

SURGICAL TREATMENT AND OUTCOME OF CONGENITAL HEART DISEASE

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Aims of surgery

The aim of surgery in congenital heart disease is to correct or palliate the heart defect to allow the child to lead as normal a life as possible. Some conditions such as atrial septal defect and ventricular septal defect can be repaired with a very low operative risk and excellent long term results. However, some more complex problems, for example, atrioventricular septal defect and single ventricles carry an appreciable operative risk and the survivors may not lead normal lives. In addition further operations may be needed after the initial repair to correct residual defects or change small or worn out artificial valves or conduits.

Indications

In general the indications for surgical intervention at any age are congestive cardiac failure, failure to thrive and cyanosis, when not ameliorated by medical therapy. However, surgery must not be delayed if there is a risk of pulmonary vascular disease developing even if the child is asymptomatic.

Some defects of moderate severity will be asymptomatic early in life, but will require elective surgery to prevent later problems developing for example arrhythmias due to atrial septal defect or hypertension due to coarctation of the aorta. Also all patients with congenital cardiac lesions are at risk of endocarditis. Whenever possible all surgery is completed in the first few years of life, ideally before schooling is started.

Surgical Method

Open heart surgery utilises the heart lung machine to maintain the circulation while the heart is repaired. The heart is protected during the operation by an infusion of cold electrolyte solution or blood containing Potassium Chloride into the coronary arteries (cardioplegia). Different centres may use different cardioplegia solutions, such as cold or warm blood plegia etc. The cardioplegia cools the heart and arrests it in diastole providing an excellent operative field for surgery inside the chambers of the heart. Incorporated in the cardiopulmonary bypass circuit is a heart exchanger which allows the patient to be cooled and rewarmed during the operation providing added protection for the brain. In infants of 5 kg or less the whole body cooling to 18°C can be used to protect the brain and this allows the circulation to be stopped for about 45 minutes providing a clearer operative field for complex intra

cardiac repairs on these very small infants. However more recently cardiopulmonary bypass without circulatory arrest is now routinely utilised down to 3 kg or less body weight to avoid the need for arrest.

New trends

There has been a continuing move from palliative procedures, for example, systemic to pulmonary artery shunt and pulmonary artery banding towards one stage corrective procedures, and these corrective procedures are being performed in children at a younger and younger age. Some 40-50% of open heart procedures in many units are now performed in children in the first year and even the first few months of life. Thus patients with symptomatic Fallot's Tetralogy can be repaired even in the first few months of life, rather than palliated by systemic to pulmonary artery shunts.

Likewise patients with transposition of the great arteries are routinely corrected by the arterial switch procedure in the first few weeks of life. Palliative procedures are not very successful in complete atrioventricular septal defect and truncus arteriosus and for corrective procedures to be successful they need to be performed in the first few months of life before the development of rapidly progressive pulmonary vascular disease.

Management of the sick neonate

In the neonatal period the closure of the ductus arteriosus may markedly reduce systemic blood flow in such conditions as coarctation of the aorta, interruption of the aortic arch, severe aortic valve stenosis and hypoplastic left heart syndrome, or it may reduce pulmonary flow, for example, severe Fallot's Tetralogy of pulmonary atresia. These conditions together with obstructive total anomalous pulmonary venous drainage may cause acute collapse with acidosis in the neonatal period requiring urgent resuscitation and intensive care unit management. Resuscitation includes ventilation, correction of acid base balance, support of the child with inotropes and the institution of prostaglandin infusion when duct closure has precipitated the crisis. Having stabilised the child, non-invasive diagnosis by echo Doppler evaluation has removed the need for the more traumatic investigation by cardiac catheter. A very sick neonate can now be resuscitated in the intensive care unit and the diagnosis made without the need for an invasive investigation. The child can be transferred to the operating room in a stable condition increasing the chance of a good operative outcome.

The adolescent and young adult

Young people with unresolved cardiac defects can present major problems in psychological and surgical management. They are anxious, worried about their heart condition, concerned about their future and need very careful counselling and psychological support. One of the difficulties is to provide continuity of care from infancy to adult life as transfer from a paediatric to an adult institution is usually required when they reach 16 years of age.

In the adolescent and young adult the cardiorespiratory problems may be very difficult to deal with. Children who underwent repair of transposition by atrial repair in infancy may present with right heart failure, tricuspid regurgitation and arrhythmias. Patients who underwent repair of Fallot's Tetralogy may present with a residual right ventricular outflow tract obstruction or aneurysm. The valve conduit which was of adequate size in the child may be too small for the adult and require changing. Sadly some complex problems may be beyond a surgical correction and other patients may have developed pulmonary vascular disease which prohibits surgery to correct the defect. In some situations consideration of a heart or heart lung transplant may provide the only hope of survival.

The spectrum of congenital heart disease

Not only are there many forms of congenital heart disease, but within each anatomical type there is a great variation in the severity and the mode of clinical presentation of the individual lesion. Isolated coarctation of the aorta is classically described as presenting with absent femoral pulses and hypertension in the upper limbs in an asymptomatic young child or adolescent, but at extremes of life this lesion can cause profound cardiac failure and death. A neonate might present with collapse and acidosis due to complete obstruction of the aorta at a severe coarctation and an adult with undiagnosed coarctation of the aorta may present in his 20s or 30s with cardiac failure due to a chronically obstructed aortic and secondary hypertensive cardiomyopathy.

The aortic valve homograft in congenital heart surgery

Valve replacement in congenital heart disease is fraught with 3 main problems:

- The size of the patient and the need to change with valve as the need grows,
- The need for anticoagulation to prevent thrombotic problems,
- The recognition that tissue valve conduits other than the aortic homograft are prone to unpredictable calcific deterioration.

Therefore valve replacement is usually deferred as long as possible to reduce the need for re-operations. When valve conduits are needed, for instance, reconstruction of the right ventricular outflow tract in pulmonary atresia, the human cadaveric homograft is the valve of choice. Fortunately it has been possible to store this valve in liquid nitrogen so that there is good supply. This valve also has the major advantage that no anticoagulation is necessary.

Individual congenital heart defects: management and outcome

Persistent ductus arteriosus

A ductus arteriosus should be closed, even if it is asymptomatic, because of the risk of endocarditis. The operation is performed through a left thoracotomy and the duct is ligated or divided. The duct can be safely occluded by the insertion of an umbrella which is plugged into the duct at cardiac catheter, and this may in the next few years, supersede direct surgical closure. In any event the operative risk is very low and providing there is no pulmonary vascular disease the outcome is excellent.

Atrial septal defect

A communication between the left and right atrium may be in the middle atrial septum, a secundum type, or high in the atrial septum adjacent to the entrance of the superior vena cava and associated with partial anomalous pulmonary venous drainage, sinus venosus type of defect. This generally does not cause problems early in life, but later may cause cardiomegaly, arrhythmias and onset of early cardiac failure. These defects are closed utilising cardiopulmonary bypass either by direct suture or by patching with native pericardium. The operative risk is very low and the outcome is excellent.

In the 1990's closure of atrial septal defects was successfully accomplished at cardiac catheter by various devices. This is an accepted method of closure and may be suitable for 40-50% of atrial septal defects. All patients with atrial septal defects should be considered and evaluated for closure by interventional cardiac catheter.

Total anomalous pulmonary venous drainage

Total anomalous pulmonary venous drainage (Fig. 1) generally presents in infancy and often in the first week of life, particularly when venous drainage is obstructed, for example, in the infra diaphragmatic type. This condition may still present as a true emergency since the child may continue to deteriorate despite resuscitative measures.

At surgery, usually utilising hypothermic circulatory arrest, the sac is anastomosed to the left atrium, the atrial communication is closed and the connecting vein is either divided or ligated. The

operative mortality is now of the order of 5% and providing there is no post-operative anastomotic stenosis the outlook is excellent.

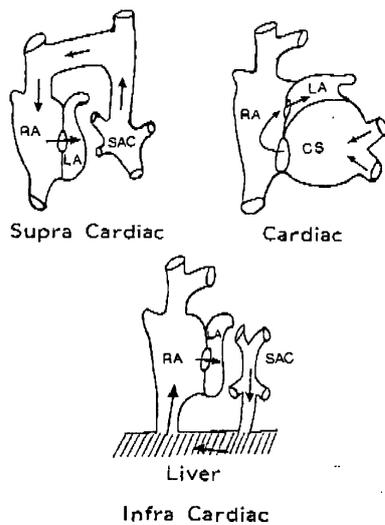


Fig 1 – Total anomalous pulmonary venous drainage.

Ventricular septal defect

A communication between the ventricles may be single or multiple. In the majority of instances (over 80%) the ventricular septal defect is single and in the perimembranous region or the septum of the heart adjacent to the tricuspid valve. Less commonly it may be in the muscular portion of the ventricular septum or in the outlet septum of the right ventricle, adjacent to the pulmonary valve. Ventricular septal defects can be closed utilising cardiopulmonary bypass at any age with a very low, less than 3% operative risk, and a very good long term outlook. There is no place now for palliative pulmonary artery banding except in the very rare complex multiple muscular ventricular septal defects and children with corrected transposition and ventricular septal defect where there is an increased risk of surgery producing heart block when the ventricular septal defect is closed.

Atrioventricular septal defect

Complete atrioventricular septal defects usually occurs in association with Down's syndrome. It is a complex abnormality where an atrial septal defect and ventricular septal defect are coalesce to form a central communication in the heart, the mitral and tricuspid valves are not individually formed and there is one large atrioventricular valve, (Fig. 2). In a partial atrioventricular septal defect there is only an atrial communication, the tricuspid and mitral valves are well formed, but there is a cleft in the mitral valve; it is not usually associated with Down's syndrome.

Partial atrioventricular septal defect can usually be repaired electively before the child goes to school. The atrial septal defect is patched with pericardium and the cleft in the mitral valve is closed. The operative risk is low and the long term outlook is excellent. However, children with complete atrioventricular septal defect usually prevent early in infancy with heart failure and they are at increased risk of rapidly progressive pulmonary vascular disease even in the first few months of life. Whilst palliative pulmonary artery banding has been tried in the past, this is a group of patients where early surgical correction in the first few months of life has been successfully introduced. Surgery involves closure of the ventricular septal defect with a dacron patch, closure of the atrial septal defect with pericardial patch and reconstruction of the mitral and tricuspid valves from the one atrioventricular valve. In the majority of instances the mitral valve can be made almost completely competent and the operative risk can be below 10%. Initially these children seem to do well in the follow up period, but long term reports of the function of this repaired mitral valve are awaited although initial results are encouraging.

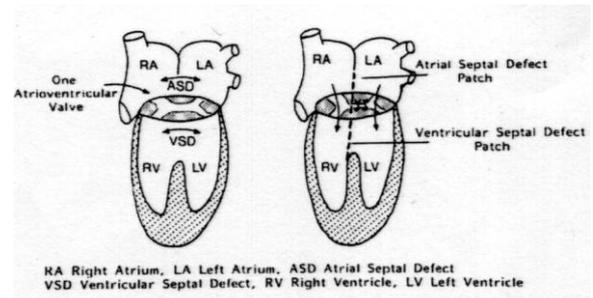


Fig 2 – Repair of complete atrioventricular septal defect.

Fallot's Tetralogy and pulmonary atresia with ventricular septal defect

In repair of Fallot's Tetralogy (Fig. 3) the ventricular septal defect is patched with dacron and the muscular infundibulum is resected and the right ventricular outflow tract is enlarged if necessary with a patch usually of pericardium. Many centres will now correct Fallot's Tetralogy when indicated at any age, whilst others would prefer to create a systemic to pulmonary artery shunt (such as the modified Blalock-Taussig shunt), (Fig.4) in the first year of life preferring to defer corrective surgery to a later age. With either approach overall operative mortalities of less than 5% are obtained and the long term outlook is excellent.

In pulmonary atresia with ventricular septal defect the right ventricular to pulmonary artery continuity is restored by insertion of a valved conduit, usually a homograft aortic valve. In this condition most centres would create a systemic to pulmonary artery shunt to tide the child over the first few years of life before correcting the lesion ideally before the child reaches school age.

In the long term, the main problems are ventricular arrhythmias, right ventricular volume loading due to pulmonary incompetence and the need to change the valve conduit as the child grows.

Truncus arteriosus

Like atrioventricular septal defects, truncus arteriosus usually presents in infancy with severe congestive cardiac failure. These infants are very susceptible to rapidly progressive pulmonary vascular disease because the pulmonary circulation is exposed to systemic pressures as well as large left to right shunt.

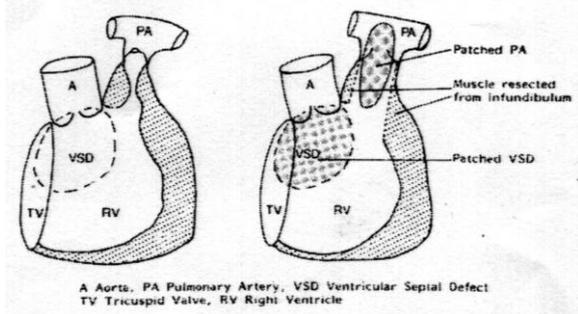


Fig 3 – Repair of Fallot's tetralogy.

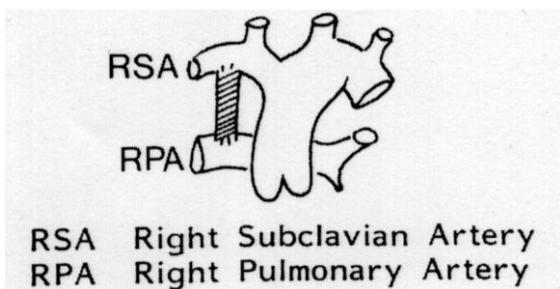


Fig 4 – Modified Blalock Taussig shunt.

Palliative pulmonary artery banding has a very low success rate in this condition and to avoid complications associated with pulmonary vascular disease surgery is advocated in the first few months of life. This involves dacron patch closure of the ventricular septal defect directing the left ventricle to the aorta and insertion of a small valve conduit, usually an aortic homograft valve (Fig. 5). The operative are still considerable in this condition, but if the child survives the correction the outlook, even with the need to change the conduit later in life is good.

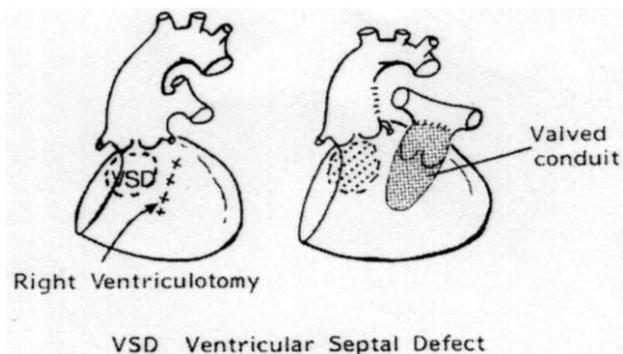


Fig 5 – Repair of Truncus Arteriosus.

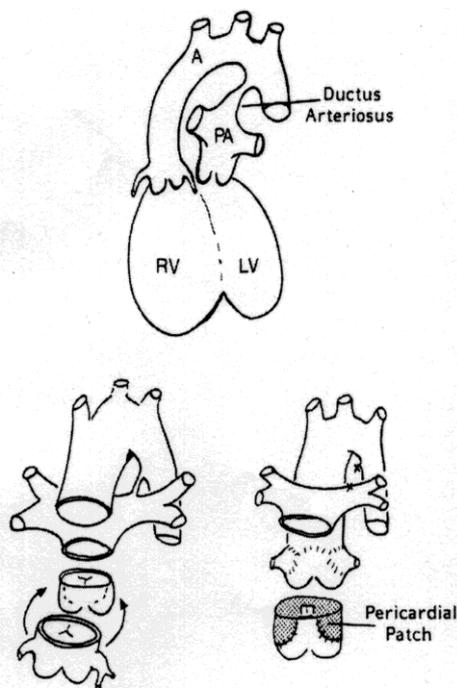
Transposition of the great arteries

Transposition of the great arteries can be conveniently divided into 3 main problems:

- simple transposition without ventricular septal defect,
- transposition with ventricular septal defect,
- more complex transpositions, for example, those with double outlet right ventricle and associated coarctation

There have been major changes in the surgical management of transposition over the last 10 years, namely the almost complete change from atrial repairs such as Mustard and Senning procedures to the arterial switch procedure which involves the relocation of the aorta to the left ventricle and the pulmonary artery to the right ventricle (Fig. 6). The surgical management of transposition is evolving in many centres but our practice is to correct transposition with or without ventricular septal defect in the first few weeks of life and more complex forms of transposition are usually corrected at this time as well. When transposition and ventricular septal defect is associated with pulmonary stenosis or atresia a systemic to pulmonary artery shunt is performed in the first instance and corrective surgery involving placement of a valve conduit from right ventricle to pulmonary artery and closure of the ventricular septal defect is deferred until the child is older.

The operative risk for the arterial switch procedure in infants with an intact ventricular septum or ventricular septal defect is currently of the order of 5% and approaches this figure even in more complex forms of transposition. The early results are excellent, however the long term follow up is not available. The main early complication is stenosis of the reconstructed pulmonary artery. Encouraging the aortic and coronary anastomoses seem to grow with the child.



A Aorta
PA Pulmonary Artery
RV Right Ventricle
LV Left Ventricle

Fig 6 – Anatomic correction of transposition of the great arteries.
Coarctation of the aorta with or without associated cardiac lesions and the interruption of the aortic arch

Coarctation of the aorta may exist as an isolated condition, be associated with a ventricular septal defect or with more complex problems. Generally when coarctation is associated with other lesions it presents in infancy with heart failure. Severe isolated coarctation may present with heart failure in infancy, but more classically presents later in life with absent femoral pulses and hypertension. When surgery intervention is necessary in infancy the coarctation may be resected followed by an end to end anastomosis, or the left subclavian artery may be divided and turned down as a flap through the coarctation to widen the narrowed area of the aorta. When there is an associated large ventricular septal defect, it may be closed at the same operation. More complex associated lesions, such as atrioventricular septal defect are treated on their merits. Surgery for isolated coarctation at any age carries a very low operative risk of less than 1%. When associated with a ventricular septal defect in an infant the risk is a little higher but when associated with more complex intracardiac morphology the risk is usually that of the underlying complex lesion.

Recurrence of coarctation of the aorta in the older patient is unusual but, in the infant it is still probably still of the order of 10-20%. Fortunately balloon dilatation of the recoarctation by cardiac catheter is very successful.

Interventional cardiac catheterisation with balloon dilatation with or without stenting the aorta is now an accepted method of treatment and should be considered in all older patients with coarctation.

Interruption of the aortic arch is a rare condition presenting in the neonate with severe congestive cardiac failure. There is a complete separation of the ascending from the descending aorta, the descending aorta being supplied by the ductus arteriosus (Fig. 7). The commonest form of interruption occurs between the left carotid artery and the left subclavian artery (Type B), but can occur beyond the origin of the left subclavian artery (Type A) or very rarely between the innominate artery and the left carotid artery (Type C). Usually the aortic interruption is associated with a ventricular septal defect and not unusually is associated with various degrees of aortic stenosis. Some 10 years ago the mortality for surgical management of this condition was very high, but now with total correction, usually in the neonatal period, operative mortalities of below 20% can be achieved.

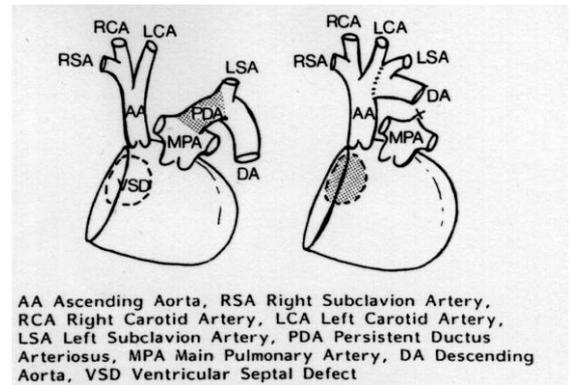


Fig 7 – Interruption of the aorta with ventricular septal defect. Repair by closure of VSD and resection of Ductus Arteriosus with direct anastomosis of descending to ascending aorta.

Fontan procedure

A sizeable group of children have complex congenital heart defects where there is essentially only 1 effective ventricle. Examples are tricuspid atresia, double inlet left ventricle and various forms of heterotaxy syndrome. The fontan type repair whereby the systemic venous return is connected directly to the pulmonary arteries has made a major impact on the management of this group of patients (Fig. 8). The requirements for a successful fontan procedure are normal ventricular function, low or normal pulmonary vascular resistance and an unobstructed pulmonary arterial supply to the lungs. Then the systemic venous pressure required to push the blood through the lungs to the ventricle need only be of the order of 10-14 mmHg and this is well tolerated.

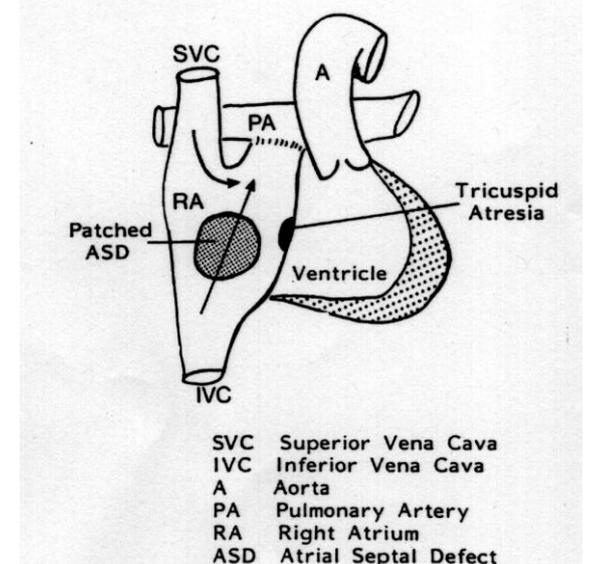


Fig 8 – The Fontan procedure

For example the tricuspid atresia palliation either by pulmonary artery banding or by systemic to

pulmonary artery shunting may be necessary early in life and thereafter an RA to pulmonary artery

connection with closure of the atrial septal defect and removal of the shunt or band can be performed before the child goes to school with the mortality of less than 10%. However, in more complex forms of single ventricle the operative mortality is higher. The fontan procedure is really an extensive form of palliation and at this time the actuarial survival curve for these patients does not approach normal.

Congenital aortic valve stenosis and hypoplasia of the left heart

Isolated congenital heart valve stenosis without associated hypoplasia of the left heart treated by open aortic valvotomy carries a good operative risk. When there is associated hypoplasia of the left heart mortality is high. All families who have a child with hypoplastic left heart syndrome diagnosed antenatally or postnatally are now given the opportunity of surgical management by variations of the Norwood procedure. Heart transplants because of lack of donors is not a viable option. Norwood procedure which involves utilising the right ventricle as a systemic ventricle and proceeding to a fontan procedure when the child is older. This approach has gained increasing acceptance in the UK and Europe over the last few years. Actuarial survival at 2 years of age can approach 60%.

In conclusion most children with congenital heart disease can be offered a primary corrective procedure when it is medically indicated with low operative risk. Because many operative procedures have only been recently introduced the long term outlook for these patients is unknown. However, the early indications are that in the majority of instance the outlook will be good. Patients who do not have 2 good ventricles and in particular those with hypoplasia of the left heart structures still present a major problem in management.

References

1. Cardiac Surgery – John W Kirklin MD, Brian G Barrett-Boyes KBE, MB, ChM. John Wiley and Sons.
2. A Colour Atlas of Cardiac Surgery, Congenital Heart Disease. James L Munro and Gerold Shore. Wolfe Medical Publications Ltd.
3. Surgery for congenital heart defects. J Stark and M de Leval Grune and Strattan.