

New ways of avoiding cardiac surgery in children

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Summary

The concept that many serious defects can be effectively treated on a day-case basis without the need for incisions has revolutionised our approach to many congenital heart defects. Interventional cardiology has undergone tremendous expansion and change in the last 20 years. Many new techniques and devices have been introduced. Balloon valvuloplasty has become the treatment of choice for pulmonary valve stenosis at all ages and is also the treatment of choice for aortic valve stenosis in neonates, children and young adults. Endovascular stent implantation has been shown to be more effective than surgery for post-operative pulmonary artery stenosis and coarctation in older patients.

Occlusion of intra-and extra-cardiac communications, such as atrial septal defects, ventricular septal defects and patent ductus arteriosus with self-expanding pre-shaped devices matched to the defect, has become routine. Electrical re-wiring of the heart, either with pacemakers or catheter ablation therapy, provides curative treatment for many arrhythmias.

Recent advances include the use of stents as a vehicle to implant pulmonary valves in failing surgical right ventricle to pulmonary artery conduits and the completion of Fontan circulations with covered stents to create intra-cardiac conduits. Hybrid procedures where the interventionalist places devices together with the surgeons through a sternotomy represent a new way of avoiding cardiopulmonary bypass of circulatory arrest during cardiac surgery.

Although interventional cardiac catheterisation has replaced surgery as the primary treatment for a number of congenital heart defects, it has also provided an additional and complementary treatment to surgery for many residual defects. Major 're-plumbing', such as the arterial switch operation for transposition of the great arteries, remains the province of the surgeon.

Introduction

The paediatric cardiac catheterisation laboratory has witnessed tremendous changes over the last 20 years such

that cardiac catheterisation in patients with congenital heart disease is now performed mainly for therapeutic rather than diagnostic reasons. The bulk of interventional cardiac catheterisation still consists of occlusion of abnormal communications and dilation of stenotic vessels or valves. Electrical therapy, either with pacemakers or catheter ablation, is another important aspect of interventional cardiology.

Basic techniques

Interventional procedures, whether it is a balloon dilation, stent implantation or device occlusion, follow a common and relatively simple methodology.

1. General anaesthesia or conscious sedation is required for most procedures on infants and children.
2. Most procedures start with percutaneous access of the femoral vessels and insertion of a valved sheath. Access can be difficult because of occluded veins or previous surgical procedures such as the extra-cardiac Fontan operation. Many of these difficulties can be surmounted using a transhepatic venous puncture or percutaneous transthoracic puncture. These newer techniques have proven in practice not to be as daunting as they might seem to be and can offer a life-saving intervention to patients with few other options.
3. Haemodynamic measurements and angiograms are taken if required to further delineate the anatomy or severity of the lesion.
4. A catheter is introduced and passed across the stenosis or abnormal communication.
5. Once the catheter has crossed the stenosis or defect, a long guidewire is passed out the end of the catheter into an appropriate location. This guidewire then provides a track over which the required therapeutic device can be passed.
6. Balloon dilation catheters can usually be threaded directly over the wire, while stents and occlusion devices must be protected or constrained within long plastic sheaths that have already been passed over the pre-

placed guidewire. Once the balloon or device has been inflated or deployed, it or its delivery system is removed.

7. The effect of the intervention is then assessed by haemodynamic, angiographic or echocardiographic measurements.
8. Finally, the sheaths are removed and pressure applied.

Dilations

Septostomy

Balloon atrial septostomy was introduced by Rashkind over 35 years ago¹ and remains in use in its original form to improve mixing in patients with transposition physiology and for patients who require venting of an atrium whose outflow is restricted. The procedure is usually performed at the bedside under echo control in the neonate with transposition of the great arteries.

A balloon septostomy catheter is passed via the umbilical vein, ductus venosus, inferior vena cava and right atrium and through the patent foramen ovale into the left atrium. The balloon is inflated in the left atrium and jerked back across the atrial septum into the right atrium. This manoeuvre tears the atrial septum to produce an atrial septal defect with improved mixing and arterial saturations. Atrial septostomy in older patients, whose atrial septum can be much tougher, can be accomplished by first cutting the atrial septum with a blade.²

Balloon valvuloplasty

Larger diameter balloon dilation catheters, along the line of those developed by Gruntzig for peripheral and coronary arteries,³ were made for the larger central vessels and cardiac valves in children. These large diameter balloon dilation catheters (valvuloplasty catheters) were initially used to treat pulmonary valve stenosis in older children⁴ and rapidly became the treatment of choice for pulmonary valve stenosis in all age groups.⁵ The aim of the procedure is to relieve the stenosis by tearing the valve, as the resultant pulmonary regurgitation is mild and well tolerated. A large valvuloplasty balloon is inflated across a stenotic pulmonary valve. A waist-like indentation in the balloon can usually be seen on fluoroscopy. Further inflation of the balloon abolishes the waist. Surgery is only employed for dysplastic valves in patients with Noonan syndrome who also have small valve rings and often require a patch to enlarge the annulus as well.

Valvuloplasty has been especially useful in neonates with critical pulmonary stenosis where surgery previously carried a high mortality.⁶ In neonates with the more extreme form of pulmonary atresia with intact ventricular septum, valvuloplasty can still be accomplished by first perforating the pulmonary valve with a hot wire.⁷ Balloon pulmonary valvuloplasty can also alleviate cyanotic spells in patients with the tetralogy of Fallot whose pulmonary arteries are not yet large enough to safely undergo primary repair.⁸

Aortic valve stenosis (which is non-calcific in infants and children) is usually treated by balloon dilation.⁹ A balloon diameter that is close to the diameter of the aortic valve annulus is chosen as overdilation (which is routinely employed in pulmonary stenosis) can result in significant aortic regurgitation and earlier valve replacement. The balloon is usually introduced in a retrograde fashion via the femoral artery and passed over a pre-placed guidewire across the aortic valve. Rapid right ventricular pacing to lower the cardiac output while the balloon is inflated may help to avoid the fluid-filled balloon being ejected by the powerful left ventricle before the valve has been split.

In neonates with critical aortic stenosis and poor left ventricular function, the balloon can be introduced in an antegrade fashion. The aortic valve is crossed from underneath by a guidewire from a transvenous catheter placed in the left ventricle via the patent foramen ovale. This technique reduces the risk of inadvertent puncture and perforation of the very soft neonatal aortic valve leaflet(s) by guidewires introduced in a retrograde fashion. It also avoids the risk of femoral artery thrombosis as the balloon is introduced from the vein. The long-term result of aortic valve dilation in neonates depends on both an effective balloon dilation of the valve and the degree of associated left heart hypoplasia.

Angioplasty

Balloon dilation of coarctation of the aorta is used for both native and post-surgical coarctation.¹⁰ In patients with re-coarctation, it is generally considered to be the treatment of choice. Its efficacy in native coarctation depends on the age of the patient and whether there is significant underdevelopment of other parts of the aortic arch. Neonates in whom the ductal tissue forms a sling around the arch have a good initial response to dilation but a high re-stenosis rate, probably because of later contraction of ductal tissue. Older patients have a good response to balloon dilation. However, overdilation may result in aneurysm formation.

Dilation of pulmonary artery stenoses, either native or acquired, is useful in a percentage of patients. Vessel recoil and the risk of overdilation of distal tapering pulmonary arteries limit the clinical usefulness of pulmonary artery angioplasty.

Stents

The problems of vessel recoil or overdistension and dissection have been addressed by the introduction of endovascular stents.¹¹ This has been a particularly important development for many post-operative and some pre-operative patients with pulmonary artery stenoses in whom the results of surgical re-operation can be disappointing.¹² The stents used in interventional cardiac catheterisation are usually much larger than those used in adult coronary

arteries as the vessels being treated are the larger central vessels that have to carry significant proportions of the entire cardiac output.

Most stents used in children are balloon expandable such that the diameter obtained relates to the diameter of the balloon used to deploy/expand them. These stents can therefore be further expanded with a larger diameter balloon at a later date to keep up with the growth of the child.¹³ It is therefore important to implant a stent that can be re-expanded at a later date to the expected normal size of the fully-grown vessel.

Implantation of stents can be technically challenging and should not be undertaken by the occasional operator. The results of stent implantation in pulmonary artery stenoses have been very good with sustained increases in vessel diameter, distal perfusion and gradient reduction. Complications can consist of misplacement and embolisation of the stent, in situ thrombosis and vessel rupture. Most of these complications can be avoided by appropriate patient and lesion selection and operator experience. The stents usually develop a neo-intima, which can be excessive, if the stent is not fully dilated throughout its length, and result in re-stenosis. Nevertheless, the increase in vessel diameter produced by stents is significantly greater than that usually produced by angioplasty.

Stents are increasingly being used to successfully treat native coarctation in older patients. The implanted stent must be capable of being dilated to a diameter of 20mm. Graded dilation of a severely stenotic segment on two separate occasions may be required to avoid overdistension and possible aneurysm formation. Covered stents are also being used in these patients to further reduce the risk of aneurysm formation, particularly in patients where there is an atretic segment that has been punctured to establish luminal continuity.

Covered stents have also been used to create an intra-cardiac-conduit between the inferior vena cava and pulmonary artery in order to complete the Fontan circulation¹⁴ in patients with single ventricles. Although this experimental approach may avoid a further surgical procedure, it requires surgical preparation at the time of the bi-directional Glenn shunt. Baffle stenoses in post-operative Mustard patients respond well to stent implantation.

The patent ductus arteriosus can also be stented as a form of palliation in patients with duct-dependent pulmonary or systemic circulations. Abandoning the duct (which one has diligently kept open with prostaglandins) and placing a surgical artificial duct (shunt) has always seemed highly illogical!^{15,16} Results with ductal stenting in the early 1990s were very poor¹⁷ because of technical problems and high re-stenosis rates and the technique fell out of favour. Recent results are much more hopeful with significantly less re-stenosis.

Stenotic collateral arteries in patients with pulmonary atresia without true central pulmonary arteries can be enlarged by stent implantation to produce a useful increase in the patient's

oxygen saturation. Recently, 'cutting' balloons have been used to pre-dilate (tear) these stenotic collaterals as they are often unresponsive to conventional high pressure angioplasty balloons.

A very exciting recent advance has been the development of percutaneous valve replacement.¹⁸ A bovine jugular vein valve is sutured to the inner aspect of a large stent and then progressively crimped onto a balloon delivery system. The stent valve is then ballooned into a previously surgically placed right ventricular valveless outflow conduit. A number of patients have been treated successfully with this system; however, clinical follow up is still very short. The technique may be limited by the nature of the previously implanted surgical conduits, which are often undersized and under significant extrinsic compressive forces.

Occlusions

Transcatheter occlusion of intra-cardiac and extra-cardiac communications has been revolutionised in the last six years by the development of the Amplatzer devices.¹⁹ These devices are made from a cylindrical nitinol wire mesh, which can be formed by heat treatment into a number of different shapes. The nitinol used in the device is of the super-elastic type that allows the device to rapidly regain its shape once it has been pushed out of the sheath. A sleeve with a female thread on the proximal end of the device allows a delivery cable with a male screw to be screwed on. The attached device can then be pulled and pushed into the loader and delivery sheath respectively. A family of devices of different shapes and diameters has been produced to occlude secundum atrial septal defects (ASD), patent foramen ovale (PFO), patent ductus arteriosus (PDA) and ventricular septal defects (VSD).

The Amplatzer ASD Occluder is a double saucer-shaped device with a central stent-like connecting cylinder. The waist in the device varies from 4mm to 40mm, allowing closure of both small and large ASDs. Very large secundum ASDs with deficient margins (other than at the aortic end of the ASD) may not be able to be closed by a device and require a surgically placed patch. The ASD is usually balloon sized and the diameter of the waist in the balloon is used to select the size of the device. The device is introduced via a long transvenous sheath in the left atrium. The left atrial disk is extruded and pulled against the defect and the long sheath is then pulled back to deploy the rest of the device (central waist and right atrial disk). The device is released after assessment of its placement with transoesophageal or intra-cardiac echocardiography. The defect is closed by the induction of thrombosis on three polyester patches sewn into the three sections of device. The device is covered over by neocardia within a couple of months. Worldwide, many thousands of patients have had their ASDs closed with the Amplatzer devices with very high occlusion rates. Complications are unusual and consist mainly of device

migration (<1%) or transient arrhythmias (1-2%). Complications such as thrombus formation with cerebral thromboembolism or aortic erosion with tamponade have also been described but fortunately are rare. Transcatheter ASD occlusion is now considered by most cardiologists to be the treatment of choice for patients with suitable defects. There are other double disk type devices available for ASD occlusion but none have the same applicability or ease of use.

Most patients with a PDA are treated by transcatheter occlusion. Premature babies or small infants with large PDAs are still treated surgically. Coil occlusion of the PDA was introduced at the beginning of the last decade and has become the most widely employed device for PDA closure.²⁰ This technique has been highly successful at closing small PDAs. However, when the minimum diameter is more than 3mm, multiple and larger diameter coils are required, which prolongs the procedure and increases the risk of encroachment on the left pulmonary artery. The Amplatzer PDA plug, which is a mushroomed-shaped nitinol frame stuffed with polyester, is used for patients with larger PDAs.²¹ The occlusion rates are close to 100% and higher than the published results of surgical ligation.

Devices have also been used for occlusion of VSDs. This has been especially useful for multiple muscular defects,²² which can be difficult for the surgeon to reach or prolong a complex cardiac reconstruction. VSD devices have also been used to occlude peri-membranous defects but devices in this location can interfere with aortic valve function. A peri-membranous VSD device with eccentric disks, which avoids interference with the adjacent aortic valve, is undergoing clinical evaluation.²³ The initial results in older infants and children appear very encouraging, with a higher rate of complete occlusion than surgery. The only discouraging complication has been a 2% risk of complete heart block requiring pacemaker insertion. This appears to be slightly higher than the 1% incidence with surgery. Hopefully, the incidence with device closure can be reduced further by softening the structure of the device which comes into contact with the adjacent conduction bundle. Small peri-membranous defects are very common and incur a 0.5% risk of infective endocarditis per year. It is still too early to know whether the risk to benefit ratio for intervention in these usually asymptomatic patients comes out in favour of intervention or not.

Coil occlusion of unwanted blood vessels such as aorto-pulmonary collateral arteries, coronary artery fistulae, arterio-venous malformations and venous collaterals is increasingly effective because of improvements in catheter and coil design allowing catheters to deliver coils to very distant parts in a controlled fashion.

Hybrid procedures

This is a new area of co-operation between interventional paediatric cardiologists and paediatric cardiac surgeons where

interventional devices are used in combination with surgical techniques. The most promising example is in the treatment of hypoplastic left heart syndrome. Neonates born with this syndrome depend on the ductus arteriosus to allow the right ventricle to provide systemic perfusion. This can be temporarily kept open with a prostaglandin E1 or E2 infusion. If the family opt for palliative surgery, then the pulmonary artery can be directly anastomosed to aorta in what is known as a Norwood operation. The now disconnected branch pulmonary arteries are then supplied with blood via a small Goretex tube from the innominate artery or right ventricle.

The main problem with this surgery is the high operative mortality (20-50%) and the significant cognitive defects in the survivors because of the need for circulatory arrest during surgery. The goals of the Norwood operation can also be achieved by a hybrid approach. Stenting open the ductus arteriosus is more logical than the surgical pulmonary artery to aorta anastomosis which requires circulatory arrest. The controlled pulmonary artery flow which is achieved surgically with a small Goretex interposition tube can be achieved by banding or placing flow restrictors at the origin of the branch pulmonary arteries. The pulmonary artery bands are placed via a median sternotomy at the same time as the ductus is stented through a small puncture in the right ventricle. This hybrid procedure can either be performed in the operating theatre with a mobile x-ray C-arm or in the cardiac catheterisation laboratory with the surgical team. Early results from this hybrid approach for hypoplastic left heart syndrome are promising.

Other hybrid approaches include peri-ventricular occlusion of VSDs in small babies with large muscular VSDs. Again, access is achieved surgically via a median sternotomy. The right ventricle is punctured and under echocardiographic control the VSD is crossed and a short sheath passed into the left ventricle. Through this, an occluder is positioned. If echocardiography demonstrates satisfactory positioning, the occluder is detached and the delivery system removed. The right ventricular puncture purse string is then tied and the sternum closed. This hybrid approach avoids both a preliminary pulmonary artery band operation and a subsequent pulmonary artery debanding and VSD closure operation.

Interventional electrophysiology

Pacemaker implantation is required for congenital and post-operative complete heart block and for sick sinus syndrome. It is now usually performed in the cardiac catheterisation laboratory using the transvenous route rather than surgical epicardial placement.²⁴ This is especially important for atrial pacing where epicardial leads generally perform poorly. Concern remains over long-term venous access if the pacing leads should have to be replaced.

The greatest advance in the last 10 years in the management of arrhythmias in infants and children has been

the introduction of radiofrequency catheter ablation.²⁵ Most arrhythmias can be cured with this technique. Most infant tachycardias are related to accessory pathways and resolve spontaneously in the first year of life. However, some tachycardias are incessant and can be very difficult to control with drugs and may induce a cardiomyopathy. These tachycardias are relatively easy to treat with radiofrequency. Large tipped ablation catheters are available for infants and are used to map the offending accessory pathway or ectopic focus and then 500kHz of radiowaves are applied to heat the subjacent tissue to around 55°C. The scars created are discrete but in experimental studies may grow with the heart, but clearly if the tachycardia was life-threatening the created scars are probably of little clinical concern.²⁶ The technique is more often applied to older children and adolescents with forms of Wolff-Parkinson-White syndrome who do not wish to have a lifetime of drug therapy.

Another important development is the implantable cardioverter defibrillator, which has now been miniaturised enough to allow transvenous implantation in children with resuscitated sudden arrhythmic death, usually due to long Q-T syndrome or hypertrophic cardiomyopathy.²⁷

Interventional cardiology versus surgery

The growth of interventional cardiology has meant that the simpler defects are dealt with in the cardiac catheterisation laboratory whilst cardiac surgeons are increasingly operating on more complex lesions such as hypoplastic left heart syndrome, which previously would not have been offered treatment.

More importantly, interventional cardiology can complement the management of these complex patients resulting in a better overall outcome for the child born with congenital heart disease. The hybrid procedures represent an example of intervention complementing surgery. Interventional cardiac catheterisation has its own set of complications: device embolisation, vessel or chamber perforation, arterial or venous thrombosis and radiation exposure. These complications can be reduced by careful patient and device selection, meticulous technique, low dose pulsed fluoroscopy and most importantly operator experience.

We have really only witnessed the beginning of interventional cardiology and await further developments in catheter and device design to improve and widen its application in the treatment of congenital heart defects.

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