Adult Congenital Heart Disease (A.C.H.D)

Study Pack

Answer book

SV/MW 2017

ACHD Study Pack Contents

Introduction

As a result of the success of pediatric cardiology and cardiac surgery since

1958, there are now more adults than children with congenital heart disease. Prior to the advent of surgery, less than 20% of children born with congenital heart malformations survived to adult life. Now, most deaths from congenital heart disease occur in adults (D.H. 2006). Today more than 96% of children born with congenital cardiac lesions who survive infancy will live until at least 15 years of age and many more into adulthood. The live incidence for congenital heart disease in the Western World is 1 in every 145 babies born and in the UK it is thought there are up to 150,000 adults with congenital heart disease. There are approximately forty congenital heart defects and many combinations but eight common ones (D.H. 2006).

This pack has been designed to assist you in the care of these adults.

How the study pack works

- Senior staff gives pack to new staff at orientation and all existing bands 5, 6 and 7.
- The Nurse completes the anonymous questionnaire about existing knowledge of adult congenital heart disease.
- After completing the pack nurses will continue to attend on-going sessions to continue their development. This will be discussed at each appraisal.

This pack and the table in the Appendix will also count towards your appraisal and re-validation.

When you see  please answer the questions or complete the exercise!

To assist you there is a Reference List and Bibliography at the end of this pack.

Learning Outcomes for this Study Pack

By completing this pack the RN will:

1) Explain the differences between the foetal heart and the fully developed human heart
2) Define and discuss eight commonly occurring congenital cardiac conditions in relation to their presentation, management, long-term care and consideration of the implications for pregnancy and contraception

The Foetal Heart

The foetal heart begins to be formed during the first few weeks after conception in its' early stages it resembles other animals' hearts. Initially the heart is just a single tube (similar to a fish heart) before the next phase whereby the two atria begin to be formed (frog heart) at this stage there is only one ventricle. After the two atria and single ventricle has formed (snake/turtle heart) the ventricle begins to separate into the four-chambered heart.

See illustration below

The normal foetal heart has three important differences, from the heart after birth, due to the reliance on the mother's blood supply via the placenta for nutrition, oxygen and blood flow:

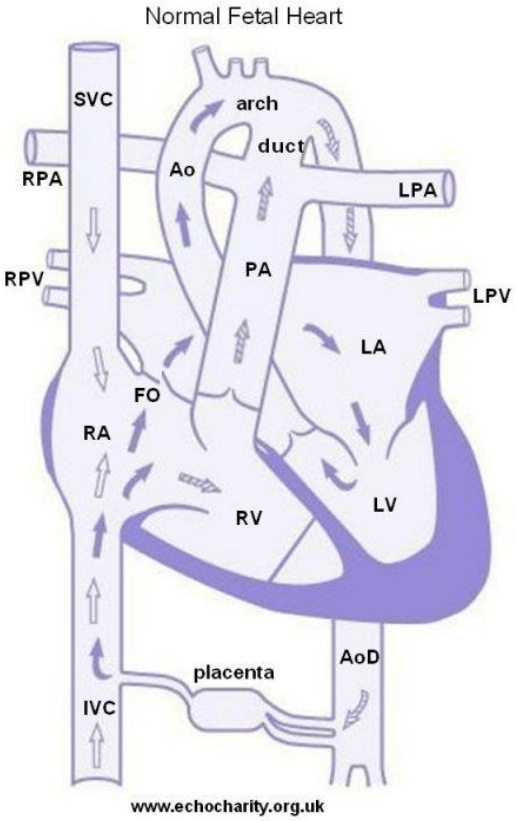
1. The **oval foramen** ("foramen ovale"), labeled **FO**, is covered by a "flap" that allows "red", oxygenated blood from the placenta to enter the left atrium (**LA**). After birth, the **FO flap** closes due to lack of flow from the placenta and high pressure in the left atrium. (the patent foramen ovale may not close).

2. The **duct** ("ductus arteriosis") is a bypass, so that "blue", less oxygen rich blood from the veins does not enter the lungs, but returns to the placenta. At birth, a baby takes its first breath, the lungs inflate and the duct slowly closes to allow the lungs to take over the oxygenation of blood. However, if there is "duct-dependent" heart disease, the closing of the duct may reduce the supply of oxygenated blood to the body causing "blueness" or breathlessness. It is important to detect these conditions before birth to prevent collapse, as the duct can be kept open the drug prostaglandin, until the problem can be corrected.

3. The **placenta** supplies nutrition and oxygen as "red", oxygenated blood from the mother and also removes waste as "blue", less oxygenated blood.

After birth the umbilical cord to the placenta is cut and baby is no longer reliant on mother's blood supply.

Note: the Foetal heart works at much lower pressure than the heart after birth and there is much more mixing of "red" and "blue" blood through the FO flap and duct. This mixing allows a baby to survive even if there is heart disease blocking a valve or a problem with the connections of the chambers and vessels.



Key to labels

- Ao: aorta AoD: descending aorta arch: Aortic arch duct: ductal arch ("ductus arteriosis")
- FO: oval foramen ("foramen ovale") IVC: inferior caval vein
- LA, RA: left, right atrium
- LPA, RPA: left, right pulmonary artery
- LPV, RPV: left, right pulmonary veins
- LV, RV: left, right ventricle
- PA: pulmonary artery SVC: superior cava vein

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1 What are the pre-disposing factors that may contribute to a child being born with congenital heart disease?

- Maternal drugs, anti-epileptics, lithium, alcohol
- Chromosomal Aberrations e.g. 1 in 700 Downs Syndrome. 40% D.S. have C.H.D. Turners/Williams Syndrome.
- Environmental Factors e.g. Radiation
- Infection/Virus e.g. Rubella
- Maternal Conditions e.g. Diabetes

2 What is the incidence of Congenital Heart Disease?

- 1 in every 145 babies born will have congenital heart disease.

3 What screening services are available for women with congenital heart disease during pregnancy?

- Foetal echo 18-22 week

4 Describe the pregnancy services that are available in the South West for women with congenital heart disease?

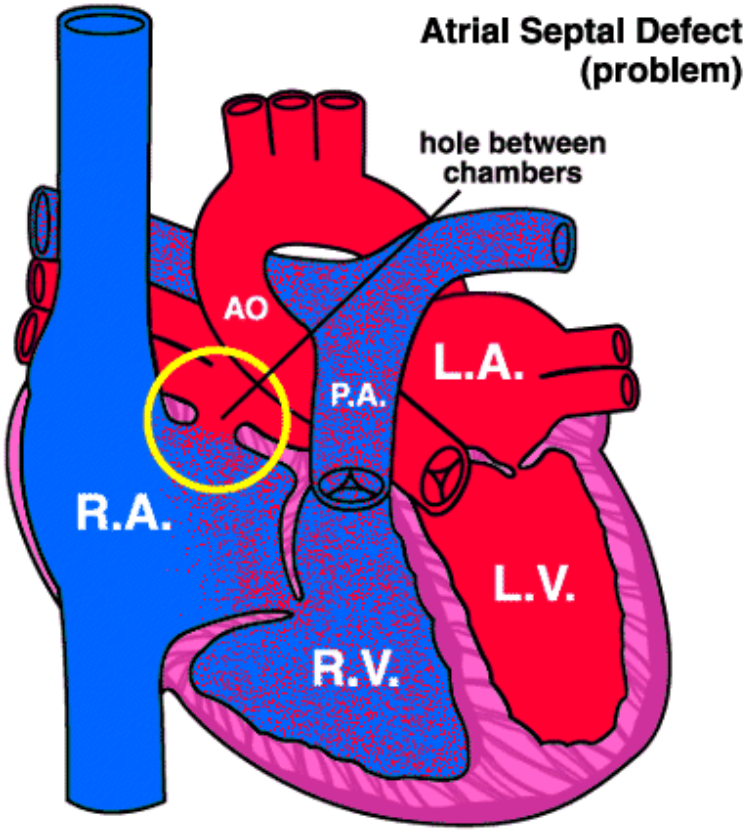
- Pre-pregnancy counselling service to discuss risk, medication and birth plan. This is not necessary for women with simple lesions.
- Pregnancy stratified into low, medium or high risk.
- Joint cardiac and obstetric clinics at St Michaels every month.
- Shared care with District General Hospital
-

5 At what age do patients with congenital cardiac disease move to the adult service? Can you think of any concerns the young person may have about this this?

Usually transfer age 16yrs.
Concerns include being scared of moving to a new hospital, new outpatient department routine, meeting a new team and being admitted to a ward with people older than themselves. Young people are also concerned about the expectations we may have of them, that they do not understand everything. They generally have poor knowledge about their cardiac condition and are at risk of being lost to follow-up at this time.

A brief overview of the most common congenital cardiac conditions

1. Atrial Septal Defect (ASD)

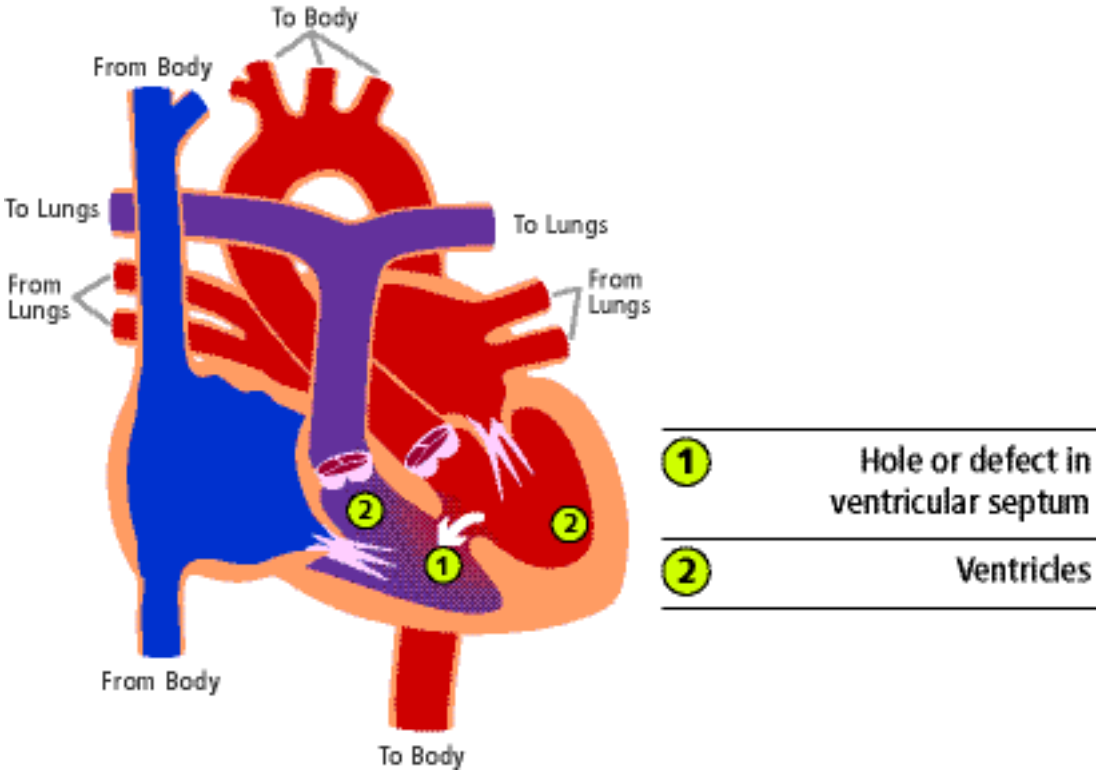


When an atrial septal defect is present, blood flows through the hole primarily from the left atrium to the right atrium. This shunting increases the blood volume in the right atrium that means more blood flows through the lungs than would normally. If left untreated, atrial septal defect may cause problems in adulthood. These problems may include pulmonary hypertension (which is high blood pressure in the lungs), congestive heart failure (weakening of the heart muscle), atrial arrhythmias (which are abnormal rhythms or beating of the heart) and an increased risk of stroke.

Please complete the following table about patients with an ASD

Presentation	May be undiagnosed for years Breathlessness, palpitations, embolus, pulmonary hypertension, murmur or dilated heart on CXR
Specific Investigations to confirm diagnosis	Trans thoracic echo ?size of RV Trans oesophageal echo ?position and size of ASD ECG? atrial arrhythmias Cardiac Cath to measure pulmonary artery pressure if PH suspected
Management (include medical and surgical)	Surgical closure (plus or minus Maze) if ASD is large or in the Sinus Venosus position Percutaneous closure if less than 4cms, away from AV valves and normal pulmonary venous drainage anatomy Anti-platelet treatment post procedure
Long term complications	Late atrial arrhythmias
Risk of endocarditis – low, *moderate or high?*	Low
Risk in pregnancy –*low, medium or high?*	Low
Suitable contraception	Progesterone only

2. Ventricular Septal Defect (VSD)



Ventricular septal defect is the most common congenital heart defect and accounts for 20-30% of children seen in paediatric cardiology clinics. The exact incidence is not known with estimates ranging from 2 to 5 out of every 1000 babies born. The cause of the problem is not well understood. A ventricular septal defect (VSD) is a defect or hole in the wall that separates the lower two chambers of the heart. These chambers are called the ventricles and the wall separating them is called the ventricular septum. A child can have single or multiple ventricular septal defects. Ventricular septal defects also occur in association with more complex heart defects such as Tetralogy of Fallot and transposition of the great vessels (explained later).

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Ventricular septal defects can be further described by 1) size of the defect, 2) location of the defect, 3) whether there is more than one defect present, and 4) the presence or absence of a ventricular septal aneurysm. The size of the defect is usually described as small, moderate, or large. Small defects cause no symptoms during infancy or childhood and often close spontaneously. Moderate and large defects are less likely to close spontaneously, may result in congestive heart failure, and more often require an intervention with a closure device. he term restrictive or non-restrictive are used to describe the size of the defect. The term restrictive describes small defects that allow little or no blood to flow from the left side of the heart to the right side of the heart. Non-restrictive defects are large defects that allow a significant amount of blood to flow from the left side to the right of the heart. This results in excessive blood flow to the lungs, high pulmonary artery pressures, extra work for the heart, and congestive heart failure. Patients with a large V.S.D which has not been closed may develop severe pulmonary hypertension and Eisenmengers syndrome which has a poor prognosis.

Different systems for describing the location of ventricular septal defects are used. Some are located in the lower portion of the septum called the muscular septum. Defects in this location are called muscular ventricular septal defects. Perimembranous ventricular septal defects (also called membranous VSD'S) are located in the membranous septum, a relatively small portion of the septum located near the heart valves. Ventricular septal defects may also be described as inlet or outlet VSDs. These terms further describe where the defect is located. Inlet VSDs are located close to where the blood enters the ventricular chamber and outlet VSDs are located close to where the blood exits the ventricular chamber.

Please complete the following table about <u>ADULT</u> patients with a VSD

Presentation	Unusual finding in adulthood. Depends on the size of the VSD. Can present in immigrant population where heath screening is not good Often asymptomatic. Survivors of large unoperated VSD may have developed pulmonary hypertension Aortic regurgitation Atrial fibrillation
Specific Investigations to confirm diagnosis	Loud murmur ECG CXR will show enlarged heart Echo determines size, location and haemodynamic consequences, number of defects and associated lesions Cardiac catheterization to calculate the size of shunt.
Management (include medical and surgical)	Indications for treatment, Limited treatment if pulmonary hypertension such as pulmonary vasodilators Aortic regurgitations due to valve prolapse Surgery Surgery must avoided in Eisenmengers Device closure Previous endocarditis? surgery
Long term complications	Aortic regurgitation, atrial fibrillation.
Risk in pregnancy low, *medium or high?*	Low High in Eisenmengers

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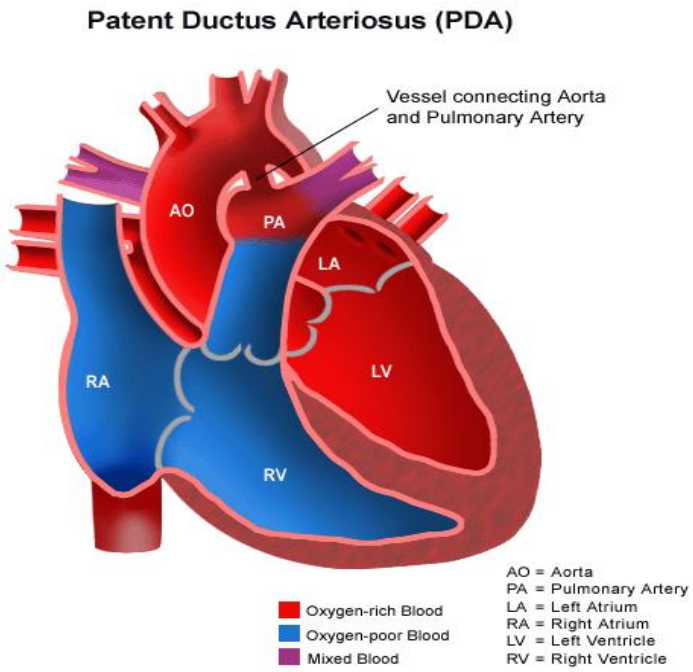
Suitable contraception	Any combined oral contraception.

3. Patent Ductus Arteriosus (PDA)



Patent ductus arteriosus (PDA) is a condition in which the connecting blood vessel between the pulmonary artery and the aorta in foetal circulation, called the ductus arteriosus, stays open in a newborn baby. Because the placenta does the work of exchanging oxygen (O2) and carbon dioxide (CO2) through the mother's circulation, the foetal lungs are not used for breathing. Instead of blood flowing to the lungs to pick up oxygen and then flowing to the rest of the body, the foetal circulation shunts (bypasses) most of the blood away from the lungs. In the fetus, blood is shunted from the pulmonary artery to the aorta through the ductus arteriosus. However, with the first breaths of air the baby takes at birth, the foetal circulation changes. A larger amount of blood is sent

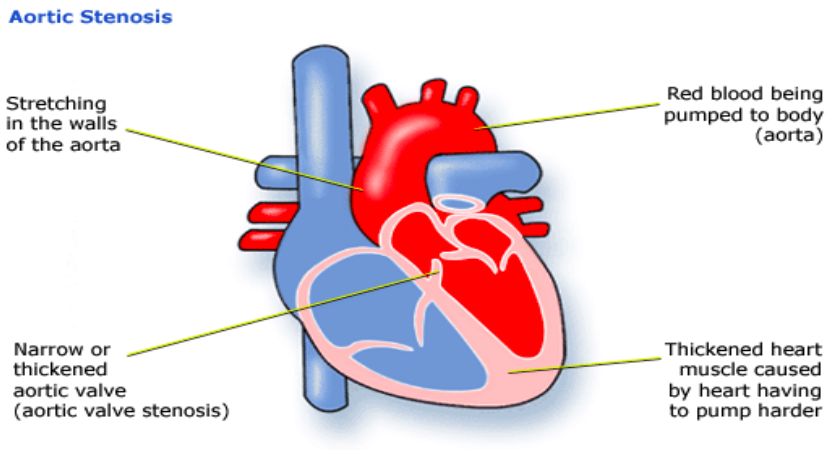
to the lungs to pick up oxygen. Because the ductus arteriosus is no longer needed, it normally begins to wither and close off. Some babies are more likely to have a PDA, especially premature babies. Babies with respiratory problems at birth may have a difficult time increasing the pressure inside the lungs and changing the blood flow. A PDA is also a common congenital heart defect. It is twice as common in females as in males.

Problems are more likely to occur if the opening of the PDA is large. It causes too much blood to flow to the lungs and not enough to the other parts of the body. There can be changes in blood pressure, this can cause heart enlargement as the heart tries to make up for the abnormal blood flow. Severe PDA can cause slow growth, and may result in heart failure. In premature babies, PDA can complicate respiratory problems, making the distribution of oxygen more difficult.

Please complete the following table about ADULT patients with a PDA

Presentation	Murmur
Specific Investigations to confirm diagnosis	Trans thoracic echo Chest x ray ECG
Management (include medical and surgical)	Closure recommended with percutaneous device if murmur detectable Ducts up to 14mm diameter usually suitable for closure Exclude PH
Long term complications	Pulmonary Hypertension Endocarditis Atrial tachycardia
Risk of endocarditis – *low, moderate or high?*	Moderate
Risk in pregnancy –*low, medium or high?*	Low High risk if Pulmonary Hypertension
Suitable contraception	In a small duct, any contraception.

4. Aortic Stenosis (AS)



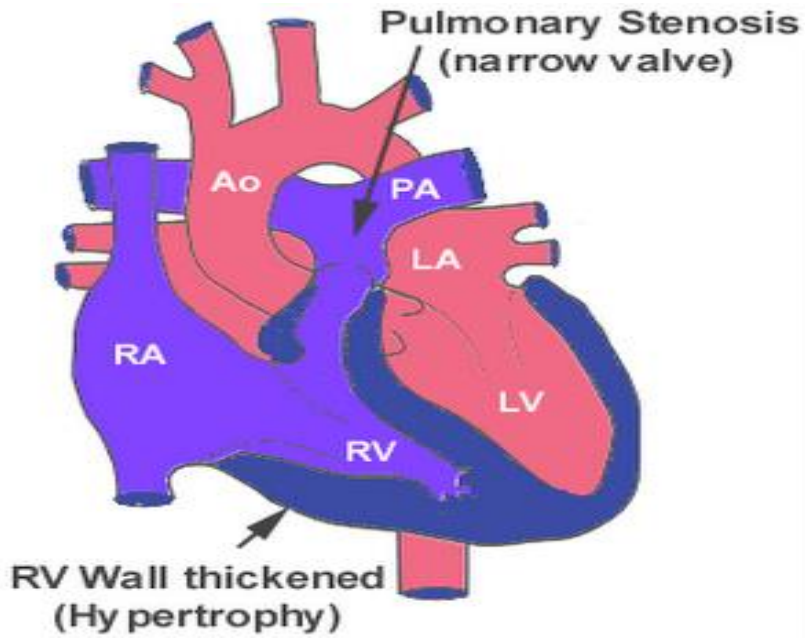
Aortic stenosis is a narrowing of the aortic valve. This means that the flow of blood from the left ventricle into the aorta is less efficient. The aortic valve itself may be too narrow, or there may be narrowing above the valve – supravalvular stenosis or below the valve – sub aortic membrane seen in patients with **Williams syndrome**. The narrowing may be mild, moderate or severe. Aortic stenosis can cause problems with the development of the left side of the heart whilst the baby is growing in the womb. Sometimes this can be seen during a pre-natal scan. After birth, the sound of the blood being forced through the narrow aortic valve can be heard as a heart murmur. In severe cases, the condition will cause a baby to become pale and have difficulties feeding properly. These symptoms are caused by not enough oxygenated blood circulating around the body. A severe narrowing in an older child may result in dizziness, fainting and pain in the chest. If a child's heart condition is complex, aortic stenosis may be only one of a number of defects affecting them.

A bicuspid aortic valve is a very common anomaly affecting 1-2% of the population with male predomination.

Please complete the following table about <u>Adult</u> patients with Aortic Stenosis

Presentation	Breathlessness. Chest pain, syncope Murmur. Systolic left ventricular dysfunction Asymptomatic to symptomatic during exercise testing
Specific Investigations to confirm diagnosis	Echo (exclude BAV). MRI (exclude Bicuspid A Valve) Exercise asymptomatic patients to assess exercise tolerance) BP and Holter monitoring.
Management (include medical and surgical)	Valvuloplasty Aortic Valve Replacement surgery
Long term complications	Ventricular arrhythmias/sudden death Dilated aortic root. Mitral regurgitation
Risk of endocarditis – *mild, moderate or severe?*	High
Risk in pregnancy –*low, moderate or severe?*	High
Suitable contraception	If stenosis is mild any contraception is suitable Progesterone only if moderate or severe.

5. Pulmonary Stenosis (PS)



Pulmonary stenosis is a condition characterized by obstruction to blood flow from the right ventricle to the pulmonary artery.

This obstruction is caused by narrowing or stenosis at one or more of several points from the right ventricle to the pulmonary artery. It includes obstruction from thickened muscle below the pulmonary valve, narrowing of the valve itself, or narrowing of the pulmonary artery above the valve. The most common form of pulmonary stenosis is obstruction at the valve itself, referred to as pulmonary valvar stenosis. The normal pulmonary valve consists of three thin and pliable valve leaflets. When the right ventricle ejects blood into the pulmonary artery, the normal pulmonary valve leaflets spread apart easily and cause no obstruction (blockage) to outflow of blood from the heart.

Pulmonary valve stenosis occurs when abnormalities of the pulmonary valve lead to narrowing and obstruction between the right ventricle and the pulmonary artery. Most commonly, the pulmonary valve leaflets are

thickened and fused together along their separation lines (commissures).
When the tissue is thickened, the leaflets become less pliable than normal and this also contributes to the obstruction. At times, the diameter of the pulmonary valve itself is small or hypoplastic. When the pulmonary valve is obstructed, the right ventricle must work harder to pump blood into the pulmonary artery.

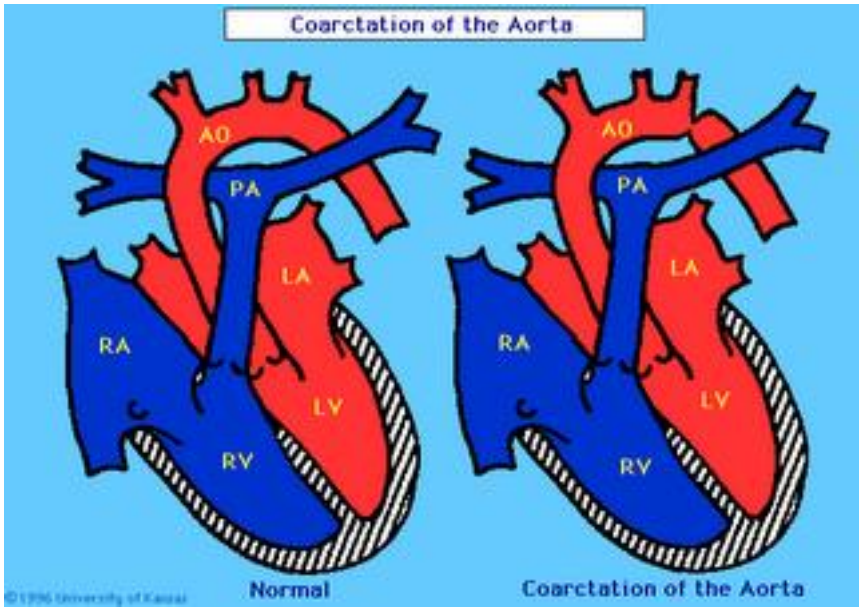
To compensate for this additional workload, the muscle of the right ventricle (the myocardium) gradually thickens to provide additional strength to right ventricular ejection. The increased right ventricular muscle, known as hypertrophy, is rarely a problem in itself, but instead is an indication that significant valve obstruction exists. When the pulmonary valve is severely obstructed, especially in newborns with critical degrees of pulmonary stenosis, the right ventricle cannot eject sufficient volume of blood flow into the pulmonary artery. In these instances, blue blood bypasses the right ventricle flowing from the right atrium to left atrium, through the foramen ovale, a communication or "hole" between these two chambers that is normally present in newborns. Newborns with critical pulmonary stenosis therefore will have cyanosis (blue discoloration of the lips and nailbeds) due to lower oxygen levels in their blood. In such cases treatment options include surgery and percutaneous pulmonary valvuplasty. Right ventricular failure can occur with pulmonary valve stenosis.

Symptoms in adults will be shortness of breath. Symptoms of breathlessness do not necessarily reflect the severity of the stenosis. Pulmonary stenosis can also be part of more complex lesions eg Fallots Tetralogy.

Please complete the following table about <u>Adult</u> patients with Pulmonary Stenosis

Presentation	Symptoms rare in mild to moderate stenosis Severe stenosis may present with exertional dyspnoea, atrial arrhythmias, fatigue and chest pain
Specific Investigations to confirm diagnosis	ECG-right ventricular hypertrophy CXR- main pulmonary artery dilated Echo-Pulmonary valve anatomy, severity of PS, RV size and function MRI-quantify RV size, identify PA stenosis
Management (include medical and surgical)	If symptomatic Percutaneous balloon valvuloplasy/ percutaneous valve replacement Surgical pulmonary valvotomy /pulmonary valve replacement
Long term complications	Depending on intervention Valvuloplasty may lead to pulmonary regurgitation. Pulmonary valve replacement may re-stenos
Risk of endocarditis – *mild, moderate or severe?*	Moderate to high depending on stenosis
Risk in pregnancy low, *medium or high?*	Low, unless severe pulmonary stenosis.

6. Coarctation of the Aorta



Coarctation of the aorta is a narrowing of the aorta, the main blood vessel carrying oxygen-rich blood from the left ventricle of the heart to the organs of the body.

Coarctation occurs most commonly in a short segment of the aorta just beyond where the arteries to the head and arms branch off, as the aorta arches inferiorly toward the abdomen and legs. This portion of the aorta is called the "juxta-ductal" part of the aorta, or the part near where the ductus arteriosus attaches. The ductus arteriosus is a blood vessel that is normally present in a fetus and has special tissue in its wall that causes it to close in the first hours or days of life. It is thought that coarctation may be caused by the presence of extra ductal tissue extending into the adjacent aorta which results in aortic narrowing as the ductal tissue contracts.

In babies with coarctation, the aortic arch may be small (hypoplastic). Coarctation may also occur along with other cardiac defects, typically involving the left side of the heart. The defects most commonly seen with

coarctation are bicuspid aortic valve and ventricular septal defect. Coarctation may also be seen as a part of more complex, single ventricle cardiac defects. Coarctation of the aorta is common in patients with some chromosomal abnormalities, such as **Turner's syndrome.**

In the presence of a coarctation, the left ventricle has to work harder, since it must generate a higher pressure than normal to force blood through the narrow segment of the aorta to the lower part of the body. If the narrowing is severe, the ventricle may not be strong enough to perform this extra work resulting in congestive heart failure or inadequate blood flow to the organs of the body.

The age at which coarctation is detected depends on the severity of the narrowing. In approximately 50 percent of cases of isolated coarctation, the narrowing is severe enough to cause symptoms in the first days of life when the ductus arteriosus closes. When the ductus arteriosus closes, the left ventricle must suddenly pump against much higher resistance which can lead to heart failure. In addition, there is impaired blood flow to the organs beyond the coarctation. Because these newborns are well until the ductus arteriosus closes, symptoms appear rapidly and are often severe. When a coarctation goes undetected in the newborn period, it may go unrecognized for many years in some cases. Since the narrowing is generally less severe or has progressed more slowly, the left ventricle has had time to thicken (hypertrophy) in order to pump against the narrowing. Adults will often be discovered to have coarctation of the aorta through investigations for hypertension which may be discovered at a work medical, ante-natal clinic or hospital admission. It can also cause headaches, nosebleeds and leg cramps, particularly with exercise. Pulses distal to the obstruction are diminished and delayed and auscultation reveals a systolic or continuous murmur.

Please complete the following table about <u>Adult</u> patients with Coarctation of the Aorta

Presentation	In in adulthood, hypertension, headache, nose bleeds, dizziness, shortness of breath, tinnitus, less commonly claudication, abdominal angina, cold feet heart failure, cerebral haemorrhage during investigation for bicuspid aortic valve Other associations, bicuspid aortic valve in 80% cases, VSD,PDA, mitral valve abnormalities, Turners syndrome, aneurysm of the Circle of Willis.
Specific Investigations to confirm diagnosis	R arm BP ECG, LVH CXR, notching, dilated aorta, prominent aortic knuckle Echo, LVH and associated lesions MRI, collaterals and related vessels (every 3-5 yrs.) Coronary angiogram
Management (include medical and surgical)	Trans catheter balloon dilatation and stenting in selected adults and older children, operated patients with residual coarctation. The procedure carries a risk of aortic dissection and aneurysm formation. Surgical repair may be required; a variety of procedures may be used. There is a risk of spinal cord injury.
Long term complications	Arterial hypertension, re-coarctation or residual stenosis, aneurysm formation, left heart failure, intracranial haemorrhage, infective endocarditis, premature coronary, cerebral artery disease, Progression of associated lesions. Haemoptysis requires urgent aortic C.T; this may indicate aortic dissection or ruptured aneurysm.

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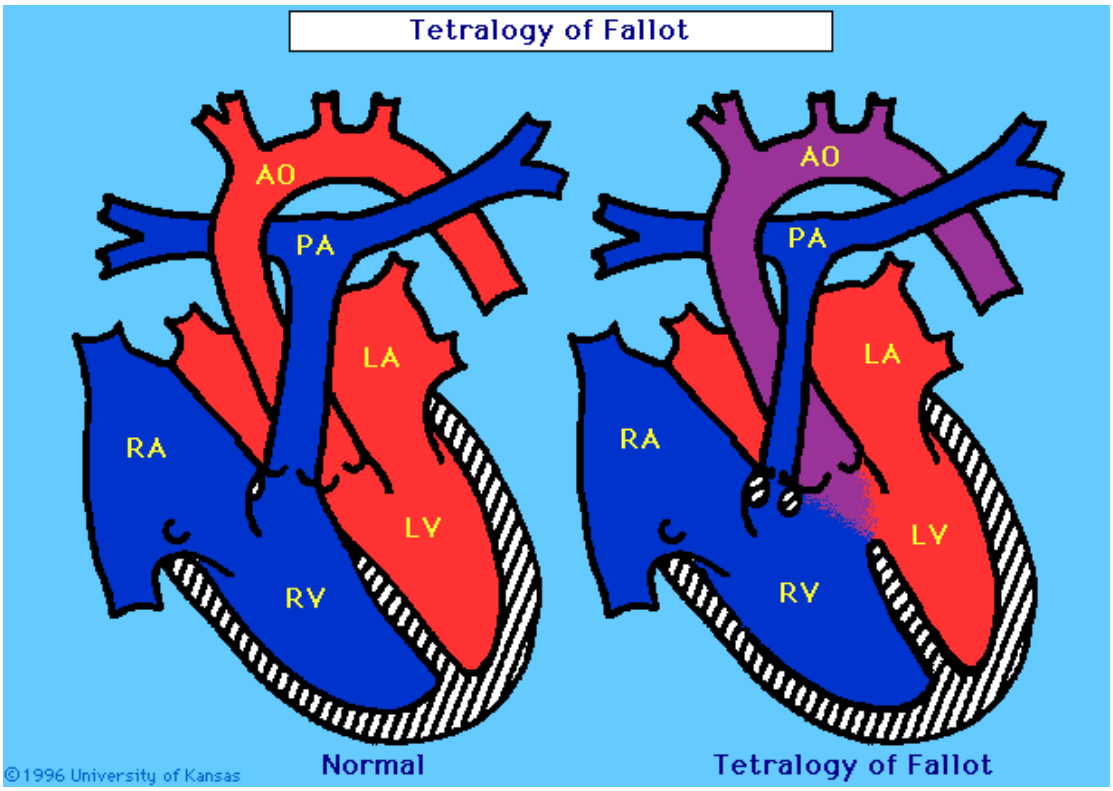
Risk of endocarditis – *mild, moderate or severe?*	High risk in patients with prosthetic material or are un repaired.
Risk in pregnancy –low, medium or high?	Moderate Risk of Aortic rupture, rupture of berry aneurysm.in those after repair or those with arterial hypertension.
Suitable contraception	Progesterone only, if hypertensive.

7. Tetralogy of Fallots



Tetralogy (Greek-four) of Fallot (named after Etienne-Lois Arthur Fallot) is a cardiac anomaly that refers to a combination of four related heart defects that commonly occur together. The four defects include:

- Pulmonary stenosis (narrowing of the pulmonary valve and

outflow tract or area below the valve, that creates an obstruction (blockage) of blood flow from the right ventricle to the pulmonary artery

- Ventricular septal defect (VSD)
- Overriding aorta (the aortic valve is enlarged and appears to arise from both the left and right ventricles instead of the left ventricle as occurs in normal hearts)
- Right ventricular hypertrophy (thickening of the muscular walls of the right ventricle, which occurs because the right ventricle is

pumping at high pressure)

A small percentage of children with tetralogy of Fallot may also have additional ventricular septal defects, an atrial septal defect (ASD) or abnormalities in the branching pattern of their coronary arteries. Some patients with tetralogy of Fallot have complete obstruction to flow from the right ventricle, or pulmonary atresia. Tetralogy of Fallot may be associated with chromosomal abnormalities, such as 22q11 deletion syndrome.

The pulmonary stenosis and right ventricular outflow tract obstruction seen with tetralogy of Fallot usually limits blood flow to the lungs. When blood flow to the lungs is restricted, the combination of the ventricular septal defect and overriding aorta allows oxygen-poor blood ("blue") returning to the right atrium and right ventricle to be pumped out the aorta to the body. This "shunting" of oxygen-poor blood from the right ventricle to the body results in a reduction in the arterial oxygen saturation so that babies appear cyanotic, or blue. The cyanosis occurs because oxygen-poor blood is darker and has a blue color, so that the lips and skin appear blue. The extent of cyanosis is dependent on the amount of narrowing of the pulmonary valve and right ventricular outflow tract. A narrower outflow tract from the right ventricle is more restrictive to blood flow to the lungs, which in turn lowers the arterial oxygen level since more oxygen-poor blood is shunted from the right ventricle to the aorta.

Tetralogy of Fallot is most often diagnosed in the first few weeks of life due to

a loud murmur or cyanosis. Babies with tetralogy of Fallot usually have a patent ductus arteriosus at birth that provides additional pulmonary blood flow, so severe cyanosis is rare early after birth. As the ductus arteriosus closes, as it typically will in the first days of life, cyanosis can develop or become more severe. The degree of cyanosis is proportional to lung blood flow and thus depends upon the degree of narrowing of the outflow tract to the pulmonary arteries. Rapid breathing in response to low oxygen levels and reduced pulmonary blood flow can occur. The heart murmur, which is commonly loud and harsh, is often absent in the first few days of life.

The arterial oxygen saturation of babies with Tetralogy of Fallot can suddenly drop markedly. This phenomenon, called a "tetralogy spell," usually results from a sudden increased constriction of the outflow tract to the lungs so that pulmonary blood flow is further restricted. The lips and skin of babies who have a sudden decrease in arterial oxygen level will appear acutely more blue. Children having a tetralogy spell will initially become extremely irritable in response to the critically low oxygen levels, and they may become sleepy or unresponsive if the severe cyanosis persists. A tetralogy spell can sometimes be treated by comforting the infant and flexing the knees forward and upward. Adults having tetralogy of Fallot who have undergone total surgical repair will have improved hemodynamics and often have good cardiac function after the operation. Ninety percent of patients with total repair as infants develop a progressively leaky pulmonary valve as the heart grows to its adult size, which will require further pulmonary valve surgery. Patients may have damage to the electrical system of the heart from surgical incisions, causing abnormalities as detected by ECG and/or arrhythmias such as ventricular tachycardia.

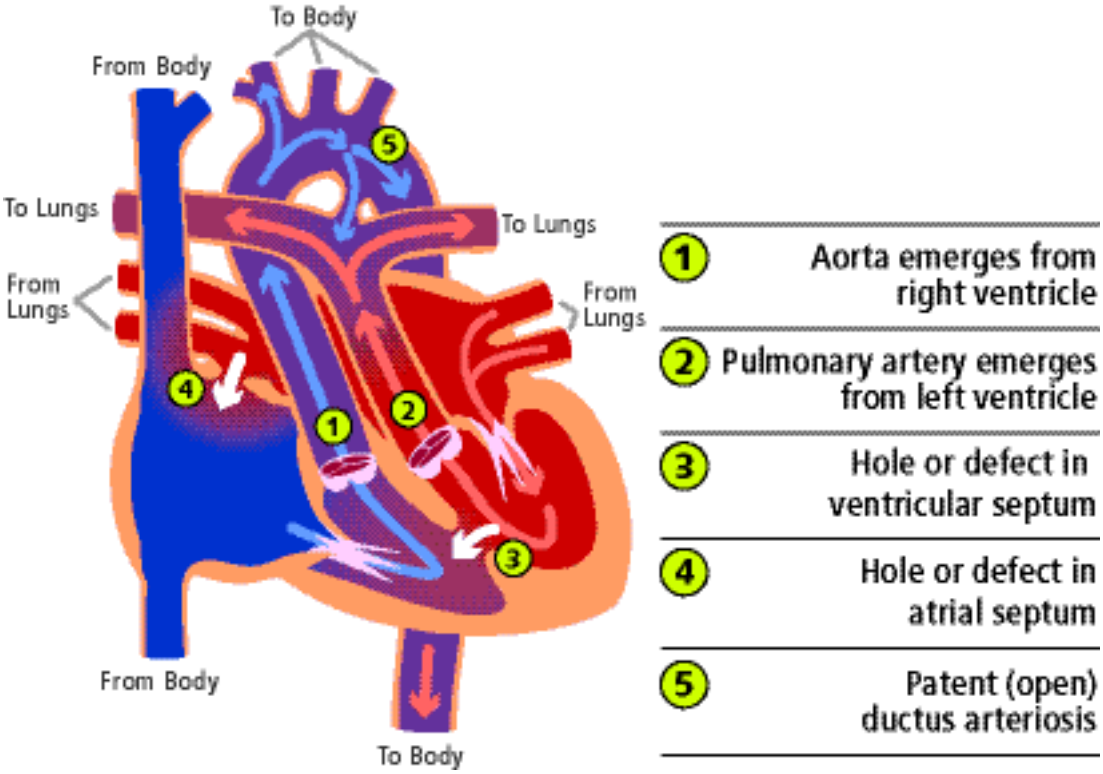
In adult life complications will include arrhythmias, right sided heart failure and leaking pulmonary valve.

Please complete the following table about patients with Tetralogy of Fallot

Presentation	Early clinical presentation is dominated by a heart murmur in infancy and progressive cyanosis Unoperated carries a poor prognosis Early management includes palliative shunts Blalock- Taussig shunt, Waterston shunt, Potts shunt **Late clinical presentation** -Pulmonary regurgitation -Residual right ventricular outflow tract obstruction (RVOTO) -RV dilatation and dysfunction -Residual VSD -Aortic root dilatation with Ao regurgitation -LV dysfunction -Atrial/ventricular tachycardia -Sudden cardiac death -Bi-ventricular failure -Endocarditis
Specific Investigations to confirm diagnosis	ECG, generally complete right bundle branch block, QRS width reflects degree of RV dilatation. A QRS ≥180ms, is a risk factor for VT and sudden cardiac death Echocardiography to assess RVOTO and PR, residual VSD, RV and LV size and function, tricuspid regurgitation, aortic root size and aortic regurgitation Cardiac MRI to assess RV volume and function, pulmonary regurgitation, assess pulmonary arteries and aortic root CT in patients with pacemaker or ICD and can assess coronary arteries and extent of calcification

	Cardiopulmonary exercise testing assesses timing of re-intervention and provides prognosis information Holter monitoring/ event recorder are required for patients with arrhythmia and those at risk of sudden cardiac death Cardiac catheterization for patients undergoing catheter based interventions and when non-invasive evaluation is inconclusive.
Management (include medical and surgical)	Late surgical/catheter surgical interventional treatment -Pulmonary valve replacement and relief of RVOTO for pulmonary regurgitation (Tissue). Residual VSD and aortic root dilatation should be addresses at this time Percutaneous pulmonary valve may be considered in selected patients Ablation
Long term complications	See above, **Late clinical presentation**.
Risk of endocarditis – *mild, moderate or severe?*	High risk if unrepaired High risk if repaired with prosthetic material
Risk in pregnancy – low, medium or high	Moderate, depends on haemodynamic status.
Suitable contraception	Progesterone only, if risk of atrial arrhythmia

8. Transposition of the Great Arteries



Complete transposition of the great arteries (TGA) occurs when the aorta, which normally leaves the left ventricle and pumps red blood to the body, arises from the right ventricle and pumps blue blood returning from the body back to the body bypassing the lungs completely. The pulmonary artery, which normally arises from the right ventricle and pumps blue blood to the lungs, arises from the left ventricle and sends red blood returning from the lungs right back to the lungs. Essentially, the great arteries are reversed from their normal connection. The cause of the problem is not understood. It is the most common form of cyanotic congenital heart disease which presents in the newborn period. It is more common in males and the babies are usually normal birth weight and size. TGA accounts for 5 to 7% of all congenital heart defects.

There are several other abnormalities that may occur along with TGA. The most common associated problem is a ventricular septal defect. This is a defect or hole in the wall that separates the lower two chambers of the heart, the ventricles. There may be narrowing of the area of the heart where blood flows out to the pulmonary artery. This is called left ventricular outflow tract obstruction LVOTO.

Many of these babies have an atrial septal defect and/or a patent ducts arteriosus. This is normal channel between the aorta and pulmonary artery present at birth that may fail to close in the presence of other heart problems.

When a baby has TGA, there are two separate circuits of blood flow instead of a connected one. Blue blood returning from the body is pumped right back out to the body and red blood returning from the lungs is pumped right back to the lungs. As a result, the baby develops cyanosis, shortly after birth. The cyanosis can be noticed in the lips or under the fingernails. In a baby with cyanosis, it does not improve with the use of oxygen.

The baby could soon die from lack of oxygen delivery to the body. The only way a baby with TGA can survive after birth is if there is a way for the red and blue blood to mix together within the heart so that some red blood gets pumped out to the body. An atrial septal defect and/or a patent ducts arteriosus will usually permit enough oxygen to allow the baby to survive until a more definitive intervention can be performed.

Some babies with TGA also have a ventricular septal defect. If this is present, enough mixing of blood may occur that the baby may not appear cyanotic at all and may actually become ill with symptoms of heart failure because of the extra blood flow to the lungs. Then the baby will have symptoms of poor feeding, poor weight gain, sweating, and fast or labored breathing.

Finally, there may be narrowing of the area leading out the left ventricle to the pulmonary artery called left ventricle outflow tract obstruction. In this situation even though there is the hole for the blood to mix, the total amount of blood flow going into the lungs is reduced. The degree of narrowing varies and can be mild at first but can get worse with time. As the narrowing increases the

baby's coloring will become more cyanotic (blue).

The severity of symptoms is dependent on how much red and blue blood mix together and the presence or absence of obstruction to blood flow out the left ventricle. The type and timing of operation depend on the combination of defects that accompany the primary problem of TGA.

Babies with TGA may develop early pulmonary vascular disease. This is an increase in the pressure in the lung blood vessels that cause changes that make it hard for them to accept low-pressure blood flow. These changes can occur as early as a few weeks of life and tend to occur more frequently in babies who have ventricular septal defects in addition to TGA. Early corrective surgery minimizes the chances of development of elevated pulmonary vascular resistance in these babies.

Please complete the following table about patients with Transposition of the Great Arteries

Presentation	In adulthood this will depend on the childhood surgical procedure. The post-operative course will depend on this. Reduced exercise capacity, poor RV function is the biggest problem. TR, tachyarrhythmia's, AF, VT, VF, sudden cardiac death Bradycardia due to sinus node dysfunction, pacemaker may be required Baffle leak Baffle may cause obstruction, may lead to Pulmonary hypertension. SVC obstruction may lead to venous congestion in the upper body half or lower body half. Residual VSD Dyspnoea Syncope

Specific Investigations to confirm diagnosis	**Atrial switch** ECG Echo or TOE if baffle leaks are suspected CMR CPET Holter monitoring Cardiac catheterization **Arterial switch** As above, include CT angio to check for narrowing of the coronary arteries See ESC guidelines (2010)
Management (include medical and surgical)	Most adults will have had surgical procedures, Mustards procedure or Sennings procedure. Young adults will have had the arterial switch operation.
Long term complications	**In patients with atrial switch** Dysfunction of RV. Tricuspid regurgitation. Heart failure. Tachyarrhythmias, atrial flutter, atrial fibrillation, SVT. VT, VF, sudden cardiac death Bradycardia due to sinus node dysfunction, pacemaker may be required Baffle leak Baffle may cause obstruction, may lead to Pulmonary hypertension. SVC obstruction may lead to venous congestion in the upper body half or lower body half. Residual VSD Dyspnoea Syncope

	In patients with arterial switch -PA stenosis, common due to stretching of PAs to reach the neo –pulmonary trunk at the time of surgery. May require balloon, dilatation, stenting or re-operation -Coronary artery abnormalities due to re-implanting of coronary arteries. ? poor myocardial perfusion, angina or sudden cardiac death. Sudden Cardiac death LV dysfunction, arrhythmias,(related to re-implanting the coronary arteries during arterial switch), dilation of proximal ascending aorta -Neo-aortic regurgitation-associated with aortic root dilatation. **In patients post Rastelli procedure** Problems with valved conduit between RV and PA and residual VSDs. Arrhythmias
Risk of endocarditis – *mild, moderate or severe?*	Atrial switch, moderate risk. Arterial switch high.
Risk in pregnancy – Low, medium or high.	Moderate to high. Risk of arrhythmias and RV dysfunction.
Suitable contraception	Risk of arrhythmias, progesterone only.

Useful Websites

www.yorksandhumberhearts.nhs.uk/default.aspx?id=271

www.bhf.org.uk

www.americanheartassociation.org

www.achd-library.com/heart_pictures/diagram3.htm

www.guch.org.uk

Further reading:

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ESC Pocket Guidelines (2010) Guidelines in Management of Grown-up congenital heart disease

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