Stent Implantation in Congenital Heart Disease: A New Therapeutic Modality

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Abstract
Vascular stenoses and hypoplasia in children are often associated with congenital heart disease or occur as a residual lesion or complications after surgical repair. Among these lesions, branch pulmonary artery stenoses are the commonest, other lesions include coarctation of aorta, stenosis of systemic veins after Fontan repairs, post-operative pulmonary venous stenosis, aortopulmonary collateral artery or shunt stenosis and post-operative conduit stenosis. They are usually associated with significant mortality or morbidity if untreated. Conventional surgical repair for these vascular stenoses may be difficult, carries a high operative risk or is associated with disappointing outcome. Some of the lesions may even be surgically inaccessible. Balloon angioplasties may be effective in relieving obstruction in some of these lesions. It can achieve successful dilation in about 60% of branch pulmonary arterial stenoses but the success rate may be lower in other lesions. Furthermore, the effects of balloon angioplasties in many lesions are often temporary and restenosis as a result of elastic recoil or subsequent scarring and fibrosis is common. Implantation of a stent provides a firm support and maintain patency of the vessels after balloon dilatation. Both balloon-expandable and self-expanding stents have been used to treat various vascular stenoses but available evidence suggests that the former is more suitable for use in children as it can be redilated to a higher diameter to keep pace with growth and is associated with fewer complications. The clinical application of stent implantation to treat various vascular stenoses and congenital heart diseases has been continuously widened since the late 1980s. It has replaced surgery as the treatment of choice in branch pulmonary arterial stenoses, postsurgical recoarctation of aorta that are unresponsive to balloon angioplasty, systemic venous obstruction and systemic venous baffle obstruction in Mustard or Senning operations. In native coarctation beyond young infancy, stenting is increasingly accepted as a primary treatment in adolescence and adults to avoid the higher surgical risks in this group of patients. In young children who do not respond to primary balloon angioplasty for native coarctation, stent implantation has also become an accepted treatment provided a stent that allows redilation to the adult aortic diameter is used. Stent implantation in conduit stenosis can prolong the life span of the conduit and delay the time of reoperation. In complex heart diseases where there is aortopulmonary shunt or aortopulmonary collateral artery stenoses, stent implantation serves as an effective alternative to surgical treatment. Stenting of the arterial duct has been increasingly incorporated into the management strategy of complex pulmonary atresias and hypoplastic left heart syndrome. Transcatheter approach using stents to complete the Fontan palliation may avoid multiple operations in patients with univentricular heart. Percutaneous pulmonary and aortic valve implantation to treat the

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Background of Development of Endovascular Stents

Vascular stenoses and hypoplasia in children are often associated with congenital heart disease or occur as a residual lesion or complications after surgical repair. These vascular obstructions can occur in arteries or veins of both the pulmonary and systemic circulations. Among these lesions, branch pulmonary artery stenoses are the commonest, other lesions include coarctation of aorta, stenosis after cavopulmonary shunt or Fontan repairs, post-operative pulmonary or systemic venous stenosis, stenosis of aortopulmonary shunts or collaterals, and post-operative conduit stenosis. Untreated vascular stenoses cause significant mortality and morbidity and will severely affect surgical outcome.

Though conventional surgical repair is effective in some of these vascular stenoses, the operations may be difficult, carries high operative risks or are associated with disappointing outcome. Recurrence of stenoses requiring reintervention is also common. Furthermore, some of the vascular stenoses may be located at surgically inaccessible sites. Therapeutic cardiac catheterisation, therefore, has been developed for more than two decades as alternatives to surgical repair.

Balloon angioplasty or dilation of the stenotic vessel is effective in many lesions. It works by tearing the intima and part of the media to allow remodeling or growth of the stenosed vessel at a higher diameter. Varying degrees of success has been reported for different lesions. Although immediate results may be satisfactory, sustained relief of obstruction can be achieved in only a certain proportion of patients. Restenosis is common. It is due to elastic recoil of the stenosed vessels, subsequent scarring and fibrosis, or external compression. This has urged clinicians and researchers to look for new interventional transcatheter procedures to relieve vascular stenoses.

The concept of using a rigid framework, like a metallic framework, to support the stenosed vessel after balloon angioplasty and thereby preventing restenosis caused by elastic recoil or scarring was first introduced by Dotter1 as early as 1969, using a coil spring endarterial tube graft implanted into a canine popliteal artery to relieve obstruction. However, because of many unresolved technical issues, progress was slow in the following decade. With subsequent advances in technology, Palmaz reported successful use of a metallic, expandable stent, the Palmaz stent (Johnson & Johnson Interventional Systems, Warren, New Jersey), to relieve obstruction in aorta, carotid, iliac and renal arteries,2 which was followed by its use in intrahepatic porto-caval shunt in dogs in 1985.3 In 1987, Schatz, from the same group of researchers, reported success of balloon-expandable intra-coronary stenting in adult dogs.4 Success in animal studies prompted a series of clinical trials. In 1988, Palmaz reported a multicentre trial of intra-luminal stent implantation in atherosclerotic iliac artery stenosis.5 Thereafter, stent application to treat coronary artery stenosis has become widespread.6-10

As for congenital heart disease, Mullins et al11 first described in 1988 successful stent implantation in pulmonary arteries and veins in an experimental model. Subsequently, O'Laughlin et al12 from the same institution reported in 1991 the first success of stent implantation in patients with congenital heart disease. Since then, stents have been used to treat a variety of cardiac lesions including complex congenital heart disease with encouraging results.

Types of Stents and Techniques of Implantation

Two basic types of stents are currently available. They are the balloon-expandable and self-expanding stents (Figure 1). As vessel diameters in congenital heart disease...
are usually larger, stents used are generally larger than those used in coronary arteries.

**The Balloon Expandable Stent**

The balloon expandable stents are made from tubular stainless steel, laser cut in a variety of designs. The prototype, the Palmaz stent (Johnson & Johnson Interventional Systems, Warren, New Jersey) is constructed of stainless steel 0.076 mm in thickness, 3 cm long and 3.4 mm in diameter before expansion. The stent has rows of staggered offset slots (Figure 1a). The nominal diameter after expansion of the stent is 8 mm but its design allows further dilation to 12-18 mm to accommodate growth. Shortening of the stent to 22 mm (27%) would occur at 18 mm diameter. After expansion, the free or open area of the metal stent is more than 90%. As its clinical application becomes widened, different sizes and stent lengths are available to suit different cardiac lesions. As these stents are designed for relatively large blood vessels in congenital heart disease, they are more rigid and after mounted onto a balloon dilation catheter, the whole unit would require use of a large long sheath, which is up to 11-12 French size, for delivery.

The procedure of stent implantation is usually performed under general anaesthesia but it can also be done under conscious sedation in cooperative patients. After the usual haemodynamic study and angiographic delineation of the stenotic vessels, the balloon-expandable stent is mounted manually by crimping onto a balloon angioplasty catheter, sized not to exceed the diameter of the vessel adjacent to the stenosis. The balloon-stent assembly is then advanced over a guide wire within a long sheath which is placed across the area of stenosis. After correct positioning of the stent, the long sheath is withdrawn, exposing the stent, and the balloon is inflated to expand the stent to the desired diameter. The balloon dilation catheter is withdrawn carefully after deflation, leaving the expanded stent to support the stenotic vessel (Figure 2).

![Figure 1](image1.png)

**Figure 1** (a) Balloon-expandable Palmaz stent (i) before mounting, (ii) after mounting onto a balloon dilation catheter and (iii) appearance when partially expanded by the balloon catheter. (b) (i) Self-expanding stent held back by a rolling membrane before expansion and (ii) appearance after full expansion on complete withdrawal of the rolling membrane.

![Figure 2](image2.png)

**Figure 2** (a) Angiogram on lateral projection showing stenosis in the LPA. (b) Fluoroscopy showing positioning of the stent across the stenotic segment of LPA. Arrow (↑) indicates position of the guidewire. (c) Expansion of the balloon-expandable stent at the stenotic segment of the LPA as indicated by the black arrows. (d) Angiogram on lateral projection showing the full expansion of the stenotic segment of LPA after stenting.

Abbreviations: LPA = left pulmonary artery; MPA = main pulmonary artery; ST = stent
Advantages of the Palmaz balloon-expandable stents include the availability of a number of stent lengths and diameters for selection and the ability to be further dilated to a larger diameter to accommodate for somatic growth in children. However, it has several shortcomings. Firstly, manual mounting by crimping may result in slippage of the stent over the balloon during its advancement in the long sheath, which may require removal of the whole stent. Secondly, the rigidity of the stent makes it difficult to negotiate sharp curves and angles. Thirdly, the need for a large delivery sheath, together with the rigidity of the stent, makes its use very difficult in young children. To overcome these limitations, various stent modifications have been made. Premounted stents like the Palmaz Corinthian stent (Cordis, Miami, FL) to reduce the risk of stent displacement over the balloon has been developed.\textsuperscript{13,14} In recent years, the Genesis stent (Cordis, Warren, New Jersey),\textsuperscript{15} which is a new low profile and more flexible stent that can be premounted and delivered through smaller (6-7 French) sheaths is available for use in young children and even infants. It can also be used in more tortuous and smaller vascular stenoses. Some other balloon-expandable stents with a different design are also available in recent years.\textsuperscript{16}

**The Self-Expanding Stent**

The self-expanding stents are of two types: a tubular mesh or a coil design stent. The tubular type self-expanding stents are available in a cobalt alloy stainless steel design or nitinol composite. The stent is mounted on a relatively smaller catheter (5-9 French) and is held behind a rolling membrane before expansion (Figure 1b).

During implantation, the stent is directly positioned across the stenotic area over a wire without the need for a long sheath. By withdrawing the rolling membrane, the stent expand itself from the distal to its proximal end.

The advantages of self-expanding stent are its smaller shaft size and flexibility which facilitate tracking through tortuous or curved vessels, as well as multiple or long stenoses. However, its major disadvantages include a greater tendency of developing obstructive in-stent intimal hyperplasia, stent migration, and the inability to be overdilated to accommodate growth, thus severely limiting its application in children.\textsuperscript{17}

**Percutaneous versus Intraoperative Stenting**

Most of the stent implantation procedures are performed percutaneously in the cardiac catheterisation laboratory. However, when vascular access is limited, using the percutaneous or surgical approach alone is difficult and risky, or when a concurrent surgical procedure is anticipated, stent implantation can be done intraoperatively. Some modifications of technique may be needed but with the cooperation of the cardiac surgeons and interventional cardiologists, good results can be achieved.\textsuperscript{18,19}

**Stent Implantation for Specific Lesions**

**Stenotic or Hypoplastic Branch Pulmonary Arteries**

Branch or peripheral pulmonary artery stenoses may occur as primary (congenital) or secondary (acquired) lesions. Congenital branch pulmonary artery stenoses may occur as a discrete stenosis or diffuse hypoplasia. They may be associated with a wide variety of congenital heart disease, like Tetralogy of Fallot, pulmonary atresia, and other complex congenital heart disease. They may also occur in Noonan, Williams, Alagille and congenital rubella syndromes.

Acquired stenosis may occur after surgical repair of Tetralogy of Fallot, complex pulmonary atresia, conduit repair of right ventricular outflow tract obstruction, arterial switch operation for transposition of the great arteries, and after cavopulmonary connection or the Fontan-like operations. They may also occur as a complication after previous aortopulmonary shunt operations like the Blalock Taussig or Waterston shunt. In addition, contraction of the arterial duct after spontaneous closure or transcatheter device occlusion may cause left pulmonary artery stenosis.

Stenosis of branch pulmonary arteries causes a lot of adverse haemodynamic effects.\textsuperscript{20} It causes underperfusion of the lungs supplied by the stenotic vessel, and hence ventilation-perfusion mismatch and poor growth of the vasculatures distal to the stenosis. Pulmonary hypertension in the contralateral pulmonary arteries may induce pulmonary vascular disease. Unrelieved obstruction would impose pressure overloading on the right ventricle and result in right ventricular hypertension, dysfunction and even failure. Arrhythmia and even sudden death is not uncommon. Overperfusion of the contralateral lung may result in pulmonary oedema. Functionally, patients have limited exercise capacity which may affect daily life. Untreated branch or peripheral pulmonary arterial stenoses is therefore associated with significant mortality and morbidity and will severely affect surgical outcomes of the underlying congenital heart disease.

Results of surgical repair of branch pulmonary artery stenoses are often disappointing. Balloon dilation provides lasting relief in only 60% of cases.\textsuperscript{21} In many instances,
although the stenotic pulmonary arteries can be acutely dilated to a satisfactory size, the natural recoil of tissue and subsequent scarring often resulted in restenosis. The need to overdilate these vessels in order to achieve a lasting effect often lead to complications like aneurysm formation or even vascular rupture causing pulmonary haemorrhage which may rarely be fatal.22

As stent implantation can overcome the problems and avoid complications associated with balloon angioplasty, it has therefore become an attractive new treatment for pulmonary arterial stenosis. Since the first clinical success reported by O’Laughlin et al in 1991,12 the application of stent implantation in branch and peripheral pulmonary artery stenoses has become widespread.20,23-25

Although both balloon-expandable and self-expanding stents have been used, the former is more popular and preferred in children because they can be further dilated to accommodate growth. Other than the most widely used Palmaz™ iliac and hepatobiliary stents (Johnson & Johnson Interventional Systems, Warren, New Jersey, USA), the Intra-stent (IntraTherapeutics, Inc.) and the Numed Cheatham Platinum stent (CP Stent) are also balloon-expandable stents available in the market.

Short-term results of stent implantation in branch and peripheral branch pulmonary artery stenoses have been very encouraging.12,19,23 The procedure has been shown to be safe and effective. Shaffer et al25 further reported in a large series excellent long-term results of endovascular stent implantation in congenital and acquired pulmonary artery stenoses. Their results showed that significant increase in the vessel diameter and decrease in the pressure gradient across the stented vessels can be maintained in most of the patients on the long-term follow-up. Right ventricular pressure and lung perfusion in both groups of patients were also shown to improve significantly after stent implantation. In the small groups of patients who had higher pressure gradient at follow-up examination, further balloon dilation of the stent was effective in decreasing the mean residual pressure gradient across the stented vessels. Early complications included stent migration, stent thrombosis and two deaths but there were no late complications. Long-term significant restenosis occurred in only a few patients. These are important results to confirm the long-term efficacy of stent implantation to treat branch pulmonary arterial stenoses and indicate that it should be the treatment of choice.

**Coarctation of Aorta**

Controversies regarding optimal treatment of coarctation of aorta exist for years. Surgical repair by end-to-end anastomosis, subclavian flap aortoplasty or synthetic patch aortoplasty has been the conventional treatment for coarctation of aorta for over five decades. The overall mortality is only 1-2% in patients 1-69 years of age. Operative mortality in infants and neonates may be slightly higher. However, complications of surgical repair are significant. They include recurrent laryngeal nerve injury, phrenic nerve palsy, chylothorax, bleeding, hypertension and spinal cord ischaemia, which may result in permanent paraplegia. The risk of developing these complications is higher in adolescence and adult patients compared with children. Long-term concern is recoarctation which occurs at a rate of about 10% in most surgical series.26 Aortic aneurysm formation after surgical repair, however, appears to be uncommon.

Interventional cardiac catheterisation has evolved since early 1980 as a non-surgical treatment for coarctation of aorta. The main stimuli behind this trend are the risk of residual or recurrent coarctation and the rare but catastrophic spinal cord infarction that follows surgical treatment. After successful experimental work of balloon angioplasty on postmortem or surgical excised coarctation and on animals,27-29 the original clinical application of the technique started in recoarctation after surgical repair30 because mortality and morbidity of reoperation in this group of patients is high and recurrent obstruction at the repaired sites is common.31-33 Success of balloon angioplasty in relieving recoarctation of aorta made it a standard treatment now and it is further extended to treat native or unoperated coarctation.34,35 It is now clear that relief of obstruction by balloon angioplasty in both recoarctation and native coarctation are similar and can be achieved in 80-90% of cases. It is generally considered safer in patients with post-surgical recoarctation because of the supporting effect of surrounding scar tissue which is absent in native coarctation. Results of balloon angioplasty are much less favourable in neonates and infants,36,37 presumably related to constriction of ductal tissue. It has been shown that balloon angioplasty in this group of young children has a much higher incidence of recoarctation (57% vs 14%) and reintervention rate compared with surgical repair. Therefore most paediatric cardiologist would recommend surgery as primary treatment of coarctation of aorta for neonates and young infants.

The commonest short-term complications of balloon angioplasty, arterial stenosis or occlusion which is particularly prevalent in young children, can usually be avoided by using smaller and low profile balloon catheters
and use of heparin during the procedure. The main long-term complications are aneurysm formation and recoarctation. Aneurysm formation occurs in 5-20% of patients and there is a remote risk of rupture (<1%). This relatively high incidence as compared with surgery is, perhaps, predictable because the mechanism that leads to effective balloon dilation is to tear the intima and part of the media. This would weaken the aortic wall and predispose the patients to aneurysm formation. In addition, over-dilation of the coarctation segment that is sometimes needed in order to reach the desired diameter is also contributing to aneurysm formation or even fatal haemorrhage resulting from acute rupture. Though many of these aneurysms remain stable and small in the medium term, a few may enlarge and require surgery or implantation of a covered stent. Recoarctation after balloon angioplasty is usually due to elastic recoil, persistent or recurrent coarctation. It occurs in about 20% of patients, who will likely require reintervention.

Given the drawbacks and limitations of balloon angioplasty, stent implantation in coarctation of aorta or recoarctation was introduced in the early 1990s and initial success has led to its application on an increasingly large scale over the past eight years (Figure 3). The principal advantage of using a stent is the fact that elastic recoil after balloon dilation is prevented by its metallic framework and the size of the dilated segment can be controlled by the size of the inflating balloon. It is also possible to relieve the coarctation in a more controlled manner – to achieve full relief in stages. These measures avoid over-dilating the coarctation and thereby help reduce the risk of aneurysm formation or aortic rupture. The pooled risk of aneurysm formation from a series of reports is only 3% and the small aneurysms can be occluded by embolisation coils.

Intermediate term follow-up in small series showed that 27-29% patients developed mild degree of intimal hyperplasia in the stents but none of them require stent redilation. Larger scale long-term studies are needed to confirm that stent implantation for native coarctation and recoarctation is genuinely superior to balloon angioplasty though the present available data suggest that it can achieve a more lasting relief of obstruction with lower risks of complications and less need for reintervention.

With the available evidence, it seems that a reasonable approach in children would be to use balloon angioplasty as the standard treatment for post-operative recoarctation. Stent implantation should be used for resistant cases. Native coarctation of aorta beyond young infancy can be treated either by surgery or balloon angioplasty as the initial treatment. Many centres, including our institution, are using balloon angioplasty as the primary treatment. However, in the presence of significant aortic arch hypoplasia, surgical repair should be recommended because balloon dilation seldom produces good relief. Stent implantation, as a primary treatment for native coarctation, should be reserved for adolescence and adults in whom surgical risk is higher and growth limitation caused by the stent is not a major concern.

**Right Ventricular to Pulmonary Artery Conduits**

The placement of extracardiac conduits between the pulmonary ventricle and pulmonary arteries has allowed correction of many complex cardiac lesions. However, multiple surgical reinterventions to replace the conduits are always required because of the inevitable conduit deterioration over time. Stark and colleagues, on reviewing 405 conduits, found an overall conduit survival at 5, 10 and 15 years of 84%, 58% and 31%, respectively, with conduit obstruction, which causes right ventricular pressure overload, being the most frequent reason for surgical reintervention. Factors contributing to conduit deterioration

![Figure 3](image-url)  
*Figure 3* The patient has Williams Syndrome. (a) Aortogram showing a long segment native coarctation of aorta with some narrowings in the cranial vessels. Note the supravalvar aortic stenosis as well. Fluoroscopy (lateral projection) showing (b) partial and then (c) full expansion of the balloon-expandable stent positioned at the narrowed aortic segment. (d) Angiogram (lateral projection) showing full expansion of the long segment coarctation after stent implantation.
include external compression by the sternum, calcification, kinking, aneurysmal change and development of a fibrotic intimal peel. Somatic growth of children also induces a relative conduit obstruction. These changes can occur in both homografts and artificial materials. Even though conduit replacement is considered a low-risk intervention, the second and subsequent conduit have a shorter survival than the original implant because adhesions and calcifications at reoperation make it more difficult to obtain an ideal fit and flow characteristics at replacement. Effects of transcatheter balloon dilation of conduit obstruction are often temporary or ineffective.

Stent implantation in the conduit, by providing a rigid support to overcome obstruction, has been found to be a reliable method to relieve conduit stenosis and postpone surgical conduit replacement. With effective reduction of pressure gradient across the stenosed conduit as well as increase in the diameter of the stenotic lesion, the haemodynamic status of the right ventricle can be improved. Ovaert et al demonstrated that with stent implantation in conduit stenosis, freedom from surgical reintervention was 86% at 1 year, 72% at 2 year and 47% at 4 year and somatic growth of children was maintained. Complications, including stent fracture, and compression of coronary artery, are rare. However, the long-term volume-overloading effect of pulmonary valve insufficiency on the right ventricle after implanting a stent at a valved conduit remains a major concern. Since the main purpose of stent implantation in stenosed conduit is to postpone, but not to eliminate reoperation, and the fact that pulmonary valve insufficiency, which may be present even before stent implantation, is usually well tolerated for a short period of time, the procedure can still be recommended, especially in young children, as a means to prolong the life span of a conduit.

**Systemic Venous and Systemic Venous Baffle Obstruction**

Systemic venous obstruction involving the superior or inferior vena cava may be congenital or occur as an acquired lesion after surgical cannulation for cardiopulmonary bypass, cavopulmonary shunt, Fontan procedures, previous transvenous pacing lead insertion and after heart transplantation at the anastomosis sites. Systemic venous baffle obstruction is also a significant complication in patients receiving the Mustard or Senning operation (atrial switch) to palliate transposition of the great arteries. Relief of obstruction in the caval veins is important for symptomatic relief as in the superior vena caval obstruction syndrome and to maintain vascular access for future intracardiac procedures.

Surgical relief of systemic venous obstruction and post Mustard procedure baffle obstruction requires general anaesthesia, cardiopulmonary bypass, is technically difficult and may be unrewarding. Likewise, balloon dilation alone offers only transient relief of obstruction because of elastic recoil or restenosis. Stent implantation has been developed as an alternate treatment since early 1990s. Both balloon-expandable and self-expanding stents have been used. Ward et al has demonstrated that the procedure is safe and effective in symptomatic relief of superior vena caval obstruction and systemic venous baffle obstruction. Sustained patency of the stented vessels could be maintained on intermediate term follow-up and none of the venous channels required readilation though a minor degree of intimal proliferation developed in some patients. No significant complications occurred on long-term follow-up. At present, balloon-expandable stent has become the treatment of choice for these systemic venous obstructions.

**Patent Ductus Arteriosus**

In ductal-dependent congenital heart disease, the use of prostaglandins to maintain short-term patency of the ductus arteriosus allows stabilisation of the critically-ill neonate before further intervention. In obstructive right heart lesions such as the complex pulmonary atresias, a systemic to pulmonary artery shunt to augment pulmonary blood flow and improve oxygenation is often required in the neonatal period. However, shunt-related complications, including damage to phrenic nerve, shunt stenosis and thrombosis leading to occlusion, pulmonary artery distortion and stenosis, differential growth of left and right pulmonary arteries, pulmonary hypertension, as well as surgical adhesions, will increase the complexity and risks of subsequent surgery. In severe hypoplasia of pulmonary arteries and unfavourable complex cardiac lesions, a shunt insertion may be difficult, if not impossible, and multiple shunt operations may be needed in infancy. A logical approach to avoid a neonatal aortopulmonary shunt would be to prevent the arterial duct from closing. Transcatheter techniques to maintain ductal patency has been explored in the past 2 decades.

While balloon dilation of the arterial duct has been shown to be unreliable or ineffective, stent implantation, on the other hand, has been shown in experimental models to be effective in maintaining ductal patency from a few months up to 21 months and the stented arterial duct can be dilated upon growth or narrowing caused by neointimal proliferation. The success was followed by clinical
application of stenting the arterial duct in various congenital heart diseases in the neonate, including critical pulmonary stenosis, pulmonary atresias with or without ventricular septal defects, and hypoplastic left heart syndrome. Balloon-expandable or coronary stents have been used. Immediate and short-term palliations were encouraging but restenosis of the stented duct due to intimal hyperplasia, necessitating balloon redilation, was common. Other procedural complications include arterial occlusion, acute stent thrombosis, stent migration, incomplete opening and ductal spasm which may be fatal. Although an earlier report on the intermediate-term follow-up results of the stented arterial duct suggested that the duration of palliation was very short, procedural risks were high and long-term survival was poor, a more recent report with a larger number of patients demonstrated more optimistic results. Procedural success was high in selected patients and stent patency can be maintained up to 3 years. Though stent restenosis was common (43%), balloon redilation was successful in 95% of cases. Growth of the pulmonary arteries can be achieved with no distortion and corrective surgery was possible in most of the patients. The overall survival rate for this group of complex cardiac lesions was 86% after 6 years.

While an aortopulmonary shunt remains the standard treatment for ductal-dependent pulmonary circulation, stenting of the arterial duct serves as an effective alternative to surgical treatment for selected cases in which the anatomy is unfavourable, surgical risk is high or a surgical shunt is undesirable. Ductal stenting is also increasingly incorporated into the complicated management strategy for hypoplastic left heart syndrome.

**Aortopulmonary Collateral Arteries and Shunt**

The management of complex pulmonary atresias, i.e. pulmonary atresia with ventricular septal defects and multiple aortopulmonary collaterals is often complicated and challenging. Ideally, patients should undergo a unifocalisation programme (multiple operations may be needed) with ultimate biventricular repair. However, some patients are unable to enter or complete such a staged repair because of severely hypoplastic pulmonary arteries or unfavourable anatomy. Pulmonary blood flow in unrepaired patients, therefore, will depend on the aortopulmonary collateral arteries or surgically created aortopulmonary shunts. However, there is a natural tendency for 58-68% of collateral arteries to progress with time to stenosis or even occlusion, resulting in underperfusion of the lung segments they supply, severe hypoxemia, and limitation in exercise capacity. Similarly, surgical aortopulmonary shunts may also become stenosed with time and patients may present with progressive cyanosis. Palliation by another surgical shunt may provide short-term palliation but it may be difficult to identify a pulmonary artery of sufficient size to put in a successful shunt. Furthermore, aortopulmonary collaterals tend to be soft and friable, making the operation risky.

Balloon dilation of aortopulmonary collaterals and shunts, like surgical palliation, only provides temporary relief or is virtually unsuccessful because of elastic recoil. Therefore, stent implantation of stenosed aortopulmonary collaterals or shunts has become an attractive alternative. Both balloon-expandable and self-expanding stents have been used with successful palliation (Figure 4). The former has the advantage of re-

Figure 4  (a) Aortogram (anteroposterior projection) showing multiple aortopulmonary collaterals in a 2-year-old girl suffering from pulmonary atresia with ventricular septal defect. Note a slightly kinked but patent modified left Blalock-Taussig shunt (arrow). (b) Selective aortopulmonary collateral contrast injection showing an isolated stenotic collateral artery as the sole supply to the left lower lobe. It did not respond to balloon dilation because of immediate elastic recoil. (c) A balloon-expandable Genesis stent was positioned and dilated at the stenotic collateral artery. (d) Angiogram to show full expansion of the stenosed collateral artery after stent implantation, thus securing blood supply to the left lower lobe.
dilation to a larger diameter to keep pace with growth in children whereas the latter, though non-dilatable, has the advantage of higher flexibility in negotiating curves in tortuous collateral arteries and avoidance of multiple stent implantations. Possible complications, the incidence of which is low, include vessel rupture leading to pulmonary haemorrhage, dissection, aneurysm formation, vasospasm and occlusion of the arteries. Significant clinical improvements with increase in arterial saturation and exercise capacity can be achieved in most patients. Redilation of the stented vessels may be needed should restenosis occur. Growth has been observed in some stented vessels. Stent implantation in aortopulmonary collaterals arteries, besides offering good palliation to un repaired complex pulmonary atresias, may also create opportunities for later surgical procedures.

**Pulmonary Venous Obstruction**

Pulmonary vein stenosis occurs as a primary lesion as in congenital pulmonary venous stenoses, total anomalous pulmonary venous drainage (especially with right isomerism) or after its surgical repair. The stenosis can be a discrete narrowing or a diffuse hypoplasia of the pulmonary vein. Multiple sites may be involved. Despite repeated surgical repair or balloon dilation, the prognosis is generally poor and mortality is high. Stent implantation into stenotic pulmonary veins, whether performed percutaneously in the catheterisation laboratory or intraoperatively, may provide short-term palliation. However, in-stent restenosis frequently occurs and responds poorly to further intervention. The long-term outlook in this setting remains dismal.

**Creating Atrial Septal Defect**

Stent implantation at the atrial septum following an atrial septostomy has been reported as a useful non-surgical means to create an interatrial communication to decompress the left atrium in patients with complex left ventricular inflow obstruction not suitable for surgical operation. Similarly, in low cardiac output syndrome occurring after the fenestrated Fontan surgery that is due to narrowing of the fenestration with resultant increased systemic venous pressures, a stent implanted across the restrictive fenestration can improve the haemodynamic status and cardiac output.

**Coronary Arteries**

Congenital coronary artery stenosis is rare but it may occur after surgery for congenital heart disease like the arterial switch operation in which transplantation of the coronary arteries to the neoaortic root (i.e. the original pulmonary trunk) is necessary. Acquired coronary artery stenosis, may occur in Kawasaki disease particularly in those with giant coronary aneurysms. These children can be treated effectively with coronary stenting if balloon angioplasty is unsuccessful. As the underlying pathogenesis of Kawasaki disease is likely to be different from atherosclerosis in coronary artery disease, it is unclear whether coronary arterial stenting in this condition will achieve long-term results comparable to that in atherosclerotic coronary artery disease.

**New Stenting Procedures in Congenital Heart Disease**

As stent implantation to treat congenital heart disease becomes widespread, new procedures using stents alone or in combination with surgical techniques have emerged. Hausdorff introduce a new method of using stent implantation instead of surgery to complete the second stage of the total cavopulmonary connection or the Fontan palliation in univentricular heart. Percutaneous ductal stenting can be combined with bilateral surgical branch pulmonary arterial banding as the initial management for hypoplastic left heart syndrome or other forms of ductal-dependent left heart obstructive lesions. A complete percutaneous approach that include ductal stenting and percutaneous implantation of the Amplatzer pulmonary artery flow restrictor into both pulmonary arteries to palliate hypoplastic left heart syndrome has also been reported recently.

In cardiac lesions that require surgical insertion of a conduit between the right ventricle and pulmonary artery, severe conduit regurgitation, either de novo or as a result of previous stenting to relieve obstruction, can be treated by percutaneous pulmonary valve implantation. The technique in which a bovine jugular valve is mounted in a balloon-expandable stent is introduced by Bonhoeffer and colleagues. Percutaneous transcatheter aortic valve replacement is also under trials.

**Restenosis and Redilation of Stents**

As stents have a fixed diameter that limits vessel growth, somatic growth in children will sooner or later create a relative restenosis. Other than growth, restenosis
of the stented vessels is mostly caused by intimal hyperplasia which reduces the lumen of the stented vessel. However, a 1-2 mm thickness of neo-intima covering the stent surface is a normal vascular response after stent implantation. Histological studies on vessels after balloon angioplasty showed that intimal hyperplasia was caused by tear of the internal elastic lamina and the migration and proliferation of smooth muscle cells and fibroblasts from the tunica media to intima. The migration and proliferation is caused by growth factors such as platelet-derived growth factor, transforming growth factor-beta, or basic fibroblast growth factor.77

Experience from stented coronary arteries showed that stent type, design, and implantation characteristics also influence the intimal hyperplasia and the degree of restenosis.78 Higher inflation pressure and overdilation of the stent, which causes greater vascular injury, will lead to more intimal hyperplasia. In this regard, self-expanding stents may be better than balloon-expandable stents. However, self-expanding stents have their inherent disadvantages. Firstly, continued exposure to its metallic surface and its sustained pressure (expanding radial force) provide a permanent stimulus for neointimal proliferation. Secondly, its alloy that is made of chromium-nickel materials is cytotoxic to the vessel wall, activating T-lymphocytes and macrophages and stimulating smooth muscle cells proliferation. In a collaborative study of our institution with another institution,17 we have demonstrated that the restenosis rate of self-expanding stents in children is much higher than that of balloon-expandable (Palmaz) stents (28% versus 3%).79

Redilation or re-expansion of stent, whether the restenosis is due to resistant stenosis, intimal hyperplasia or somatic growth,80 has been shown to be effective and safe.75,79,80 The effects can also be sustained in the long-term without further complications. Morrow et al81 has demonstrated very nicely in animal studies that stent re-expanding was accompanied by plastic deformation of the neo-intima without neo-intimal dissection. Where the neo-intima was thin, areas of localised neo-intimal abrasions were observed with focal fibrin and platelet adherence to the stent struts. The absence of medial and adventitial haemorrhage or dissection further confirmed that stent redilation, even at a higher diameter, does not create additional injury to the blood vessel.

Future Stents of Congenital Heart Disease

The future stent design will be directing towards developing a low profile stent with increase in flexibility and deliverability to track through tortuous stenotic vessels and small vessels in young children. A wider range of diameters and lengths should be available to suit the great varieties of vascular structures encountered in congenital heart disease and to eliminate the need for multiple implantations.

The second major issue for new stents to address is to decrease restenosis that are due to intimal hyperplasia. Previous attempts to decrease intimal hyperplasia, including coating stents with various medications (medicated or drug-eluting stents), covering a stent with polytetrafluoroethylene (covered stents), radioactive stents and intraluminal radiation therapy after stenting, have been investigated with varying degree of success. As most of these experience come from coronary stents used in adults, the efficacy and safety of these new devices and methods for congenital heart disease and in children remains unknown. However, if the mechanism of restenosis is the same for both coronary arteries and other vessels, the devices may still be future options for children with congenital heart disease. Increase in the radial strength of stents can also help the vessel to resist restenosis that are due to stent recoil or extrinsic compression.

Covered stents have been used uncommonly to treat a few congenital heart disease, like the transcatheter approach to complete the Fontan circuit or as a bailout device when life-threatening haemorrhage caused by rupture of stenotic vessels during balloon dilation occur.82-84 The major drawback at present is the need to use a very large delivery sheath (14 French size) on the femoral artery, thus highlighting the need to miniaturize such devices for use in children.

Conclusions

Stent implantation is safe and effective in relieving a wide variety of vascular stenoses in congenital heart disease. It has become the treatment of choice and provides a cure for many vascular stenoses. In others, where surgical treatment or balloon angioplasties are ineffective, good palliation can be achieved by stent implantation. Patients can, therefore, avoid the high risks of repeated operations.
It has become an important adjunct in the management of complex congenital heart disease. Restenosis caused by intimal hyperplasia remains an important long-term complication but it can be treated effectively and safety by stent redilation and long-term relief of vascular obstruction can be maintained in most patients. Future development and new stent design are expected to overcome the present limitations of stent implantation and widen the scope of its application. Stent implantation is a new therapeutic modality that has added to the armamentarium of paediatric cardiologists and cardiac surgeons in treating congenital heart disease.

References


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