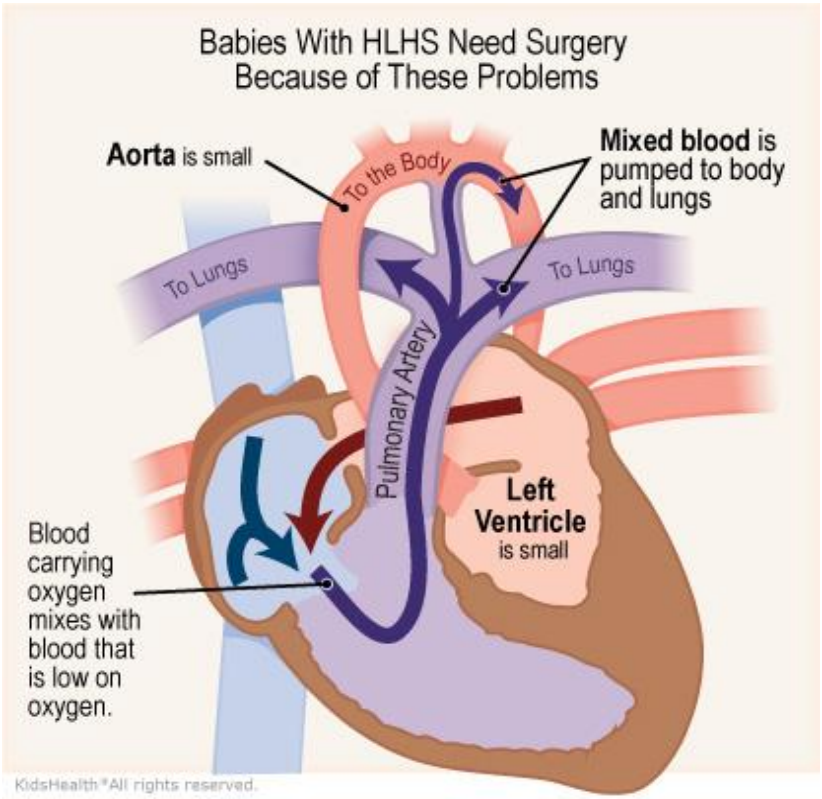
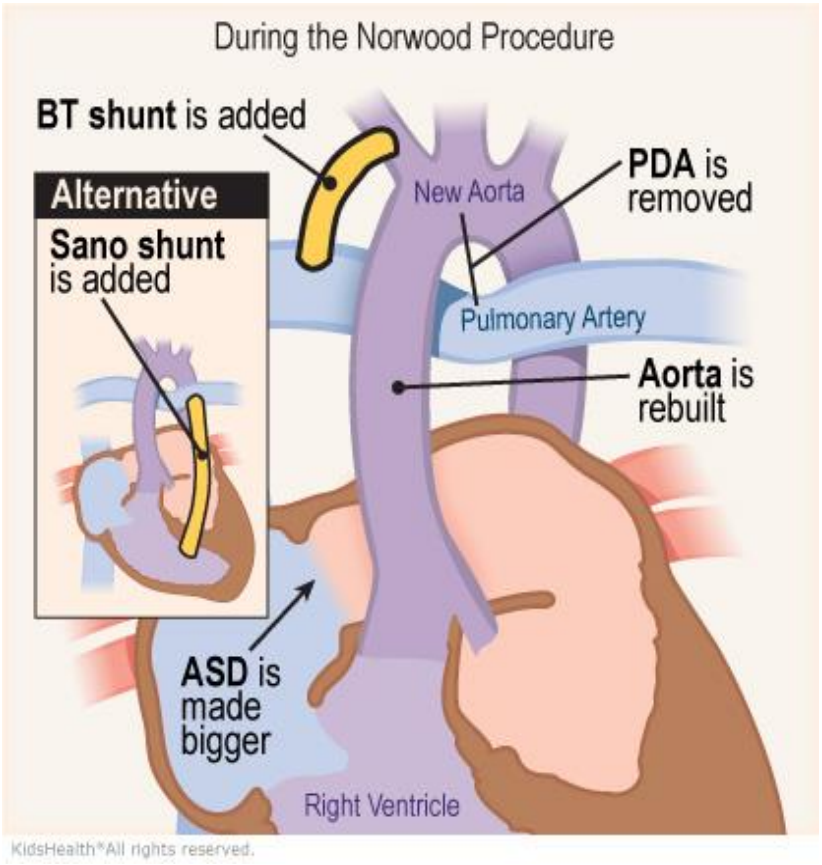
Norwood Procedure continued

- **Building a new, larger aorta.** The bottom part of the pulmonary artery is joined with the baby's weak, undeveloped aorta. This new aorta, or neoaorta, becomes the path for blood from the right ventricle to the body.
- Because the bottom of the pulmonary artery is used to make the new aorta, a new path from the heart to the lungs has to be built. A round tube (shunt) routes blood either from a large vessel off the aorta (with a Blalock-Taussig, or BT shunt) or directly from the right ventricle (with a Sano shunt) to the pulmonary artery.

What a Norwood operation does





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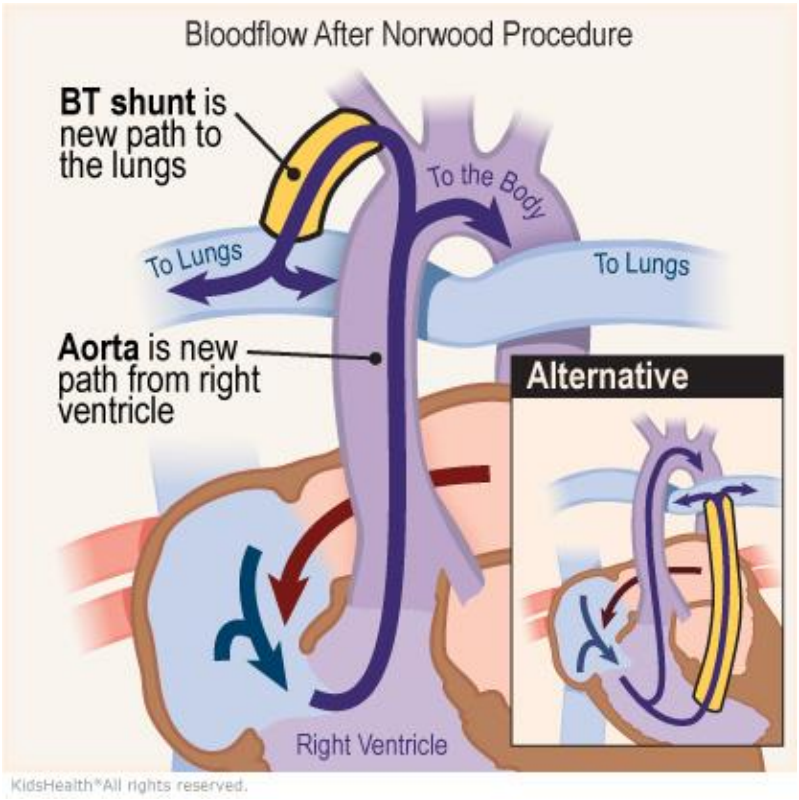
Blood flow after Norwood Procedure



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Care of child between Stage 1 & Stage 2

While the baby is recovering, the nursing team teaches parents how to care for their baby at home. Babies usually can go home when they are feeding well, growing well and gaining weight.

- At home, the baby needs to be watched closely. If you are caring for a baby who has had this procedure, stay in close contact with the hospital and community nursing team and keep an eye on:
- Weight gain and growth
- Oxygen levels

Call the CCNS team right away if the baby:

- Has difficulty feeding
- Has difficulty breathing
- Seems very irritable
- Just doesn’t seem quite right
- Has lower oxygen levels than usual (goals will be given on discharge of what saturations are acceptable)

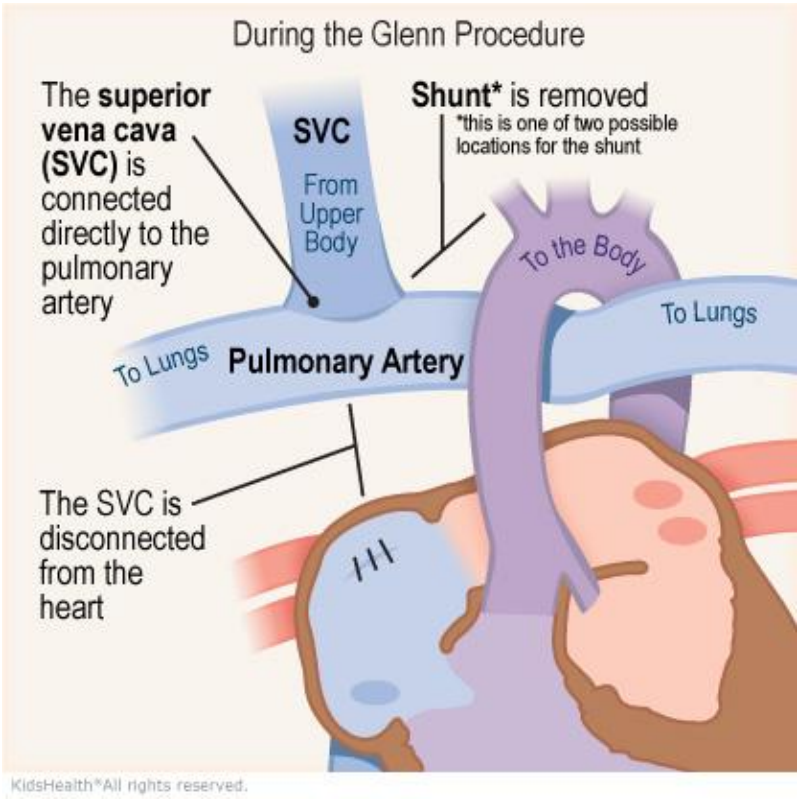
Glenn Procedure

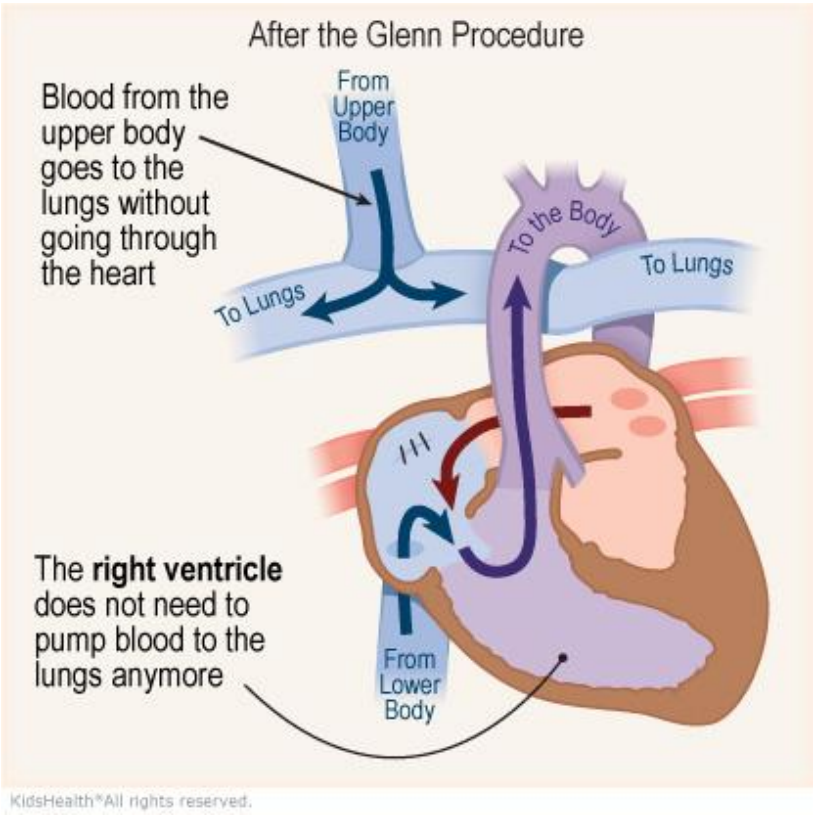
The second surgery is called the Glenn procedure. It's usually done when a baby is 4 to 6 months old.

Goals of a Glenn Procedure

- Make blood from the upper part of the body (head, neck and arms) go directly to the lungs. This lets the blood pick up oxygen without passing through the heart.
- Take some of the extra work away from the single ventricle. Until now, the only working ventricle in a baby with an HLHS single ventricle lesion has been pumping blood to both the lungs and the body.
- To redirect blood flow from the upper body to the lungs. Surgeons disconnect the superior vena cava (the vein that brings blood back from the upper part of the body) from the heart and reattach it to the pulmonary artery.
- If present, they also remove the shunt that may have been placed during an initial operation, such as the Norwood procedure. A shunt is no longer needed because blood from the upper body will now go to the pulmonary artery and on to the lungs. Also, the single ventricle now has a single job: pumping blood to the body.

During and After Glenn Procedure





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Fontan Procedure

The Fontan procedure is the final surgery in a series of operations to help the heart function better in children born with complex single-ventricle conditions such as hypoplastic left heart syndrome (HLHS). The procedure doesn't cure the congenital heart defect. It allows the heart to work better in the absence of a second functioning ventricle.

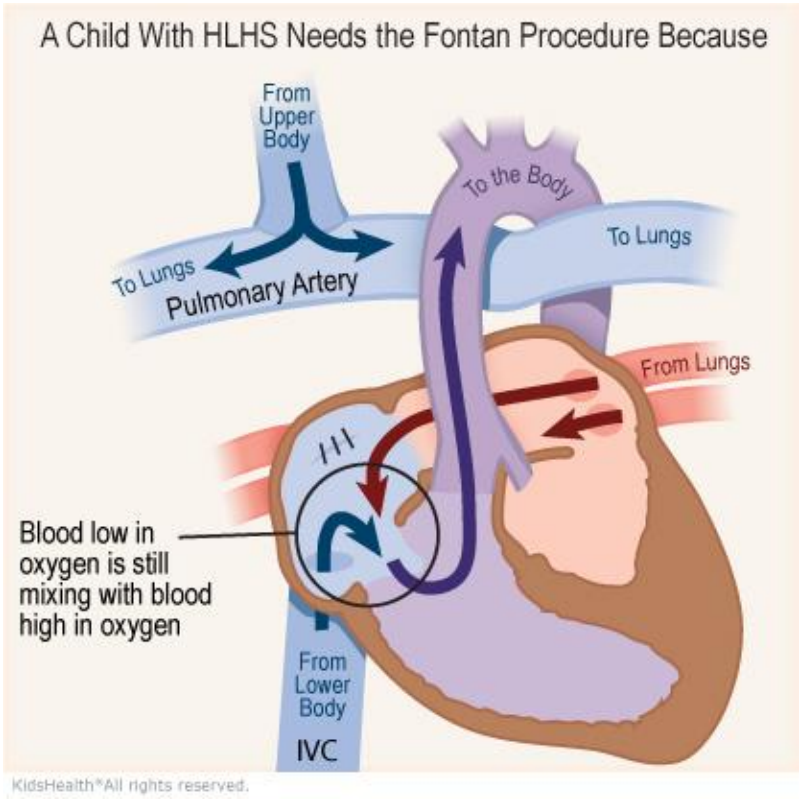
- Advances in treating babies born with just one ventricle have allowed more children to live full lives. HLHS was considered inoperable and fatal before surgeons developed the three-surgery series.
- Fontan procedure is done when a child is approx 18-36 months old.
- With just one ventricle, freshly oxygenated blood is diluted by mixing with deoxygenated blood, completing its trip throughout the body.
- The Fontan procedure reconfigures the veins to direct blood from the lower part of the body directly to the lungs. The route bypasses the heart, preventing deoxygenated blood from mixing with oxygenated blood.
- The aim of the Fontan procedure is to redirect blood flow so the heart isn't overworked and more oxygen-rich blood is distributed throughout the body.

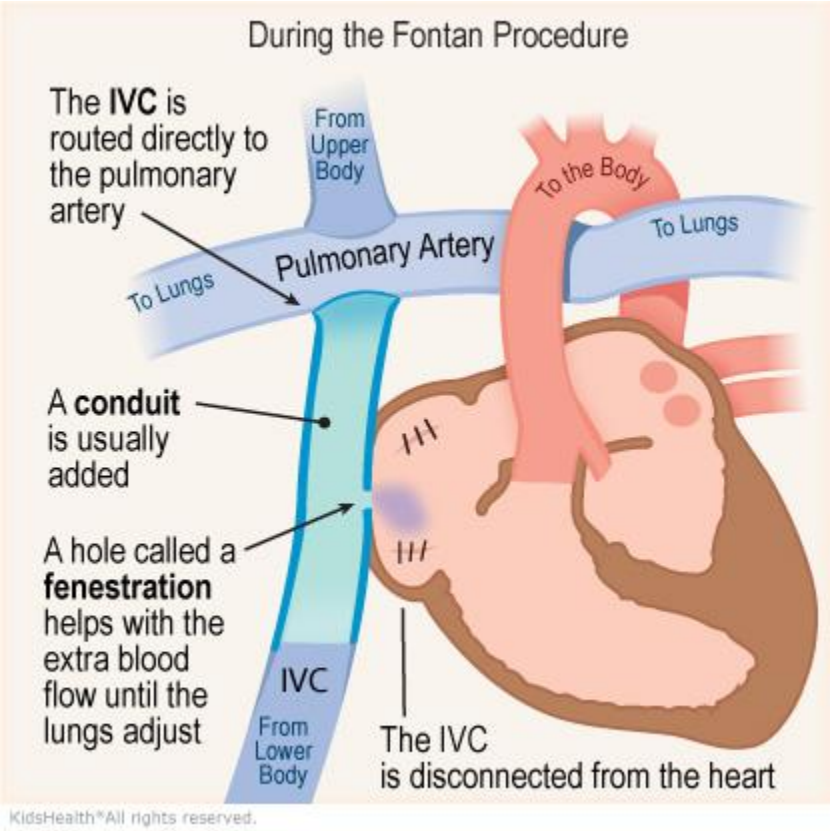
Fontan operation

Surgeons performing the Fontan procedure disconnect the inferior vena cava (the vein that brings blood back from the lower part of the body) from the heart and connect it to the pulmonary artery.

- Often, a small hole, called a fenestration, is created between the conduit and the right atrium. This allows some blood to flow directly back to the heart. It acts as a relief valve as the lungs get used to the extra flow from the lower part of the body. The fenestration can be closed later in life with a minimally invasive cardiac catheterization procedure.
- With all the connections made, blood from the **lower body** now goes to the pulmonary artery, and then to the lungs, without passing first through the heart. The patient's single ventricle now pumps oxygen-rich blood from the lungs out to the body and all the way back through the lungs.
- With the high- and low-oxygen blood no longer mixing in the heart because of the defect, more oxygen gets out to the body.
- Children who have the Fontan procedure usually spend one to two weeks in the hospital to recover.

Before and During Fontan procedure





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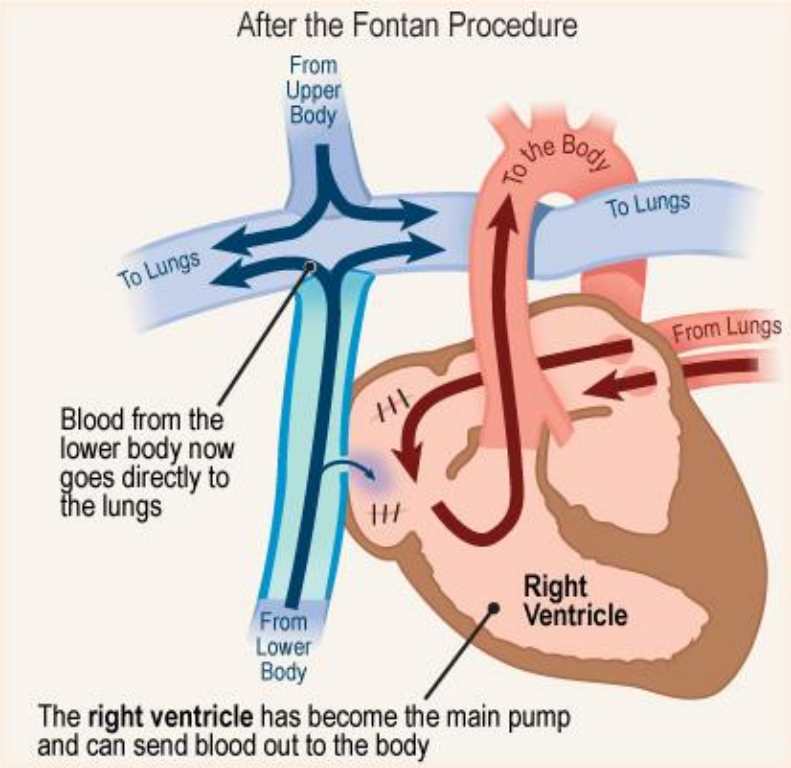
After the Fontan procedure



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Living with Fontan circulation

While the Fontan procedure and other treatments have saved lives, children with this unique blood circulation pattern are at risk for complications in other parts of the body, such as the digestive tract and liver complications as they grow into adulthood.

- Patients are closely monitored as they grow and to catch developing complications.
- Even in hemodynamically well compensated Fontan patients, **cardiac output is impaired** resulting in chronic subacute cardiac insufficiency that may eventually lead to end organ damage particularly affecting the liver and the kidneys.
- Some think that most single ventricle hearts will not work well after **30 to 40 years**. Improvements in surgical technique and medical care may increase this age significantly.
- Patients with failed Fontan circulation make up an increasing proportion of those presenting for heart

Complex diagnosis

- Hypoplastic left heart variant
- Dysplastic mitral valve with single papillary muscle and small orifice
- Small aortic valve
- Hypoplastic aortic arch Functionally bicuspid pulmonary valve with mild flow acceleration
- Small fistula from branch RPA to RA
- This patient underwent Norwood procedure at 5 days old

Palliative Care – key points

Surgical palliation is used when a heart defect cannot be fully corrected but can be modified to improve a condition.

- Palliative care has generally been considered as an end-of-life treatment but can also be provided at the same time as cure-directed treatments.
- Many children who had surgical palliation are now young adults being followed by doctors who specialize in adults with congenital heart disease. How long their surgical palliation will extend their lives can only be determined through excellent follow-up in specialized cardiac units.

Joint care between hospital and community services

- Greater collaboration between CCNS, Palliative care teams and community nursing teams.
- What can we do to improve quality of care for this cohort of patients? - Questions and answers?

Study of CHD Patients requiring Palliative care

- **Design** A national survey including closed-ended and open-ended questions as well as clinical scenarios designed to capture referral practices, attitudes towards palliative care, confidence delivering key components of palliative care and perspectives on for whom to provide palliative care. Responses to closed-ended questions and scenarios were analysed using descriptive statistics. Open-ended responses were analysed thematically.
- **Participants** Paediatric cardiac and palliative care professionals caring for children with complex cardiac conditions in the UK.
- **Results** 177 professionals (91 cardiac care and 86 palliative care) responded. Aspects of advance care planning were the most common reasons for referral to palliative care. Palliative care professionals reported greater confidence than cardiac colleagues with such discussions. Clinicians agreed that children with no further surgical management options, comorbid genetic disorders, antenatal diagnosis of a single ventricle, ventricular device in situ, symptomatic heart failure and those awaiting heart transplantation would benefit from palliative care involvement.

Resources

Tiny Tickers –British Heart Foundation (BHF)

Little Heart Matters (dedicated to those with single ventricle condition)

Somerville Heart Foundation

Children's Heart Federation

Many more charities and resources available online

References and Further resources

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