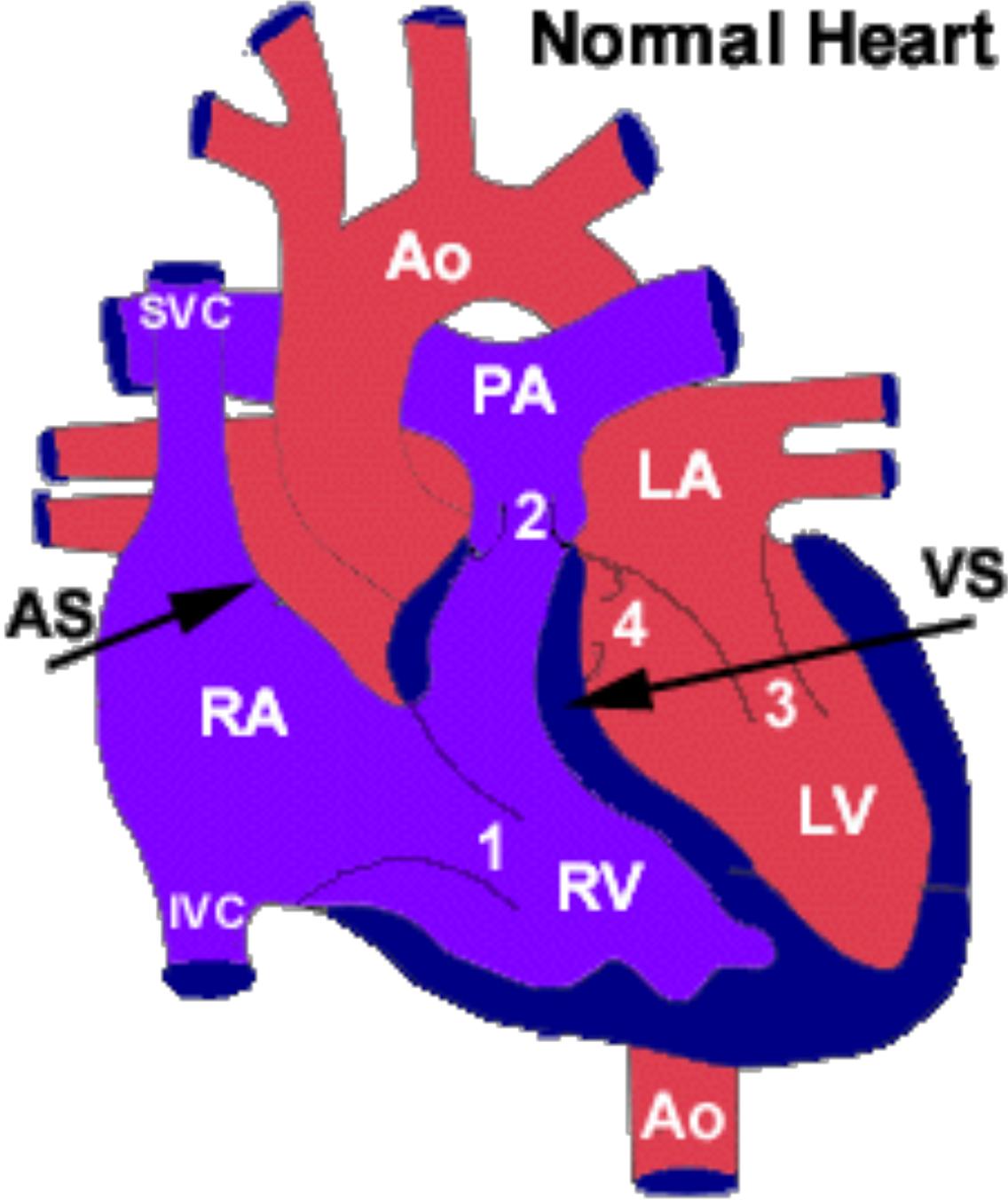


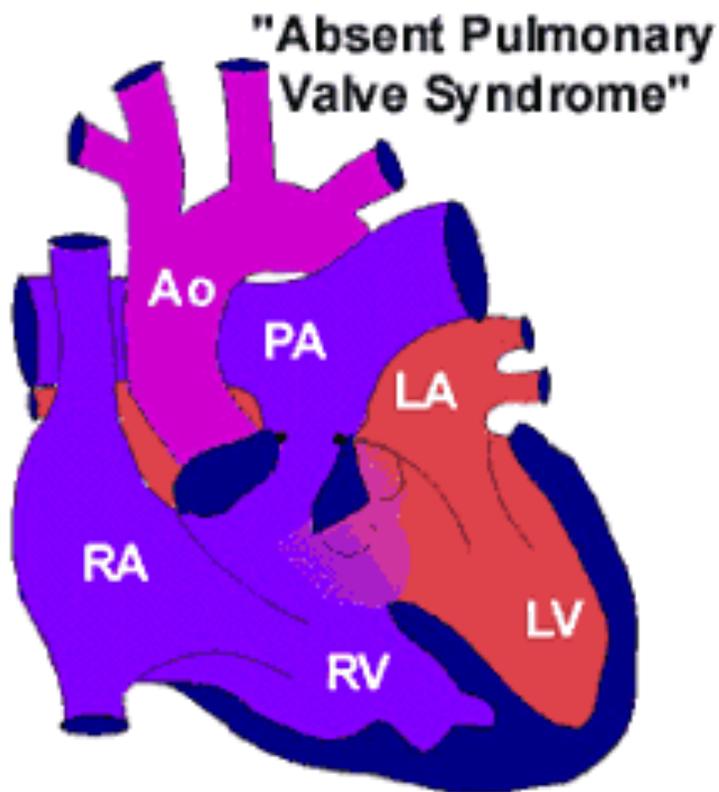
Normal Heart



Absent Pulmonary Valve Syndrome

- Fact sheet on Absent Pulmonary Valve Syndrome

In this condition, which has some similarities to Fallot's Tetralogy, there is a VSD with narrowing at the pulmonary valve. However the Pulmonary Valve leaflets (flaps) are rudimentary and non functional, resulting in severe Pulmonary Incompetence. The Pulmonary Arteries tend to be enlarged and may cause problems by compressing and narrowing air passages in the lungs, resulting in respiratory difficulty.

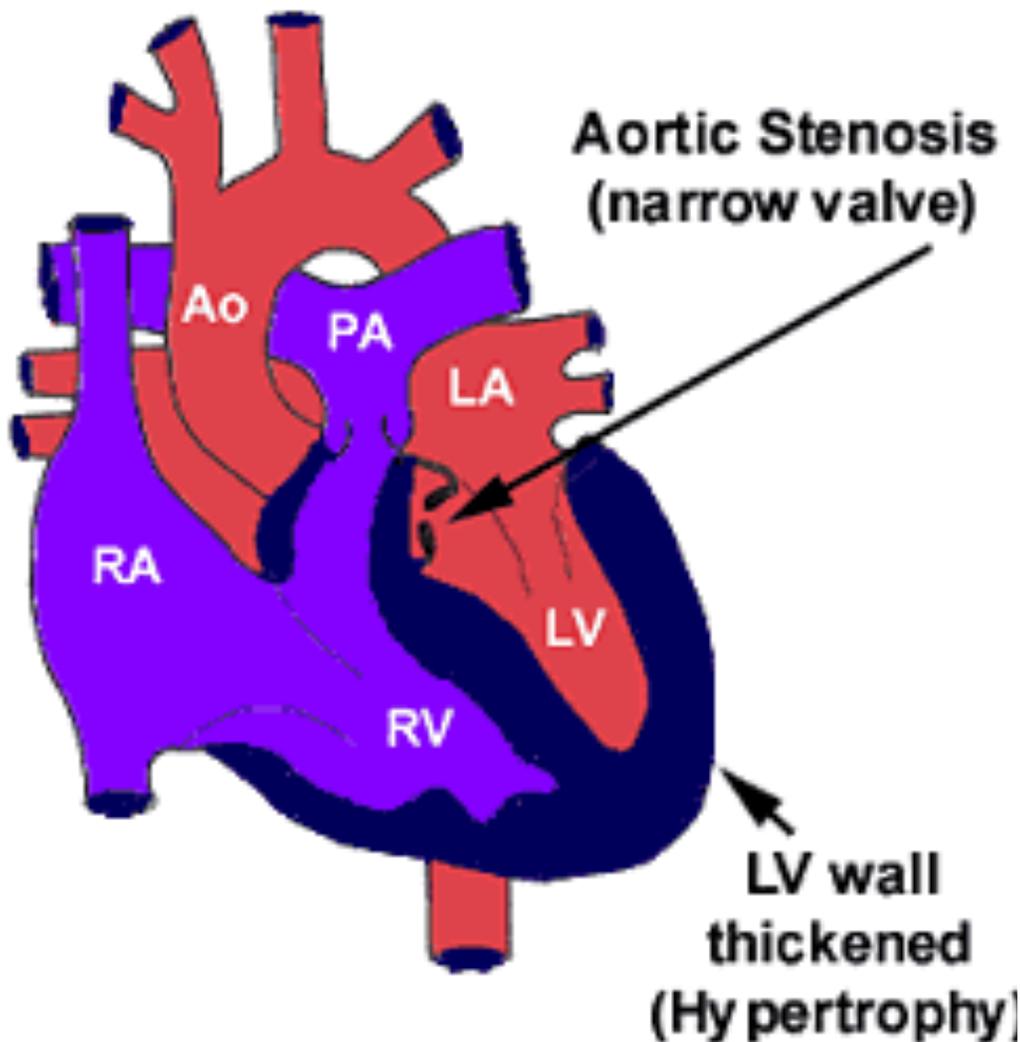


Aortic Stenosis (AS)

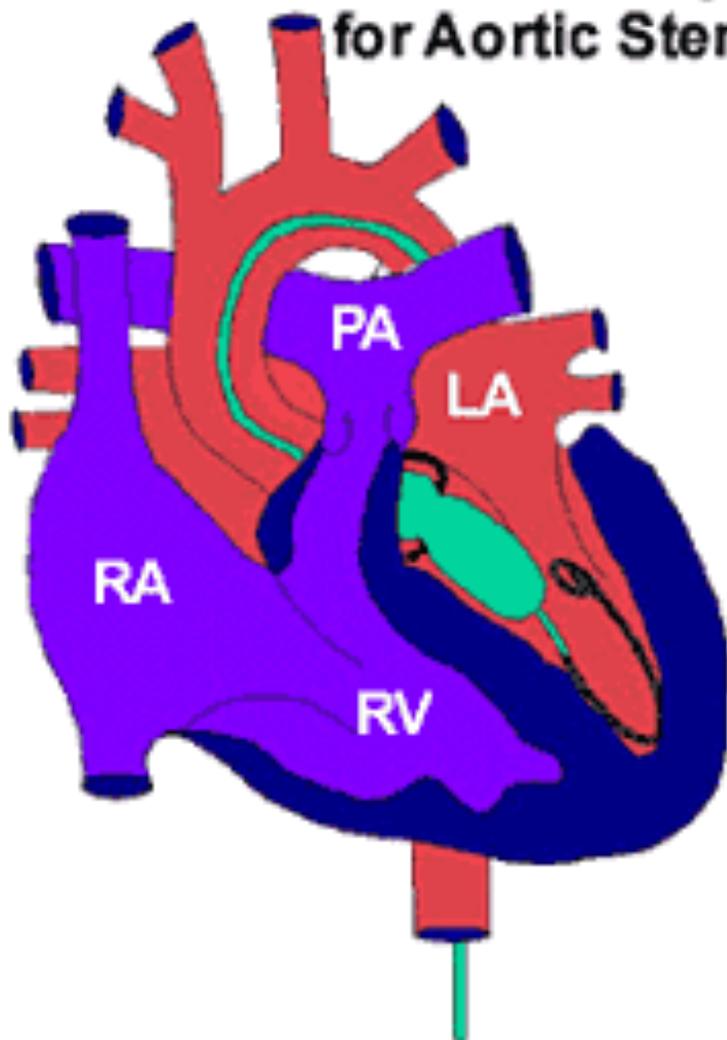
The Aortic Valve is thickened and narrowed leading to the development of abnormally high pressure in the left ventricle. The left ventricular wall becomes thickened ("Hypertrophied"). If the problem is severe it may require treatment, which usually involves surgery in younger patients, though it may be possible to stretch the valve with a balloon catheter (Balloon Valvuloplasty), in older children. The catheter is passed from an artery in the leg. When the tip is through the valve the balloon is inflated to open the valve. Treatment does not completely cure the problem and the valve sometimes tends to develop further problems with time, sometimes needing reoperation or further balloon stretching.

If the valve is severely abnormal a valve replacement may be required.

Subaortic Stenosis - In this condition the obstruction is below the valve

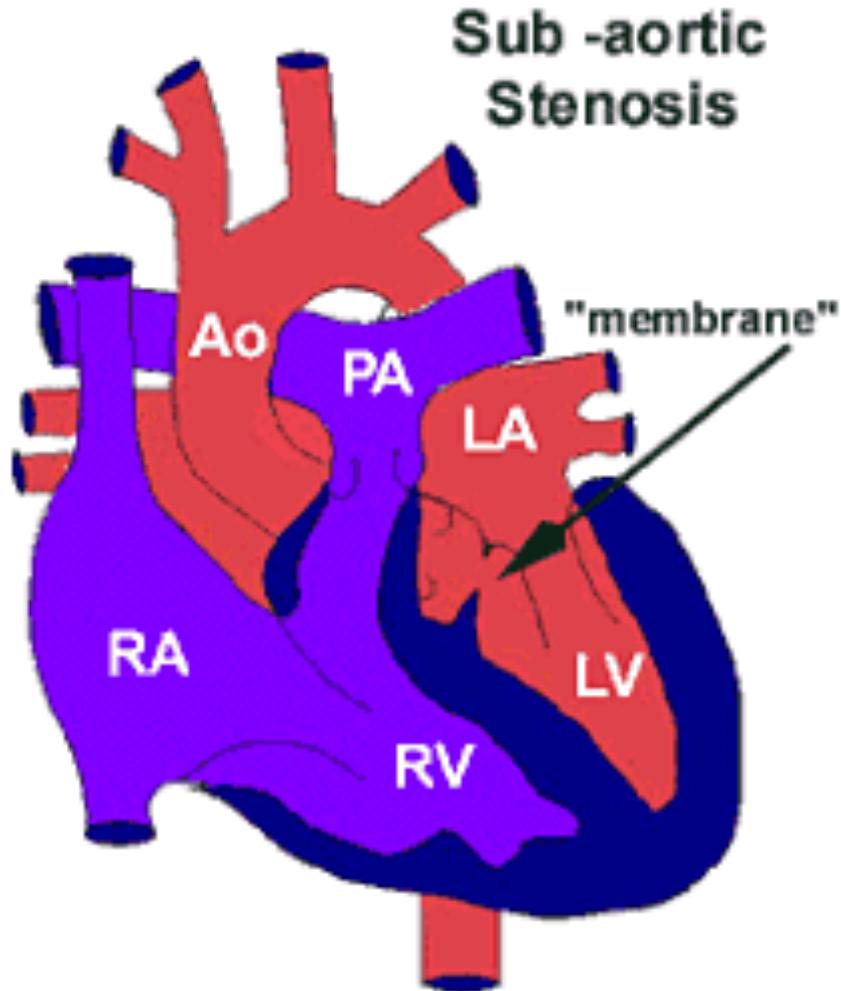


Balloon Valvuloplasty for Aortic Stenosis



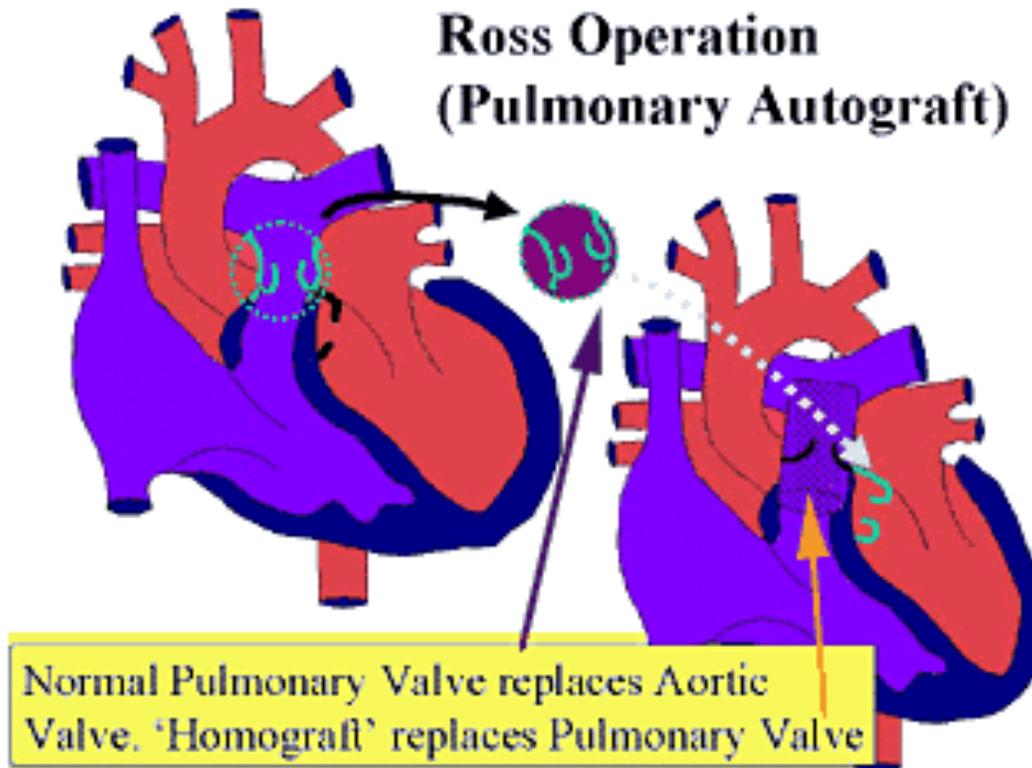
Subaortic Stenosis

In this condition the narrowing is below the aortic valve (indicated by arrow). The effect on heart function is similar to aortic valve stenosis. In many cases the obstruction is produced by a 'membrane', but other types of subaortic stenosis also occur - notably a 'muscular' type (also called "Hypertrophic Obstructive Cardiomyopathy" (HOCM) or "Idiopathic Hypertrophic Subaortic Stenosis" (IHSS)).



Aortic Valve Replacement

When the aortic valve is very abnormal and if it cannot be effectively repaired a valve replacement operation may be recommended. This may involve the use of an artificial valve, but in many cases the patient's own normal Pulmonary valve can be used. This is called the 'Ross Operation' (or Pulmonary Autograft).



Ross Operation

The healthy Pulmonary Valve is removed and sewn into the position of the damaged Aortic Valve. The Pulmonary Valve itself is then replaced with a 'Homograft Valve'. The advantage of this operation is that the new aortic valve will grow with the child and the Homograft Valve, which can be large enough to allow for growth, is not subjected to high pressure and can last much longer in the position of the low pressure Pulmonary Valve - though it is likely that it will eventually need to be replaced at a further operation.

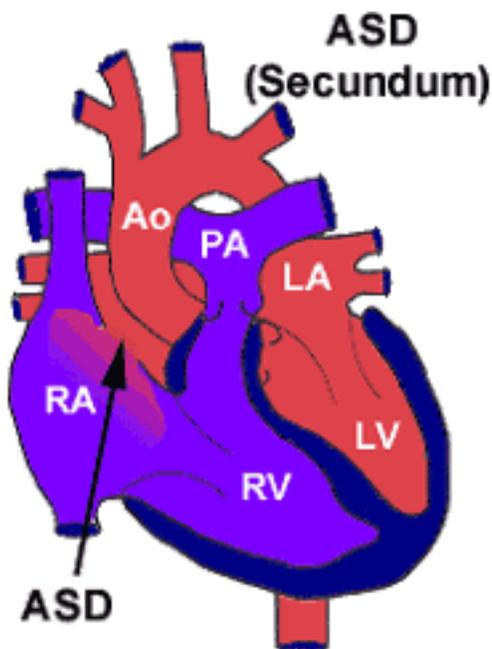
Atrial Septal Defect

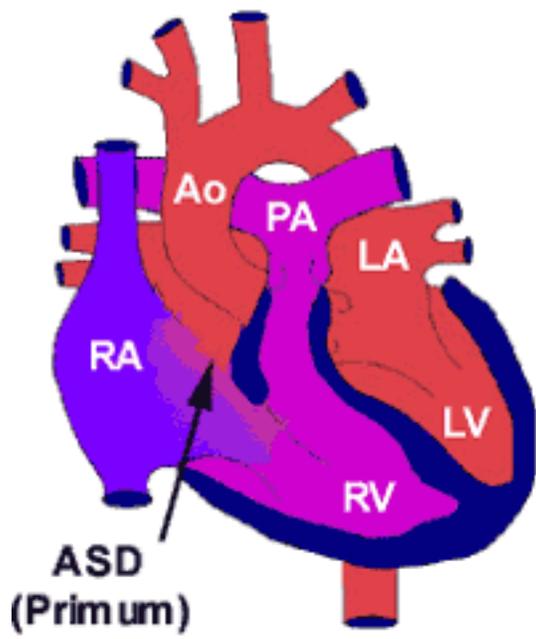
The commonest form of this defect (so called Secundum ASD) is a defect in the central part of the "Atrial Septum" (the partition separating the Atria). This allows red blood to pass through into the right side of the heart, leading to enlargement of the right ventricle and excessive flow in the lung circulation. Most affected children are free of any major symptoms, but the risk of heart failure developing later in life makes closure desirable, unless the defect is very small. These defects may be repaired surgically or by using an expanding plug ("Device"), which can be inserted through a heart catheter without an operation (Device Closure)

Other types of ASD affect different parts of the atrial septum. Defects in the lower part of the septum, close to the Atrioventricular Valves (e.g. Tricuspid valve) are called 'Primum ASD' (or Partial Atrioventricular Septal Defect).

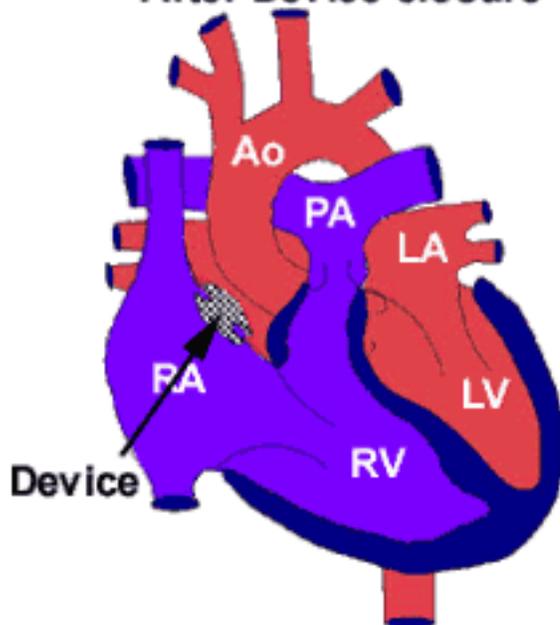
Primum ASD

Defects of this type may lead to symptoms in infancy or childhood and need surgical repair. They are not suitable for device closure. The Mitral Valve is usually abnormal and is often Incompetent.





After Device closure



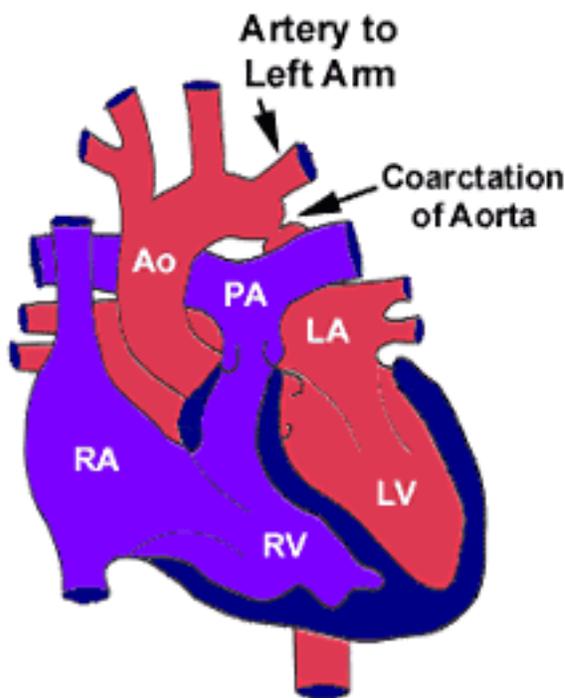
Coarctation of the Aorta HD

- Fact sheet on Coarctation

A narrow area (stricture) is present in the aorta and leads to restricted blood flow to the lower part of the circulation. Blood pressure in the arms and head is high, whilst that in the legs is low. Heart failure may develop. In most cases surgical repair is needed, sometimes in the early weeks of life.

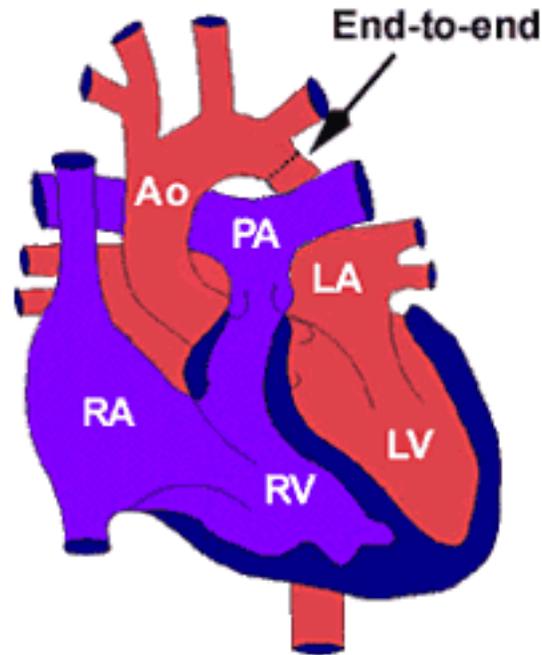
Repair may involve surgical removal of a short segment of aorta, including the stricture (with the ends sewn back together) or use of the artery to the left arm to create a flap, which is turned down to enlarge the narrow section.

Alternatively it may, in some cases, be possible to deal with the problem with a balloon catheter.



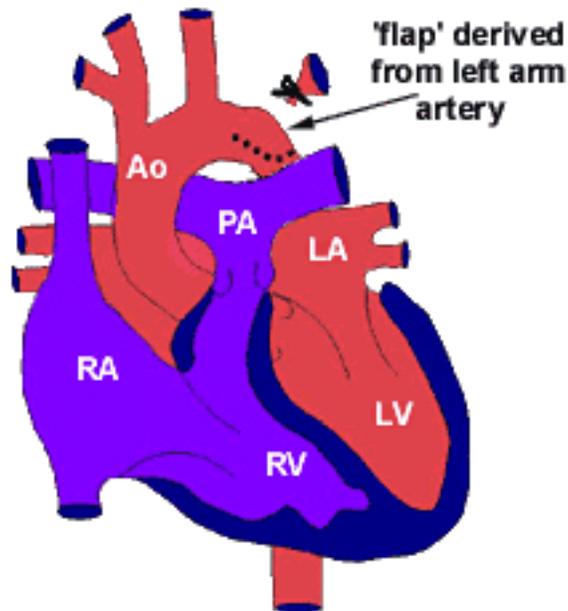
Coarctation Repair

Conventional repair involves the removal of the narrow segment with the ends being sewn together ("end to end").



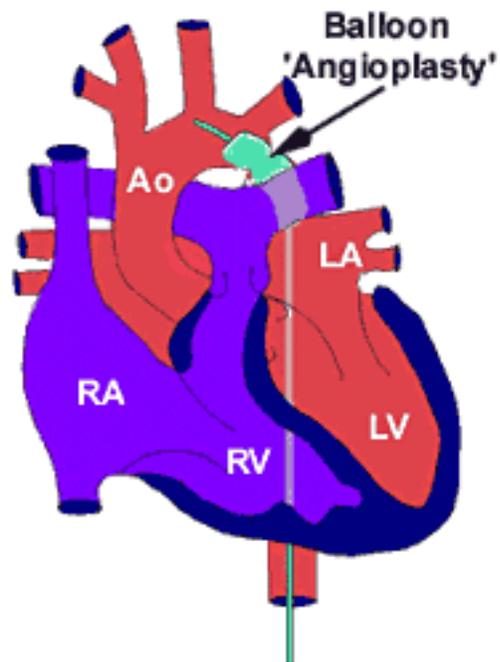
Subclavian Flap

With this type of repair the left arm artery (Subclavian Artery) is used to produce a flap to enlarge the Aorta and repair the Coarctation.



Balloon Angioplasty

A catheter with an inflatable balloon is introduced from an artery in the leg. The balloon is inflated to enlarge the narrow area.



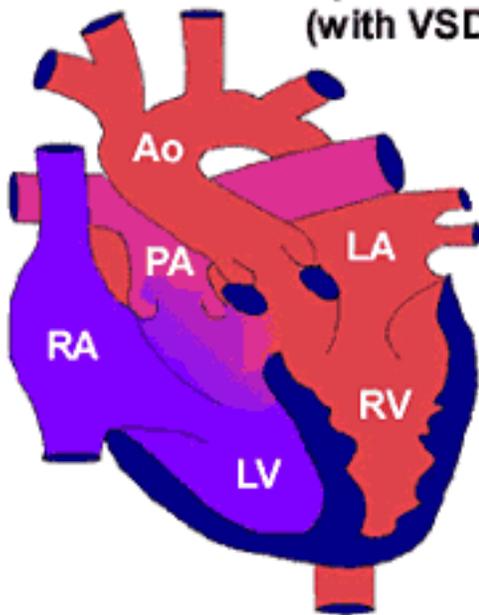
Congenitally Corrected Transposition of the Great Arteries

In this complex malformation the ventricles are on the opposite side of the heart to the usual. The atriums are connected to the incorrect ventricles and the great arteries also come from the wrong ventricles (Transposition). Despite these problems, blood from the lungs passes to the aorta, so the child is not blue. In individuals who have no other heart defects the heart may function well enough for the affected person to survive to adult life without symptoms

In many cases an associated VSD is present (as shown in the illustration). This allows excessive blood flow and pressure in the lung circulation. Other heart defects are commonly present (eg valve abnormalities, coarctation, pulmonary stenosis). Many patients develop 'Heart Block', which may necessitate a pacemaker.

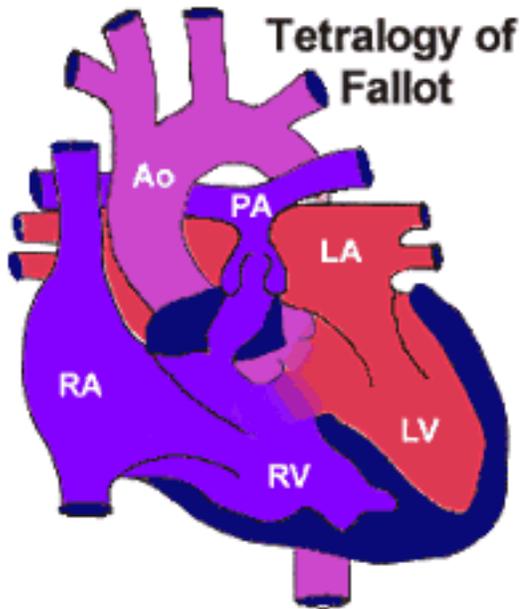
A related abnormality is "Double Inlet Left Ventricle" (DILV).

**'Corrected' Transposition
(with VSD)**

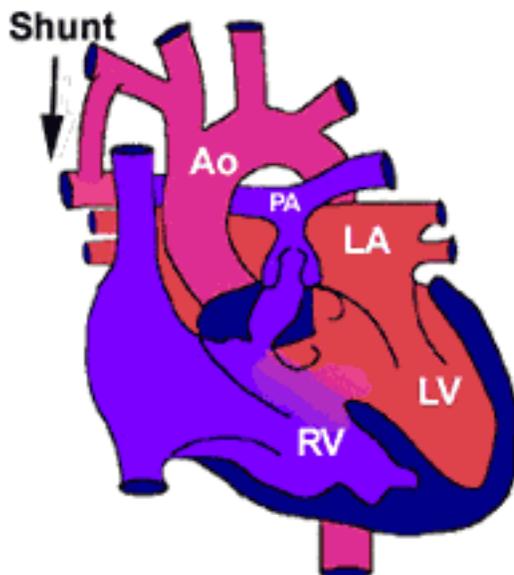


Fallots Tetralogy

The combination of a VSD with Pulmonary Stenosis, with the Aorta "Overriding" (sitting 'astride') the VSD and with RV Hypertrophy is termed "Tetralogy of Fallot". The obstruction to flow into the lungs leads to blood being diverted through the VSD to the aorta. Flow in the lung circulation is reduced and the child appears 'Blue' (Cyanosed).

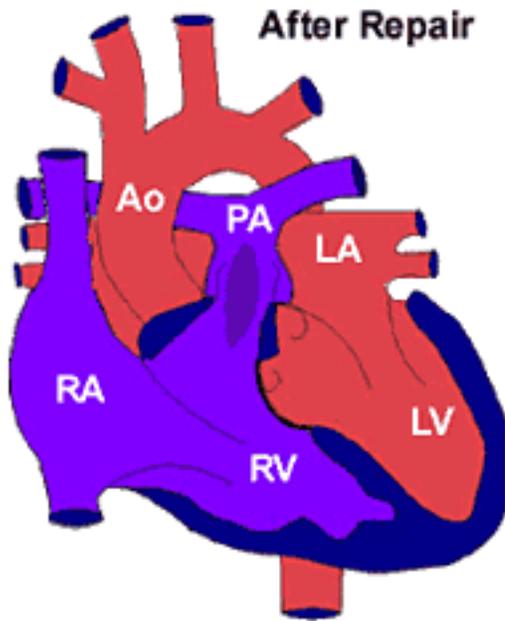


Some affected patients, who are severely blue, need a temporary operation (called a shunt operation), which is carried out in infancy to increase lung blood flow and improve cyanosis. This involves insertion of a tiny piece of artificial tube (made from Goretex) between the Aorta, or a branch (usually one of the arm arteries), and one of the branch Pulmonary Arteries. Corrective surgery is usually performed at about six months. Correction involves closure of the VSD with a patch and enlargement of the narrow area of the right ventricle and pulmonary artery (pulmonary stenosis), often requiring a further patch. (Complete repair).



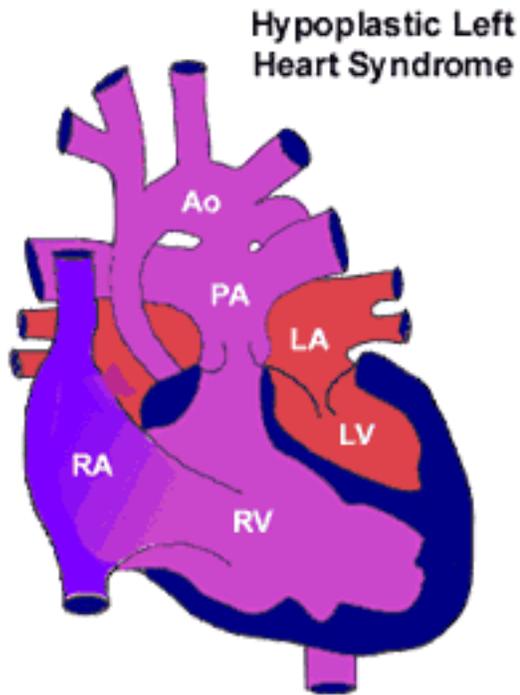
Complete Repair

Corrective surgery is usually performed at about six months. Correction involves closure of the VSD with a patch and enlargement of the narrow area of the right ventricle and pulmonary artery (pulmonary stenosis), often requiring a further patch.



Hypoplastic Left Heart Syndrome HD

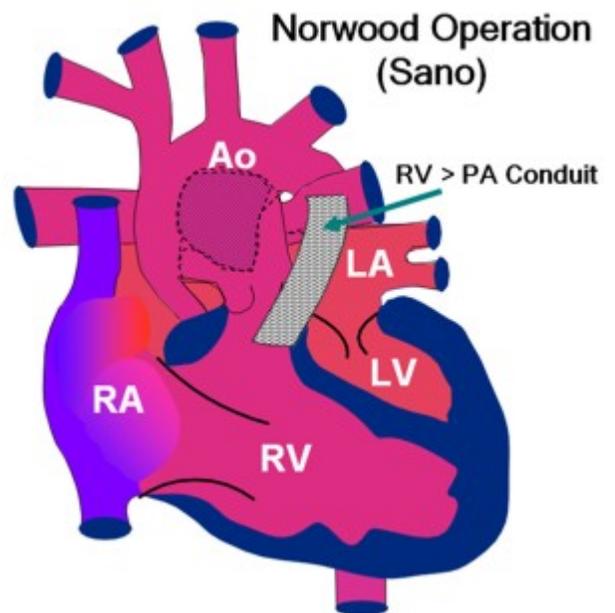
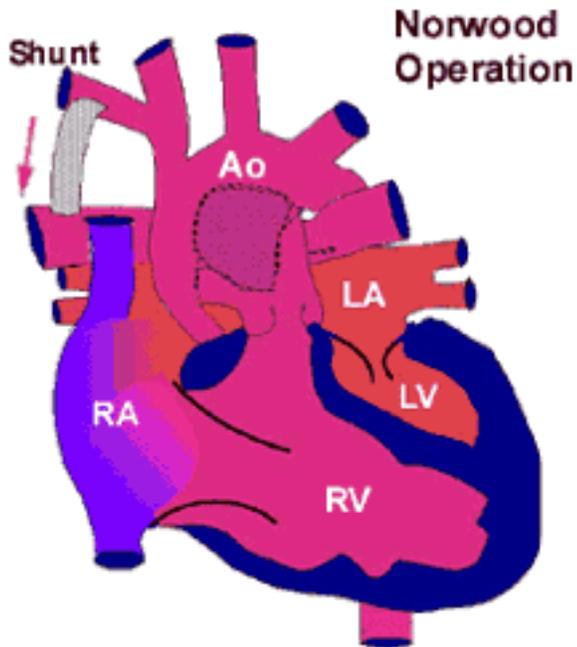
The left side of the heart is very poorly formed and cannot support the main circulation (round the body). The left ventricle and aorta are abnormally small (hypoplastic). This is amongst the most severe forms of heart defect. Most babies are very ill in the early days of life and need urgent surgery to survive. The first stage of surgery is called a 'Norwood' operation.



Norwood Operation

The Norwood operation involves connecting the origin of the pulmonary artery to the aorta, to allow the right ventricle to pump blood to the main circulation and a 'Shunt' operation, involving insertion of a tiny piece of artificial tube (made from Goretex) between the right arm artery and the right pulmonary artery, to maintain blood flow to the lungs. Later in childhood it may be possible to carry out a modified Fontan operation.

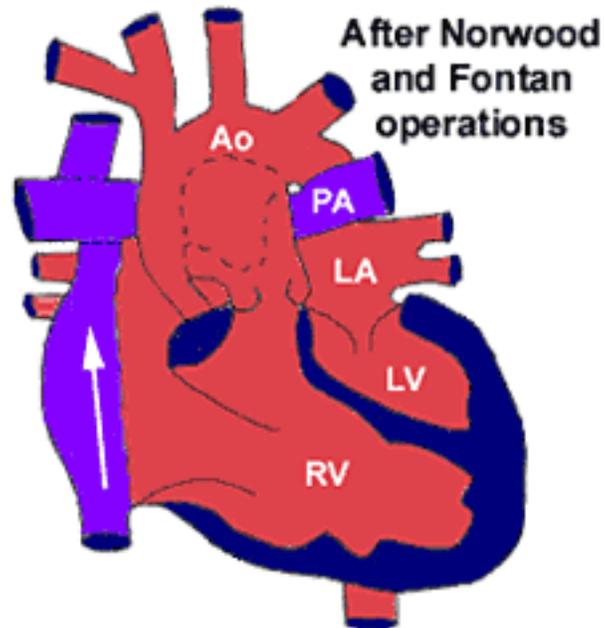
Another option that is sometimes used, involves a Goretex tube (called a "Conduit") from the RV to the pulmonary artery instead of the "Shunt".



A second operation follows after about three months and is called a "Cavo-Pulmonary Shunt". At a later stage (around three to four years old) a Fontan Operation may be performed.

Fontan Operation

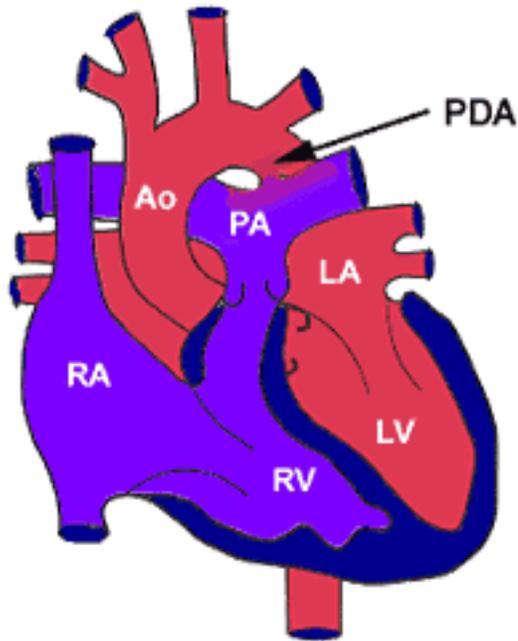
This involves connecting the veins from the main circulation (SVC & IVC) directly to the pulmonary arteries. Blue blood is thus directed into the lungs rather than to the left atrium. A patch is placed to prevent blood passing from the RA to the LA - though sometimes a small hole (a 'Fenestration') is deliberately left.



Patent Ductus Arteriosus (PDA)

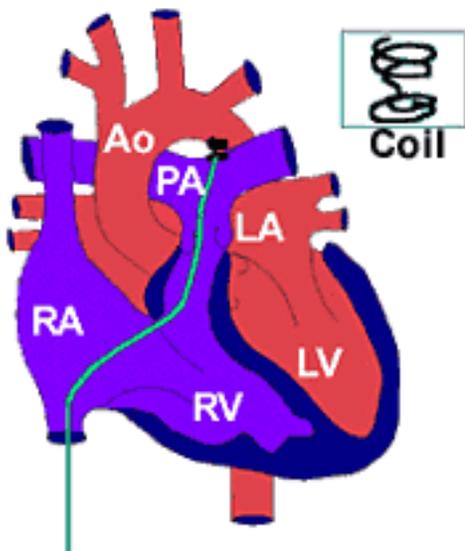
Failure of the ductus to close in the early weeks of life, as normally occurs, results in a "Patent" (or "Persistent") Ductus Arteriosus (PDA). This allows blood to flow between the aorta and the pulmonary artery, leading to an increase in flow in the lung circulation. If the PDA is large the pressure in the lungs may also be elevated. Affected babies may develop heart failure in the early weeks of life.

Children with a small PDA are often asymptomatic, but may develop Infective Endocarditis.



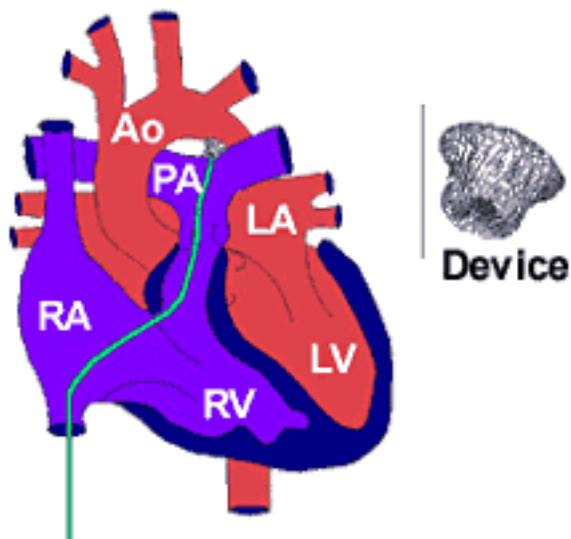
Surgery may be needed when the ductus is large, but in many cases the ductus can be closed using a spring coil introduced with a heart catheter.

Coil closure of PDA



With some larger PDAs a 'Device' (similar to that used for closure of ASD) may be employed.

Device closure of PDA



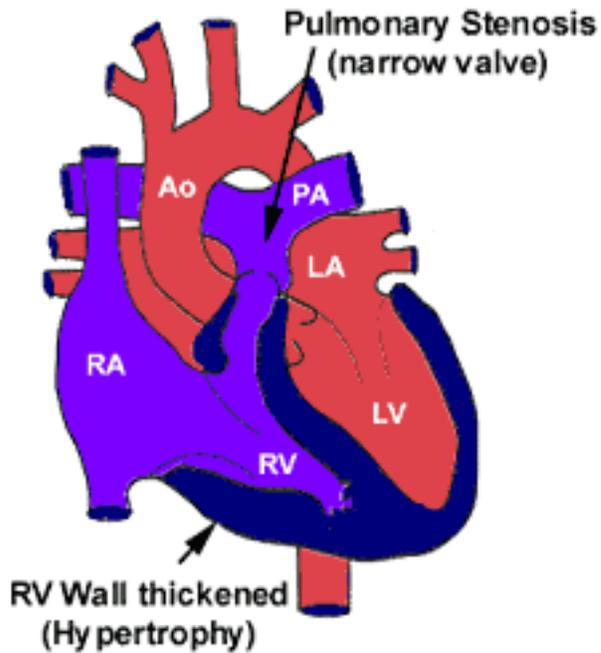
Device closure of PDA

Various devices have been used in recent years. They are introduced through a heart catheter, usually under a general anaesthetic.

This technique is not suitable for premature babies with a PDA and cannot be used for all affected children, some of whom still need an operation.

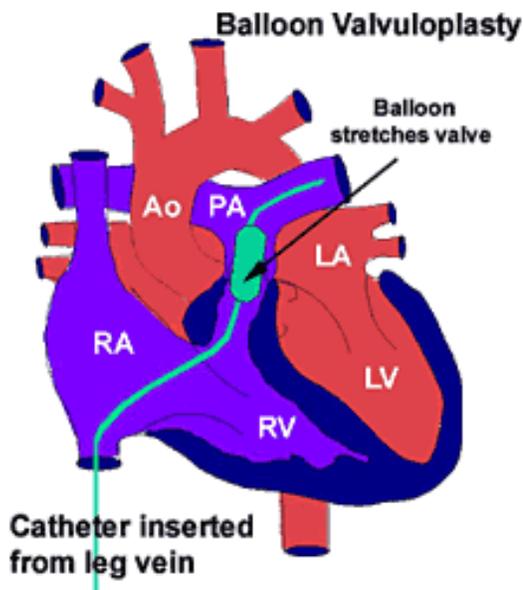
Pulmonary Stenosis

The Pulmonary Valve is thickened and narrowed leading to the development of abnormally high pressure in the right ventricle. The right ventricular wall becomes thickened ("Hypertrophied"). If the problem is severe it may require treatment which usually involves stretching the valve with a balloon catheter (Balloon Valvuloplasty)



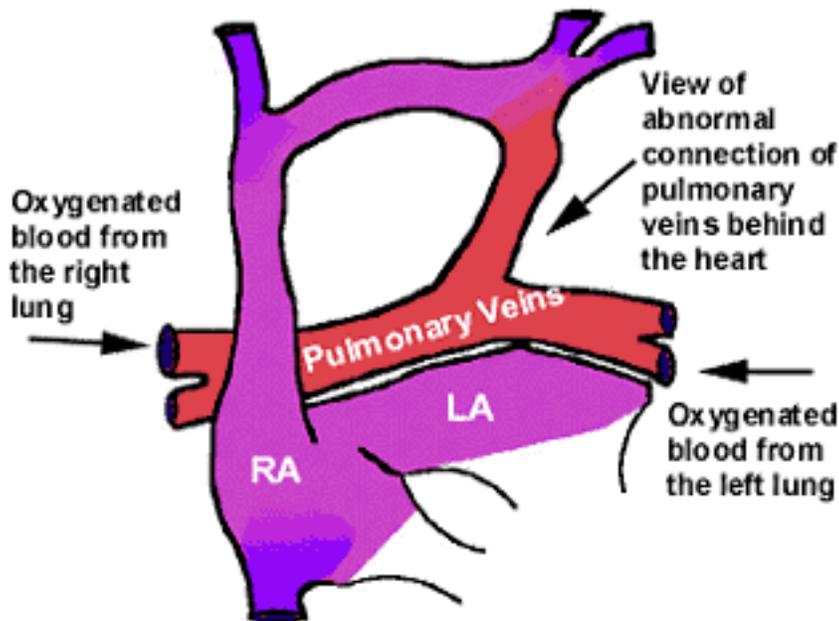
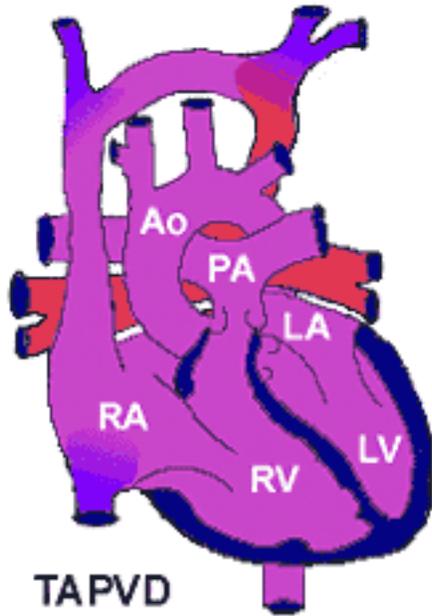
Balloon Valvuloplasty

A heart catheter, with an inflatable balloon at the tip, is passed from a vein in the leg. When the tip is through the valve the balloon is inflated to stretch open the valve. Usually this produces effective longterm relief of the obstruction. Some patients who are unsuitable for this type of procedure, or in whom it is unsuccessful, may require surgery to correct the problem.



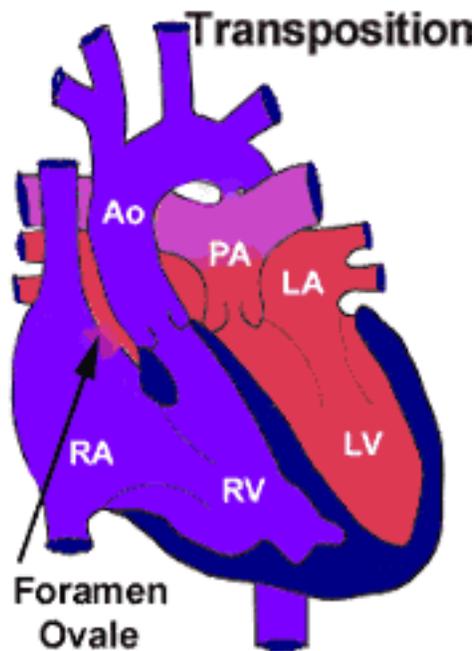
Total Anomalous Pulmonary Venous Drainage

The Pulmonary Veins, which carry blood back to the heart after it has circulated through the lungs, are not connected to the left atrium. Instead they are connected to one of the veins from the main circulation so that the blood returning from the lungs drains back to the right side of the heart. The affected babies may be blue or show signs of heart failure. Most of them require surgical repair in the newborn period.



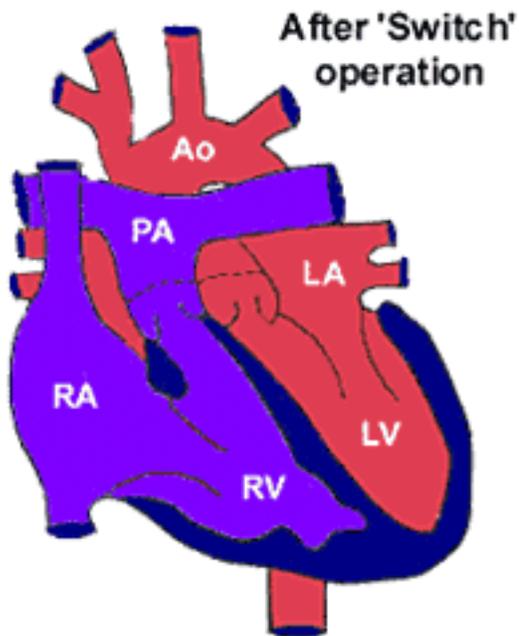
Transposition of the Great Arteries

The Aorta arises from the right ventricle and receives "blue" blood, whilst the Pulmonary Artery arises from the left ventricle. The baby becomes blue immediately after birth and needs urgent treatment. Survival depends on the ductus or the Foramen Ovale remaining open in the early days of life until treatment can be applied. The Foramen Ovale can be enlarged with a catheter procedure, called Balloon Septostomy, which is performed in the first few days of life. This involves a catheter with a balloon at the tip, which is passed from a leg vein until the balloon is in the left atrium (across the Foramen Ovale). The balloon is then inflated and the catheter is pulled back to the right atrium.



Early surgery is essential and involves the " Arterial Switch Operation", which is carried out in the first week or two of life and corrects the abnormality. The small coronary arteries, which feed the heart muscle with blood, need to be transferred as well as the two Great Arteries (Aorta and Pulmonary Artery).

Until around 1980 this delicate surgery could not be performed safely and an alternative procedure was used, redirecting blood within the atriums. This was called a "Senning" operation. (The "Mustard" operation was very similar.)

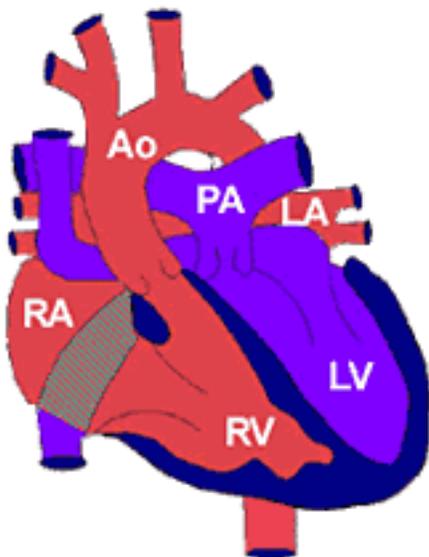


Senning Operation.

In this operation the blood from the SVC and IVC was redirected to the left atrium and ventricle, while blood from the pulmonary veins was channelled to the right atrium and ventricle. This relieved Cyanosis but did not correct the original abnormality, leaving the Right Ventricle pumping blood to the Aorta.

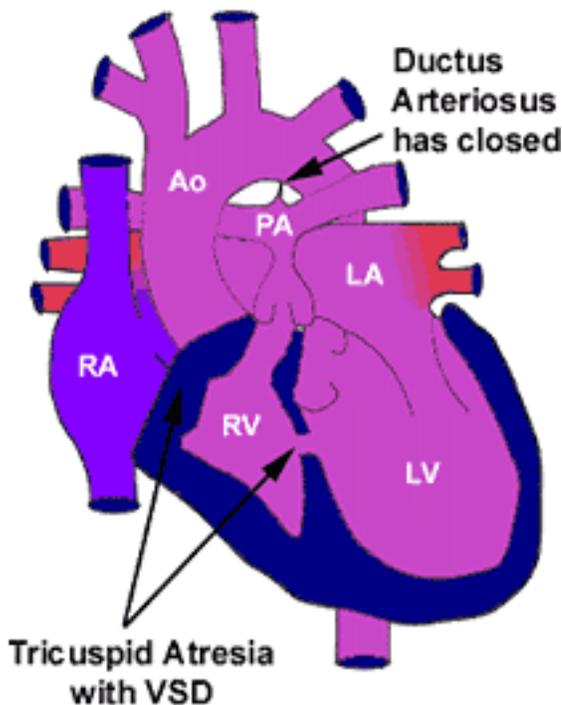
Eventually some patients may develop heart failure or other problems after this type of surgery, which is now seldom used.

Senning Operation



Tricuspid Atresia

Absence of any connection between the right atrium and the right ventricle leads to blood being diverted from the right atrium to the left atrium. The right ventricle (RV) is usually small (Hypoplastic). Survival depends on an associated VSD (often quite small), in order for blood to reach the lungs, or on the ductus remaining open in the early days of life. Most babies will need a 'Shunt' operation during infancy, involving insertion of a tiny piece of artificial tube (made from Goretex) between the Aorta, or a branch (usually one of the arm arteries), and one of the branch Pulmonary Arteries.



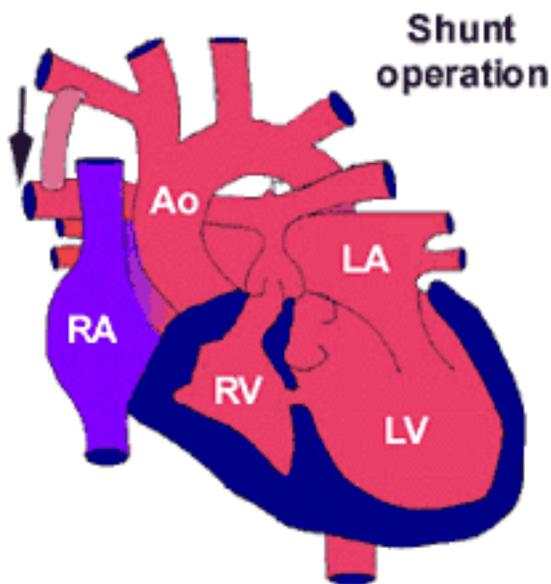
At a second operation some blood from veins in the upper part of the body may be connected directly to the lung arteries (BCPC). Definitive surgery is usually delayed until the age of at least two years. It involves the so called Fontan operation. This involves connecting the veins from the main circulation (SVC & IVC) directly to the pulmonary arteries. Blue blood is thus directed into the lungs rather than to the left atrium. A patch is placed to prevent blood passing from the RA to the LA - though sometimes a small hole (a 'Fenestration') is deliberately left.

Bidirectional Cavo Pulmonary Connection (BCPC)

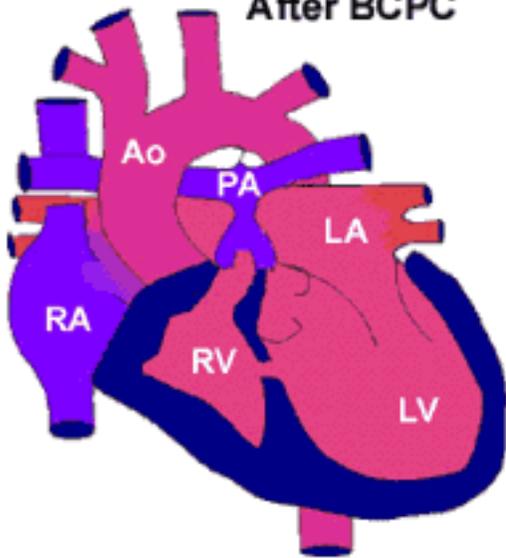
A Cavo-Pulmonary shunt involves connecting the main vein from the upper circulation (the SVC) to the right pulmonary artery (RPA), in order to direct the blood from the upper part of the main circulation into the lung circulation. The main vein from the lower circulation (the IVC) still carries blood into the right atrium. This operation will often improve cyanosis (blueness), but does not eliminate it completely. Many children will go on to have a Fontan operation later, when the IVC blood will also be channelled into the pulmonary arteries and the cyanosis will be completely, or almost completely, relieved.

Fontan operation

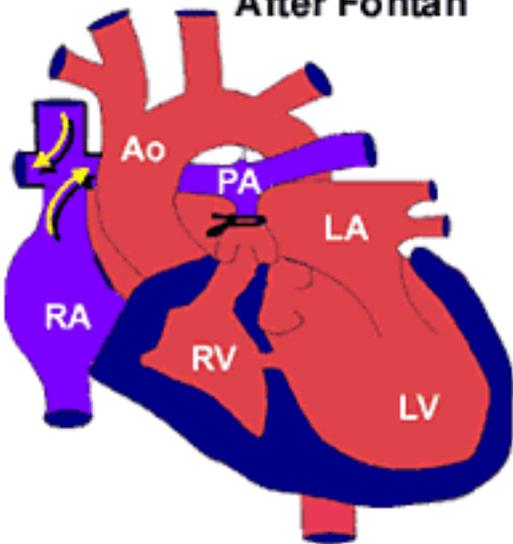
The Fontan operation involves connecting the veins from the main circulation (SVC & IVC) directly to the pulmonary arteries. Blue blood is thus directed into the lungs rather than to the left atrium. A patch is placed to prevent blood passing from the RA to the LA - though sometimes a small hole (a 'Fenestration') is deliberately left.



**Tricuspid Atresia
After BCPC**

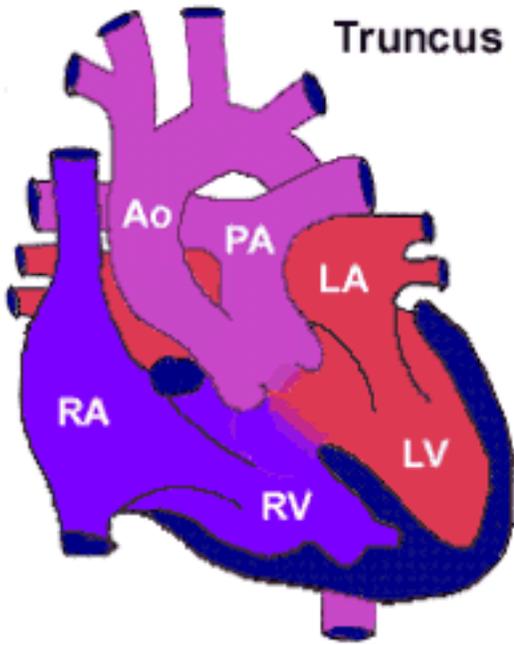


**Tricuspid Atresia
After Fontan**

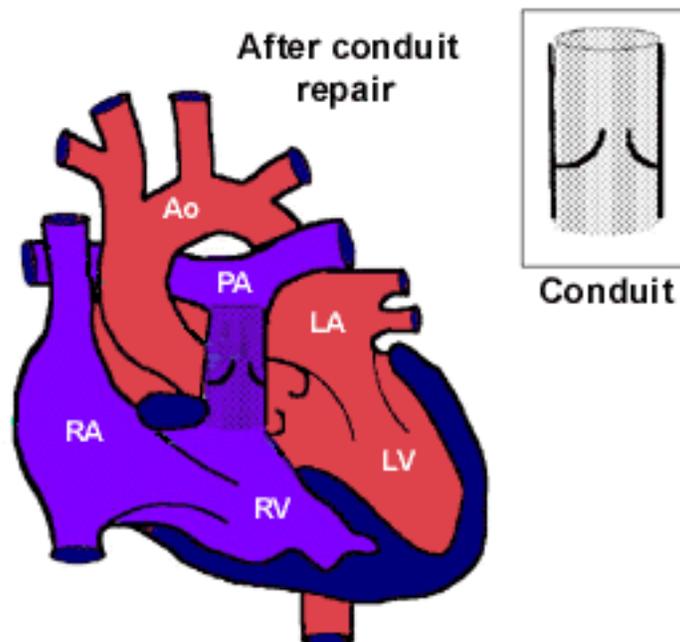


Truncus Arteriosus

The two Great Arteries (Aorta and Pulmonary Artery) have a single origin from the heart and blood from both ventricles passes across a VSD into the single arterial trunk. The lung circulation is exposed to very high pressure and increased blood flow (as with a large VSD). Heart failure often develops in the early weeks of life.



Complete repair for Truncus necessitates the insertion of a 'Conduit', which is a tube containing a valve placed to connect the right ventricle to the pulmonary artery. The VSD is also closed with a patch. Operation needs to be performed in the first six months of life



Ventricular Septal Defect (VSD)

The commonest heart defect ("Hole in the heart"). When small, such defects cause little in the way of problems and often "heal" (close) on their own. Blood flows from left ventricle to right ventricle at high pressure, often producing a loud "murmur" with each heart beat. The effect on heart function depends on the size of the defect, but may be very minor. Surgery may not be required, if the heart shows no evidence of "strain".

Larger VSDs allow more blood to flow from the left ventricle to the right ventricle and lead to increase in pressure and flow in the lung circulation. This places significant strain on the heart and affected babies usually need surgical repair of the defect. An operation may be required in the first three to four months of life, though some defects, which are causing less trouble, may be left for a few years, in the hope that they may get smaller, only needing surgery if they remain large or are associated with other problems.

