Pediatric interventional catheterization: reasonable expectations and outcomes

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Interventional cardiology, that is the practice of using catheters to treat heart disease, began 4 decades ago with William Rashkind’s development of the balloon septostomy to alleviate cyanosis in transposition of the great arteries [1]. The field expanded explosively in the final 2 decades of the twentieth century, and now there are few defects in which this treatment plays no role. Although most past discussions of the subject have been organized by the type of intervention performed (balloon dilation, device placement, and others), this article considers the role transcatheter therapy plays in the treatment of each major type of congenital heart lesion. The focus is on the clinical ramifications of the interventions rather than their technical aspects.

Congenital heart defects (CHD) can be classified by physiology, complexity, or severity. Complexity and severity often—although not always—go hand in hand: complex defects are most likely to require intervention within the first year of life. The impact of interventional cardiology is most evident in simple defects. Technologic advances have been impressive, allowing pediatric cardiologists to enlarge vascular narrowings, close holes, and occlude vascular connections by using catheters, but it will be some from time before it will be possible to create complex anastomoses and vascular reconstructions by endovascular therapy. Thus, transcatheter treatments have replaced surgery only for simple shunt defects. In complex disease, catheter interventions have a supporting rather than a
lead role. Although surgery is always required, the addition of transcatheter interventions to the therapeutic armamentarium has permitted innovative treatment strategies with lower mortality and improved outcome.

**Simple heart defects**

Approximately 6 to 8 per 1000 individuals is born with a heart defect severe enough to require the care of a cardiologist. Although some of these lesions may result in a critically ill newborn or symptomatic infant, more often patients are asymptomatic and are diagnosed after evaluation of physical findings such as murmurs or hypertension; approximately three fourths of all congenital heart defects are simple [2]. In these lesions, transcatheter treatments are less invasive than surgery, less disfiguring, and result in a more rapid recovery with less discomfort. The procedures are not necessarily safer or more efficacious, and it is important to evaluate the data when making recommendations to patients and their families.

**Atrial septal defects**

Atrial septal defects (ASD) are among the more common defects and account for about 10% of all CHD. Children with these defects are usually diagnosed after referral to a cardiologist for the evaluation of an ejection murmur. Only rarely can symptoms such as failure to thrive be attributed to the presence of an isolated ASD in infants. Usually, when symptoms are present, there are associated lesions that lead to excessive atrial left-to-right shunting (for example mild abnormalities of the mitral valve). All ASDs of consequence need to be closed, but because the impact of ASD is evident only after the first few decades of life, the timing of closure is elective. Practice dictates that most children undergo closure between about 2 and 6 years of age or at the time of diagnosis if this occurs later. The first device for ASD closure was used almost 30 years ago. Device technology matured in the 1980s and 1990s. Now most ASDs are closed by placement of a transcatheter device. Patients with ASD are screened by echocardiography to determine whether they are candidates for the catheter approach. Closure using a device is possible only for secundum defects, not sinus venosus or atrioventricular canal–type (so-called “primum”) ASDs. The defect must have adequate rims and be of a size that permits placement of the device. Although devices to close very large defects (up to 40 mm in diameter) are available, the larger
devices fit only in the heart of an adult. There are three devices currently in use in
this country for closure of ASD: the Amplatzer Septal Occluder (AGA Medical
Corporation, Golden Valley, California), which is used in the vast majority of
implants, the Cardioseal Occluder (NMT Medical, Boston, Massachusetts), and
the Helex Occluder (W. L. Gore and Associates, Inc., Flagstaff, Arizona) (Fig. 1).
Only the Amplatzer device is approved for this use. The Cardioseal device is ap-
proved for other purposes, and the Helex device is still considered investigational.

Patients usually undergo ASD closure as outpatients. Device placement is
performed under a combination of fluoroscopic and echocardiographic guidance.
In children of school age and younger, a general anesthetic is administered to
permit transesophageal echocardiography. In older patients intracardiac ultra-
sound is performed using a specialized catheter (Fig. 2), and the procedure may
be performed with sedation and local anesthesia. Patients typically remain supine

Fig. 2. Images of Amplatzer Occluder device (AGA Medical Corporation, Golden Valley, California) after placement in a patient with atrial septic defect. (A) Radiographic image of the device in lateral view. (B) The appearance of the device on intracardiac ultrasound imaging. The ultrasound catheter sits in the right atrium and aims toward the left. Thus the apex of the sector at the top of the image is right atrium. The device (arrow) is oriented across the atrial septum.
for several hours after the procedure but may ambulate normally by the time they leave hospital. Subsequent care is straightforward, with abstention from vigorous athletics for several days followed by resumption of all normal activities without restriction. Patients receive low-dose aspirin and adhere to bacterial endocarditis prophylaxis precautions for 6 months. At that time a follow-up echocardiographic examination is performed to document effective and complete ASD occlusion, after which prophylaxis is no longer needed.

Several studies have documented the efficacy of closure using a device in comparison to surgery. In general these studies show no or little difference in efficacy rates between the two strategies (95%–98%), with longer hospital stay and higher rate of complications after surgical closure [3,4]. The actual numbers reported for complications and the length of stay vary substantially, the former as the result of variability in ascertainment, and the latter as the result of regional differences in practice. Several other issues are important in evaluating published results. The first is patient selection: whereas all ASDs are closeable by surgery, the same is not true for device closure procedures; thus with better patient selection, the efficacy of device closure improves. The risk of major complications is low after both surgical or device closure; therefore statistical comparisons of major complications are meaningless. The main difference in complication rates is attributable to postprocedural pericardial effusion (post-pericardiotomy syndrome), which is common after surgery but is rare after closure using a device. Treatment of postpericardiotomy syndrome is straightforward with anti-inflammatory medications, but pericardial drainage may be necessary, and rapidly progressive tamponade may be life threatening. Device integrity has not been an issue with the newer generations of devices, and other problems, such as formation of subacute thrombus on the device, have been rare [5].

**Ventricular septal defect**

Ventricular septal defect (VSD) is the single most common heart defect, accounting for almost 40% of all defects [2]. Patients with large VSDs present with overt congestive heart failure in infancy. Moderate defects may have more subtle manifestations of heart failure, such as failure to thrive, and patients with smaller defects are typically asymptomatic. VSD closure is indicated in infants with heart failure, pulmonary hypertension, or failure to thrive. In the absence of these manifestations, indications for closure are less clear, but most concur that closure is appropriate when the shunt through the defect exceeds 2:1; that is, when the pulmonary blood flow is at least twice as much as the systemic cardiac output [6,7].

In contrast to ASDs, surgical closure remains the mainstay of treatment for VSD; however, device closure is being performed with increasing frequency as the technique and technology advance. Like ASD, device closure of VSD is performed under both fluoroscopic and ultrasound guidance. The catheter course required for the procedure is much more complex than that for ASD (Fig. 3).
Fig. 3. Closure of ventricular septal defect (VSD) with device. (A) Catheter course for closure of VSD. The catheter enters the heart from the superior vena cava (upper arrow), loops into the right ventricle, crosses the VSD, and enters the left ventricle (arrowhead). The wire then passes through the mitral valve into left atrium, right atrium, and down the inferior vena cava (lower arrow). A transesophageal echo probe is seen on the same image. (B) Angled angiographic view of the same patient after deployment of a Cardioseal device (NMT Medical, Boston, Massachusetts) in the VSD. There is contrast outlining the ventricular septum (arrows), and the device is seen occluding the defect.
need to manipulate relatively large and stiff catheters through the heart increases the risk of this procedure, especially in very small infants. Another option for small infants with VSD is a combined surgical-catheter (hybrid) approach. In this method, a midline sternotomy is performed, the heart is exposed, and (without the need for cardiopulmonary bypass) the device-delivery catheter is advanced through the free wall of the right ventricle and across the VSD. The device is then opened under echocardiographic guidance. The major advantages of this technique are the avoidance of cardiopulmonary bypass and, in particular, the ability to close VSDs that are in portions of the heart that are difficult to reach by standard open surgical technique (eg, apical VSDs).

The main determinants of candidacy for device VSD closure are the type and location of the VSD and the size of the patient. The first-generation devices for VSD closure were developed to close muscular defects. Patients who are of sufficient size to permit placement of the delivery catheter and in whom location of the VSD allows device placement without valve impingement are candidates for the procedure. Currently one device is approved for VSD closure (Cardioseal, NMT Medical), and one is investigational (Amplatzer, AGA Medical Corporation). Although the devices were originally designed for muscular VSDs, the most common, hemodynamically significant VSDs are of the membranous type. Until recently, the proximity of membranous VSDs to the aortic valve precluded device closure. The Amplatzer Membranous VSD Occluder (AGA Medical Corporation), however, has recently been designed specifically for these defects. Early studies with this device have been encouraging, although longer-term follow-up is necessary.

Published data are much more limited for VSD closure than for ASD closure. Several reports have demonstrated that the procedure is efficacious, reporting success rates from 86% to 100%. These reports include patients with VSDs in whom surgical closure was unsuccessful or who were considered poor surgical candidates [8–12]. Follow-up care of children after device occlusion of VSD is essentially the same as after ASD closure. In summary, VSD closure by device is currently performed for muscular defects and for membranous defects at specialized centers under investigational protocols. It is likely that the procedure will become more widespread as experience with these devices grows, and they are approved for general use.

**Patent ductus arteriosus**

Patent ductus arteriosus (PDA) accounts for 10% of CHD [2]. Larger PDAs may lead to congestive heart failure or pulmonary vascular disease; however, this condition is uncommon other than in the premature neonate. Most often children with a PDA have a small left-to-right shunt, are asymptomatic, and present with a continuous murmur. Thus, the most common indication for closure of the PDA is the prevention of bacterial endocarditis. In larger PDAs closure is also indicated to alleviate overload of the left ventricular volume resulting from the left-to-right shunt. In the current era it is common for PDA to be diagnosed serendipitously on
echocardiograms performed for other reasons. These so-called “silent PDAs” represent a therapeutic dilemma: these lesions are of no hemodynamic consequence, and although there are reasons to believe the risk of endocarditis may be substantially lower than in larger PDAs, no data exist to confirm this supposition.

Transcatheter closure is standard treatment for PDAs other than in the premature newborn population. The procedure is performed on an outpatient basis under sedation and local anesthesia. Closure may be performed using generic embolization coils or the specifically designed Amplatzer PDA Occluder device (AGA Medical Corporation) (Fig. 4). Follow-up care after these procedures is identical to that for other device closures.

Transcatheter closure of PDA has been extensively studied. A European registry series reported an overall efficacy of 95% in more than 1200 procedures

Fig. 4. Amplatzer device closure of patent ductus arteriosus (PDA). (A) Frontal fluoroscopic image demonstrating the occluder device in place (arrow). (B) Lateral projection of an angiogram with occluder in place. The descending aorta (arrowheads) filling the ampulla of the PDA can be seen on the anterior aspect of the aorta with the device occluding it (arrow).
performed between 1994 and 2001 [13]. In this series, successful occlusion was less likely in larger PDAs. Since that large European report, the Amplatzer PDA Occluder has been approved in the United States. The Amplatzer device has proven efficacious, with a closure rate of 100% in one report [14], and is particularly useful for closure of larger PDAs. In current practice the procedure should have an efficacy of greater than 97% (defined by successful device placement with complete occlusion of flow on follow-up ECG study) with a low complication rate.

**Valvular stenosis**

In combination, stenosis of the pulmonary and aortic valves has a prevalence of 0.6 to 1.5 per 1000 population, accounting for about 10% of CHD [2]. Other than newborns with critical stenosis, children with valve stenosis are almost never symptomatic and are diagnosed after cardiologic evaluation for an ejection-type murmur. The indications for intervention in these lesions are primarily the severity of obstruction, first as estimated by clinical examination and echocardiography and then confirmed by measurements obtained at cardiac catheterization. Data from more than 30 years ago, obtained from a sequential follow-up study, suggest that patients with pulmonic stenosis with a gradient across the stenotic valve of more than 50 mm Hg had a better outcome at 20 years’ follow-up when treated with surgery (compared with no intervention) [15]. Similar data for aortic stenosis suggested a gradient of more than 50 mm Hg as the degree of obstruction at which intervention is preferable to observation and medical management [16].

Balloon dilation at catheterization has become the standard first-line treatment for aortic and pulmonic stenosis. The procedures are performed under sedation and local anesthesia. Patients are usually observed in hospital the night of the procedure and then discharged home the following day. Procedural care is minimal, with exercise restriction for a short while to allow healing of catheter entry sites. In pulmonic stenosis, balloon dilation is effective in most patients. The main exception is the subset of patients with so-called “dysplastic pulmonary valve” (common in Noonan’s syndrome), where the valve is thick and often has associated supravalvular narrowing. Follow-up for this procedure now extends to nearly 25 years. Efficacy is long lasting in most cases, although about 8% of individuals require repeat dilation for restenoses [17]. Although dilation results in (planned) valvular regurgitation, because pulmonary artery resistance is normally quite low, the physiologic consequences of the insufficiency are rarely significant in childhood and adolescence. Thus the main long-term issues are the need for observation for the rare cases of restenosis, the continued need for endocarditis precautions, and the unknown long-term (eg, 30–60 years) effects of the pulmonary regurgitation.

Aortic stenosis represents a very different circumstance. All known forms of therapy—balloon dilation, surgical valvotomy, or replacement of the valve—are palliative in the sense that, in all cases, further surgery will probably be needed at
some point because of valve failure. Balloon dilation of the aortic valve relieves obstruction either by separating fused leaflets or by creating small tears that increase the size of the valve orifice. In either case, it is common for the procedure to result in some degree of aortic insufficiency. Attempts to alleviate obstruction completely by using overly large balloons result in an unacceptable amount of regurgitation [18]. Thus, after successful balloon dilation, it is common for patients to have residual obstruction or insufficiency. In most cases the residual physiologic abnormality is mild, with average residual gradients of 22 to 35 mm Hg, but moderate or more severe aortic insufficiency occurs in about 10% of patients [19–21]. The benefit achieved after aortic valve dilation is of variable durability: in one recent report the intervention-free survival was 50% to 60% at 10 years after dilation [22]. The need for subsequent intervention may result from the development of recurrent obstruction, insufficiency, or both. In the first case, repeat valve dilation is an option. When aortic insufficiency becomes severe, surgery is required. Thus follow-up care of patients after intervention for aortic valve disease is quite important. Depending on the severity of residual stenosis and insufficiency, patients may be limited from competitive athletics; all require ongoing assessment of residual valve disease and scrupulous adherence to endocarditis prophylaxis precautions.

Coarctation of the aorta

Coarctation of the aorta occurs in 0.04% of individuals and accounts for 5% of heart defects [2]. Like aortic stenosis, critical coarctation presents in the newborn period with severe heart failure or shock when the ductus arteriosus closes. Older patients are most often asymptomatic and come to attention when hypertension or a murmur is identified. The examination reveals decreased femoral pulses and a blood pressure gradient from upper to lower extremities, and the diagnosis is confirmed by noninvasive imaging of the aorta (echocardiogram or MR imaging). All patients with significant coarctation require treatment. Balloon dilation has been used as a treatment for coarctation since the 1980s, but there is still substantial variation in practice. Surgery is the benchmark against which interventional catheter treatments must be judged. Most agree on the choice of therapeutic approach at the extremes of age. In newborns, the preferred treatment is surgical because there is a high rate of recurrent obstruction after balloon dilation [23]. In patients who present after adolescence and into adulthood, stenting of the coarctation at catheterization is gaining wide acceptance as first-line treatment or at least as an acceptable alternative to surgery. Stents result in effective relief of the obstruction in 92% to 100% of cases. The reported rate of complications for this procedure is difficult to assess accurately, because all reports include relatively small numbers of patients. Many of these reports include one or more patients with major complications, including myocardial infarction and aortic dissection. At follow-up, recurrent coarctation is infrequent but clearly can occur [24–27].
In infants and children with coarctation, balloon angioplasty is widely used as the primary form of therapy, but opinion is divided, and surgery remains quite common. The explanation for this variability in approach becomes clear when one considers the reported results. Balloon dilation is reported as “successful” in 88% to 100% of cases, but recurrent obstruction after a successful dilation has been reported to occur between 0 and 30% of the time, with the highest rates occurring in the very young [28–30]. The most worrisome complication of the procedure is aneurysm formation, and the incidence aneurysm formation has been variously reported as ranging from less than 2% to 40% of cases. The former percentage is as much an underestimate as the latter is an overestimate of the true incidence of this complication. In a more recent single-center series with relatively long follow-up of 69 patients, the freedom from reintervention after coarctation angioplasty was 90% at 1 year and 87% at 5 years with two deaths during the follow-up period. One aneurysm was documented in 69 cases. The median age of the patients in this series was almost 6 years with no patient younger than 6 months [31]. A contemporary surgical series in young infants reports a freedom from reintervention of 88% at 1 year and 82% at 5 years. There were two deaths (2% of patients) [32]. The incidence of recoarctation after surgery clearly decreases after early infancy. Thus it is probably true that in younger children recoarctation is more prevalent after catheter intervention. The variable approaches to treatment are justified by the current state of the state of the literature.

Follow-up care of patients after coarctation angioplasty or stenting includes adherence to endocarditis prophylaxis precautions as well as blood pressure screening. Patients should also undergo follow-up noninvasive imaging studies to evaluate the coarctation site for aneurysm formation. Either cine CT or MR imaging is an acceptable method (the latter can be used only after angioplasty because of the artifact cast by stents on MR imaging). There is growing evidence of endothelial dysfunction in patients with coarctation that persists even after successful therapy. Patients in whom arch obstruction was longstanding because of a delayed diagnosis are at particularly high risk for persistent hypertension even after effective treatment. When present, persistent hypertension typically responds to standard pharmacologic treatment, such as β-blockade, diuretics, or inhibition of angiotensin-converting enzyme. Recommendations concerning activity after interventional therapy of coarctation are individualized. If the arch obstruction has been effectively eliminated, imaging studies have confirmed the integrity of the aorta, and the blood pressure is normal at rest and on exercise testing, it seems appropriate to allow sports participation, although it may be prudent to limit high-intensity isometrics. More restrictive regimens are required when blood pressure control is suboptimal [33].

**Severe and complex heart defects**

Severe heart disease occurs in about 3 of 1000 live-born infants [2]. Most of these patients require treatment as newborns or within the first year of life.
Between 1 and 1.5 per 1000 have complex heart defect (roughly 20%–25% of all CHD). The remaining are patients with severe manifestations of simple defects. Examples of the latter include the newborn with critical left heart obstruction from aortic stenosis or coarctation and the infant with a large VSD and heart failure. Valve disease leading to symptoms of critical obstruction in the newborn is so different from that presenting later in life that it is more appropriately discussed as part of the spectrum of complex lesions.

**Critical pulmonic stenosis**

When an infant is born with critical pulmonic stenosis, the obstruction to flow at the pulmonary valve is so severe that very little blood can cross; instead, the majority of the venous return to the heart shunts from right to left across the foramen ovale. Left-to-right shunting at the ductus arteriosus maintains pulmonary blood flow. As the normal constriction of the ductus arteriosus occurs, the infant becomes progressively and severely hypoxemic. The hypoxemic and cyanotic infant is treated with prostaglandin E₁ (PGE₁) infusion to maintain ductal patency. After ultrasound imaging makes the diagnosis, the patient is catheterized, the diagnosis is confirmed, and the pulmonary valve is dilated. The procedure is more technically demanding than in older children. In early series technical failures and serious complications were common [34]. Refinements in technique and technology have resulted in progressively improved outcome. In current practice technical success (ie, successful dilation of the valve) is achieved in almost all cases. Clinical success, which is defined as relief of significant hypoxemia with no more than mild residual valvular obstruction, occurs in 94% of patients [35]. In rare cases, after successful dilation, patients develop dynamic right ventricular outflow obstruction caused by vigorous contraction of the hypertrophic infundibular muscle. This obstruction may be severe enough to cause significant and persistent hypoxemia, which will require reinstitution of PGE₁ for a time or the short-term use of β-blocker therapy. Overall, the prognosis of patients with isolated critical pulmonic stenosis is excellent through childhood; potential long-term findings await on-going follow-up.

**Pulmonary atresia**

Newborns with pulmonary valve atresia present in a fashion identical to those with stenosis, and the initial medical management is identical. The subsequent management of these patients varies enormously because the disorder is very heterogeneous. Some patients are born with an otherwise normal right heart with the exception of the atretic pulmonic valve, whereas others are born with severe right heart hypoplasia including a tiny tricuspid valve and right ventricle (Fig. 5). Patients with a hypoplastic but patent tricuspid valve may also have communications between the cavity of the right ventricle and the coronary arteries (coronary cameral fistulae). These fistulae are often associated with coronary artery stenoses so that coronary artery flow depends on maintaining a high
pressure in the right ventricle. A variety of treatment approaches are used to deal with the heterogeneity of this disorder. When the right heart is well developed, the treatment may be much like the treatment for pulmonic stenosis, but surgical management is required when there is significant underdevelopment of any of the right heart structures. Surgery may be directed at achieving a biventricular repair or toward single ventricle palliation, depending on the severity of anatomic abnormality [36,37].

Cardiac catheterization is performed in almost all patients with pulmonary atresia. The first goal of the procedure is to complete the diagnostic assessment: angiography is the only way to determine the extent of coronary artery involvement. Patients with a well-developed right heart are candidates for transcatheter treatment. Innovative strategies have been developed that allow the interventionist to perforate the atretic valve plate safely using radiofrequency energy [38,39]. A guide-wire can then be passed through the perforation, and sequential balloon dilation of the valve can be performed. In accomplished hands, successful outcome after transcatheter intervention of pulmonary atresia probably depends more on patient selection than any other factor. Selection criteria continue to evolve, and there is significant institutional variation. The approach at the Children’s Hospital of Philadelphia is to perform catheter intervention in patients with an adequate-sized tricuspid valve and well-developed right ventricular outflow tract (infundibulum) in whom right ventricular–dependent coronary circulation has been ruled out. Early published results of this strategy reflect the evolution of technique; valve dilation was unsuccessful in many cases, and pericardial tamponade from inadvertent perforation of the heart was sometimes fatal [35]. Contemporary series highlight the difficulties with patient selection. Technical problems are much less frequent, but it remains common for patients to require surgical shunts for augmentation of pulmonary blood flow or outflow tract patches to achieve complete relief of outflow obstruction [38–40].
Follow-up concerns in infants with pulmonary atresia vary depending on initial anatomy and the type of intervention performed. After transcatheter perforation and dilation, oxygen saturation is assessed for evidence of atrial right-to-left shunting, and echocardiography is performed to quantify residual obstruction. In patients without residual hypoxemia, the prognosis through childhood tends to be quite good, although some will require repeat intervention for restenoses, and, as is true for almost all therapies for CHD, the truly long-term (decades) outcomes require on-going study.

Critical aortic stenosis

Critical aortic stenosis in the newborn remains a challenging and high-risk lesion. The designation “aortic stenosis” belies the complexity of this disorder in the neonate. Some patients have a left ventricle that is anatomically and functionally normal after relief of the obstruction, others have what is essentially hypoplastic left heart syndrome (HLHS), and still others have a left ventricle of adequate size but with such severe myocyte injury that the ventricle is not recoverable. When the ductus arteriosus closes after birth in a neonate with critical aortic stenosis, the patient develops the classic picture of critical left heart obstruction: heart failure and shock, followed by circulatory collapse. Initiation of PGE1 infusion allows restitution of systemic blood flow and medical resuscitation.

Therapeutic decision-making for patients with critical aortic stenosis is clear at the ends of the spectrum of the disease: patients with a very hypoplastic left ventricle are palliated like those with HLHS, whereas those with a normal-appearing left ventricle undergo balloon valvuloplasty. The optimal treatment strategy for patients with some degree of left ventricular hypoplasia or evidence of myocardial damage is less clear. Options include the two procedures discussed previously as well as surgical enlargement of the left ventricular outflow tract with autograft aortic valve replacement (Ross-Konno operation) [41]. The latter approach results in a biventricular circulation and is ideal for infants with a very small aortic annulus but an otherwise adequate left ventricle.

The efficacy of balloon valvotomy in newborn critical aortic stenosis is comparable to that of surgical valvotomy and thus has largely replaced the surgical procedure. Patients are intubated and mechanically ventilated for the procedure, commonly on inotropic support and PGE1. Before dilation of the valve, diagnostic information from catheterization is used to supplement the previously obtained echocardiographic data to confirm that valve dilation is the optimal strategy for the patient. If this treatment is deemed appropriate, the valvuloplasty is performed. After balloon aortic valvotomy, most infants need several days of continued intensive care with inotropic support to allow recovery of left ventricular performance.

Reported early survival of newborns after balloon dilation of critical aortic stenosis is 88% in patients without significant left ventricular hypoplasia; most deaths are procedure-related. Subsequent mortality was negligible when followed for up to 8 years. In marked distinction to patients with pulmonary valve disease,
however, the need for reintervention was common—46% [42]. In the authors’ recent experience at the Children’s Hospital of Philadelphia, procedural mortality is lower than in past reports (4%), but subsequent intervention remains quite prevalent. The actuarial freedom from reintervention is only 45% at 3 years [43]. If reintervention is necessary in childhood, it is likely to be necessary in the first year of life. Recurrent aortic stenosis, insufficiency, or both necessitate re-intervention. Patients with isolated restenosis may benefit from repeat balloon dilation, but many of these patients will undergo autograft root replacement (Ross procedure).

To summarize, aortic valve disease is a lifelong disease, and patients who require dilation of aortic stenosis as newborns are likely to have significant residual disease and on-going cardiovascular concerns. Although all patients with aortic stenosis require cardiac follow-up, newborns with critical aortic stenosis require frequent evaluation during growth and are virtually certain to need subsequent aortic valve interventions during infancy, childhood, and possibly throughout adulthood. With appropriate follow-up management, however, the majority of these children remain asymptomatic [42,43].

Complex cardiac defects

Although the day may come when even complex defects can be treated by endovascular therapy alone, with current techniques catheter therapy of these disorders is an adjunct (albeit an important one) to surgery. It is not possible to give an exhaustive account of the role of catheter interventions for each defect in this review. Rather, one major lesion, HLHS, is used as an example to highlight some of the ways in which interventional cardiology adds to the treatment of complex defects.

The most widely applied treatment strategy for HLHS is a staged surgical approach: the Norwood operation is performed in the newborn period to establish unobstructed blood flow to the body and provide regulated pulmonary blood flow. The Norwood operation is followed by a bidirectional Glenn operation at around 6 months of age. In this procedure, the shunt placed during the Norwood procedure is closed, and the superior vena cava is anastomosed to the pulmonary artery. This operation reduces the volume load on the single ventricle, converting the source of pulmonary blood flow from a systemic arterial shunt to a cavo-pulmonary connection. Symptoms of heart failure typically resolve rapidly. To separate the pulmonary and systemic circulation more completely, these children then undergo the Fontan operation, which diverts the inferior vena caval blood to the pulmonary circulation, sometime between 18 months and 3 years of age. At every stage of this process situations can arise that require catheter intervention. In current practice patients almost always undergo multiple interventional procedures over the course of their treatment. The procedures themselves can best be understood in the context of the child’s stage of reconstruction and the physiologic and anatomic abnormality being addressed. Table 1 provides an outline based on this scheme.
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Catheter interventions after first-stage palliation (Norwood operation)

Recoarctation is one of the most common causes of heart failure or ventricular dysfunction after Norwood operation and is also the most common problem requiring catheter treatment. When data at Children’s Hospital Philadelphia were reviewed, 56 (9%) of 605 early survivors of the Norwood procedure developed distal arch obstruction. Most underwent subsequent intervention, although in some the diagnosis of arch obstruction was made at postmortem examination. Patients with recurrent arch obstruction typically present between 6 weeks and 4 months after surgery, although obstruction may first manifest at any time from the immediate postoperative period to a year of age or older. The clinical spectrum of presentation for this entity varies depending on the severity of obstruction and other aspects of the patient’s cardiovascular function. In the most severe cases, patients present with shock, and sudden death has occurred in this group [44]. More commonly, patients have signs of congestive heart failure with echocardiographic evidence of ventricular dysfunction. In many instances, no symptomatology is present, and the diagnosis is based on a combination of clinical signs and noninvasive evaluation. Physical examination may underdiagnose obstruction. Shunt runoff tends to accentuate femoral pulses even in the presence recoarctation, and blood pressure assessment can be misleading when arch obstruction coexists with distortion of the brachiocephalic arteries.

All patients with evidence of distal arch obstruction require catheterization. Patients with low arterial oxygen saturation before angioplasty must be carefully monitored after the procedure. Dilation results in a decreased perfusion pressure to the more proximally placed shunt, and occasionally such children develop severe hypoxemia after relief of arch obstruction [45].

Published reports document immediate success of angioplasty for HLHS arch obstruction of 90% or greater, with periprocedural mortality rates of 15% to 25%. Recurrent obstruction has been noted in a significant but variable number of cases [46,47]. At the Children’s Hospital of Philadelphia, 39 patients underwent balloon dilation between 1986 and January 2001. Periprocedural mortality was zero. Procedures were all considered successful in that no patient undergoing dilation required surgical arch revision, although three patients had gradients between 15 and 20 mm Hg after dilation. Consistent with other series, repeat dilation was required in 25% of the patients in this group [44].

Inadequate pulmonary blood flow causes hypoxemia in the palliated HLHS patient. After the Norwood operation, inadequate pulmonary blood flow may be caused by obstruction of the shunt from stenosis or thrombosis or by restriction of the atrial septal defect—the venous return from the lungs must cross the atrial septum to get to the systemic ventricle. An inadequate atrial septal defect may be enlarged by a number of catheter techniques including balloon dilation and stenting of the atrial septum. Although the ASD must usually be enlarged by subsequent surgery, successful catheter intervention allows this surgery to be delayed until the patient’s next planned cardiac surgical procedure.

Successful outcome after Norwood operation depends in part on a precise balance of systemic and pulmonary blood flow: too little pulmonary flow results
in unacceptable hypoxemia, whereas too much leads to heart failure from excessive work of the right ventricle. To provide this balance, the systemic-to-pulmonary shunt is typically a Gore-Tex (W.L. Gore and Associates, Inc., Flagstaff, Arizona) tube 3 to 4 mm in diameter if the shunt arises from the innominate or subclavian artery or 5 to 6 mm in diameter if the shunt arises from the right ventricle (Sano modification). These Gore-Tex tubes may become narrowed from suture line stenosis, kinking, or, more dramatically, thrombosis. Shunt narrowings can be treated by catheter intervention, avoiding the need for reoperation on fragile infants. Catheter techniques include thrombectomy or stenting (Fig. 6). After such procedures the patients are probably at risk for recurrent thrombosis and are typically treated with antiplatelet and or anticoagulants to decrease this risk.

An important goal of catheterization before the second stage of palliation is the identification of veins that would result in hypoxemia after surgery by allowing superior venous return to bypass the pulmonary vascular bed to the pulmonary venous atrium. The most common of these veins is a persistent left superior vena cava (LSVC) that drains inferiorly to the coronary sinus. When a normal innominate vein and right superior vena cava are present, the LSVC is occluded by using a catheter to deposit small metal coils into the vessel. These coils promote thrombosis of the vein.

Interventions after Glenn and Fontan procedures (stages II and III)

After Glenn and Fontan procedures, patients undergo catheter intervention either to decrease the risk of complications by treating anatomic abnormalities or to alleviate symptoms. Patients are at risk for a wide range of complications after the Fontan operation, but most can be categorized into one of two main symptom
complexes: cyanosis or heart failure (see Table 1). Pulmonary blood flow in these patients is passive from the systemic veins to the pulmonary arteries. If a connection allows the systemic venous return to bypass the lungs, the blood will preferentially go through this lower-resistance pathway, resulting in right-to-left shunt and hypoxemia. These connections may take many forms: they can be the result of venous collaterals, intertrabecular pathways from one atrium to the other, or baffle leaks. The venous connections can be closed by inserting coils, as previously described [48]. Baffle leaks and other short connections are amenable to closure with one of the devices designed for occlusion of ASD or PDA.

Heart failure after Glenn and Fontan operations manifests as edema, ascites, and pleural effusion. Because there is no ventricle to pump blood through the lungs, anything that increases the resistance to flow through the pulmonary circuit will cause these symptoms, including obstruction of the Fontan baffle, pulmonary artery stenosis, and elevated pulmonary resistance. Also, if there is pump dysfunction or significant valvular regurgitation, the elevated filling pressures will be reflected through the pulmonary circuit and result in an elevated systemic venous pressure, with the same result. Causes include pressure load on the ventricle from residual coarctation and volume load on the ventricle from systemic-to-pulmonary collateral arteries. Narrowings of the so-called “Fontan circuit” (baffle stenosis and pulmonary artery stenosis) are usually best treated with large stents. Elevated pulmonary resistance is difficult to treat directly, but creating a connection between the systemic and pulmonary venous chambers can alleviate the resultant symptoms of right heart failure. This surgically created Fontan baffle leak (Fontan fenestration) causes some arterial desaturation but has the benefit of augmenting cardiac output [49–52]. These hemodynamic changes often result in clinical improvement (eg, weaning from mechanical ventilatory support, cessation of chest tube drainage, hospital discharge) [51]. Finally, recoarctation can be treated with balloon dilation or stents, and collateral arteries can be closed by coil embolization as described previously. In summary, catheter interventions, when creatively applied to the wide variety of problems that arise in the care of patients with complex disease, can have a dramatic impact on mortality as well as on quality of life in these patients.

References


