The Emerging Role of Percutaneous Intervention in Adults With Congenital Heart Disease

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Adults with congenital heart disease (CHD) constitute a patient population that has grown rapidly, due to advances in diagnosis and treatment of children with CHD. Though ideally served by cardiologists with advanced training in congenital conditions, adults with CHD often receive the majority of their care from primary care physicians and general cardiologists. These patients often have unique clinical presentations. An understanding of available therapeutic modalities can assist clinicians in the timing of subspecialty referral. This review focuses on the unique challenges of cardiac catheterization and the role of percutaneous interventions in adults with CHD.
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Numerous medical and surgical pioneers have made advances in the field of congenital heart disease (CHD) and, as a result of their accomplishments, survival to adulthood has become the norm for most patients with these disorders.[1] According to recent estimates, there are now nearly three quarters of a million adults with CHD,[2] and these numbers will continue to rise with further advance in diagnosis and treatment. The purpose of this review is to illustrate

the important role that diagnostic cardiac catheterization continues to play in the evaluation of an adult with CHD, and to review the most commonly performed percutaneous therapies.

Basic Approach

An organized approach to diagnosis and management is particularly important in patients with CHD, and the critical first step should be the gathering of historical data. Review of pediatric records, if available, is essential in understanding the complexities of the cardiac and vascular anatomy and defining the outcomes of previous diagnostic studies and surgeries. Surgical procedures have changed considerably over the last several decades, and anatomic presumptions based on current practice may not apply.

Certain signs and symptoms should prompt an extensive evaluation of adults with CHD, particularly syncope and progressive exertional dyspnea. Arrhythmias are not uncommon in adults with CHD, and often originate near the myocardial scars of previous surgeries. Supraventricular arrhythmias, such as atrial flutter or fibrillation, are often poorly tolerated due to a dependence on atrial mechanical function.[3] Ventricular arrhythmias, which can result in sudden death, appear to be more common in certain populations, such as those with a corrected tetralogy of Fallot with a widened QRS interval on a surface electrocardiogram.[4]

Hemodynamic derangements can be quite subtle, such as pulmonic regurgitation following a patch outflow repair of tetralogy of Fallot. Because of the low-pressure pulmonic insufficiency, this can be overlooked during auscultation and routine echocardiography, and can result in right ventricular enlargement[5] and increased risk of sudden death.[6] Thus, knowing the precise surgical procedure can guide what needs to be done at cardiac catheterization.

Diagnostic imaging is a critical adjunct, and less invasive modalities such as echocardiography should always be the first choice for evaluation.[7] The use of echocardiography can be limited by difficult windows due to excessive scar tissue from previous surgeries, concomitant lung disease, and obesity. Subsequent computerized tomography (CT) scanning or magnetic resonance imaging (MRI) may add substantially to the anatomic description, especially in patients with unclear great vessel or pulmonary vascular anatomy.[8,9] The use of MRI has expanded with more widely available scanners and simplified scanning protocols. CT scanning is complicated by the need for intravenous contrast, and MRI is generally not compatible with current implantable cardiac devices.

Diagnostic Cardiac Catheterization

Diagnostic cardiac catheterization, though generally performed later in the diagnostic workup of CHD patients than in the past, remains the gold standard for pressure measurement, output calculation, and vascular resistance determinations. The relative size of shunt lesions can be assessed using oximetry, and the hemodynamic consequences of additional blood flow can be assessed. Most importantly, cardiac catheterization affords the opportunity to intervene and palliate or repair anatomic defects or to clarify the suitability of further surgical intervention.

Assessment of Intracardiac Shunting

Anatomic shunting can be quantified in the catheterization laboratory by examining blood/oxygen saturations in the respective chambers.[10] The mixed venous (MV) saturation is the saturation of blood returning to the right atrium (RA) with contributions from the inferior vena cava (IVC), superior vena cava (SVC), and coronary sinus (CS). IVC saturation is normally higher than that of the SVC, due to high renal blood flow and less oxygen extraction by the kidney. CS saturation is very low, but its volume of contribution is negligible and usually ignored. To normalize MV saturation, 3 times the SVC saturation is added to the IVC saturation and the sum is divided by 4.

Because so much mixing of blood with differing saturations occurs in the RA, an 11% increase in oxygen step-up (saturation increase from a chamber to its successive chamber) is required to diagnose a shunt lesion between the SVC and the RA. A 7% increase is necessary to detect a shunt between the RA and right ventricle (RV) and a 5% increase to detect a shunt between the RV and pulmonary artery (PA). A quick and simple measure of the overall size of a left-to-right shunt ratio can be obtained by using the formula: (aortic saturation – MV saturation)/ (PV saturation – PA saturation). The PV saturation, if not directly measured, can be assumed to be 95%.

Generally, a shunt is considered significant when the shunt ratio is greater than 1.5:1.0. However, the simplicity of this formula is frequently lost in adults. As pulmonary hypertension develops and RV compliance falls, a left-to-right shunt that was 3-to-1 for 30 years may become less than 1.5-to-1 due to the gradual reversing of the shunt. In fact, the left-to-right shunt may totally reverse at some point, and result in arterial desaturation, the so-called Eisenmenger's Syndrome.[11] The significance of a shunt in the adult must, therefore, be examined in the context of other hemodynamics,

chamber sizes, and the history of the defect over time.

Assessment of Pulmonary Hypertension

Pulmonary hypertension is a frequent complication of certain CHDs. It can be secondary to pulmonary venous hypertension from elevated left-sided filling pressures, or the result of systemic-to-pulmonary artery shunting. For unclear reasons, shunts proximal to the tricuspid valve (atrial septal defects or partial anomalous pulmonary venous return) infrequently result in pulmonary hypertension (~ 15% of cases) despite high pulmonary blood flow.[12] The development of pulmonary hypertension from shunts distal to the tricuspid valve, however, is highly dependent on pulmonary blood flow. For example, a large unrestricted ventricular septal defect (VSD) may not result in pulmonary hypertension if the pulmonary circuit is protected by concomitant pulmonary valvular or subvalvular obstruction.

To help differentiate the cause of pulmonary hypertension, the pulmonary vascular resistance should be determined: (mean PA pressure – mean pulmonary capillary wedge pressure [mmHg])/(pulmonary blood flow [liters/min]). Higher resistances (> 7 Wood units or a ratio of the pulmonary-to-systemic vascular resistance of > 0.5) have also been associated with considerably higher perioperative mortality.[13] In addition, assessment of pulmonary vascular reactivity with endothelium-dependent vasodilators, such as inhaled nitric oxide or intravenous adenosine, may provide additional prognostic information in these patients by confirming whether any of the observed pulmonary hypertension has a vasoconstrictor component.[14]

Percutaneous Interventions

Percutaneous interventions in patients with CHD may generally be broken down into 3 types: percutaneous occlusion of shunt lesions, percutaneous balloon valvotomy or valvuloplasty, and balloon angioplasty and stenting of vascular structures. Special attention should be taken in approaching these patients because adults with CHD have well described hematologic and metabolic disturbances that can influence periprocedural care.[15] By the time these patients reach adulthood, they are likely to have undergone diagnostic and therapeutic angiography multiple times. Patients may have memories of needing to be restrained and sedated, and as a result, fear the cardiac catheterization laboratory. Indeed, the adult cardiac catheterization may be the first they have had without general anesthesia. Education and preparation of these patients prior to their procedure is critical. Depending on the expected duration of the procedure and the level of patient anxiety, involving an anesthesiologist in the procedure may also improve patient comfort and facilitate case flow.

Vascular access must be carefully thought through to ensure all cardiac structures can be entered. If the patient has had multiple cardiac catheterizations, routine access sites, such as the femoral artery and vein, may not be available due to injury from childhood invasive procedures. Regardless, the cardiac catheterization should be planned and focused on obtaining data not available by noninvasive methods, and then by proceeding with the appropriate intervention in as expedient a manner as possible.

Percutaneous Occlusion of Shunt Lesions

Intracardiac shunts are the most common forms of congenital heart lesions and are often diagnosed in otherwise healthy adults. These are associated with increased pulmonary blood flow that can lead to right heart enlargement and arrhythmias. In post-TV lesions, they also frequently result in pulmonary hypertension. The surgical correction of many of these lesions has been previously determined to be safe and efficacious. Percutaneous alternatives are becoming more common as a way to avoid the morbidity and mortality of surgery.

Atrial Septal Defect

Atrial septal defect (ASD) is the most common congenital heart defect encountered in adults (excluding mitral valve prolapse and bicuspid aortic valve), accounting for up to 15% of all adults with CHD. An ASD is a direct communication between the atria that allows for shunting of blood between the atria. It results from improper embryologic development of the atrial septum.

Figure 1 outlines the embryologic formation of the atrial septum. Initially, the septum primum migrates from the superior atrial wall to the ventricular cushions. The gap preceding the migrating septum is termed the ostium primum. If this gap persists after birth, an ostium primum defect is present. Normally the septum continues its migration and separates the right and left atria into 2 chambers. Fenestration(s) then form in the middle of the septum primum (the ostium secundum). A second septum (the septum secundum) on the RA side of the first septum then begins forming and migrates downward past the ostium secundum, eventually covering it over and fusing with the septum primum. If this second septum fails to cover the ostium secundum, an ostium secundum defect is present (Figure 1C).

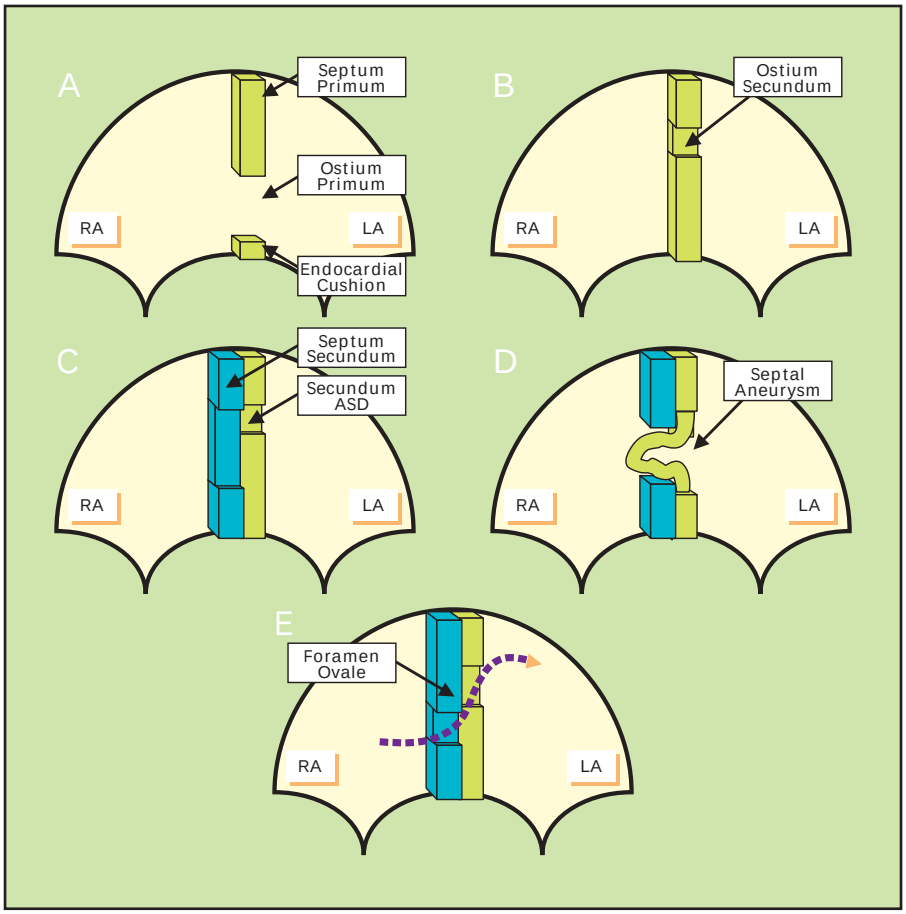

Figure 1. *Anatomic formation of the interatrial septum and the most common abnormalities encountered in practice (see text for details). ASD, atrial septal defect; LA, left atrium; RA, right atrium.*

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If the septum is redundant, a septal aneurysm is said to be present (Figure 1D). If there is failure of the fusion of the 2 septi, then a potential pathway for blood flow remains, which is referred to as a patent foramen ovale (PFO; Figure 1E).

In the adult, the PFO is the most common abnormality seen, occurring in up to 25% of all patients.[16] It is usually of no clinical consequence, but may be associated with cryptogenic stroke. The most commonly occurring atrial septal lesion that results in significant shunting is the secundum ASD, as explained above, lying in the middle of the atrial septum. More than 1 fenestration may co-exist. Less common variants include defects in the fusion of the vena cava to the left atrium (sinus venosus defects), the aforementioned primum ASDs, and defects in the coronary sinus roof with shunting into the left atrium. At this time only the PFO and secundum ASD have successfully been approached by percutaneous methods. Sinus venosus ASD almost always has a persistent, partially-anomalous pulmonary vein present and requires surgical intervention.[17]

Indications to repair an ASD have historically included evidence of right heart volume overload (resulting from the ASD) or the presence of a hemodynamically significant defect (classically a Qp/Qs of 1.5:1). The timing of closure of an ASD appears important. Closure after the age of 40 is associated with an increased incidence of arrhythmias (i.e. atrial fibrillation) compared with closure before age 40.[18] Epidemiologic evidence also suggests that long-term survival is worse with unrepaired defects.[19]

The classic surgical approach to closure of a secundum ASD is a median sternotomy and primary suture closure of the defect or closure with a pericardial or synthetic patch. The procedure can also be done with a mini-thoracotomy. Though in younger patients the quoted mortality is less than 1%, the morbidity of a median sternotomy remains considerable and many patients opt for percutaneous closure.

There are a number of different types of percutaneous devices available that have been studied for closure of an ASD (Figure 2). Whereas many secundum defects are oval in shape, there should ideally be an adequate rim around the defect (at least 4 mm) to hold any of these devices in place. Although both fluoroscopy and transesophageal echocardiography have traditionally been utilized in the placement of the device, the ability to use intra-cardiac echocardiography (ICE) during percutaneous closure has been a substantial advance (Figure 3), and precludes the need for general anesthesia.[20,21] Though both the CardioSeal® (NMT Medical; Boston, MA) and the Amplatzer® Septal Occluder (AGA Medical; Golden Valley, MN) are being used clinically for ASD occlusion, only the legless Amplatzer® device has received US Food and Drug Administration approval for this indication. To achieve proper apposition of the device, the maximal stretched diameter is measured either with fluoroscopy or echocardiography by

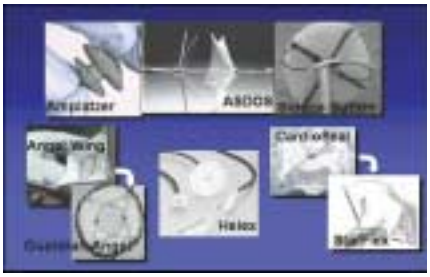

Figure 2. *Recent designs of atrial septal occluders. Only the Amplatzer® septal occluder (AGA Medical; Golden Valley, MN) is currently FDA-approved for occlusion of atrial septal defects.*

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inflating a soft balloon positioned across the defect (Figure 3B). An Amplatzer® device with a waist 2 to 4 mm larger than the diameter is then selected for the closure. A percutaneous closure usually takes under an hour, and the patient can be discharged on the following morning.

Du and colleagues[22] recently reported the results of a prospective study where participating centers had the option of referring patients for percutaneous occlusion with an Amplatzer® device or surgical repair of the ASD. Successful occlusion was the result in 96% of percutaneous procedures and 100% of surgeries ($P = .006$). Mortality was 0% in both groups and complications occurred in 7.2% of the device group and 24.0% of the surgical group ($P < .001$). The mean length of hospital stay was reduced from 3.4 ± 1.2 days for the surgical group to 1.0 ± 0.3 days for the device group.

Reported adverse outcomes with percutaneous occlusion include device embolization, device thrombosis, mitral regurgitation, and pulmonary vein obstruction. The subsequent anticoagulation regimen is controversial, though we usually prescribe aspirin and a thienopyridine for at least 6 months (by this time endothelialization of the device should be complete). Though unsubstantiated, patients should also receive endocarditis prophylaxis for at least the same duration of time and possibly for life after percutaneous closure.

Following percutaneous closure, right heart hemodynamics can improve quite dramatically.[23] Regression of right heart enlargement typically occurs over weeks to months and some reductions in pulmonary artery pressure can occasionally be seen immediately. Follow-up transthoracic echocardiography is recommended by the manufacturer and recent data suggest a possible role for screening transesophageal echocardiography to exclude device-related thrombus during clinical follow-up.[24,25]

Patent Foramen Ovale

The role of PFO in cryptogenic stroke remains unclear. Certainly there are data showing that in patients less than 55 years of age, who have either a transient ischemic attack (TIA) or stroke, paradoxical emboli may be at work.[26] It also appears that the concurrent presence of an atrial septal aneurysm may further increase the risk of a future event.[27] Several current studies are directly addressing the proper usage of closure devices in this situation. Currently, the CardioSeal® and the Amplatzer® PFO occluders are available through a US Food and Drug Administration Humanitarian Device Exemption for patients with a recurrent event (TIA or cardiovascular accident) while on any type of anticoagulant (aspirin or warfarin) and in whom no other explanation for the central nervous system event is obvious. All patients should be screened for hypercoagulability first (including a check of the levels of platelets, protein C and S, factor V Leiden, and prothrombin antibodies).

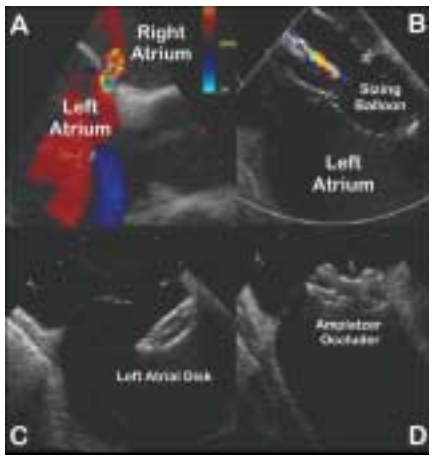
Other potential indications for PFO closure include the presence of platypnea orthodeoxia (hypoxia by shunting through the PFO upon upright position), hypoxemia in certain patients with elevated RA pressures and right-to-left shunting, and those who have had decompression illness and want to continue to scuba dive.[28] There are also anecdotal reports of the advantage of PFO closure in patients with migraine headache (on the presumption that some humoral factor may be able to cross into the arterial circulation that ordinarily would be broken down by the lung parenchyma).[29]

The Amplatzer® PFO occluder is similar to the Amplatzer® ASD device, with the exception that the larger disk is on the right rather than the left side of the septum and the connector between the disks is narrower. Deployment techniques and complications are similar as well.

Ventricular Septal Defects

VSDs can now be approached by percutaneous techniques.[30] At this time the CardioSeal® device is approved for muscular VSDs[31] and post-infarction VSDs. An Amplatzer® device is also under investigation and the results of a registry of 75

Figure 3. *Intracardiac echocardiographic images obtained at the time of atrial septal defect occlusion using an Accunav imaging probe (Accuson, Mountain View, CA). Color flow demonstrates a left to right shunt **(A)**. The stretched diameter of the defect is measured to allow proper sizing of the occluder **(B)**. Release of the left atrial disk followed by the right atrial disk of the Amplatzer® septal occluder **(C, D)**.*



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patients with muscular VSDs has recently been published.[32] Devices were successfully implanted in 87% of cases. Procedure-related major complications occurred in 11% of 75 patients including 2 device embolizations, a cardiac perforation, and 2 deaths. The complete closure rate was 47% at 24 hours, 70% at 6 months, and 92% at 1 year.

VSD closure differs substantially from ASD closure in that the RV trabeculae are quite prominent, and the RV side of the device has difficulty opening completely.

VSD closure differs substantially from ASD closure in that the RV trabeculae are quite prominent, and the RV side of the device has difficulty opening completely. There are probably too little data at this time to analyze outcomes, but certain patients, especially those with congenital muscular VSDs appear to do very well. Post-infarction VSD patients have a tremendously high rate of mortality, regardless of course of treatment,[33] and the use of the occluder device in this setting is still being evaluated.

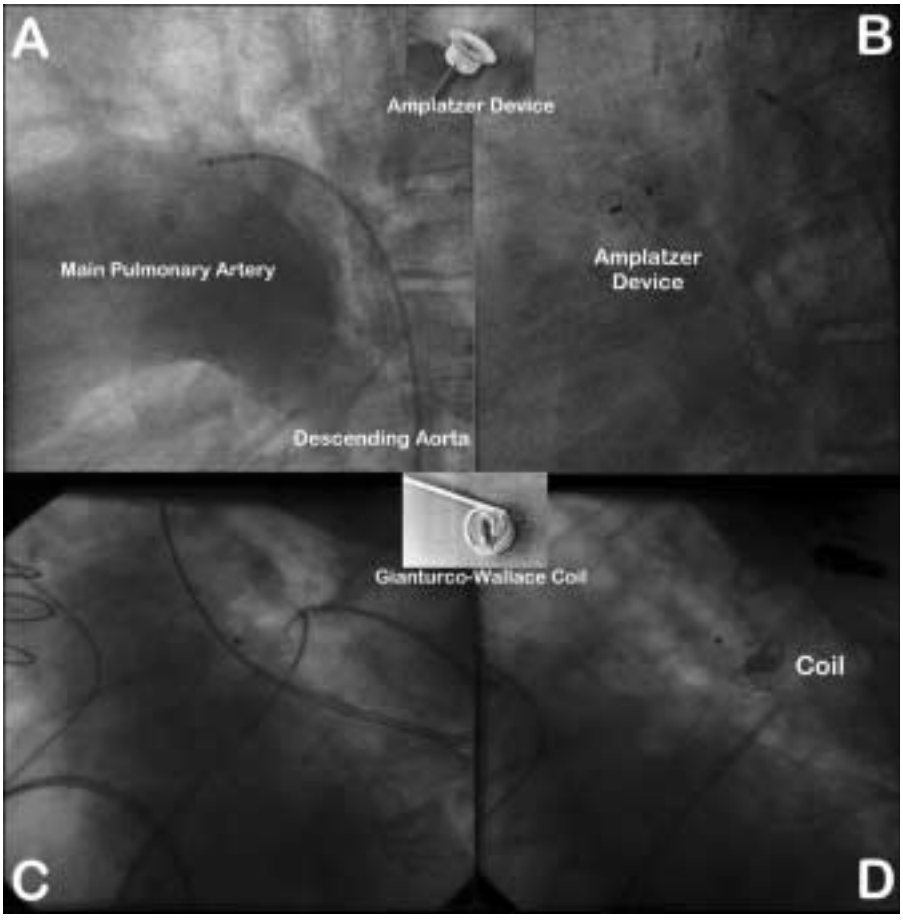
Fenestrated Fontan

The Fontan procedure is a congenital heart surgery that connects the right atrium to the pulmonary arteries in patients born with inadequate pulmonary flow often related to tricuspid or pulmonary atresia or single ventricle physiology.[34] Recently, total cavopulmonary conduits (connecting the IVC to the right pulmonary artery) have been popularized. This is almost always done either following SVC to pulmonary artery anastomosis (Glenn procedure) or concurrent with the Glenn operation.[35] Often the pulmonary circuit is unprepared for this increase in pulmonary flow, and high conduit pressure results. To decompress this high pressure, the Fontan conduits are now often left with a fenestration into the native RA.[36] When desaturation becomes problematic, closure of this right-to-left shunt can be accomplished by either surgical closure or by use of an occluder device.[37] The Food and Drug Administration recently approved the Amplatzer® Occluder device for the percutaneous occlusion of these fenestrations.[31] Goff and colleagues[37] published the results of a registry of 154 patients following successful fenestration occlusion with either a Clamshell or a CardioSeal® device. Patients experienced improved oxygenation, a reduced need for diuretics and digoxin, and a low risk of decompensation (3.2%) or death (1.3%).

Patent Ductus Arteriosus

Patent ductus arteriosus (PDA) is the second most common congenital heart defect seen in adults (~10-15% of all CHD in adults). It is present in isolation in 75% of adults, unlike in children where it is frequently associated with more complex heart defects. As in patients with VSD,

Figure 4. *Two techniques of closing a patent ductus arteriosus (PDA) with devices pictured in insets. Left lateral angiogram of a PDA via retrograde passage of a catheter from the descending aorta through the defect and into the pulmonary artery* ***(A)****. Final position of the Amplatzer® PDA occluder* ***(B)****. The snare technique used to position a detachable, Gianturco-Wallace coil (Cook, Inc.; Bloomington, IN) in the defect* ***(C, D)****.*



patients with a large uncorrected PDA can develop pulmonary hypertension,[12] and we have witnessed rapid reversibility following percutaneous closure. Some feel that any residual PDA should be occluded to prevent endarteritis and to remove any excess flow to the pulmonary circuit and left heart that may result in volume overload over time.

There are currently 2 options available for closure: an occluder device (Amplatzer® PDA occluder) or spring occlusion coils (Figure 4). Either strategy appears favorable with relatively little morbidity, though coils are generally not recommended for defects in excess of 4 mm in diameter because of the higher risk for arterial embolization.

In the coil occlusion technique, the defect is first sized angiographically to optimize coil size selection. A catheter (often a right coronary catheter) is then used to cross the defect in retrograde fashion from the aorta. A snare is advanced from the pulmonary side. A detachable coil is pushed out the end of the aortic catheter and the tip of the coil snared in the pulmonary artery after at least 1 major coil loop is evident in the PA (Figure 4C-D). The aortic catheter is then withdrawn and the proximal coil loops are allowed to expand into the ductus itself. Several coils may be used. Nondetachable coils are now available that allow positioning and observation of the results before detaching the coil from the delivery wire. Most recently, PDA closure has been accomplished by delivery of a PDA occluder device from the PA side of the ductus.[38]

Incomplete occlusion can be associated with hemolysis due to the damage to red blood cells within the residual shunt jet lesion.[39] Rare cases of endocarditis have also been reported. Follow-up echocardiography is useful to detect residual shunt. Coil occlusion is successful in 75% to 90% of patients; a smaller ductus tends to improve results. Occasionally, recanalization of the defect can occur during follow-up, and may require further intervention. A strategy of coil occlusion has been compared head-to-head with surgical therapy and been found to be cost-beneficial.[40]

In the coil occlusion technique, the defect is first sized angiographically to optimize coil size selection.

Faella and Hijazi reported the results of an international registry of 316 patients who underwent occlusion of a PDA with an Amplatzer® occluder.[41] Complications occurred in 15 patients including a single device embolization. Echocardiography in this cohort demonstrated complete occlusion of the defect in 56% immediately after the procedure, 76% at 24 hours, 95% at 6 months, and 100% at a year. Given these promising results, it is likely that the occluder method will predominate over the coil method in the near future.

Bronchial Collaterals

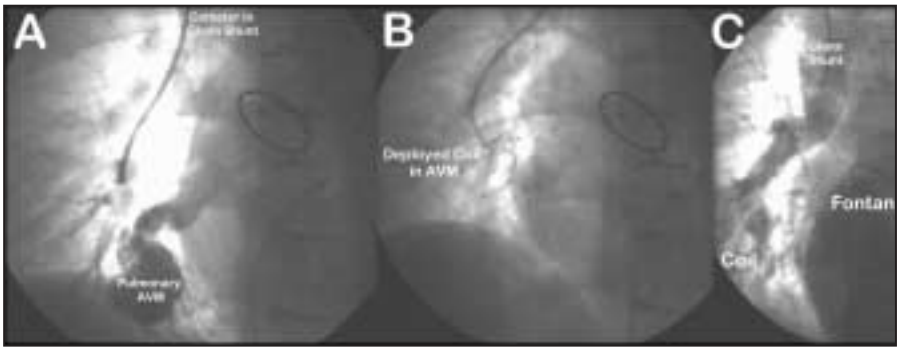
In patients with inadequate pulmonary perfusion, bronchial collaterals may enlarge and provide arterial blood flow to the lungs. These vessels may arise from the aorta, the mammary arteries, or even below the diaphragm. They can become quite large and lead to hemoptysis. These vessels can be coil occluded with high rates of success.[42] If they are recognized and prominent, some physicians have advocated coil occlusion prior to lung transplantation to reduce the risk of post-operative thoracic bleeding.

Gradients across the pulmonary outflow tract can involve the valvular level, but may also involve the infundibulum and/or the peripheral pulmonary arteries.

Atrioventricular malformations

Coil occlusion of atrioventricular malformations, especially those that result in a significant right-to-left shunt, can be achieved if they are

Figure 5. *Occlusion of a large pulmonary arteriovenous malformation (AVM) in the lung of a patient with a Glenn shunt and Fontan repair. An injection of the Glenn anastomosis (inferior vena cava to right pulmonary artery), which fills a large AVM* ***(A)****. After a detachable coil is used to occlude the AVM* ***(B)****, simultaneous angiography of the Glenn and Fontan demonstrate no further evidence of right-to-left shunting* ***(C)****.*



localized, and an appropriate coil or coils can be placed.[43,44] CHD patients in whom these are symptomatic include those who have undergone a classic Glenn procedure (SVC to right lung only via right pulmonary artery) and those with high SVC pressure leading to the opening of systemic venous channels, often to the pulmonary veins. Closure of 1 of these channels with detachable coils is demonstrated in Figure 5.

Balloon Valvuloplasty of Congenital Valve Disease

Aortic Valve Stenosis

Bicuspid aortic valve is the most common congenital cardiac disorder, occurring in approximately 2% of the population. A percutaneous approach to the stenotic bicuspid valve is often used in infants and children,[45] as there is frequently commissural fusion that can be relieved. In adults with calcific aortic stenosis, the leaflets are virtually weighted down with calcium deposition and commissural fusion is not the hallmark. In adults the clinical results following percutaneous aortic valvuloplasty have been poor,[46] with improvement seen only in those with a preserved left ventricle. Usually this type of patient does well with surgery regardless of age. Aortic valvuloplasty is typically accomplished with a single balloon delivered across the aortic valve from the femoral artery. The inflated balloon diameter never exceeds the aortic root size at the valve plane. Aortic insufficiency is uncommon following the procedure. The use of aortic valvuloplasty is now mostly restricted to infants and children as a palliative procedure before eventual surgical aortic valve replacement, and in the very elderly who have preserved left ventricular function, yet are not operative candidates for reasons of comorbidity.[47]

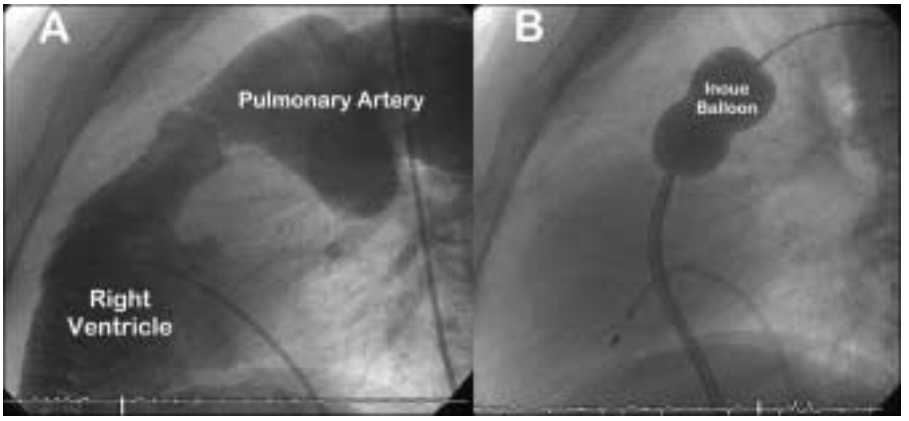


Figure 6. *Technique of pulmonic valvuloplasty using the Inoue balloon® (Toray; Houston, TX). The right ventricular angiogram **(A)** demonstrates a doming, stenotic pulmonic valve which is ideally approached using the hourglass-shaped, single balloon **(B)**.*

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Pulmonary Valve Stenosis

The most common congenital valve lesion that can necessitate therapy in adults is pulmonic valve stenosis (PS). Gradients across the pulmonary outflow tract can involve the valvular level, but may also involve the infundibulum and/or the peripheral pulmonary arteries. Careful tracking of any gradient is critical for decision making. Generally, an intervention is warranted when the transvalvular gradient exceeds 50 mm Hg (moderate or greater PS), though patients with lesser gradients may benefit if it can be clearly shown that exertional symptoms (typically exertional dyspnea) accompany elevated gradients during provocation.

Use of either single- or double-balloon techniques is acceptable[48] and successful procedures are the norm, with immediate improvement in valvular gradient and regression of right ventricular hypertrophy over time.[49] For the best results, an effective balloon diameter of 1.2 to 1.4 times the measured pulmonic orifice should be chosen, as the pulmonary artery is quite elastic. Recently the use of the Inoue® mitral valvuloplasty balloon (Toray, Houston, TX) has been shown to be quite effective in performing percutaneous balloon pulmonic valvuloplasty (Figure 6). In 53 adolescents and adults, Chen and colleagues[49] reported a 13% incidence of pulmonic insufficiency after the procedure, which was no longer present during later follow-up. Results appeared to be durable with little in the way of restenosis, even at 10 years.

Physicians performing pulmonic valvuloplasty should be aware of the possibility of "suicide right ventricle." This can occur as a result of dynamic subpulmonic obstruction following the procedure[50] and can be prevented by pretreatment with hydration and a negative inotropic agent such as a ß-blocker or calcium blocker. An excellent and permanent long-term result can be achieved when the final valvular gradient is reduced to less than 30 mm Hg.

Angioplasty and Stenting of Congenital Heart Lesions

Coarctation of Aorta

Coarctation of aorta (CoA) is a common congenital heart defect, accounting for approximately 8% of all congenital defects. Anatomically it can occur before the ductus

arteriosus, at the duct or after the duct. It likely results from extraneous ductal tissue that contracts following birth. Adults with previously undiagnosed CoA will almost always have post-ductal lesions. The most common presentation in adults is the fortuitous discovery during secondary work-up for systemic hypertension. Lower extremity and renal hypoperfusion leads to a hyperrenin state that may not abate even after repair of the coarctation. In most patients there is upper extremity hypertension and the development of collateral vessels around the coarctation to the lower extremity. These collateral channels often create a continuous murmur heard in the back, and involvement of the intercostal arteries leads to the familiar rib notching noted on chest x-rays. An associated bicuspid aortic valve is present in up to 85% of coarctation, and a significant aortic gradient is particularly important to exclude when deciding on definitive therapy. Also of note is an association with berry aneurysms in the circle of Willis, which can be seen in up to 10% of patients and can lead to CNS bleeding.[51]

Echocardiography with a focus on the descending aorta is an excellent noninvasive manner in which to make a clinical diagnosis in patients with suspicious clinical findings, with a resting peak systolic velocity of 3.2 m/s or greater or a diastolic velocity of 1.0 m/s or greater suggestive of significant coarctation. Echo also allows interrogation of the aortic valve and assessment of the ascending aortic root. MRI has become the preferred imaging modality pre- and post-operatively to size the aorta and the coarctation region and to follow patients. In the event of a contraindication (pacemaker or severe claustrophobia) or lack of availability, CT is a reasonable alternative.

Symptomatic patients with a peak gradient over 30 mm Hg on invasive measurement, as well as those with a similar gradient who are asymptomatic but have upper extremity hypertension that becomes severe with exercise or is associated with left ventricular hypertrophy, should be considered for therapy. Other evolving indications for treatment include the presence of aortic aneurysms and symptomatic aneurysms of the circle of Willis. Young women who wish to bear children are also at risk, as there may be inadequate placental flow should they become pregnant.

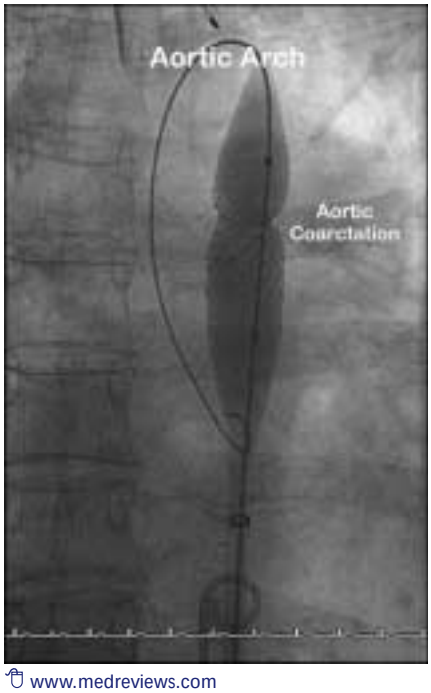
Post-coarctation aneurysm formation appeared commonly when angioplasty alone was used to treat native coarctation, but appears to be less of a concern with the wider use of stents.

Surgery has previously been the mainstay in the approach to a native CoA, with available options including resection and end-to-end anastomosis, prosthetic patch aortoplasty and interposition (tube bypass) grafting. Recently some success has been achieved with the use of percutaneous angioplasty in young patients with native coarctation.[52] Older patients, however, should still be considered for surgery if they are viable candidates. Percutaneous angioplasty has been performed since 1982, and the availability of stents has recently led to improved outcomes, to the extent that percutaneous intervention is now considered the procedure of choice in patients with recoarctation following surgery (Figure 7). Stents are particularly effective in preventing complications from recoil of the aorta following angioplasty. The size of the stent should never be larger than the native aorta. Intravascular ultrasound has been very useful in ensuring that there is adequate apposition of the stent against the aortic wall.

Several large series of angioplasty/stent patient studies suggest that success rates of 65%-100% with a complication rate of approximately 13% are the norm. Problems to watch for include recoarctation and aneurysm formation at the site of intervention, blood pressure problems including hypo- and hypertension, and paraplegia. Endocarditis prophylaxis remains important.

Post-coarctation aneurysm formation appeared commonly when angioplasty alone was used to treat native coarctation, but appears to be

Figure 7. *Primary stenting of a recurrent aortic coarctation at the site of previous surgical anastomosis.*



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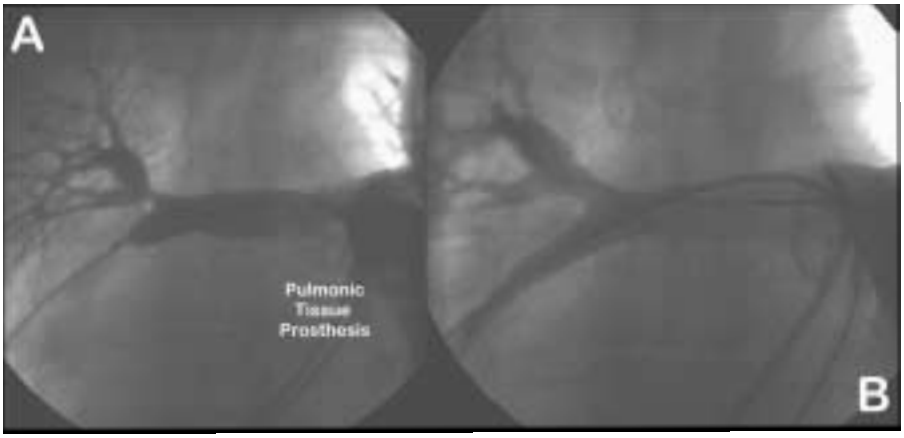

Figure 8. *Angioplasty and stenting of peripheral and branch pulmonary artery stenoses in a patient with previous pulmonic valve replacement.* www.medreviews.com

less of a concern with the wider use of stents.[53] It is important to re-emphasize that the resetting of the renin-angiotensin system that occurs in patients with coarctation may not resolve after repair. Most patients with residual systemic hypertension respond to β-blockers, angiotensin-converting-enzyme inhibitors, and/or diuretics.

Branch and Peripheral Pulmonic Stenoses

Branch and peripheral pulmonic stenoses (PS) may be discovered as isolated lesions or seen in association with valvular pulmonic stenosis. The presence of pulmonary hypertension (mean PA > 35 mm Hg) is usually an indication to attempt to stent a branch or peripheral PS. Because of the high elasticity of the pulmonary vessels, rebound is frequent. Often in peripheral PS, there are many lesions and only the largest non-occluded vessels can be approached. Stenting has clearly proven better than angioplasty alone.[54] A representative patient is shown in Figure 8. Interventions on branch lesions are generally more successful than on peripheral lesions, though neither is as successful as treatment of valvular PS. Long-term reductions in pulmonary pressures following branch or peripheral interventions are not common.

Systemic Venous Stenosis

As mentioned previously, balloon angioplasty alone has generally not proven very effective as a treatment for systemic venous stenosis. Representatives of this include SVC stenosis (related to the long-term use of intravenous catheters or pacemakers) or either tumor or sclerosis (ie, due to histoplasmosis). These can all be approached, with the use of stents, with reasonable success.[55] Stenotic venous baffles in patients who have received the Mustard (atrial switch) procedure for transposition of the great vessels can also be relieved with stenting.[56]

Conclusion

In conclusion, far too many adults with CHD now exist to permit for all of them to receive their care from a trained specialist. It therefore behooves the general practitioner and the practicing cardiologist to have a general understanding of the most common problems they may encounter in daily practice. After carefully reviewing the clinical record and imaging studies, referral for diagnostic or therapeutic cardiac catheterization may be indicated in adults with CHD. Percutaneous approaches to occlusion of shunt lesions, significant pulmonic stenosis, and aortic coarctation are now mainstays in the treatment of these patients; and in most cases these procedures are associated with significantly lower morbidity and mortality than the surgical alternatives. As clinical experience with newer treatment modalities, including percutaneous heart valve delivery and repair of regurgitant valves, continues to grow, the role of prompt diagnosis and referral will become ever more essential. ■

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Main Points

- The population of adults with congenital heart disease (CHD) continues to rapidly expand and provides important clinical challenges to the general cardiologist.
- Arrhythmias, syncope, and progressive dyspnea should prompt a thorough evaluation of the adult with CHD.
- Diagnostic cardiac catheterization, though generally performed later in the diagnostic workup than in the past, continues to serve an important role in the evaluation of patients with CHD.
- Defects of the atrial septum are the most commonly encountered in adults, and percutaneous closure of the classic secundum defect with suitable anatomy and clinical indications for repair is now standard practice.
- Other shunt lesions, including ventricular septal defect, patent ductus arteriosus, Fontan fenestrations, and pulmonary arteriovenous malformations, can also be approached percutaneously.
- Balloon valvuloplasty has demonstrated success in adults with pulmonic stenosis and in children and adolescents with bicuspid aortic stenosis.
- Balloon angioplasty and stenting play a growing role for the management of recurrent aortic coarctation, as well as peripheral and branch pulmonary artery stenosis.

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