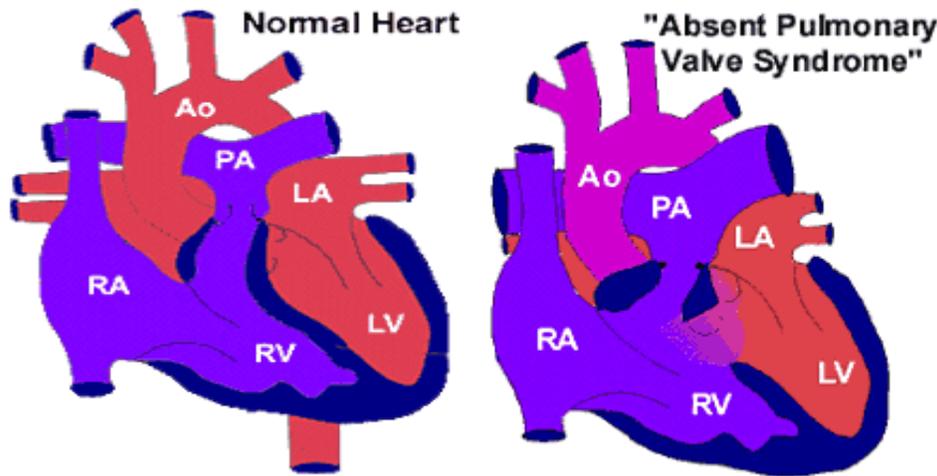


Cardiac Defects

Absent Pulmonary Valve Syndrome

In this condition, which has some similarities to Fallot's Tetralogy, there is a Ventricle Septal Defect (VSD) with some degree of narrowing of the channel carrying blood to the lungs. 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.



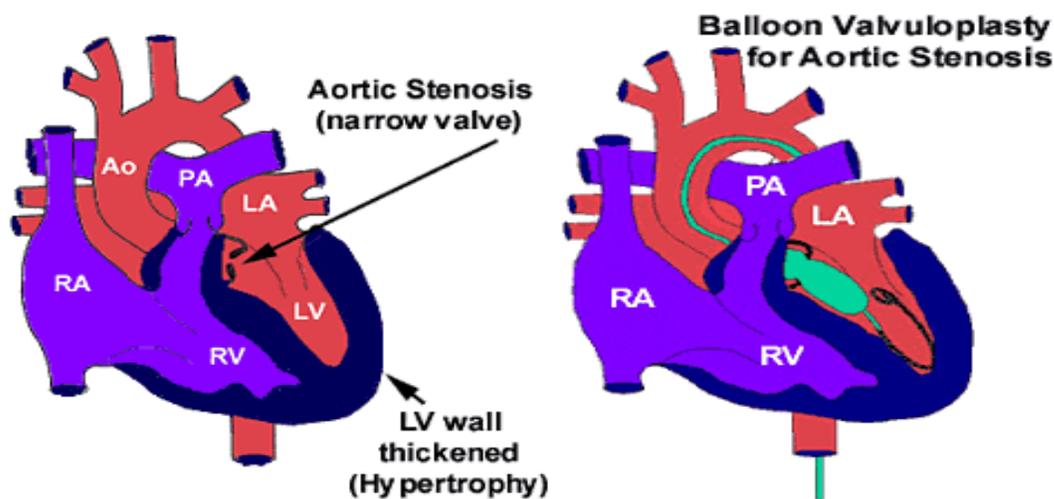
Absent Pulmonary Valve Syndrome

This is a similar combination of problems to that seen in Fallot's Tetralogy. A VSD with "overriding" of the Aorta coincides with a narrow pulmonary valve. The difference is that the pulmonary valve flaps (leaflets) are very rudimentary (poorly formed). As a result the valve is severely incompetent as well as being narrow (stenotic). The pulmonary arteries are characteristically enlarged (rather than being small, as is the case with Fallot's Tetralogy). The large pulmonary arteries may put pressure on the adjacent air tubes (bronchi) within the lungs, leading to breathing problems. Affected babies sometimes have severe respiratory difficulties in early infancy and may need early surgery to repair the heart problems and to reduce the size of the enlarged pulmonary arteries. Surgical repair is essentially similar to that of Fallot's Tetralogy.

Aortic Stenosis (AS)

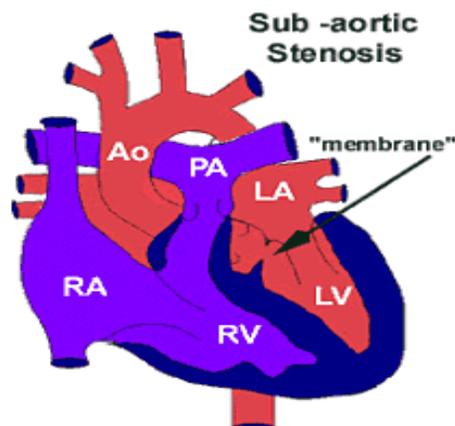
The Aortic Valve is thickened and narrowed leading to the development of abnormally high pressure in the left ventricle. The left ventricular wall becomes thickened ("Hypertrophied"). If the problem is severe it may require treatment, which usually involves surgery in younger patients, though it may 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 re-operation or further balloon stretching.

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



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).

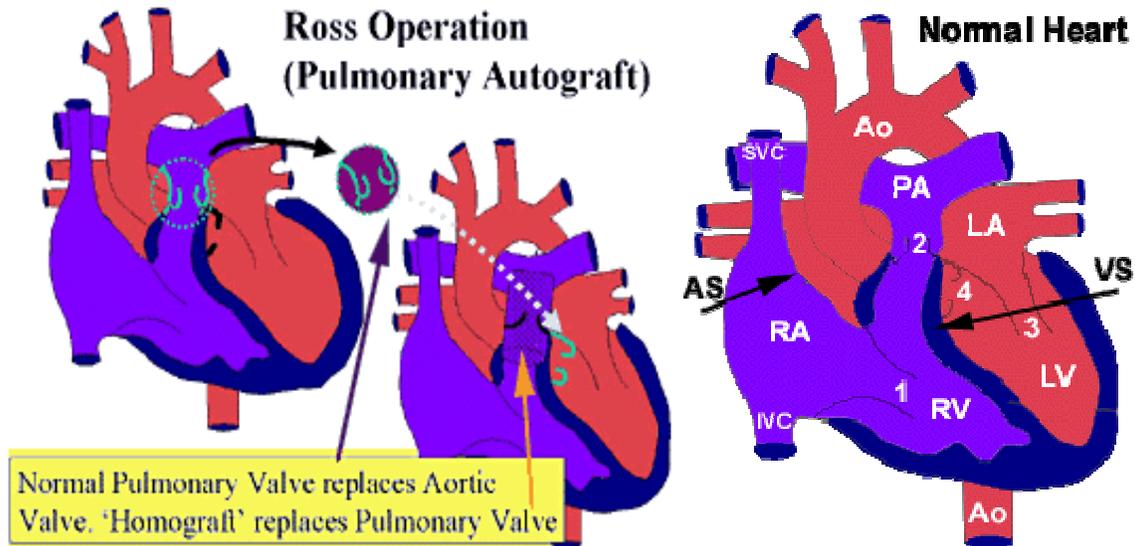


Aortic Valve Replacement

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'. (Pulmonary Autograft)

Ross Operation

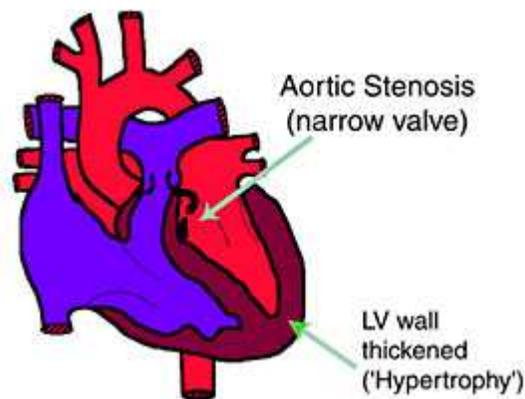
The healthy Pulmonary Valve is removed and sewn into the position of the damaged Aortic Valve. The Pulmonary Valve itself is then replaced with a "Homograft Valve." The advantage of this operation is that the new aortic valve will grow with the child and the Homograft Valve, which can be large enough 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.



Aortic Stenosis

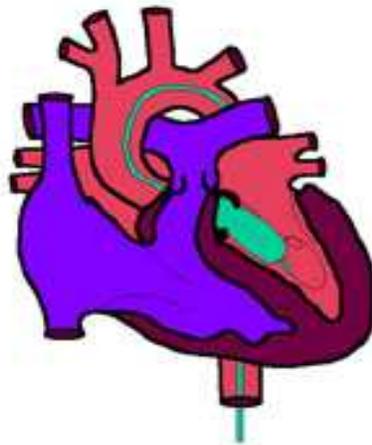
The Aortic Valve is thickened and narrowed leading to the development of abnormally high pressure in the left ventricle. The left ventricular wall becomes thickened (Hypertrophied).

Stenosis (narrowing) of the aortic valve restricts flow into the aorta. This leads to the presence of a heart "murmur". Often the narrowing is mild and does not put significant strain on the heart. However the narrowing frequently worsens with growth. If the obstruction is severe, symptoms may develop, or the heart may show evidence of "strain." The valve may require treatment to open it up. This may be surgical or with the use of a "balloon catheter" procedure.



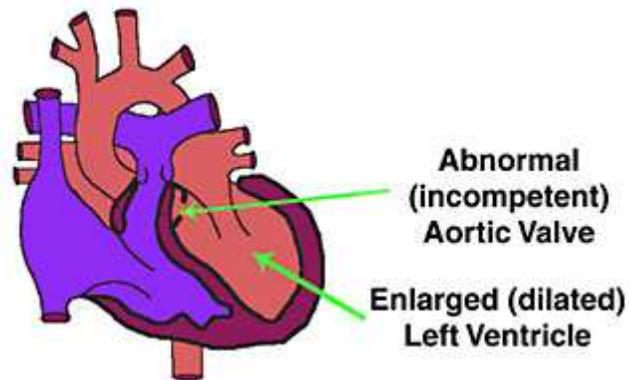
Balloon Valvuloplasty for Aortic Stenosis

Balloon Valvuloplasty is a procedure done using a Heart Catheter threaded up an artery from the leg. A balloon at the tip of the catheter is placed across the valve and is then inflated to "Open Up" the narrow valve. This procedure may allow surgery to be delayed until the patient is older, but will usually not remove the need for an operation at a later stage.

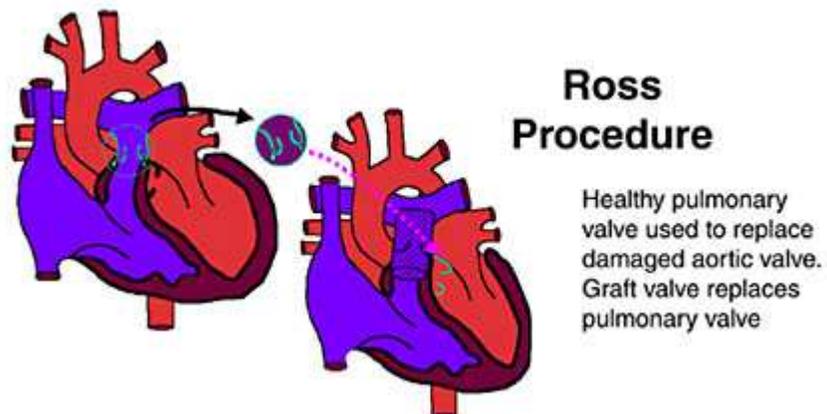


Aortic Incompetence and Ross procedure

The Aortic valve may become incompetent as a result of a congenital abnormality of the valve, the presence of a VSD just below the valve or for other reasons. In some cases this problem follows earlier surgery or balloon valvuloplasty. The regurgitation of blood across the valve leads to progressive enlargement of the left ventricle. Symptoms may include increasing breathlessness with exertion. If the degree of leakage (incompetence) is severe and the valve cannot be repaired then a valve replacement may be recommended.



Replacement of the Aortic Valve may be achieved by using an artificial valve or a graft valve. However in many patients it is possible to replace the damaged aortic valve with the normal pulmonary valve, which will often function well and can grow with the child. The pulmonary valve has to be replaced itself with a graft valve. The grafted valve that is used will last longer in the low-pressure pulmonary position and a larger valve can often be fitted in to allow for growth- though further surgery is likely eventually. This type of surgery is called a Ross Operation.



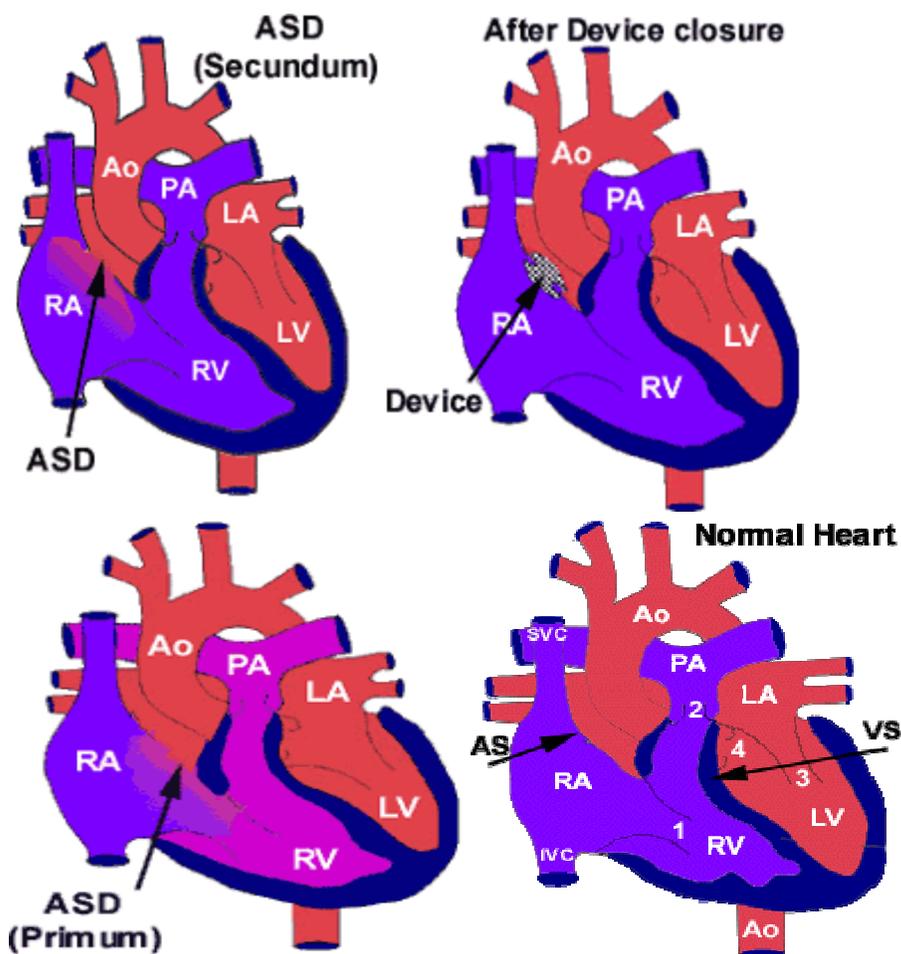
Atrial Septal Defect

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.



Atrial Septal Defect Secundum

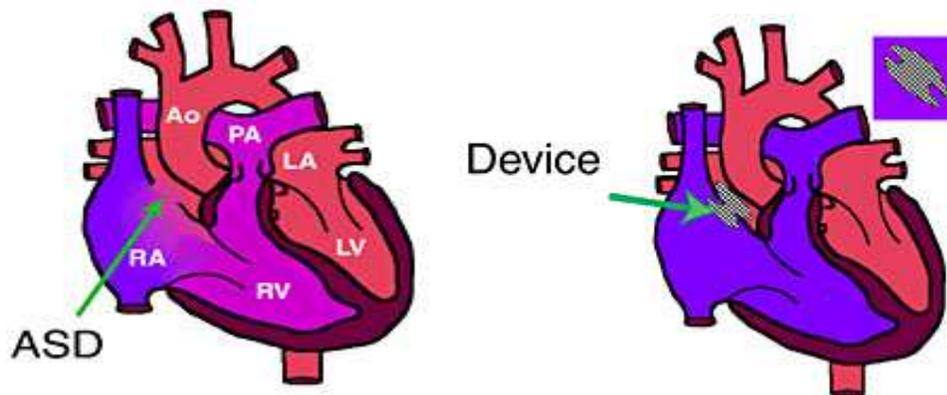
A defect is present in the central part of the Atrial Septum (the partition separating the Atria). This allows red blood to pass through into the right side of the heart, leading to enlargement of the Right Ventricle and excessive flow in the lung circulation.

Defects of this type are common. They seldom lead to symptoms during childhood and may not be detected until school age or later. Symptoms are likely to develop by early to middle adult life.

After Device Closure

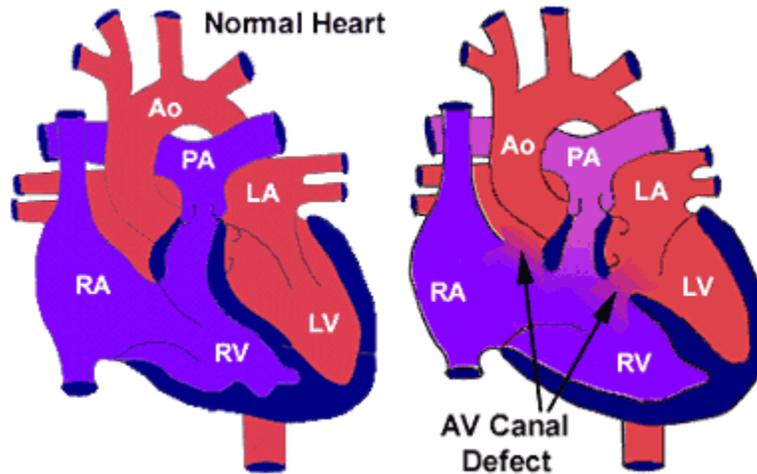
Defects of this kind may be repaired surgically during childhood (before they are likely to lead to damage to the heart or lungs). Alternatively they can be closed, in suitable cases, with a catheter procedure (Device Closure).

The Device is introduced through a Heart Catheter, which has been passed up from a vein in the leg. The Device is made of collapsible metal mesh, with thin membranes inside. Once inside the ASD it will become covered by natural tissue within a few weeks.



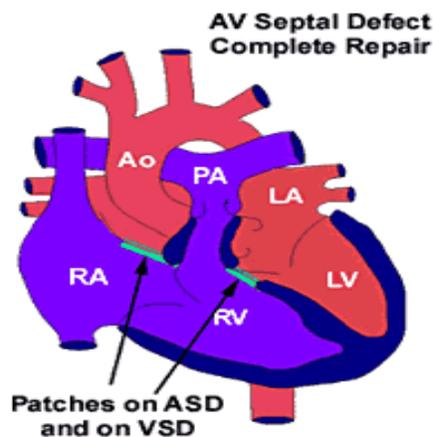
Atrioventricular Septal Defect (AV Canal defect)

This is usually a large defect involving both the atrial (ASD) and the ventricular (VSD) septums, which allows blood to pass freely between the two ventricles and the atriums. The valve apparatus at the junction between atriums and ventricles are shared there being effectively only one valve instead of the normal two. Blood flow and pressure in the lung circulation is substantially increased. Early surgical repair is needed in most cases (in the first four to six months).



Repair of Atrioventricular Septal Defect (AV Canal defect)

Surgical repair usually involves two patches. One closes the Atrial Septal Defect and the other repairs the Ventricular Septal Defect. The Atrioventricular valve is carefully separated into two valves, which are attached to the two patches. This allows the two sides of the heart to function independently, each now having its own individual valve - as in the normal heart. The repaired valves are not completely normal and may sometimes allow blood to leak back through the valve. (Valve Incompetence)

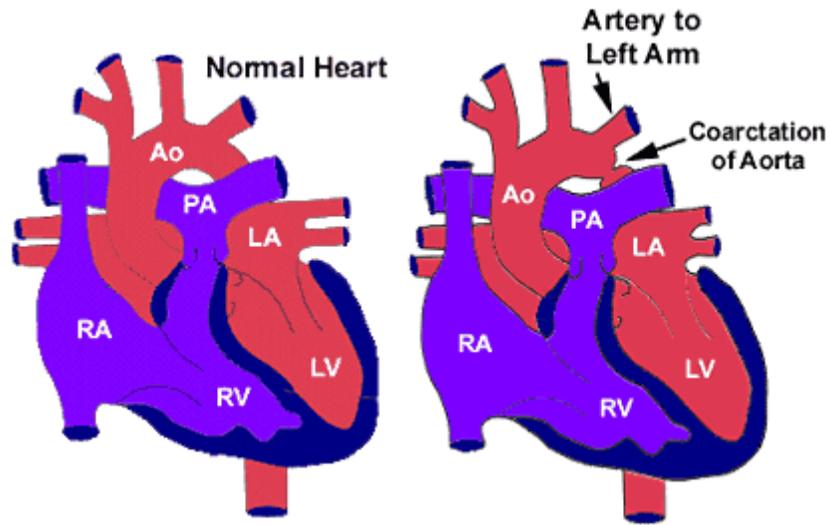


Coarctation of the Aorta HD

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.

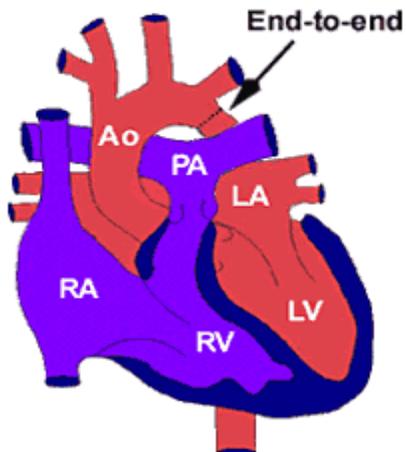
Repair may involve surgical removal of a short segment of aorta, including the stricture (with the ends sewn back together) or use of the artery to the left arm to create a flap, which is turned down to enlarge the narrow section.

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



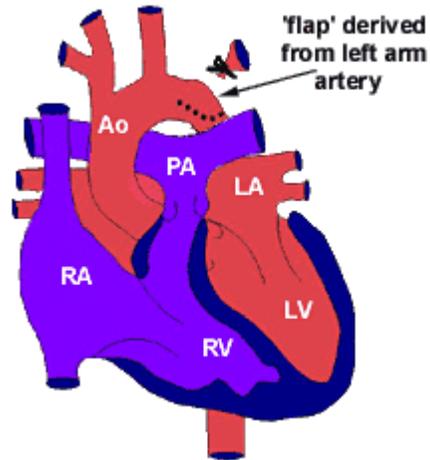
Coarctation Repair

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



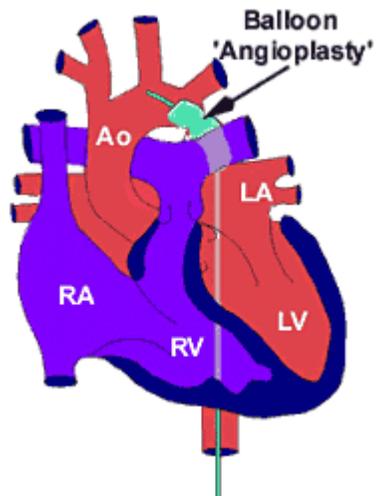
Subclavian Flap

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.



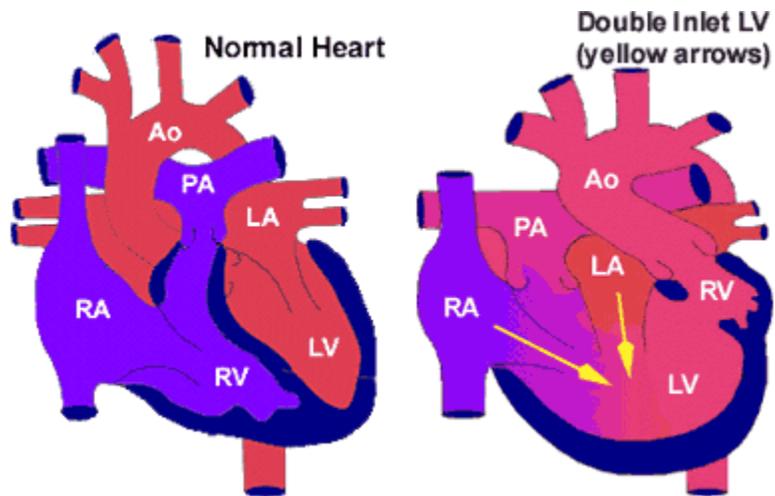
Balloon Angioplasty

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



Double Inlet Left Ventricle (DILV) (Sometimes called Single Ventricle)

In this abnormality both atriums are connected to the Left Ventricle. Usually there is a small (hypoplastic) right ventricle (RV), which may be on the opposite side of the heart to the usual (as in “Corrected Transposition”). The arteries usually arise with the aorta from the RV and the pulmonary artery from the LV (Transposition). Many other defects are often present (e.g. Pulmonary Stenosis or Atresia; Coarctation of the aorta).



Double Inlet Left Ventricle (Sometimes called Single Ventricle)

This is one of a group of very complex defects where both Atriums connect to the same ventricle and/or one ventricle is absent or very tiny. In most cases the infant develops symptoms in the early weeks of life- either with “Cyanosis” (Blueness of the skin) or with breathlessness and failure to gain weight normally. Many affected patients have associated defects in the heart or main arteries, including such problems as Pulmonary Stenosis (PS), Pulmonary Atresia, other valve abnormalities or Coarctation of the Aorta, etc. Such problems may cause severe obstruction to blood flow to the lungs or into the main circulation and may require urgent surgery.

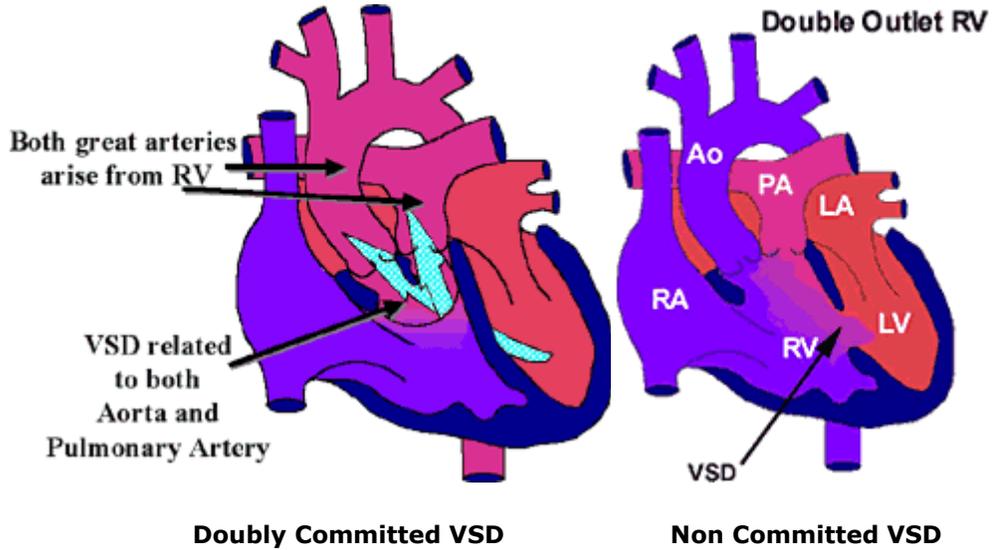
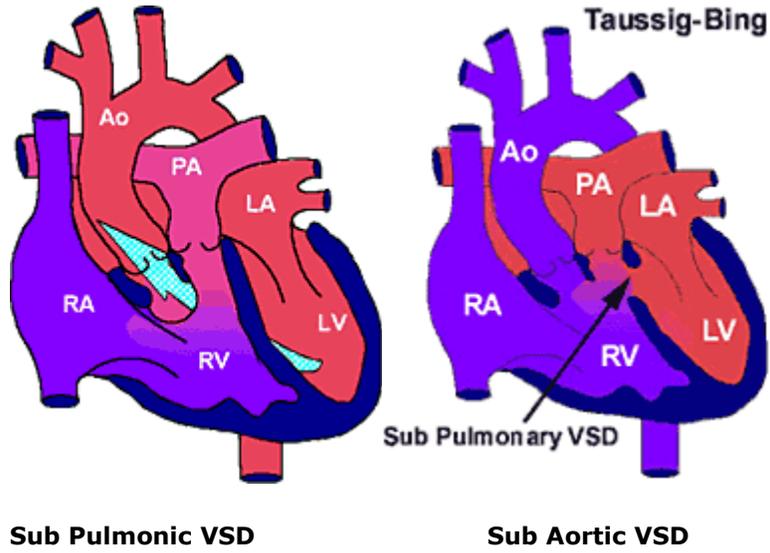
The RV (usually very small) may be on the opposite side of the heart to normal. This is similar in some respects to the situation with Corrected Transposition.

Treatment is aimed at repairing the associated problems, where that can be done and performing staged surgery (often with several operations being needed) to make the heart work effectively. The basic defect cannot be repaired, but in many cases the heart can be made to function sufficiently well to eliminate cyanosis and to allow normal schooling and near normal activities.

For many children the final operation is called the Fontan Operation. This leads to blue blood (low in oxygen) being channeled through the lungs, without any “pump” driving it. The large ventricle then pumps the red blood (high in oxygen), round the body. This operation makes the child “Pink”- but does not correct the original problem. Not all affected children need to have a Fontan operation. If they are well, with less drastic

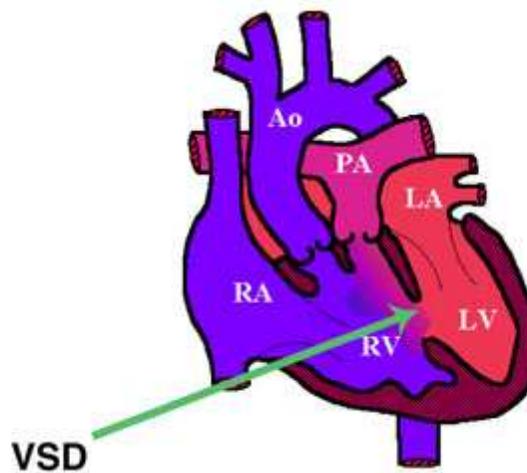
Different types of DORV

The site of the VSD may vary and can affect the clinical manifestations and the options for surgery. The commoner variations are shown below.



DORV - Double Outlet Right Ventricle

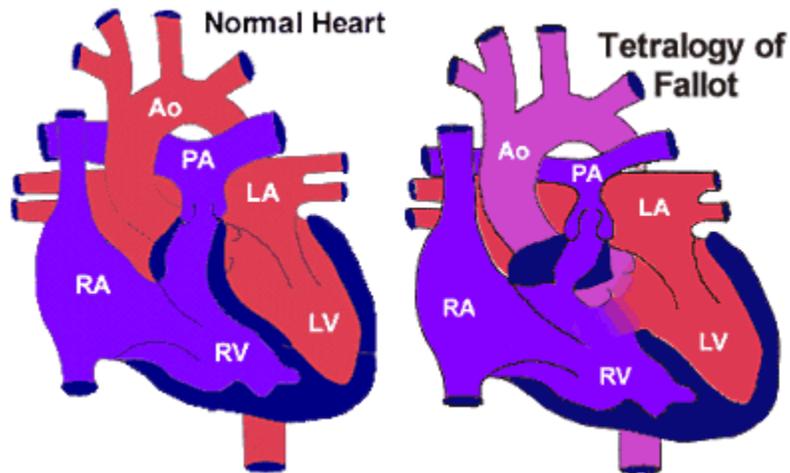
In Double Outlet Right Ventricle (DORV) the two Great Arteries (Aorta and Pulmonary Artery) both originate from the right ventricle and blood from the left ventricle passes across a VSD into the RV to reach the great arteries. The lung circulation is often exposed to very high pressure and increased blood flow (as with a large VSD). In most cases the infant develops symptoms in the early weeks of life- either with Cyanosis (Blueness of the skin) or with breathlessness and failure to gain weight normally. There are many different varieties of this abnormality. Many affected patients have associated defects in the heart or main arteries, including such problems as Pulmonary Stenosis (PS), Pulmonary Atresia, other valve abnormalities or Coarctation of the Aorta, etc. Such problems may cause severe obstruction to blood flow to the lungs or into the main circulation and may require urgent surgery.



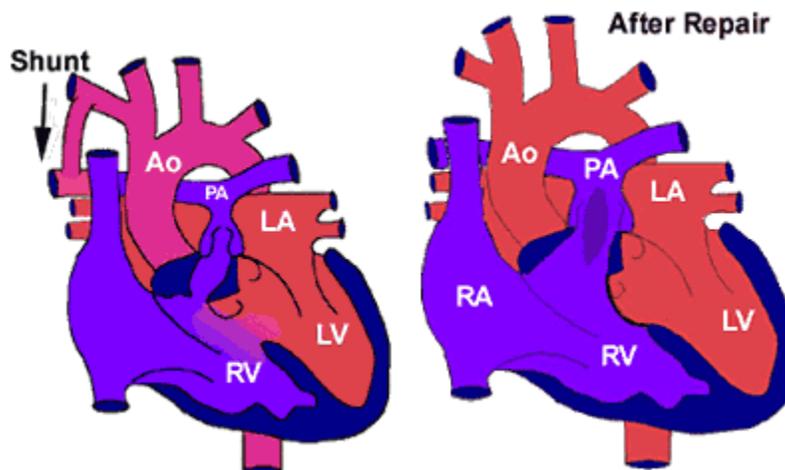
Treatment, which is surgical, is aimed at repairing the defect and any associated problems where that can be done. In many cases the heart can be made to function sufficiently well to eliminate symptoms and to allow normal schooling and near normal activities. Most cases are amenable to complete repair; though more than one operation may be needed. In a minority of the more complex cases complete repair may not be possible. In such cases treatment aims to make the heart work as effectively as possible. This may involve several operations. For some children the final operation is called the Fontan Operation. This leads to blue blood (low in oxygen) being channeled through the lungs, without any “pump” driving it. The ventricles then pump the red blood (high in oxygen), round the body. This operation makes the child “Pink” but does not correct the original problem.

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).



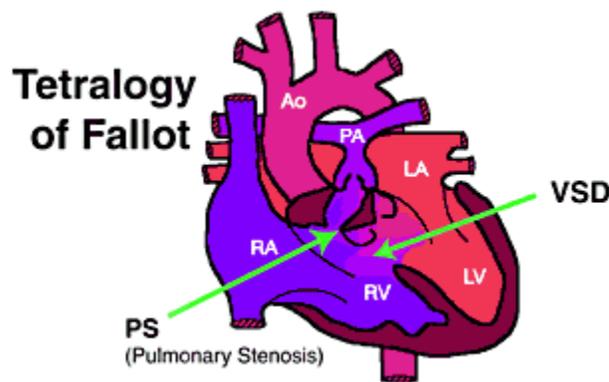
Complete Repair

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.

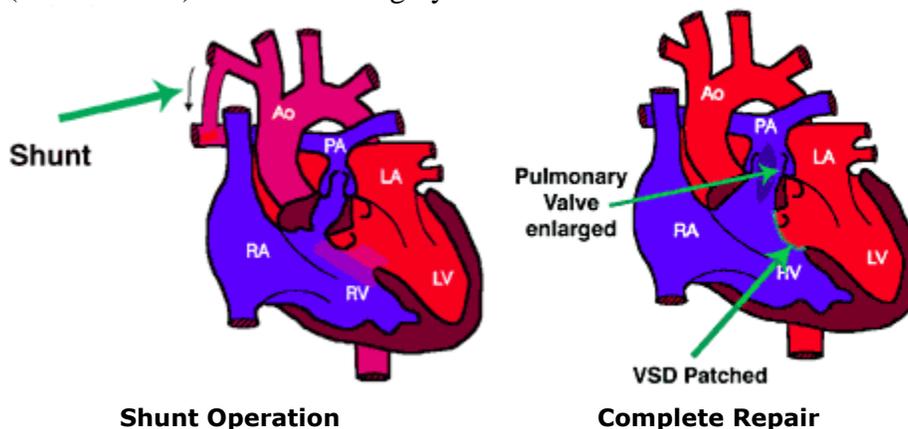
Fallot's Tetralogy

The combination of a VSD with Pulmonary Stenosis, with the Aorta "Overriding" (sitting astride) the VSD and with Right Ventricle Hypertrophy (thickening of the muscle) 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).

Affected infants are often relatively free of symptoms in the early weeks of life. Cyanosis (Blueness) gradually appears, sometimes with a tendency to intermittent severe exacerbations ("Spells"). A heart murmur is usually heard early in infancy even before symptoms develop.

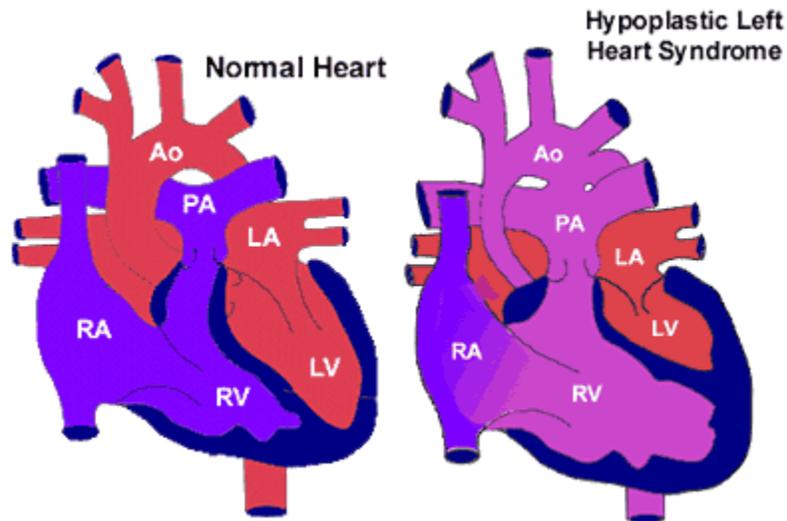


Surgery is required during infancy or early childhood. If severe symptoms develop in the first few months the baby may require early surgery (a so-called "Shunt" operation). This involves insertion of a tiny piece of artificial tube (made from Goretex) between the Aorta, or a branch (usually one of the arm arteries), and one of the branch Pulmonary Arteries (LPA or RPA). Corrective surgery is carried out at around six months.



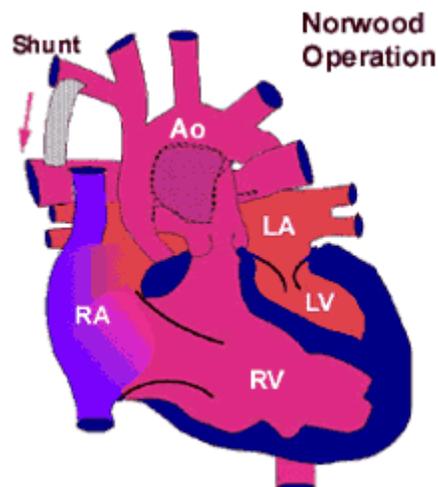
Hypoplastic Left Heart Syndrome HD

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.



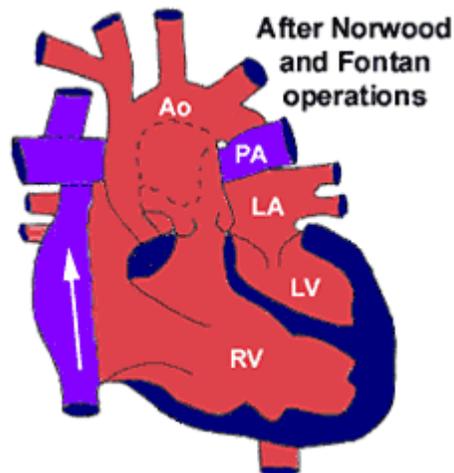
Norwood Operation

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.



Fontan Operation

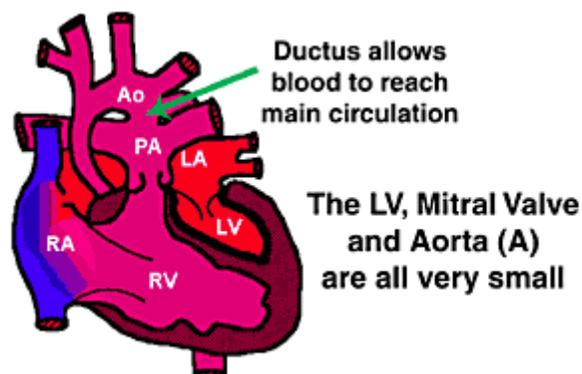
This involves connecting the veins from the main circulation (SVC & IVC) directly to the pulmonary arteries. Blue blood is thus directed into the lungs rather than to the left atrium. A patch is placed to prevent blood passing from the RA to the LA- though sometimes a small hole (a Fenestration) is deliberately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



Hypoplastic Left Heart Syndrome

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.

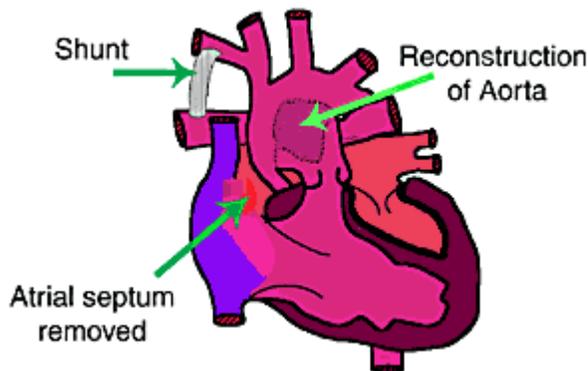
Affected infants usually become severely symptomatic soon after birth as the ductus closes. This is one of the most serious cardiac malformations and leads to death in the newborn period in almost all affected babies, unless surgery or Heart Transplantation can be offered.



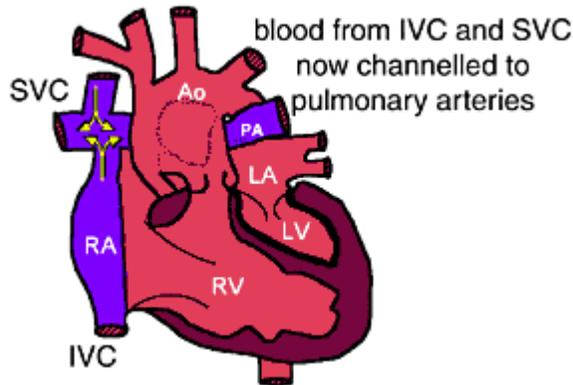
Surgery consists of a “Norwood” operation. This involves connecting the origin of the pulmonary artery to the aorta, to allow the right ventricle to pump blood to the main

circulation and a “Shunt” operation. The atrial septum is removed to allow blood to pass freely from the left atrium to the right side of the heart. The “Shunt” involves 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.

Other surgery is carried out later in childhood and may involve several operations. For many children the final operation is called the Fontan Operation. This leads to blue blood (low in oxygen) being channeled through the lungs, without any “pump” driving it. The right ventricle then pumps the red blood (high in oxygen), round the body. This operation makes the child “Pink”- but does not correct the original problem. Not all affected children need to have a Fontan operation. If they are well, with less drastic surgery, they may not have this “Final” big operation. Some children may be unsuitable for a Fontan operation and alternative forms of treatment may be offered.



'Norwood' Operation



After 'Fontan' Operation

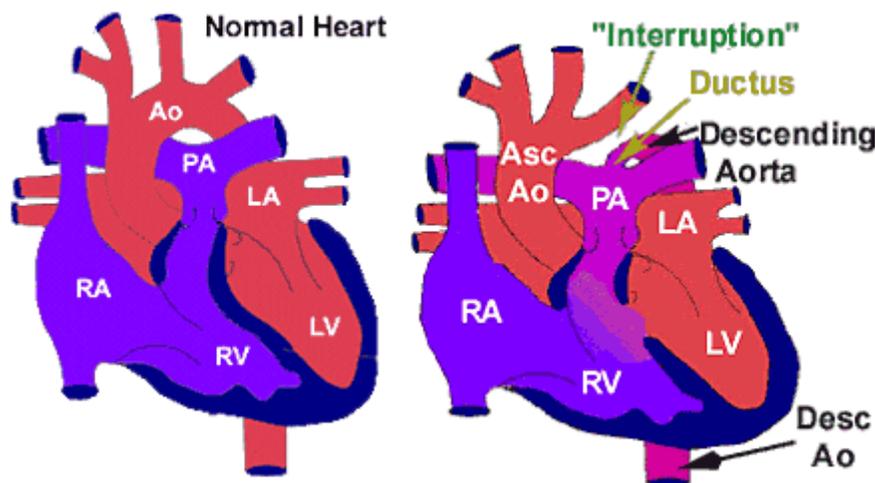
Interrupted Aortic Arch

Part of the Aorta is absent and this leads to severe obstruction to blood flow to the lower part of the body.

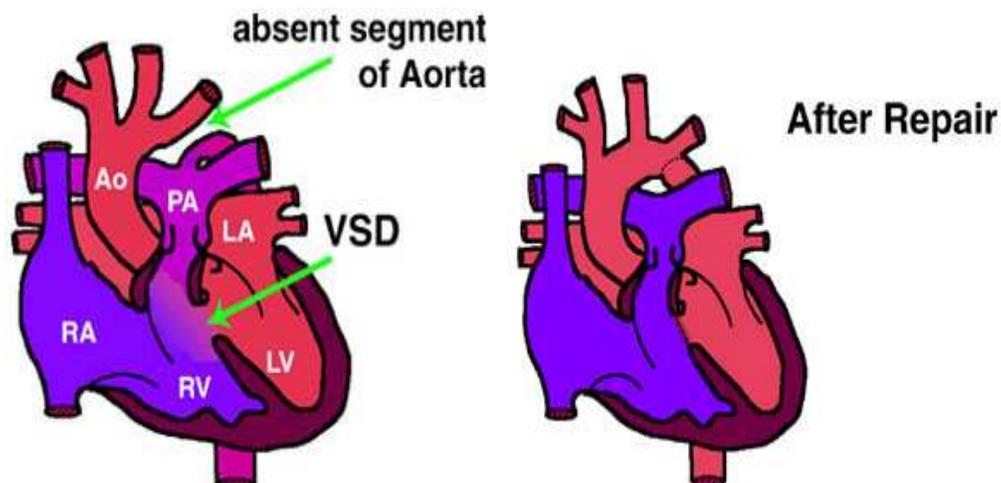
In the immediate newborn period blood flows through the Ductus into the Descending Aorta and hence reaches the lower part of the circulation.

As the Ductus closes after birth blood pressure in the lower circulation becomes inadequate and severe symptoms develop.

Most affected infants develop severe symptoms: difficulty breathing and impaired kidney function in the first week of life and need urgent surgery. Most affected infants also have a large VSD. Sometimes other defects may be present.

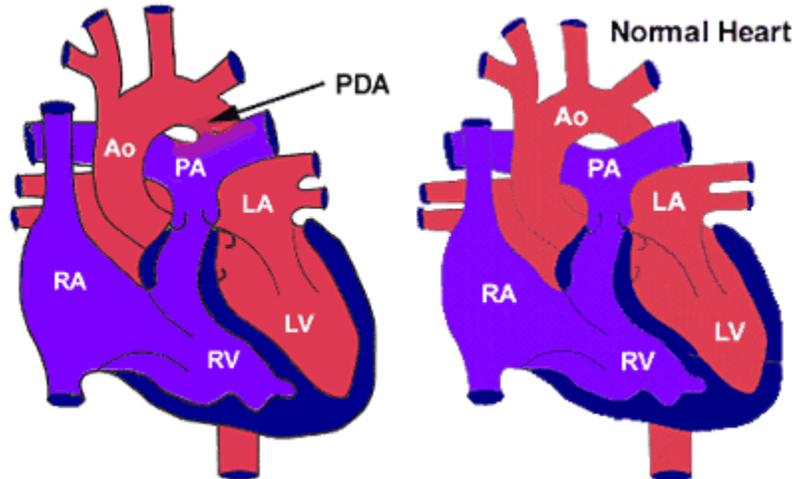


Surgery involves repair of the Aorta and of the associated VSD and other defects. This usually needs to be carried out in the first weeks of life and is difficult and major surgery, though fortunately in most cases the results are now very good.



Patent Ductus Arteriosus (PDA)

Failure of the ductus to close in the early weeks of life, as normally occurs, results in a 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.



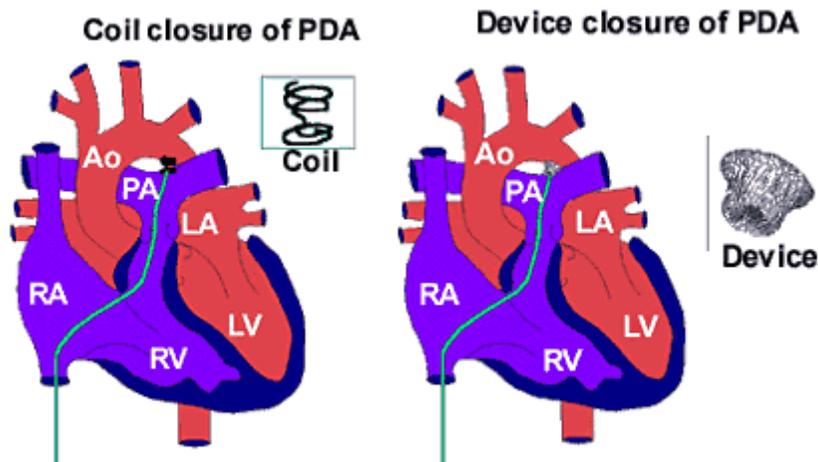
Surgery may be needed when the ductus is large, but in many cases the ductus can be closed using a spring coil introduced with a heart catheter.

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

Device closure of PDA

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

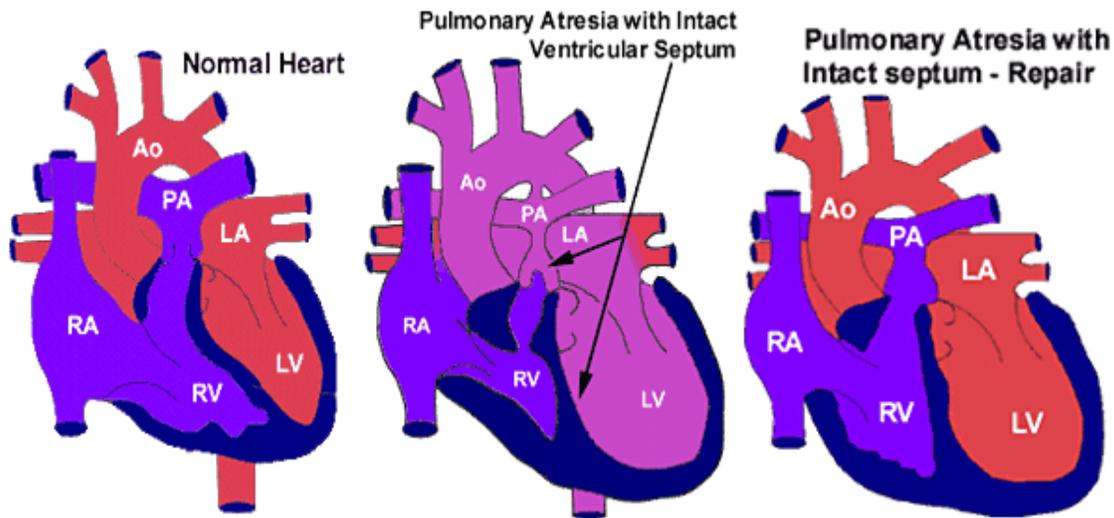
This technique is not suitable for premature babies with a PDA and cannot be used for all affected children, some of whom still need an operation.



Pulmonary Atresia with intact ventricular septum

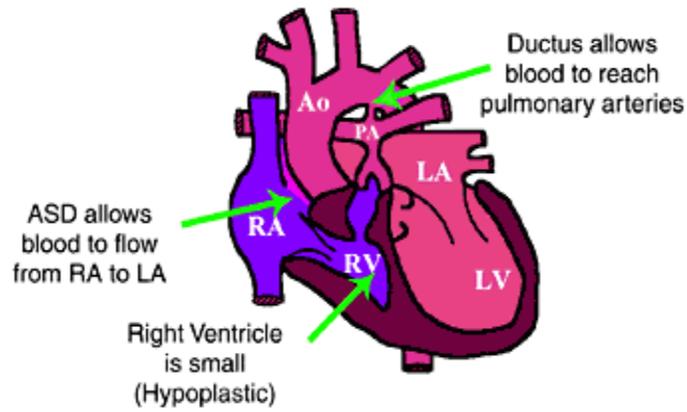
As in Pulmonary Atresia with VSD, this defect is also associated with complete obstruction of the Pulmonary Artery. However, as there is no associated VSD, blood is diverted from the right atrium to the left atrium via the Foramen Ovale. The right ventricle (RV) is usually small (Hypoplastic), though its wall may be thickened (Hypertrophied). Survival depends on the ductus remaining open in the early days of life (in order for blood to reach the lungs), 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.

Complete repair may be possible, but often necessitates several operations.

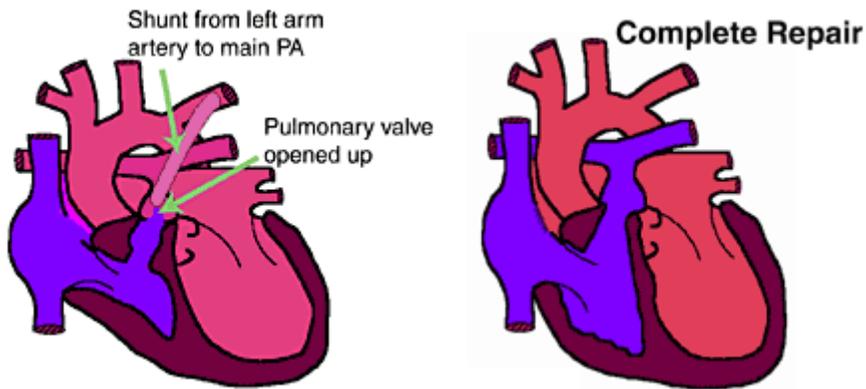


Pulmonary Atresia with Intact Septum

This defect is associated with complete obstruction of the Pulmonary Artery. However, as there is no associated VSD, blood is diverted from the right atrium to the left atrium, via the 'Foramen Ovale' or a defect in the Atrial Septum (ASD). The right ventricle (RV) is usually small (Hypoplastic), though its wall may be thickened (Hypertrophied). Survival depends on the ductus remaining open in the early days of life (in order for blood to reach the lungs). Affected infants usually become blue soon after birth (as the ductus closes). Early surgery often involves opening the Pulmonary Valve and a "Shunt" operation. Corrective surgery is carried out later in childhood if the right ventricle is adequate. In some infants the RV is too small to allow complete repair and other forms of surgery may be required. (e.g. Fontan operation)

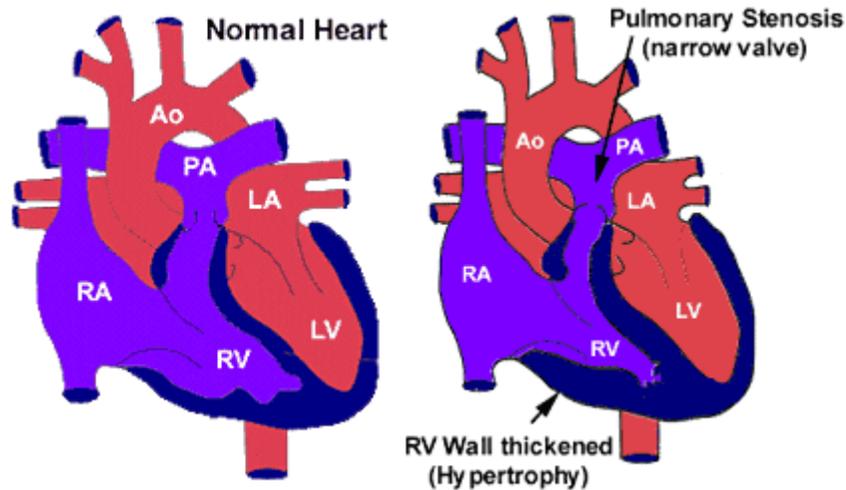


Surgery is often needed urgently, in the first week or two of life and involves a “shunt” operation (often combined with opening the obstructed valve at the same time). Complete repair may be possible later in childhood, if the right ventricle is large enough.



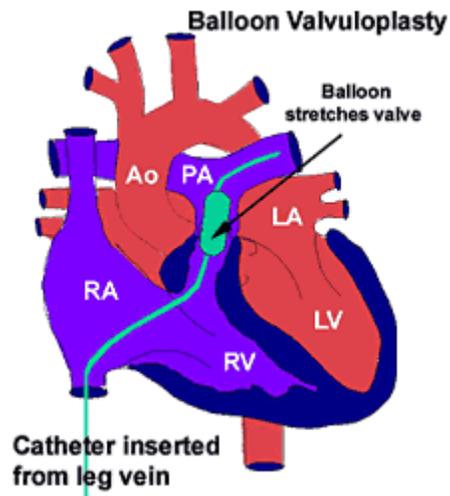
Pulmonary Stenosis

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)



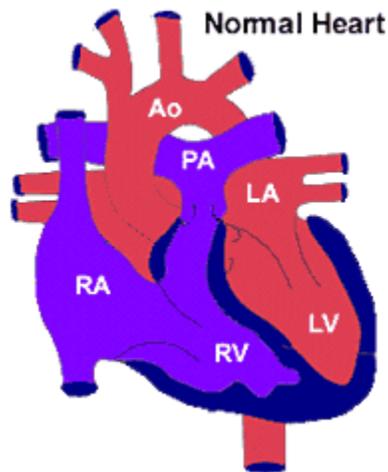
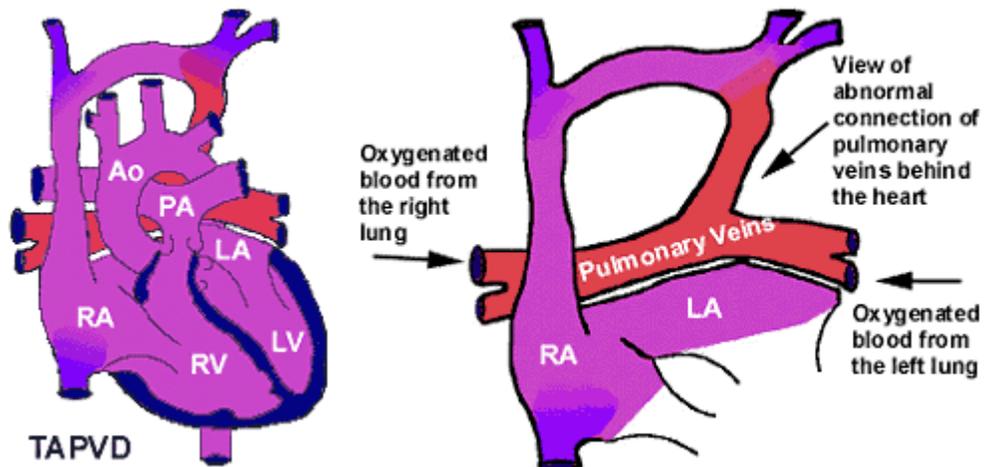
Balloon Valvuloplasty

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 long-term 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.



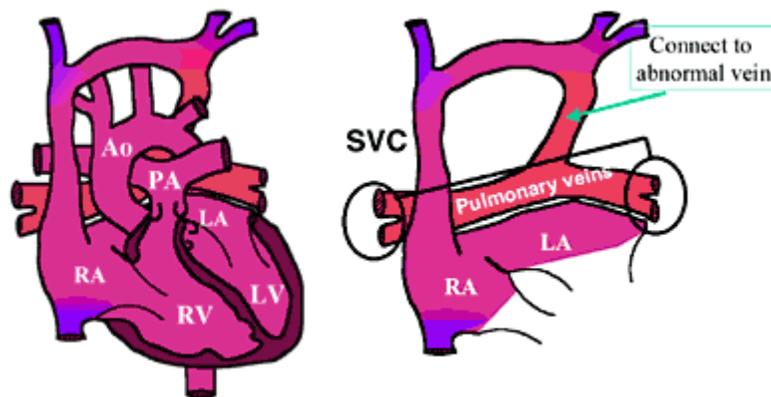
Total Anomalous Pulmonary Venous Drainage

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.



Total Anomalous Pulmonary Venous Drainage (TAPVD)

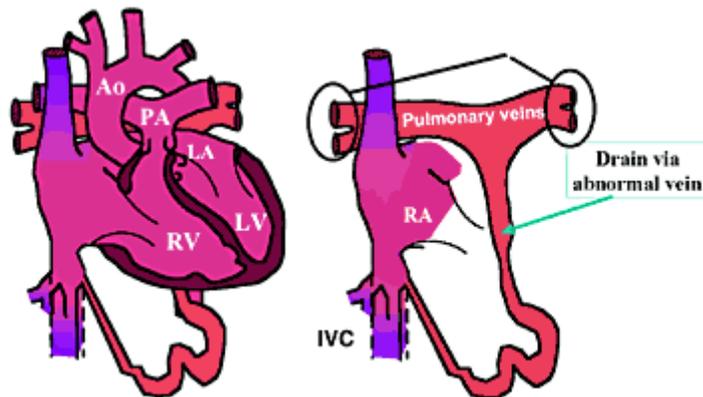
The Pulmonary Veins, which carry blood back to the heart after it has circulated through the lungs, are not connected to the left atrium. Instead they are connected to one of the veins from the main circulation so that the blood returning from the lungs drains back to the right side of the heart. In the example illustrated here all the pulmonary veins are connected to the Superior Vena Cava (SVC). This is referred to as Supracardiac TAPVD.



Pulmonary veins not connected to LA

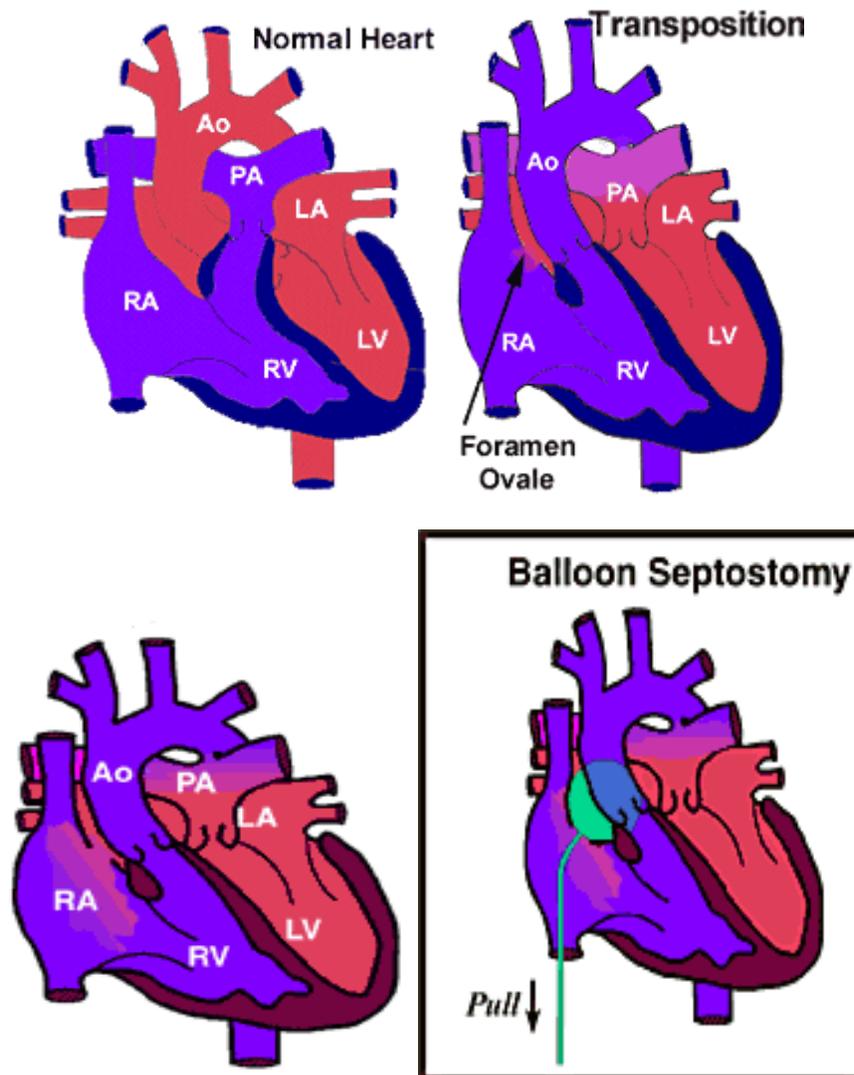
Infracardiac TAPVD

In this form of the defect the blood returning from the lungs returns through the Inferior Vena Cava and is often 'obstructed', leading to early onset of severe symptoms.



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.



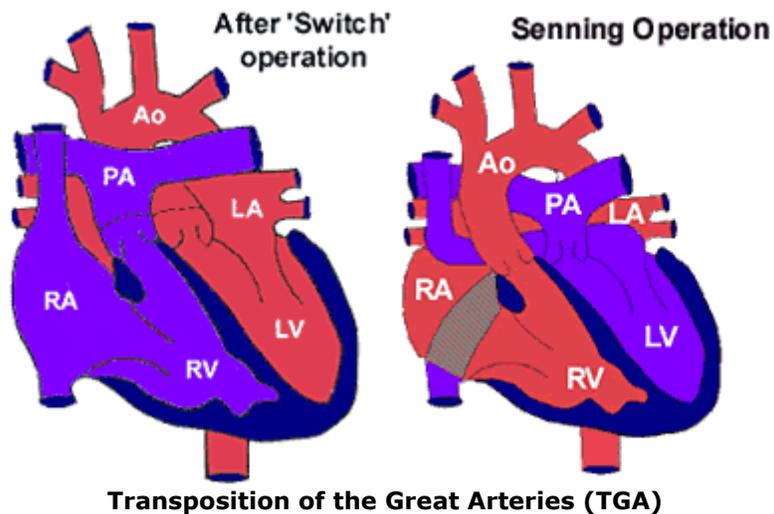
Early surgery is essential and involves the Arterial Switch Operation, which is carried out in the first week or two of life and corrects the abnormality. The small coronary arteries, which feed the heart muscle with blood, need to be transferred as well as the two Great Arteries. (Aorta and Pulmonary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

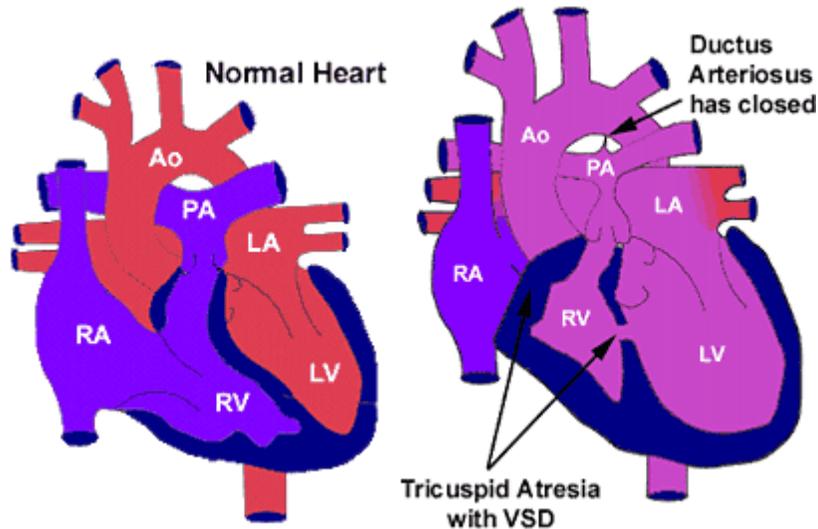
In this operation the blood from the SVC and IVC was redirected to the left atrium and ventricle, while blood from the pulmonary veins was channeled to the right atrium and ventricle. This relieved Cyanosis but did not correct the original abnormality, leaving the Right Ventricle pumping blood to the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

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



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.



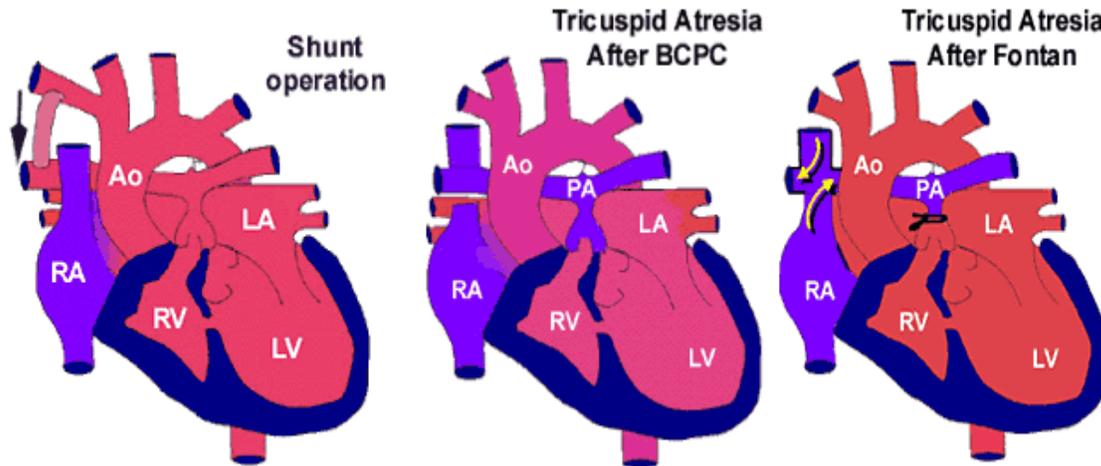
At a second operation some blood from veins in the upper part of the body may be connected directly to the lung arteries. (BCPC)
Definitive surgery is usually delayed until the age of at least two years. It involves the so-called Fontan operation. This involves connecting the veins from the main circulation (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 channeled into the pulmonary arteries and the cyanosis will be completely, or almost completely, relieved.

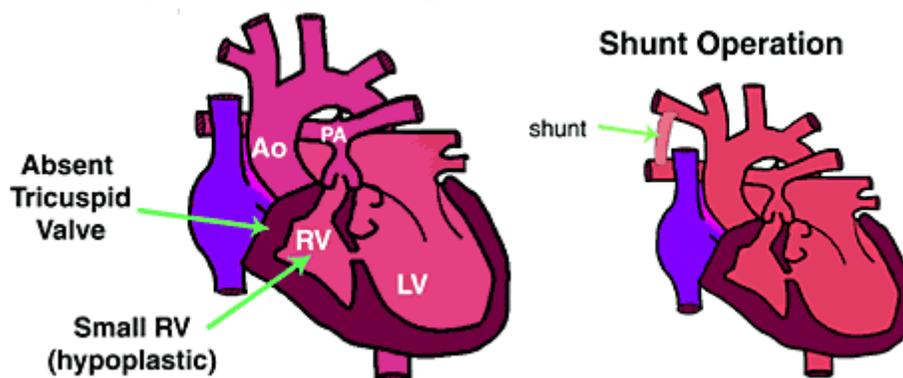
Fontan operation

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.

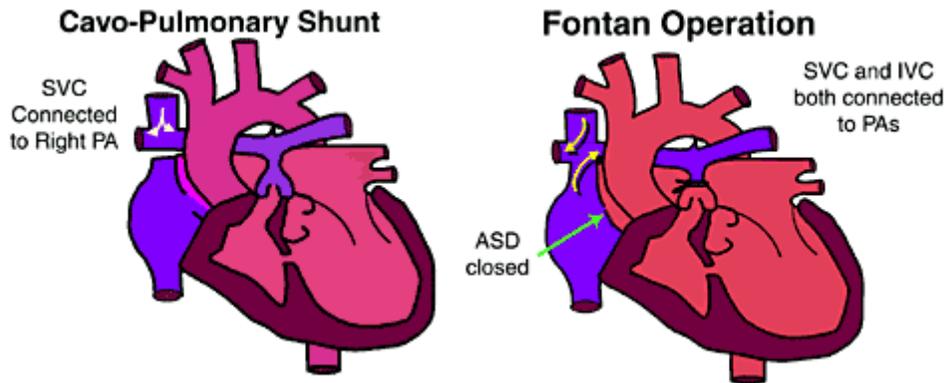


Tricuspid Atresia

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. Affected infants may be well in the early weeks of life, but gradually become blue, as they get older. Early surgery often involves a “Shunt” operation, involving insertion of a tiny piece of artificial tube (made from Goretex) between the right or left arm artery and the pulmonary artery to the respective lung.

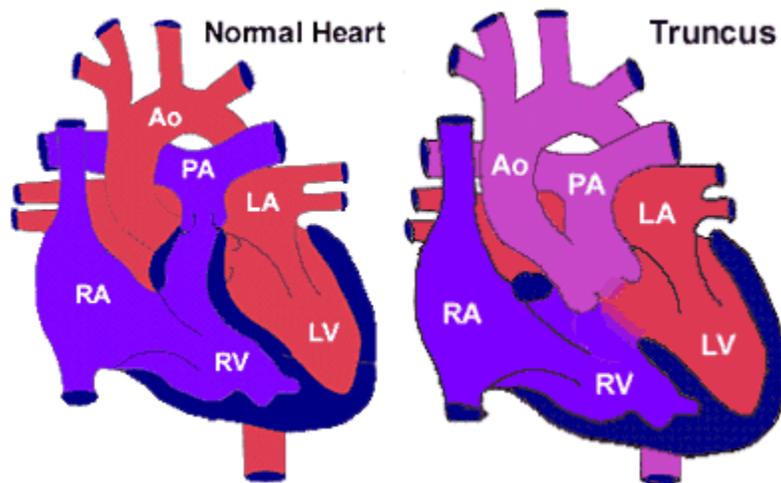


Later a Cavo-Pulmonary shunt may be performed. This involves connecting the Superior Vena Cava (SVC) to the right Pulmonary Artery, to direct blood from the upper part of the circulation into the lungs without returning to the heart. Corrective surgery, a Fontan operation, is carried out later in childhood. This results in the blood flow from the IVC also passing directly to the lungs. After this procedure the lung circulation operates without a separate ventricle (as the right ventricle is too small to perform its usual job). This only works satisfactorily when the lung arteries are well developed and have not been damaged by the effects of high pressure or other factors.

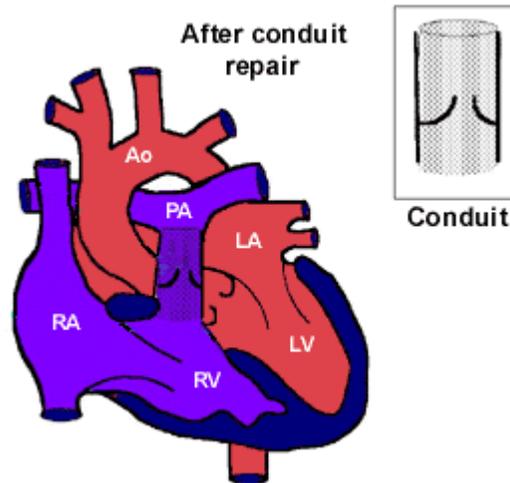


Truncus Arteriosus

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) (“Hole in the heart”)

The most common of all heart defects. 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 heartbeat. 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.

