

CONGENITAL HEART SURGERY: WHAT WE DO TO OUR PATIENTS

Postoperative management of children who have undergone congenital heart surgery is critically dependent on the understanding of the surgery by the referring paediatrician.

John Hewitson, FCS (SA)(Cardthor)

Cardiothoracic surgeon, Red Cross War Memorial Children's Hospital, Cape Town

John Hewitson has been in charge of the Red Cross Cardiothoracic Unit since 1994, and has a special interest in developing paediatric cardiac services in Africa. He believes time is an illusion.

Rik De Decker, MSc, MB ChB, DCH, FCPaed (SA), Cert Med Genetics (Paeds)

Senior specialist paediatric cardiologist, Division of Critical Care and Children's Heart Diseases, Red Cross War Memorial Children's Hospital, Cape Town

Rik is a paediatric cardiologist at Red Cross Children's hospital with special interests in the genetic control of heart development and interventional cardiac catheterisation. He panics when in big flat spaces.

Correspondence to: John Hewitson (john.hewitson@uct.ac.za)

You are about to leave your practice after a hectic Friday, and are looking forward to a quiet weekend off. Just as you reach the door, the phone rings, and your first instinct is to ignore it. But you answer it anyway. It is a Mrs Harmse, in a panic. Then you recall: you had sent her 8-month-old son, Karl, who has tricuspid atresia, to Red Cross Hospital 6 weeks ago. He had become very blue when his Blalock-Taussig shunt, inserted 7 months ago, blocked. Now, she says, he has had diarrhoea for 3 days, he has become very blue again, and his face is horribly swollen. His discharge letter from the hospital says something about a heart operation 4 weeks ago with a Glenn shunt... Who is this Glenn bloke, you wonder, and why did he make Karl's head swell up?

Congenital heart defects are responsible for more deaths in the first year of life than any other birth defects. However, for those with access to good paediatric cardiac surgical services, most of these lives can be saved through timeous surgery. Even then, not all defects can be fully corrected; about 20% of patients either require further staged surgery or have a permanent palliative solution.

Congenital heart defects are responsible for more deaths in the first year of life than any other birth defects.

The postoperative management of children who have undergone congenital heart surgery does not end at discharge at the door of the referral centre, but is critically dependent on the broader understanding of the surgery by the referring paediatrician, as well as a working knowledge of potential late complications. This article addresses these issues without dwelling on the technicalities of the surgery *per se*.

The common operations we do can be grouped into 5 categories by their longer-term outlook:

- temporary palliation for defects that cannot initially be repaired
- operations for heart defects that are fully corrected at the first procedure
- heart defects that *might* require further surgery or intervention after repair
- heart defects that *will* require further surgery after the initial procedure
- long-term palliation of uncorrectable lesions: functionally univentricular hearts.

Temporary palliation for defects that cannot initially be repaired

Palliative operations provide symptomatic relief but leave the basic pathophysiology uncorrected. The two main palliative approaches are

directed at either increasing inadequate flow to the lungs, or restricting excessive flow to the lungs.

Systemic to pulmonary shunts

In cyanosed children with insufficient pulmonary blood flow, a shunt is formed from the systemic to the pulmonary circulation, to provide more pulmonary blood flow. This is usually in situations of obstructed right ventricular outflow, for example in tetralogy of Fallot, or pulmonary atresia.

The commonest form of shunt used is a modified Blalock-Taussig shunt (Fig. 1), usually done through a thoracotomy, whereby a synthetic (Goretex®) conduit is used to create an aorto-pulmonary connection to supply sufficient blood to the lungs. These conduits connect the subclavian artery and the main branch pulmonary artery (on either side of the chest). Alternatively, should the branch pulmonary arteries be too small, through a sternotomy, the aorta or brachiocephalic artery may be connected to the pulmonary artery via a central shunt. These children are kept on 5 mg/kg aspirin daily to maintain patency of the conduit until corrective surgery can be done.

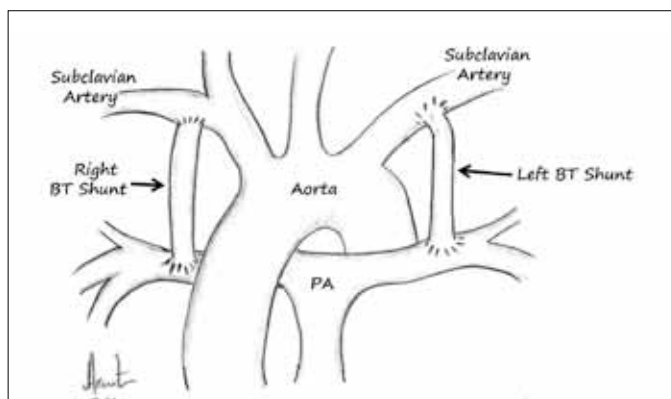


Fig. 1. Blalock-Taussig shunt.

Pulmonary artery band

When there is a large left-to-right shunt, consequent flooding of the lungs predisposes to repeated infections as well as cardiac failure and failure to thrive. A constricting band is placed around the main pulmonary artery to restrict this excessive pulmonary flow. The aim is to make the cardiac defect more tolerable and allow for growth until a complete correction can be done. Obviously correction of the defect is preferable, so banding is only done when surgical correction is not possible for one of a variety of reasons. As advancements have been made, more and more lesions are primarily corrected at a younger age, and PA banding is consequently done less often. Banding is done through either a left thoracotomy or a sternotomy.

Operations for heart defects that are fully corrected at the first procedure

The most common corrective operations can be considered definitive once-off repairs, with the necessity for further surgery unlikely. Examples are described below.

Patent ductus arteriosus (PDA)

PDAs are commonly closed by transvascular catheter approaches nowadays. This is not always possible, especially in neonates, when closure is done through a left thoracotomy.

Coarctation of the aorta

Surgical correction of isolated coarctation is done through a left thoracotomy by either resection and direct end-to-end anastomosis, or a subclavian flap aortoplasty. The latter procedure involves using the proximal left subclavian artery as a turned-down patch, and the left arm is then supplied via collaterals (usually resulting in a diminished left radial pulse). In an older patient primary transcatheter stenting may be considered.

About 10% of coarctations will recur due to scarring at the repair site. Most of these can be successfully dilated with a balloon catheter, and even stented if necessary.

Atrial septal defects (ASDs)

Isolated ASDs are often closed with a device implanted by a transvascular catheter approach. If this is not possible, usually due to the size or position of the defect, surgical closure is typically done through a midline sternotomy.

Ventricular septal defects (VSDs)

Isolated VSDs are the most common recognised congenital heart lesion (2 per 1 000 live births), and about 30% of these patients will require surgery. The remaining defects will close spontaneously with growth, or are haemodynamically insignificant. There is a possibility that these may also be addressed by a catheter approach in the future, but this technology is under development. Surgical closure is done through a midline sternotomy with minimal morbidity. VSD repairs often form part of the correction or palliation of more complex lesions.

Atrioventricular canal defects (AVSD)

This lesion is not merely the co-occurrence of an ASD and VSD in one patient, but a complex defect at the junction of the atrial and ventricular septae. Instead of two atrioventricular valves, a single AV valve with five or six leaflets forms, which may or may not be competent. The most important associated anomaly is Down syndrome; approximately 50% of children with Down syndrome have a canal defect, while 75% of children with a canal defect have Down syndrome. These children should have reparative surgery before 6 months of age to avoid the development of irreversible

pulmonary hypertension (through excessive pulmonary flow). In some cases the reconstructed AV valves will be incompetent to a degree, and a few of these children need valve repair surgery at a later stage.

Total anomalous pulmonary venous drainage (TAPVD)

In TAPVD all pulmonary venous blood returns to the right atrium, either directly or via the SVC or IVC, creating a large left-to-right shunt. These children typically require surgery very early in life, done through a midline sternotomy, with an associated mortality of about 5%. Those that are successfully repaired may have persistent pulmonary hypertension, though typically do well.

Palliative operations provide symptomatic relief but leave the basic pathophysiology uncorrected.

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA)

The left coronary artery arises directly from the main pulmonary artery, rather than from the aorta. Collaterals from the right coronary bed support the left coronary bed, but with low post-neonatal pulmonary artery pressures, a 'steal' occurs with retrograde bloodflow into the PA away from the left coronary bed. Cardiac function deteriorates rapidly after birth, with myocardial ischaemia and myocardial infarcts. Repair is an emergency, and consists of reimplanting the left coronary into the aorta. After the repair, myocardial function gradually improves (often to normal) over months and even years as the child grows.

Heart defects that *might* require further surgery or intervention after repair

Some children, having had corrective operations, might need repeat surgery in the future because the repair does not last through the growing years, or because further problems arise. Examples are discussed below.

Tetralogy of Fallot (TOF)

TOF represents 5% of all congenital heart defects. The tetralogy (a VSD, RV outflow obstruction, aorta positioned towards the right and overriding the septum, and RV hypertrophy) is in fact the result of a single embryological defect, the displacement of the anterior part of the infundibular septum.

Surgical repair – which is ideally completed before 12 months of age – usually comprises patch closure of the VSD and widening of the obstructed RV outflow. If this widening includes the pulmonary valve anulus,

causing pulmonary regurgitation, RV volume overload will make pulmonary valve replacement necessary in the decades to come because of declining RV function.

In some patients the pulmonary arteries are hypoplastic and unable to carry the full cardiac output, and repair must then be delayed until they are large enough. In the meantime, symptomatic patients require a systemic to pulmonary shunt as palliation and to enhance pulmonary arterial growth.

Transposition of the great arteries (TGA)

The aorta arises from the RV and the PA arises from the LV. TGA is seen in 1 in 4 000 live births and is the commonest cyanotic lesion (3%). Without intervention, most patients will die within 3 months. Initial palliation is by transcatheter balloon septostomy to improve mixing between the two circulations. This is soon followed by an arterial switch repair to correct the defect, typically done within the first 3 or 4 weeks of life. Ten-year survival is about 80%, and most patients have normal exercise tolerance. About 10% require repeat surgery or catheter intervention for pulmonary stenosis due to scarring. Dilatation of the new aortic root can also occur with associated aortic regurgitation in a few patients.

TGA may also occur in association with more complex lesions, such as TGA with pulmonary stenosis and a VSD. These patients may undergo a Rastelli procedure, whereby the VSD is closed in such a way as to direct the LV outflow into the aorta; the pulmonary valve is oversown and an artificial valved conduit is used to construct a new RV outflow.

Heart defects that *will* require further surgery after the initial procedure

Some corrective operations are temporary solutions. This may be because materials used or areas of scarring will not grow, or because materials wear out.

Pulmonary atresia, truncus arteriosus, transposition of the great arteries with pulmonary stenosis

These are all examples of defects that require reconstruction of the RV outflow tract as part of the surgical correction. This is typically done using a conduit containing a tissue valve, which may be homograft (human donor) or xenograft (animal) valve which has been treated to destroy allergenicity, and as such it is non-viable tissue and will not grow. In addition, the tissue gradually scars and calcifies, and valve function is lost over 5 - 10 years, necessitating replacement of the valve and conduit. Tissue valves last much longer in adults, so that repeat operations will be less often necessary as the child grows up.

Congenital aortic stenosis and LV outflow tract obstructions

These are complex problems that commonly require repeat surgery. Obstruction may be at the subvalvar, valvar and supra-valvar levels. Obstruction above or below the valve is more likely to be permanently relieved by surgery, but can recur through scarring. Valvar narrowing, on the other hand, almost always eventually leads to valve replacement (Box 1 and Fig. 2).

Box 1

The South African-trained cardiac surgeon, Donald Ross, developed an ingenious procedure in the 1960s whereby a patient's defective aortic valve is replaced with his or her own pulmonary valve, the pulmonary valve then being replaced with a homograft tissue valve (Fig. 2).

The reasoning behind this is that replacement aortic valves do not last long. The patient's own pulmonary valve will grow and adapt to the severe stresses and wear in the aortic position, whilst the homograft replacement will not do too badly in the low pressure pulmonary position.

This is a complex surgical procedure, but is particularly suited to children with congenital aortic stenosis, because their pulmonary valve is typically normal, and when moved to the aortic position it will grow along with general somatic growth.

Permanent pacemaker

Placement of a permanent pacemaker may be required for patients with congenital heart block, or after acquired heart block, usually following congenital heart surgery. These patients will require occasional replacement of pacemaker leads or the pacemaker itself when the batteries run out. In children, the pacemaker leads are placed epicardially (not

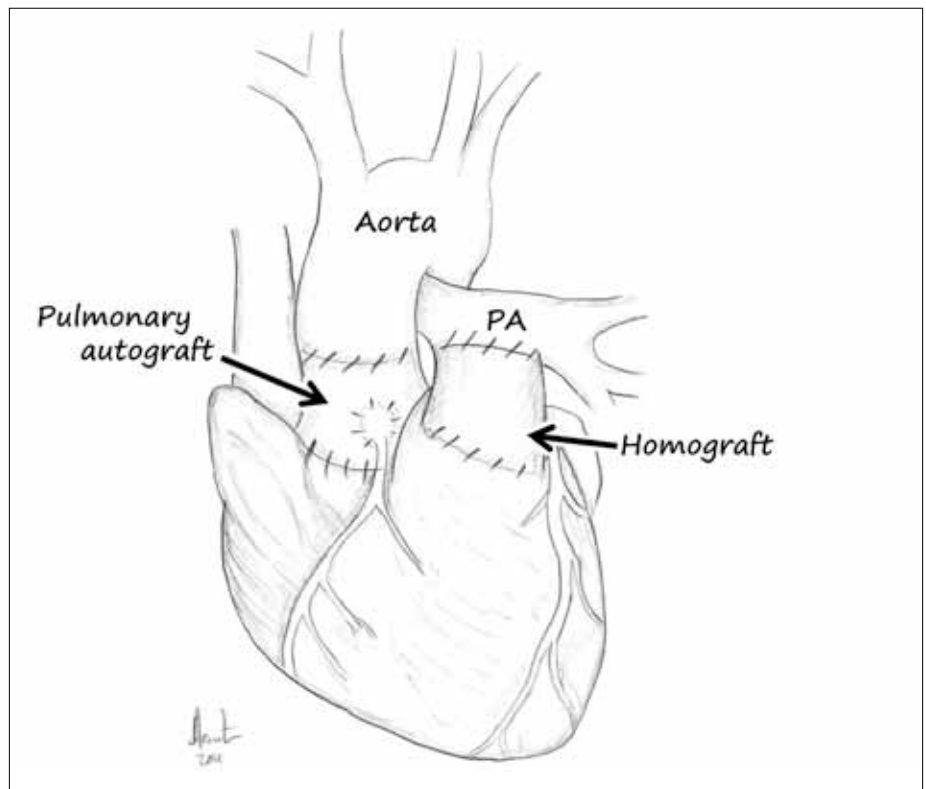


Fig. 2. Ross' procedure.

transvenously) and the pacemaker is inserted in a pouch fashioned below the diaphragm in the left upper quadrant of the abdomen.

Long-term palliation of uncorrectable lesions: functionally univentricular hearts

The commonest uncorrectable lesion is a heart which has only one functional ventricle, or which has two that cannot be separated to form separate systemic and pulmonary circulations. The commonest example of such a functional univentricular heart is tricuspid atresia (TA). TA is the congenital absence of any identifiable tricuspid valve tissue, thus the RA and RV have no direct connection.

Some corrective operations are temporary solutions. This may be because materials used or areas of scarring will not grow, or because materials wear out.

In order to minimise the intracardiac mixing of oxygenated and deoxygenated blood, the surgical solution is to redirect the systemic venous return directly to the pulmonary arteries before entering the heart. The heart then fills with and pumps only oxygenated blood. This palliative solution to the functionally univentricular heart was developed by Francois Fontan in the 1970s

and is known as the Fontan procedure. The systemic and pulmonary circulations are again in series, but the pulmonary pump is excluded. Surprisingly, this arrangement works quite well in carefully selected patients!

Typically the Fontan procedure is done in two stages: the SVC is disconnected from the heart and connected directly end-to-side to the right pulmonary artery at 6 months to 2 years of age (known as a bidirectional Glenn shunt). A few years later the IVC is connected via an extracardiac conduit, completing the right heart bypass (aka TCPC, or *total cavopulmonary connection*).

This is an imperfect palliation, and many complications may be seen over the ensuing decades (arrhythmias, emboli, stroke, polycythaemia, valvular insufficiency, ventricular dysfunction, protein-losing enteropathy, and end-stage heart failure), but nevertheless has a 90% 15-year survival rate with reasonable quality of life in most. Careful patient selection is the key to good outcomes; the main requirements for selection relate to good pulmonary circulation with no pulmonary hypertension, and good function of the dominant ventricle.

In conclusion

Congenital cardiac surgery is a technically complex field to understand, and the attending physician may be reluctant or struggle to make management decisions in light of the altered anatomy created by many operations. A broad understanding of these operations is essential for correct management and appropriate follow-up.

Table I. A glossary of some procedures in congenital heart disease surgery

Operation	What does it do?	Possible late complications
Arterial switch	Swapping the MPA and Aorta in a TGA	Pulmonary or aortic stenosis
Atrial switch (rare)	Redirection of right atrial blood to LV and left atrial blood to right ventricle in a TGA; the RV remains the systemic ventricle	Arrhythmias, baffle leak, RV failure; LV involution
Blalock-Taussig shunt	Connection of subclavian to pulmonary artery	Blockage with cyanosis
Brock procedure	Tetralogy of Fallot repair without complete closure of the VSD	Pulmonary regurgitation, residual pulmonary stenosis, excessive left to right shunt via VSD
Central shunt	Connection of innominate artery or aorta to MPA	Blockage (unusual), with cyanosis
Damus-Kaye-Stansel	Division of MPA from branch PAs with connection to aorta to bypass subaortic stenosis in univentricular heart	Outflow tract stenosis, pulmonary and/or aortic regurgitation
Fontan (TCPC)	Anastomosis of IVC to pulmonary artery	PLE, cyanosis, CCF, chylothorax, arrhythmias, stroke
Glenn shunt	Anastomosis of SVC to pulmonary artery (usually first stage of Fontan)	Thrombosis with SVC syndrome
Homograft insertion	Insertion of valved homograft in RV outflow tract position	Homograft obstruction, pulmonary regurgitation, RV failure
Kawashima procedure	Glenn shunt when absent IVC, where lower body drains via azygous to the SVC (equivalent to Fontan)	As for Fontan
Konno procedure	Enlargement of the LV outflow tract, often including aortic valve replacement or Ross procedure	LV outflow tract stenosis, aortic regurgitation, LV failure
Norwood procedure stage 1	Creation of a single outflow from the RV to the descending aorta in hypoplastic left heart or mitral atresia; a Blalock-Taussig shunt is also done	Multiple – high morbidity
Norwood procedure stages 2 and 3	Completion of Glenn (stage 2) and Fontan (stage 3) with removal of the BT shunt	As for Glenn and Fontan
PDA ligation	Closure of a PDA	Exposure of coarctation of aorta
Pulmonary artery band	Constriction of the MPA to reduce flow	Cyanosis (too tight) or CCF (too loose) Branch PA obstruction
Rastelli	Formation of a new RV outflow tract with a valved homograft, and closure of VSD	Homograft complications, branch PA stenosis
Ross procedure (see box)	Placement of patient's own pulmonary valve in aortic position; and placement of a valved homograft in RV outflow tract	Homograft complications, aortic regurgitation, LV outflow tract stenosis
Septostomy	Creation of a large hole in the intratrial septum to improve interatrial mixing	Minimal; stroke
Takeuchi procedure (rare now)	Repair of ALCAPA by creating a baffle inside the MPA to direct blood from the aorta to the anomalous LCA	Pulmonary stenosis, baffle leak, LCA stenosis, LV failure

CCF = congestive cardiac failure; IVC = inferior vena cava; LA = left atrium; LCA = left coronary artery; LV = left ventricle; MPA = main pulmonary artery; PA = pulmonary artery; PDA = patent ductus arteriosus; PLE = protein-losing enteropathy; RV = right ventricle; SVC = superior vena cava; TGA = transposition of the great arteries; VSD = ventricular septal defect.

IN A NUTSHELL

- Congenital heart disease is by far the commonest birth defect, occurring in 8/1 000 live births (i.e. just below 1%), and is therefore also the commonest cause for early death due to a birth defect.
- Affected children who have access to paediatric cardiac surgical services have a good chance of a satisfactory long-term outcome.
- Congenital heart surgery ranges in complexity from simple repairs achieving near-normal cardiac physiology to the creation of long-term palliative solutions for extremely complex cardiac lesions.
- In some instances, temporising palliative surgery assures the survival of the patient to an age when more definitive surgery becomes possible.
- It is impossible for the general paediatrician to keep abreast of developments in congenital heart surgery, so for perspective, this article offers a brief overview of the commoner procedures, their indications and possible longer-term complications.
- This perspective may assist with the urgent management of late postoperative complications.

Further reading available at www.cmej.org.za