Indications for Cardiac Catheterization and Intervention in Pediatric Cardiac Disease

A Scientific Statement From the American Heart Association

Endorsed by the American Academy of Pediatrics and Society for Cardiovascular Angiography and Intervention

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Preamble
Since publication of the last American Heart Association (AHA) scientific statement on this topic in 1998, advances in interventional techniques, and an innovative spirit have opened the field of congenital heart therapeutic catheterization. Unfortunately, studies testing the safety and efficacy of catheterization and transcatheter therapy are rare in the field because of the difficulty in identifying a control population, the relatively small number of pediatric patients with congenital heart disease (CHD), and the broad spectrum of clinical expression. This has resulted in the almost exclusive “off-label” use of transcatheter devices, initially developed for management of adult diseases, for the treatment of CHD.

The objective of the present writing group, which included representatives of the AHA and endorsemes from the Society for Cardiovascular Angiography and Interventions and the American Academy of Pediatrics, was not only to provide the reader with an inventory of diagnostic catheterization and interventional treatment options but also to critically review the literature and formulate relative recommendations that are based on key opinion leader expertise and level of evidence. The writing group was charged with the task of performing an assessment of the evidence and giving a classification of recommendations and a level of evidence to each recommendation. The American College of Cardiology/AHA classification system was used, as follows:

Classification of Recommendations
- Class I: Conditions for which there is evidence and/or general agreement that a given procedure or treatment is beneficial, useful, and effective.
- Class II: Conditions for which there is conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of a procedure or treatment.
  - Class IIa: Weight of evidence/opinion is in favor of usefulness/efficacy.
  - Class IIb: Usefulness/efficacy is less well established by evidence/opinion.
- Class III: Conditions for which there is evidence and/or general agreement that a procedure/treatment is not useful/effective and in some cases may be harmful.

Level of Evidence:
- Level of Evidence A: Data derived from multiple randomized clinical trials or meta-analyses.
- Level of Evidence B: Data derived from a single randomized trial or nonrandomized studies.
- Level of Evidence C: Only consensus opinion of experts, cases studies, or standard of care.

For the practice recommendations provided in this statement, the classification of recommendations and the level of evidence determinations were taken from data available from clinical trials or registries about the usefulness/efficacy in different subpopulations. A recommendation with level of evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the indications do not lend themselves to clinical trials. Although randomized trials may not be available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective. In 2003, the American College of Cardiology/AHA Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers’ comprehension of the indications and allow queries at the individual recommendation level.

In pediatrics, there are not likely to be any randomized controlled trials for devices, which posed a challenge for the writing group in compiling recommendations. Therefore, many of the indications are based on consensus opinion. In cases of strong consensus that an intervention be considered as standard-of-care practice with scientific evidence, interventions were designated as Class I indications. In these cases, the wording often used was “is indicated.” The writing group acknowledges that these recommendations may not exclude surgical management of the cardiac condition. Therefore, we opted to use the term “is indicated” versus “should be performed” in recognition of the fact that individual centers must assess their own capabilities and limitations. A device use may have a Class I indication but not necessarily be preferred at an individual center. Cardiac lesions, for instance, do not always exist in isolation, and patients with solitary or multiple lesions may be deemed better served by surgery. Cardiologists and surgeons must work together to formulate an interventional approach to patients based on individuality of the program. The writing group adhered to conflict of interest rules set by the AHA, and members were obliged to reveal all potential conflicts of interest. The writing group was chaired by a member of the Congenital Cardiac Defects Committee of the AHA who had no conflicts related to the topic of this scientific statement.

1. Preparation for Cardiac Catheterization
Cardiac catheterization has long served as the “gold standard” for the anatomic and physiological assessment of patients with CHD. Real-time fluoroscopy with contrast injection coupled with rapid digital angiography has provided the high-resolution images of the heart necessary for successful surgical management of these patients. The direct measure-
ment of pressures within cardiac chambers and great vessels helps to stratify patients according to risk, assists in evaluation of medical therapy, and helps to indicate a need for intervention.

Although it still serves a valuable role in this respect, cardiac catheterization has its limitations. Angiographic imaging for the most part is limited to 2 simultaneous imaging planes, and 3-dimensional reconstruction from angiographic data is still in development. The invasive nature of cardiac catheterization and common need for anesthesia forces the clinician to at least consider noninvasive alternatives for data collection. Fortunately, advances in noninvasive imaging have allowed cardiac catheterization to become increasingly a catheter-based therapeutic option rather than a diagnostic tool. Two-dimensional echocardiography and 3-dimensional imaging by echocardiography, magnetic resonance imaging (MRI), and computed tomography (CT) in many cases has replaced the need for cardiac catheterization.2–7 These same modes of noninvasive imaging can improve the success of catheter-based therapies and help to minimize the risks of invasive procedures by helping to target the data collection or therapy intended by catheterization. Thus, dialogue between the interventionalist, surgeon, and managing cardiologist is paramount to the provision of quality care for the patient with CHD.

General Recommendations for Cardiac Catheterization Preparation

Class I
1. Complete cardiac echocardiographic imaging or alternative noninvasive imaging modes such as MRI or CT are indicated before invasive cardiac catheterization to facilitate planning of data collection and performance of an intervention (Level of Evidence: A).

2. Indications for Diagnostic Catheterization
Cardiac catheterization and angiography have transformed the care of children with CHD and have greatly increased the safety and efficacy of surgery for CHD. As a result of advances in noninvasive imaging as outlined in section 1, “Preparation for Cardiac Catheterization,” diagnostic catheterization is no longer indicated in the routine preoperative evaluation of most congenital defects, such as ventricular septal defects (VSDs), atrial septal defects (ASDs), atrioventricular canal, tetralogy of Fallot, double-outlet right ventricle (RV), coarctation of the aorta, hypoplastic left heart syndrome (HLHS), and other complex CHD. Details of anatomy, including situs, venous, and arterial connections; septal integrity; severity of valvular stenosis or insufficiency; size of pulmonary arteries; coronary artery origins; and aortic arch anatomy are easily established with echocardiography to the degree of certainty required for surgical intervention. Assessment of patient hemodynamics and, when necessary, assessment of anatomy by angiography should be accomplished before interventional catheterization to confirm congenital or acquired heart disease in infants and children.

The availability of adequate noninvasive imaging is not uniform at all centers. At centers where MRI and CT imaging for congenital diagnosis are not state-of-the-art and where echocardiography does not provide sufficient detail, diagnostic catheterization is indicated. However, diagnostic catheterization should not be considered routine for diagnosis of congenital defects, and the performance of complete right- and left-sided heart studies may subject patients to unnecessary risk and exposure to radiation. All diagnostic catheterizations may lead to the necessity for an interventional procedure. As a consequence, the capability to proceed with the interventional procedure should be a requirement of the individual and institution that will perform the diagnostic portion of the catheterization. Individual centers must assess their abilities as they relate to this ability when determining which catheterization cases they might perform.

It is not the purpose of the present statement to provide a comprehensive list of all indications for diagnostic catheterization; however, there are certain broad categories in which the need for diagnostic catheterization is well established in current practice. In most instances, these indications have been developed empirically, and there are no prospective studies, controlled or otherwise, that prove the superiority of catheterization over other diagnostic modalities. Therefore, most indications are supported by patient series, retrospective case reviews, or opinion of authorities. Diagnostic catheterization is or may be indicated in the circumstances outlined in the following section.

Cardiac catheterization should be used in any circumstance in which the anatomy of a child with CHD is inadequately defined by noninvasive means. Although many congenital cardiac lesions are diagnosed and referred directly to surgery on the basis of noninvasive studies, there are some occasions, particularly in very complex lesions, on which more specific details about the anatomy or hemodynamics are necessary. The diagnostic cardiac catheterization remains the “final authority” for this definitive anatomic and hemodynamic information for many of the very complex lesions. Some circumstances that suggest the need for diagnostic catheterization include the following:

1. High-flow or low-flow physiology associated with semilunar valve stenosis can lead to inaccurate gradients determined by noninvasive Doppler assessment. Combined aortic stenosis (AS) and insufficiency and pulmonary stenosis and insufficiency, especially with multiple levels of obstruction of both the systemic and pulmonary circulation, can be confusing when examined on the basis of echocardiographic studies alone. A diagnostic catheterization is necessary to determine true peak-to-peak gradient measurements, which still represent the standard by which the need for both surgical and catheter interventions is determined in some conditions. In many cases, noninvasive determination of pressure gradients is sufficient to proceed with surgical intervention; however, catheterization may be required to resolve conflicting noninvasive data.

2. Low-flow lesions for which calculation of pulmonary arteriolar resistance can be misleading may be best studied by combining angiographic assessment of pulmonary artery anatomy and transit time with pulmonary artery pressures. Details of anatomy alone without corresponding pressure data can be misleading. This is often the case in the assessment of patients after...
cavopulmonary anastomosis and after Fontan completion. Diagnostic catheterization is useful in these patients in the evaluation for proceeding with completion of Fontan, revision of Fontan, or transplantation. In addition, assessment of a fenestration Fontan with trial occlusion is useful in patients with excessive cyanosis and in making the decision to proceed with closing the fenestration. The ability to proceed with closure of fenestrations is required. Similar to the above category of patients, the postoperative patient who is not following an anticipated clinical course merits strong consideration for cardiac catheterization (further discussed in section 10, “Unique Catheter/Interventional Care Considerations”).

There are many potential problems that can occur after surgery for congenital defects for which diagnostic catheterization is indicated because of the inability of noninvasive tests to provide the diagnosis. As noted above, the diagnostic procedure may become an interventional procedure, and cardiologists performing these studies should be prepared to proceed with intervention. Examples of some but certainly not all of the more common indications in this setting are provided below:

1. In situations in which excessive desaturation occurs after a systemic-to-pulmonary artery shunt has been created, catheterization is useful to exclude branch pulmonary artery stenosis and shunt stenosis or occlusion. Such a situation may arise if the patient is too unstable to tolerate an MRI or MRI/magnetic resonance angiography (MRA), which can readily show pulmonary artery stenosis and shunt occlusion.
2. If excessive systemic desaturation is present after cavo-pulmonary anastomosis, catheterization is indicated to exclude venovenous, venoatrial, or pulmonary arteriovenous malformations. The ability to proceed with venous collateral occlusion is a requirement in this setting. State-of-the-art MRA and CT angiography may help to detect and define the presence of these lesions.
3. If an excessive residual left-to-right shunt is suspected after closure of a VSD that included atrioventricular canal repair, catheterization is indicated to calculate shunt size and severity.
4. If the presence of excessive aortopulmonary collateral flow is suspected, diagnostic catheterization should be performed. The ability to proceed to occlusion of the offending systemic-to-pulmonary collateral arteries is required.
5. When there is suspected RV outflow tract obstruction after surgery for tetralogy of Fallot or double-outlet RV and MRI is either not available or the patient is unable to tolerate MRI, diagnostic catheterization is indicated.
6. If there is an unexplained need for extracorporeal membrane oxygenator support or failure to wean without obvious reasons, catheterization should be performed to identify problems and direct care.

In the past, the utility of diagnostic catheterization for providing a definitive diagnosis in the setting of unexplained heart failure with diminished systolic function was limited. Endomyocardial biopsy with routine light microscopic examination is useful when positive findings of myocarditis are found, but in most cases, light microscopic findings of myocarditis or specific criteria by light and electron microscopy that suggest a specific diagnosis of cardiomyopathy are absent. The Dallas criteria for myocarditis are rarely met. MRI with assessment of myocardial edema and delayed enhancement is useful in this situation but does not provide a definitive diagnosis. Although there may be situations in which the clinical condition of the patient precludes catheterization, catheterization with biopsy should be considered whenever possible, and if performed, appropriate samples of myocardium should be obtained for histological diagnosis and viral polymerase chain reaction. In addition, in some instances, examination of biopsy material for abnormalities in contractile proteins and for other molecular abnormalities may enable a definitive diagnosis of cardiomyopathy. The latter should occur in the setting of a comprehensive evaluation of potential heritable causes of cardiomyopathy.

Cardiac catheterization continues to play an important role in the assessment of pulmonary hypertension and its responsiveness to medical therapy. This may be the case in patients with CHD in whom an accurate assessment of pulmonary resistance is needed to make surgical and medical decisions. In addition, catheterization to assess resistance is essential in the management of patients with pulmonary vascular disease to assess response to pharmacological agents. Laboratories that perform catheterization to assess reactivity of the pulmonary vascular bed must have the capability to deliver inhaled nitric oxide or other pulmonary vasodilators.

A preoperative evaluation of pulmonary resistance is commonly performed in patients undergoing a staged Fontan approach to single-ventricle palliation. Some authorities do not recommend catheterization in this setting, but for most, catheterization to assess patients with single-ventricle physiology before stage 2 and 3 palliation remains common practice. However, the presence of elevated pulmonary arteriolar resistance, unsuspected pulmonary venous obstruction, and elevation of ventricular filling pressures all impact the outcome of second- and third-stage Fontan palliation. Catheterization in this setting is justified on this basis until prospective studies demonstrate the safety of its omission from preoperative assessment of these patients.

In specific cases, such as patients with pulmonary valve atresia, the pulmonary arterial supply of the lungs may be complex. Catheterization in patients with pulmonary valve--VSD or pulmonary atresia and complex ventricular anatomy is directed toward determining details of the aortopulmonary collateral supply to the lungs and the size of the true pulmonary arteries. Accurate characterization of dual supply by collaterals and true pulmonary arteries versus single supply by collaterals is important in planning the surgical approach to be used. The ability to perform selective catheterization of collaterals, balloon occlusion angiography, and pulmonary venous wedge angiography is essential. Noninvasive imaging such as MRI/MRA may be helpful in delineating these vessels.

Likewise, the coronary artery anatomy in patients with pulmonary valve atresia and an intact ventricular septum can influence the success of the intervention (catheter-based or surgical) and patient survival. Although the presence of RV-to-coronary artery fistulae in patients with pulmonary
intravascular ultrasound can be performed safely in children. Published as the definitive procedure in adults, its use in children remains limited by inexperience and lack of training and, in part, because of the size of the catheters and sheaths that are required. Yet intravascular ultrasound should be used increasingly as part of the routine annual or semiannual evaluation of pediatric heart transplant recipients, especially given the progressive nature of coronary disease, the potential for sudden death or need for retransplantation, and increasing evidence that changing or augmenting immunotherapy may slow the process of intimal proliferation in the early stages of graft vasculopathy.

Despite promising new diagnostic methods, such as surveillance that includes biomarkers (brain natriuretic peptide), catheterization with endomyocardial biopsy is the mainstay of rejection identification. Endomyocardial biopsy does not diagnose rejection in all cases of unexplained deterioration in graft function but remains the “gold standard.” Samples obtained at catheterization should be studied for both cellular and antibody-mediated rejection. Criteria for the diagnosis of both are well developed at this time, including the International Society for Heart and Lung Transplantation revised criteria for cellular rejection and newly published criteria for diagnosis of humoral rejection. Diagnosis of antibody-mediated rejection by biopsy requires both light microscopic findings and fluorescent microscopy for recognition of deposition of complement fragments C4D and C3D.

Risks/Complications
Cardiac catheterizations are not without risk to the patient. The following is a listing of the more common complications. The reader is referred to one of the cited references for more information.

- Exposure to ionized radiation (decreasing with newer equipment)
- Risk of general anesthesia (when used)
- Hypothermia (especially in small infants)
- Aggravation of hypoxia
- Arrhythmias (temporary instability or even permanent, as in heart block)
- Vascular injury/perforations/tears
- Cardiac perforation
- Cardiac valve injury
- Blood loss that requires transfusion
- Allergic reactions to contrast, drugs, or anesthetics
- Renal insufficiency caused by contrast material
- Diffuse central nervous system injury
- Stroke
- Death

Recommendations for Diagnostic Catheterization

**Class I**

1. It is recommended that hemodynamic and anatomic data be obtained (via angiography when necessary) at the time of a planned interventional cardiac catheterization (Level of Evidence: A).

2. It is recommended that cardiac catheterization be used to assess pulmonary resistance and reversibility of pulmonary hypertension in patients with CHD or primary pulmonary hypertension when accurate assessment of pulmonary resistance is needed to make surgical and medical decisions (Level of Evidence: B).

3. Cardiac catheterization is indicated in patients with complex pulmonary atresia for the detailed characterization of lung segmental pulmonary vascular supply, especially when noninvasive imaging methods incompletely define pulmonary artery anatomy (Level of Evidence: B).

4. Cardiac catheterization is indicated in determination of coronary circulation in pulmonary atresia with intact septum (Level of Evidence: B).

5. Cardiac catheterization is indicated in patients being assessed for cardiac transplantation unless the patient’s risk for catheterization outweighs the potential benefit (Level of Evidence: C).

6. Cardiac catheterization is recommended for surveillance of graft vasculopathy after cardiac transplantation (Level of Evidence: B).
**Class IIa**

1. It is reasonable to perform a cardiac catheterization to determine pulmonary pressure/resistance and transpulmonary gradient in palliated single-ventricle patients before a staged Fontan procedure (Level of Evidence: B).

2. Cardiac catheterization is reasonable in any CHD patient in whom complete diagnosis cannot be obtained by noninvasive testing or in whom such testing yields incomplete information (Level of Evidence: C).

3. Cardiac catheterization is reasonable for the assessment of cardiomyopathy or myocarditis (Level of Evidence: B).

4. Cardiac catheterization is reasonable for the assessment of coronary circulation in some cases of Kawasaki disease in which coronary involvement is suspected or requires further delineation or in the assessment of suspected congenital coronary artery anomalies (Level of Evidence: B).

5. Cardiac catheterization is reasonable to perform for the assessment of anatomy and hemodynamics in postoperative cardiac patients when the early postoperative course is unexpectedly complicated and noninvasive imaging techniques (eg, MRA, CT angiography) fail to yield a clear explanation (Level of Evidence: C).

### 3. Opening of Atrial Communications

#### 3.1. Transseptal Techniques

An atrial transseptal approach is indicated whenever access to the left side of the heart, and in particular the left atrium, is desired and a preexisting communication between the right and left heart is not present or, when present, cannot be crossed readily from the right side of the heart. An atrial transseptal puncture/perforation from the right atrium into the left atrium provides dependable, direct access to the left atrium in the presence of a previously intact atrial septum. With the use of an atrial transseptal approach in the investigation of the entire left side of the heart, the potential for further compromise of an artery from a retrograde study is reduced or even eliminated, because most of the “left heart” information can be obtained by way of the transseptal procedure while arterial pressure is monitored simultaneously through a small-caliber, indwelling arterial line. In the event a retrograde study is necessary for specific aortic root or arterial information, collection of data from the left side of the heart by the transseptal approach may allow the retrograde study to be performed with a smaller-diameter catheter and requires a much shorter arterial cannulation time. For assessment of mitral valve disease, the transseptal approach may be used. Although the widespread use of pulmonary capillary wedge pressure with simultaneous left ventricular end-diastolic pressure monitoring usually proves sufficient for the assessment of mitral valve disease, atrial pressures obtained by the transseptal approach are generally of higher fidelity and therefore provide optimal waveforms. The final decision about this approach rests with the experienced interventionalist.

After the performance of complex congenital heart surgical repairs, such as a Mustard or Senning venous switch procedure for transposition of the great arteries or the lateral tunnel Fontan procedure for single ventricle, the most reasonable access to critical locations in the pulmonary venous circulation of these patients is by a transseptal puncture through the baffle. Biplane fluoroscopy is essential for the safe, dependable performance of a transseptal atrial puncture/perforation, particularly in small patients. Biplane fluoroscopy allows a constant 3-dimensional mental reconstruction of the intracardiac structures that provides depth, as well as a simultaneous side-to-side relationship within the heart during the puncture/perforation. The use of single-plane fluoroscopy, even with a rotating C-arm, is considered only under extenuating circumstances, only for operators who have a thorough knowledge of the atrial septum and its variations, and only for those who are experienced and skilled in the transseptal technique. A variety of adjunct procedures, including various types of simultaneous echocardiograms, special angled views in a single plane, and the use of marker catheters in the aorta, have been proposed as substitutes for the use of biplane fluoroscopy, but none of these provide an equal substitute. A single-plane system should not even be considered in a very small patient or, conversely, in larger patients in the presence of either a very large or a very small left atrium, a large dilated aortic root, no inferior vena cava access to the atrial septum, or in the presence of any abnormal cardiac chamber or great vessel positional abnormalities, all of which add to the hazards of a transseptal technique.

The most common puncture/perforation of the atrial septum is performed with the Brockenbrough transseptal needle. These are used in conjunction with the Mullins long transseptal sheath/dilator sets (Medtronic or Cook Medical). The tips of the dilators of the original system have a very tight acutely (as much as 90°) to conform to a circuitous approach to the perforating wire to be positioned against the septum by rotating C-arm, is considered only under extenuating circumstances, only for operators who have a thorough knowledge of the atrial septum and its variations, and only for those who are experienced and skilled in the transseptal technique. A variety of adjunct procedures, including various types of simultaneous echocardiograms, special angled views in a single plane, and the use of marker catheters in the aorta, have been proposed as substitutes for the use of biplane fluoroscopy, but none of these provide an equal substitute. A single-plane system should not even be considered in a very small patient or, conversely, in larger patients in the presence of either a very large or a very small left atrium, a large dilated aortic root, no inferior vena cava access to the atrial septum, or in the presence of any abnormal cardiac chamber or great vessel positional abnormalities, all of which add to the hazards of a transseptal technique.

Radiofrequency transseptal perforation is a technique that was developed relatively recently. Radiofrequency energy is used to perforate the atrial septum; this technique is particularly useful in very small patients with small left atria (such as the newborn with a hypoplastic left heart) or when there is no direct femoral approach or ability for a needle to impinge on or be pushed forcefully through the atrial septum. The use of radiofrequency energy for the perforation allows the perforating wire to be positioned against the septum by use of a preformed guiding catheter, which can be curved acutely (as much as 90°) to conform to a circuitous approach to the septum. This adjunct for perforation of the septum is especially useful when the approach to the atrial transseptal procedure is from the jugular vein and there is no enlargement.
of the left atrium or no bulge of the atrial septum into the right atrium. A special radiofrequency generator is used for septal perforation in contrast to the generators used for electrophysiological radiofrequency ablation. The energy necessary for perforation is a low-power (5 W), high-intensity (150 to 180 V) electric current, which is administered for a very short time (0.4 second) through a very tiny-diameter (1.3F) electrode. This energy causes breakdown and perforation of the tissues that are immediately in front of and in contact with the electrode. Essentially no “force” is required to accomplish the perforation with the radiofrequency wire, but at the same time, some side-to-side control over the tip is lost compared with a needle puncture. Once the radiofrequency wire has perforated through the atrial septum, a fine coaxial catheter is advanced over the radiofrequency wire, which enables the radiofrequency wire to be exchanged for stiffer and more supportive wire. A fine-tipped dilator or a Mullins Transseptal Introducer Sheath (Medtronic, Inc) can then be introduced into the left atrium over this wire. To overcome the side-to-side control issues with the standard radiofrequency wire perforation, several recent reports have appeared that report the use of radiofrequency or electrosurgical cautery energy in combination with a transseptal needle. This energy causes breakdown and perforation of the tissues that are immediately in front of and in contact with the electrode. Essentially no “force” is required to accomplish the perforation with the radiofrequency wire, but at the same time, some side-to-side control over the tip is lost compared with a needle puncture. Once the radiofrequency wire has perforated through the atrial septum, a fine coaxial catheter is advanced over the radiofrequency wire, which enables the radiofrequency wire to be exchanged for stiffer and more supportive wire. A fine-tipped dilator or a Mullins Transseptal Introducer Sheath (Medtronic, Inc) can then be introduced into the left atrium over this wire. To overcome the side-to-side control issues with the standard radiofrequency wire perforation, several recent reports have appeared that report the use of radiofrequency or electrosurgical cautery energy in combination with a transseptal needle. These are still isolated reports, and the technique requires further investigation before it can be considered as a recommended approach.

Risks/Complications
It should be understood that transseptal puncture procedures are not without risk. Cardiac perforation (especially if the left atrium is of small volume), entry into the aorta, and air embolization associated with catheter and needle exchanges are all potential complications of this procedure.

Recommendations for Atrial Transseptal Puncture or Radiofrequency Perforation

**Class I**
1. Transseptal puncture is indicated for any patient for whom a transcatheter intervention is optimally performed from a left atrial approach (eg, pulmonary vein dilation or stenting, closure of a paramitral valve leak) and in whom no interatrial communication exists (Level of Evidence: B).
2. Transseptal puncture is indicated for the hemodynamic assessment of patients with suspected left ventricular outflow tract obstruction when retrograde crossing of the aortic valve is difficult or cannot be performed and no interatrial communication exists (Level of Evidence: B).
3. Transseptal puncture is indicated for the hemodynamic assessment of patients with clinically significant mitral valve stenosis when simultaneous pulmonary artery wedge pressure and left ventricular end-diastolic pressure are considered inadequate (Level of Evidence: C).
4. Transseptal perforation is recommended when the atrial septum is intact and entry to the left atrium is necessary for electrophysiological study or therapy (Level of Evidence: B).

**Class IIa**
1. It is reasonable to consider transseptal puncture for hemodynamic assessment of patients with hypertrophic cardiomyopathy and in whom no interatrial communication exists (Level of Evidence: C).
2. It is reasonable to consider transseptal puncture for aortic valvuloplasty and in cases in which no interatrial communication exists (Level of Evidence: C).
3. It is reasonable to perform transseptal puncture in the hemodynamic assessment of patients with or suspected of having mitral stenosis, pulmonary vein stenosis, or pulmonary hypertension if no interatrial communication exists (Level of Evidence: C).
4. It is reasonable to perform a radiofrequency perforation of the atrial septum when either the hemodynamics of the left atrium or access to the left side of the heart for diagnostic or therapeutic purposes cannot otherwise be obtained by routine transseptal puncture (Level of Evidence: B).
5. It is reasonable to perform a transseptal puncture to obtain a pulmonary venous saturation in the patient being considered for lung, heart, or heart-lung transplantation (Level of Evidence: C).

**Class IIb**
1. It may be reasonable to consider transseptal puncture when performing balloon angioplasty/stent implantation for coarctation of the aorta if no interatrial communication exists (Level of Evidence: B).

### 3.2. Atrial Septostomy
The survival of patients with certain forms of CHD (eg, tricuspid valve atresia, hypoplastic left heart) depends on unrestricted communication between the left and right atria. The presence of an interatrial shunt may be important to augment cardiac output in obstructive lesions of the right side of the heart (eg, tricuspid atresia, severe pulmonary valve stenosis or atresia); to enhance mixing in patients with transposition of the great vessels; to off-load the right side of the heart in pulmonary vascular obstructive physiology; to relieve left atrial hypertension in left-sided obstructive lesions (eg, HLHS); and to decompress the right atrium in postoperative RV failure.

Few techniques have been developed over the years to create or enlarge an interatrial communication. These include balloon atrial septostomy, blade atrial septostomy, static balloon dilation of the septum, and radiofrequency perforation or transseptal puncture of the septum that then leads to 1 of the above procedures. These techniques provide a temporary solution; for longer-lasting mixing/relief of obstruction, stent implantation in the septum may provide a more durable solution.

The creation of an atrial communication is typically performed in the cardiac catheterization laboratory by trained interventional pediatric cardiologists. The exception is routine balloon atrial septostomy in newborns performed at the bedside with echocardiographic guidance. The creation of an atrial communication should include surgical and circulatory support backup. The surgical backup does not necessarily mean a standby operating room but rather the availability of surgeons and anesthesiologists who can manage neonates and infants in case of complications from the procedure.
Balloon atrial septostomy is perhaps the oldest interventional cardiac procedure, first performed by Rashkind and Miller in 1966 on a neonate with transposition of the great vessels and severe desaturation. Access is obtained from either the umbilical vein or the femoral vein by use of an appropriate-sized sheath. If the baby is stable, routine hemodynamic assessment may be performed, followed by the septostomy. Currently, there are 4 different catheters that can be used for this purpose: Miller catheter (Edwards Lifesciences), Rashkind balloon catheter (USCI-CR Bard), Fogarty (Paul) balloon catheter (Edwards Lifesciences), and the NuMED septostomy catheter (NuMED). The latter catheter is the only one with an end hole that enables the operator to advance it over a guidewire and to confirm position by injecting contrast in the left atrium. Once the balloon is positioned in the left atrium and the position is confirmed (by fluoroscopy and/or echocardiography), the balloon is inflated with the appropriate volume of saline/contrast mixture (80%/20%), and a forceful jerk/pull is applied to the catheter to bring the balloon to the junction of the right atrium and inferior vena cava. Care must be exercised as to how vigorously the balloon is pulled into the inferior vena cava. This process is repeated at least once until there is no resistance to passage of the full balloon across the defect. A gradient across the septum may be measured, and if still significant, a balloon atrial septostomy may be repeated as above. Alternatively, 2-dimensional echocardiography along with Doppler assessment of the residual gradient may be used to determine the adequacy of the septostomy.

The indications for a blade atrial septostomy are the same as for the balloon atrial septostomy except that blade septostomy is used when the patient’s atrial septum is either intact or thick and resistant to a pull-through type of procedure. This includes newborn patients in whom there is an anticipated in infants >4 to 6 weeks of age or any older infants/children who require the creation of an atrial communication. When the atrial septum is totally intact, a long-sheath transseptal technique is indicated to introduce the blade catheter into the left atrium. In patients with these indications, the blade atrial septostomy should be performed before any attempt at balloon septostomy is made. In the presence of a thickened septum in all of the aforementioned circumstances, the septum often can undergo a balloon septostomy with only a ballooning (pull-through or dilation), but in that circumstance, the septum is stretched rather than torn, and in turn, only a temporary defect is created. Once an initial stretched opening is created in the septum with a balloon pull-through or dilation, a subsequent blade septostomy only elongates the defect to accommodate the width of the opened blade, so that the opened blade apparatus slides through the stretched opening and does not incise the septal tissues at all. In addition, by first making 1 or more small incisions in the septum with the blade, less wall tension on the balloon is needed to create a “tear,” and there is more control over the opening created with a dilation balloon.

The small “incisions” in the septum created by the blade withdrawal through a small, tight septal opening become the starting point for more controlled linear tears in the specific directions of the cuts as they are split or “torn” further with the balloon septostomy/dilation. As a consequence, a controlled tear using less force is accomplished instead of the whole circumference of the opening in the septum being stretched until it forcefully “explodes” in any or multiple directions. A dilation septostomy after an initial blade septostomy is very effective when a very controlled diameter of the opening is desired. Controlled static balloon dilation of the atrial septum or stenting of the atrial septum may be necessary in clinical situations in which the atrial septum is thickened and subject to recoil restriction with conventional balloon septostomy or when blade septostomy is difficult.

Atrial septostomy as a palliative measure for the treatment of pulmonary hypertension has been advocated for patients who are nonresponsive to medical therapy. Septostomy is regarded as a “last resort” procedure, akin to lung transplantation. Atrial septostomy is not without risk in these patients but may afford temporary improvement in functional class.

**Risks/Complications**

The potential complications that can be encountered during atrial septostomy include the following:

1. Balloon rupture and embolization of balloon fragments: Retrieval can be attempted, and if unsuccessful, surgical retrieval may be necessary.
2. Failure of balloon deflation: This is a rare complication. One can attempt passage of a stylet wire in the balloon lumen to clear it from any material that may be obstructing the lumen.
3. Misjudgment of the position of the balloon: This can be avoided by use of echocardiographic or biplane fluoroscopic guidance to confirm position. Contrast can be injected into the left atrium if an end-hole catheter is used.
4. Cardiac perforation or damage that might include rupture of the atrial appendage, which may occur in patients with juxtaposition of the atrial appendage: This is a serious complication that can be avoided by use of biplane fluoroscopy or echocardiography to confirm position before the septostomy. Another example of this type of complication would be mitral valve injury.
5. Vascular injury: This complication is rare, especially with use of the umbilical vein or small catheters. Nonetheless, vascular injuries have been reported with balloon atrial septostomy and include pulmonary vein avulsion and tear of the inferior vena cava.
6. Embolic complications such as stroke may be encountered.

**Recommendations for Atrial Septostomy**

**(Including Balloon Atrial Septostomy, Blade Atrial Septostomy and Static Balloon Atrial Septal Dilation, and Atrial Septal Stenting)**

**Class I**

1. Atrial septostomy is indicated to enhance atrial mixing (eg, transposition of the great vessels with restrictive/intact atrial communication) or to decompress the left atrium (*Level of Evidence: B*).

2. Atrial septostomy is indicated for the decompression of left atrial hypertension, for example, in patients on extracorporeal membrane oxygenation support.
and with evidence of severe pulmonary edema or if there is poor cardiac return from the extracorporeal membrane oxygenation circuit and low venous saturations (Level of Evidence: C).

Class IIa

1. Atrial septostomy is reasonable to perform in an effort to decompress a hypertensive pulmonary venous chamber in HLHS with an intact or restrictive atrial communication (Level of Evidence: B).

2. It is reasonable to attempt transseptal puncture and static balloon dilation of interatrial synthetic or bioprosthetic material (eg, Gore-Tex) if there is a need to decompress the left atrium (Level of Evidence: C).

Class IIb

1. Atrial septostomy may be considered for decompression of a hypertensive pulmonary or systemic venous chamber (eg, tricuspid atresia with restrictive atrial communication; pulmonary atresia with intact ventricular septum and restrictive atrial communication; total anomalous pulmonary venous connection with restrictive atrial communication) if needed before surgery (Level of Evidence: B).

2. In select patients with pulmonary arterial hypertension who have been unresponsive to medical management, atrial septostomy may be considered to allow preservation of cardiac output in severe pulmonary vascular disease at the expense of increasing systemic desaturation (Level of Evidence: C).

4. Transcatheter Device Closure of Septal Defects

4.1. Secundum ASD

ASDs account for 7% of all congenital heart defects. The most common ASD is a secundum defect versus defects located in the septum primum, sinus venosus defects, or unroofed coronary sinus. If left untreated, these defects may result in right-sided heart failure, arrhythmia, and pulmonary hypertension. Although surgical closure of ASDs is safe, effective, and time-tested, it still requires open heart surgery and hospitalization.46,47

Transcatheter closure of secundum ASD was first described in 1976 by Mills and King, who successfully treated 5 patients with defects as large as 26 mm using a “double umbrella device.”48 A 27-year follow-up report showed 4 of the 5 patients to be alive and well with no residual shunt or long-term sequelae.49 The large delivery system and somewhat cumbersome nature of this device precluded its widespread application; however, the ensuing 30 years saw a plethora of innovative developments in devices designed to treat secundum ASD. All currently existing transcatheter ASD closure devices are designed for use only for secundum ASDs, and all other anatomic subtypes of ASD continue to require surgical repair.

Several studies have shown outcomes from transcatheter device closure of secundum ASD to be comparable to surgical outcome in carefully selected adult and pediatric patients.46,47,50 Device closure of secundum ASD is associated with low complication rates, short anesthetic times, and short hospitalizations. When conditions are favorable, transcatheter secundum ASD closure has become the treatment of choice rather than surgery in many institutions.

Echocardiography, either intracardiac or transesophageal (TEE), plays a significant role in the guidance of these procedures and in the assessment of the final result. Research efforts are ongoing to examine other imaging modalities, such as MRI, as a means of guiding transcatheter ASD closure.

In the United States, currently only the AMPLATZER septal occluder (AGA Medical) and HELEX septal occluder (WL Gore and Associates) are approved for secundum ASD closure. Both devices were approved after prospective clinical trials comparing outcomes with traditional surgical closure revealed favorable results.33,51 The AMPLATZER septal occluder is composed of a nitinol wire mesh encasing Dacron fabric. The device is available in a wide range of sizes and features a left and right atrial disk connected by a self-centering connecting waist. This device is suitable for all subtypes of secundum ASD and has successfully closed defects as large as 38 mm in diameter. Before release from the delivery cable, the device can be repositioned or removed easily. The Gore HELEX occluder comprises a single strand of nitinol bonded to a piece of microporous expanded polytetrafluoroethylene. When deployed, the device forms 2 opposing disks that are locked into place across the atrial septum. The HELEX occluder is suitable for closure of small-to moderate-sized defects (≤18 mm in diameter) and is easily repositionable or removable even after release from the catheter delivery system. Both devices can be implanted successfully in children <2 years of age, although common practice suggests that a weight >15 kg may offer some technical advantages and simplify the procedure. With these newer devices, it is no longer necessary to have a septal rim present along the entire margin of the defect. Several reports describe successful closure of secundum ASD with deficiencies of the inferior, posterior, and superior rims.

Risks/Complications

The following risks are associated with this technique: Device migration; device malposition; cardiac erosion/perforation leading to tamponade and death; atrioventricular block; and the complications encountered from a cardiac catheterization, including air embolism, infection, and hematomas. The overall risk of complications with transcatheter device closure of secundum ASDs with the AMPLATZER device was 7.2% (32 of 442 cases).44,52 The major complication rate was 1.6% (7/442), including device embolism with surgical removal in 3 cases, cardiac arrhythmia requiring major treatment in 2 patients, marker band (located at the tip of the delivery sheath) embolism with surgical removal in 1 patient, and cerebral embolism with extremity numbness in 1 patient. There were no patient deaths. The minor complication rate was 6.1% (27/442), including cardiac arrhythmia that required minor treatment in 15 patients, thrombus formation in 3, allergic reaction to drug in 2, device embolism with percutaneous removal in 2, headaches with possible transient ischemic attack in 2, marker band embolism in 2, and urinary tract disturbance in 1 patient. The HELEX device has not been associated with device erosion; however, there have been numerous cases of wire-frame fracture, with a quoted
incidence between 5% and 7%. Although these fractures typically cause no clinical sequelae, rare cases of damage to the mitral valve have been reported.

Currently, there are no transcatheter devices designed for closure of sinus venosus, primum, or coronary sinus ASDs.

**Recommendations for Transcatheter Device Closure of Secundum ASD**

**Class I**

1. Transcatheter secundum ASD closure is indicated in patients with hemodynamically significant ASD with suitable anatomic features (Level of Evidence: B).*

**Class IIa**

1. It is reasonable to perform transcatheter secundum ASD closure in patients with transient right-to-left shunting at the atrial level who have experienced sequelae of paradoxical emboli such as stroke or recurrent transient ischemic attack (Level of Evidence: B).
2. It is reasonable to perform transcatheter secundum ASD closure in patients with transient right-to-left shunting at the atrial level who are symptomatic because of cyanosis and who do not require such a communication to maintain adequate cardiac output (Level of Evidence: B).

**Class IIb**

1. Transcatheter closure may be considered in patients with a small secundum ASD who are believed to be at risk of thromboembolic events (eg, patients with a transvenous pacing system or chronically indwelling intravenous catheters, patients with hypercoagulable states) (Level of Evidence: C).

**Class III**

1. Transcatheter secundum ASD closure is not indicated in patients with a small secundum ASD of no hemodynamic significance and with no other risk factors (Level of Evidence: B).
2. Transcatheter ASD closure should not be performed with currently available devices in patients with ASDs other than those of the secundum variety. This would include defects of septum primum, sinus venosus defects, and unroofed coronary sinus defects (Level of Evidence: C).
3. Transcatheter ASD closure is contraindicated in the management of patients with a secundum ASD and advanced pulmonary vascular obstructive disease (Level of Evidence: C).  

**4.2. Ventricular Septal Defects**

VSDs account for ≈20% of all forms of CHD. The ventricular septum can be divided into 4 regions: Membranous, inlet, trabecular, and outlet. VSDs can be single in any of the mentioned regions or multiple (“Swiss cheese”) in the muscular part of the septum.

Perimembranous VSD is the most common type, accounting for ≈80% of all VSDs, followed by the muscular type, which accounts for 5% to 20% of all defects. Supracristal (subarterial) VSD is a rare type, accounting for fewer than 5% of all defects, except in the Asian population, in whom the incidence is as high as 15% to 20%. Inlet VSDs account for ≈5% of all defects. Transthoracic echocardiography is the best diagnostic imaging tool with which to characterize the type, size, and number of defects and can estimate the pulmonary artery pressure. Surgical closure for patients with perimembranous VSD is safe and effective and is considered the therapeutic option of choice in neonates, infants, and children <3 years of age. Currently, there are no approved devices with which to close perimembranous VSD in the United States, but the utility of membranous VSD occluding devices is being tested internationally. In the absence of device availability, the writing committee has no recommendations on the use of transcatheter device closure for perimembranous VSDs at the present time.

Patients with hemodynamically significant muscular VSDs (MVSDs) may be offered percutaneous or hybrid-approach device closure. Children ≥5 kg in weight and with favorable anatomy are considered candidates for percutaneous closure. For infants who weigh <5 kg or for patients with abnormal septal wall planes, percutaneous MVSD device closure carries additional risk beyond the procedure- and device-related adverse events. Therefore, surgery or an alternative method of device delivery (ie, hybrid perventricular) is typically considered. This topic is discussed further in section 9, “Hybrid Procedures.”

We would like to emphasize that there are wide variations in practice between centers in the approach to MVSD intervention. Many surgeons prefer to repair midmuscular VSDs in the operating room using traditional techniques. However, for apical MVSDs, some surgeons prefer a hybrid approach. Patients are selected for MVSD occlusion device on the basis of transthoracic echocardiography. Exclusion criteria include weight <3.0 kg (unless the hybrid perventricular approach is used in this case); distance of <4 mm between the VSD and the aortic, pulmonic, mitral, or tricuspid valves; pulmonary vascular resistance >7 indexed Wood units; sepsis; and patients with conditions that would be expected to be exacerbated by the use of aspirin, unless other antplatelet agents could be used for 6 months. MVSDs most amenable to device closure are those located in the mid, apical, posterior, or anterior muscular septum.

**Risks/Complications**

Risks encountered during device closure of MVSD include device migration/embolization, tricuspid and mitral valve regurgitation, hemolysis, transient ischemic attack/stroke, ventricular tachycardia, and the rare chance of atrioventricular heart block.

**Recommendations for Device Closure of MVSDs**

**Class IIa**

1. It is reasonable for infants who weigh ≥5 kg, children, and adolescents with hemodynamically significant (left ventricular or left atrial volume over-
load or pulmonary-to-systemic blood flow ratio \(\geq 2:1\) MVSD to undergo percutaneous VSD device closure (Level of Evidence: B).

**Class IIb**
1. Neonates, infants who weigh \(< 5\) kg, and children with hemodynamically significant (left ventricular or left atrial volume overload or pulmonary-to-systemic blood flow ratio \(> 2:1\)) MVSD and associated cardiac defects requiring cardiopulmonary bypass may be considered for performance of hybrid percutaneous closure of the VSD off bypass, followed by surgical repair of the remaining defects or device placement during cardiopulmonary bypass (Level of Evidence: B).

**Class III**
1. Neonates, infants, and children with hemodynamically significant (left ventricular or left atrial volume overload or pulmonary-to-systemic blood flow ratio \(> 2:1\)) inlet MVSDs with inadequate space between the defect and the atrioventricular or semilunar valves should not undergo device closure (hybrid or percutaneous) (Level of Evidence: B).
2. Neonates, infants, and children with a small to moderate-sized MVSD (without symptoms or evidence of pulmonary hypertension) in whom there is a reasonable expectation that the defect will become smaller over time should be followed up expectantly and do not need closure of the VSD (Level of Evidence: B).

4.3. Fontan Fenestration and Baffle Leak Closure

**Fontan Fenestration**
Lateral tunnel Fontan baffle fenestration and subsequent catheter closure was first described in 1990 as a method of increasing operative survival and decreasing postoperative morbidity in patients identified as being at high risk for conversion to this type of circulation. The concept behind this strategy is that the creation of a right-to-left shunt or “pop-off” in patients with borderline Fontan physiology or anatomy (eg, diminished ventricular function, pulmonary artery distortion, elevated pulmonary vascular resistance) allows cardiac output to be maintained, albeit at the cost of systemic oxygen saturation.

Although debate continues about which patients might benefit from fenestration creation, possible benefits include improved operative survival for high-risk patients, decreased chest tube drainage in the postoperative period, and improved cardiac output and a lower incidence of arrhythmia in late follow-up. The ultimate goal in patients with a single ventricle, however, remains the partitioning of the systemic and pulmonary circulations. Additionally, there is some concern that patients left with patent fenestrations long-term may be at increased risk for paradoxical thromboembolic phenomenon secondary to an obligatory atrium-level shunt in the setting of cyanosis and a low-flow venous situation. To this end, numerous techniques have been developed to close these surgically created communications using devices primarily created for other indications, including ASD occluders, patent foramen ovale occluders, VSD occluders, embolization coils, vascular plugs, and covered stents. Typically, after a complete right and left hemodynamic catheterization and angiography are performed, the fenestration is crossed from a transvenous approach and temporarily occluded for a period of time (typically between 10 and 20 minutes). If hemodynamics are deemed acceptable with the fenestration temporarily occluded (improved systemic oxygen saturation associated with only a modest rise in Fontan pressure or fall in cardiac output), a device is selected to use for permanent fenestration closure. The type of device chosen may be influenced by the type of Fontan operation performed (intracardiac versus extracardiac), patient size, specifics of the anatomy, and operator preference. Reported results have been encouraging, with a high rate of procedural success and a low rate of complications. In follow-up, increased systemic oxygen levels and improved clinical symptomatology tend to be sustained; however, data are mixed on whether objective exercise testing improves after fenestration closure.

**Recommendations for Transcatheter Closure of Fontan Fenestrations**

**Class IIa**
1. It is reasonable to consider transcatheter closure of a chronic (outside of the immediate postoperative period) Fontan fenestration if the patient has favorable hemodynamics and tolerates test occlusion (Level of Evidence: C).

**Fontan Baffle Leaks**
Fontan baffle leaks typically occur at the suture margins that anchor an intracardiac baffle to the atrial wall and result in an obligatory right-to-left shunt, which in turn results in cyanosis. Numerous reports have described successful catheter therapy for this problem with use of a variety of occluders, coils, and covered stents; the reader is referred to these articles for technical details.

**Risks/Complications**
Because of the location of these baffle leaks, there is a higher likelihood that there will be incomplete closure of the baffle leak, and there is a small potential risk for vascular embolization or valvular compromise. Such risk needs to be assessed at the time of device placement.

**Recommendation for Transcatheter Closure of Fontan Baffle Leaks**

**Class IIa**
1. It is reasonable to consider transcatheter closure of a chronic (outside of the immediate postoperative period) Fontan baffle leak for the purpose of relieving cyanosis if the patient has favorable hemodynamics and tolerates test occlusion (Level of Evidence: C).

5. Transcatheter Balloon Dilation of Cardiac Valves

5.1. Pulmonary Valvuloplasty
First reported in 1982, balloon valvuloplasty remains the treatment of choice for valvar pulmonary stenosis in patients
of all ages. The subgroup of patients with thickened, dysplastic pulmonary valves, as commonly seen in Noonan syndrome, has shown a lower success rate with balloon valvuloplasty, but given the low incidence of complications, the procedure is usually attempted. Indications for balloon pulmonary valvuloplasty are similar to surgical indications, namely, symptoms or a resting gradient ≥40 mm Hg with the patient sedated in the catheterization laboratory. Many will choose to wait for an echocardiographic peak instantaneous gradient of 40 mm Hg or higher in asymptomatic patients before referring them for valvuloplasty. Echocardiographic peak instantaneous gradients have reliably correlated well with catheterization peak-to-peak gradients. Patients with mild pulmonic stenosis rarely progress or require intervention.

Since the initial report, multiple studies have proven the effectiveness and safety of valvuloplasty. Restenosis after balloon dilation is rare, with few children ever requiring repeat dilation. Pulmonary regurgitation after dilation is common, occurring in 10% to 40% of patients. Previously, it was believed the insufficiency was not an issue, but increasingly, children are being referred for pulmonary valve implantation as the RV shows signs of dilation and dysfunction, as seen with transannular patch repair of tetralogy of Fallot. Compared with surgical valvotomy, however, the incidence and severity of regurgitation were lower in the group treated with valvuloplasty. Some have now chosen to use balloons smaller than the previously recommended 120% to 140% of the annulus to try to reduce this incidence of pulmonary insufficiency.

Balloon pulmonary valve dilation has been used successfully in patients with tetralogy of Fallot and other forms of cyanotic heart disease with clinically significant valvular pulmonic stenosis. These procedures may cause hypercyanotic episodes, but in some rare situations, they may be a reasonable option for palliation. Balloon dilation is not useful for treatment of infundibular pulmonary stenosis unassociated with pulmonary valve stenosis.

Taking the process one step further, multiple techniques have been used to perforate the valve in selected patients with pulmonary atresia. Using the stiff end of a guidewire, transseptal needles, lasers, and, most recently, radiofrequency perforation catheters or wires, the atretic pulmonary valve can be crossed. This procedure requires great caution, and some institutions have incorporated a hybrid strategy to the procedure.

Risks/Complications
Complications, specifically perforation of the RV outflow tract, were fairly common early in the pulmonary valvuloplasty experience, but recent reports are encouraging, with higher success rates and lower numbers of complications.

Once a communication is made between the RV and the pulmonary arteries, a wire can be placed across the valve, and the valve can be dilated in the usual fashion. It is not yet clear which patients with pulmonary atresia and an intact ventricular septum will potentially benefit from this procedure. One group that clearly needs to be excluded are patients with RV-dependent coronary circulation, because they have shown higher mortality rates after surgical decompression.

Recommendations for Pulmonary Valvuloplasty

Class I
1. Pulmonary valvuloplasty is indicated for a patient with critical valvar pulmonary stenosis (defined as pulmonary stenosis present at birth with cyanosis and evidence of patent ductus arteriosus dependency), valvar pulmonic stenosis, and a peak-to-peak catheter gradient or echocardiographic peak instantaneous gradient of ≥40 mm Hg or clinically significant pulmonic valvar obstruction in the presence of RV dysfunction (Level of Evidence: A).

Class IIa
1. It is reasonable to perform pulmonary valvuloplasty on a patient with valvar pulmonic stenosis who meets the above criteria in the setting of a dysplastic pulmonary valve (Level of Evidence: C).

Class IIb
1. Pulmonary valvuloplasty may be considered as a palliative procedure in a patient with complex cyanotic CHD, including some rare cases of tetralogy of Fallot (Level of Evidence: C).

Class III
1. Pulmonary valvuloplasty should not be performed in patients with pulmonary atresia and RV-dependent coronary circulation (Level of Evidence: B).

5.2. Aortic Valvuloplasty
First described in the early 1980s, balloon dilation has replaced open surgical valvotomy as the treatment of choice for children with moderate to severe congenital valvar AS in the majority of centers. A relatively large literature now exists documenting the safety and effectiveness of balloon dilation for congenital AS. This literature consists primarily of single-institution case series, with few multicenter reports and no randomized clinical trials comparing balloon dilation with alternative therapies. Nevertheless, the technique is widely regarded as the therapy of first choice for children with valvar AS of sufficient severity to warrant intervention.

In infants, children, and adolescents with congenital aortic valve stenosis, a technically adequate balloon dilation will typically reduce the catheter peak-to-peak systolic valve gradient to 20 to 35 mm Hg. Severe aortic regurgitation occurs uncommonly. The VACA (Valvuloplasty and Angioplasty of Congenital Anomalies) Registry reported the results of aortic balloon dilation in 606 children who underwent the procedure between 1984 and 1992 at 23 institutions. For the group as a whole, the procedure achieved a 60% reduction in the peak systolic aortic valve gradient. Identified risk factors for a suboptimal outcome (defined as a residual systolic gradient ≥60 mm Hg, major morbidity, or mortality) included patient age <3 months, a higher predilation valve gradient, a ratio of balloon-annulus diameters <0.9, the presence of an un repaired coarctation, and an earlier procedural date. In this data set, it appeared that the optimal ratio...
of balloon-annulus diameters was 0.9 to 1.0. Larger-diameter ratios were associated with a significantly greater risk of aortic regurgitation after the procedure.

The longer-term outcome after a successful balloon dilation of congenital aortic valve stenosis in childhood is good, but late restenosis and valve regurgitation eventually necessitate reintervention in the majority of children.\textsuperscript{113,117,122,123,125} For example, Pedra and colleagues\textsuperscript{122} reported the late outcomes after aortic valve dilation in 87 children \(>6\) months of age at the time of the procedure (median age 6.9 years) and who were followed up for an average of 6.3 years. The freedom from reintervention was 86% after 1 year, 67% after 5 years, and 46% at 12 years. For newborns who undergo valve dilation for critical AS, the reintervention rate is higher during follow-up, as pointed out by McCrindle et al\textsuperscript{120} and McElhinney et al\textsuperscript{124} in 2 reports that both described a reintervention-free survival rate of 48% at 5 years in this population. These studies underscore the palliative nature of balloon dilation for congenital aortic valve stenosis.

Risks/Complications
Complications of the procedure include suboptimal relief of valve obstruction (more common with a balloon-annulus diameter ratio \(<0.9\), smaller annulus Z values, or valve leaflet calcification) and creation of significant aortic valve regurgitation (which occurs more often with a balloon-annulus ratio \(>1.0\)). Other possible adverse events include femoral vascular injury, thromboembolic stroke, injury to the mitral valve, and in newborns, myocardial perforation. Mortality is uncommon in children and adolescents. Infants represent a higher-risk group, and individual risk-benefit analysis of surgery versus catheterization must be weighed, especially in the infant who is ductal dependent and has significant left ventricular dysfunction.\textsuperscript{120,124,125,127}

A diagnostic cardiac catheterization typically precedes the valve dilation procedure and is intended to document the valve gradient, the degree of valve regurgitation, the aortic valve annulus dimension, and left ventricular function. Patients are heparinized unless a medical contraindication exists. Aortic valve dilation can be performed with an antegrade venous approach or a retrograde approach from the femoral, umbilical, or carotid artery. In newborns with critical AS, care must be taken when crossing the valve in a retrograde direction to avoid inadvertent valve leaflet perforation. If an antegrade technique is used, care must be taken to avoid wire injury to the anterior mitral valve leaflet during balloon inflation. A balloon is chosen with an inflated diameter 80% to 100% of the aortic valve annulus diameter; the use of a balloon larger than the annulus diameter risks causing important valve regurgitation. A single balloon inflation will generally suffice if the balloon is well positioned across the aortic valve. In children with good ventricular function, rapid RV pacing can be helpful to stabilize the balloon position by minimizing balloon ejection from the ventricle during inflation. After dilation, repeat hemodynamics are measured, and an aortic root contrast injection is performed to document the degree of aortic regurgitation that may have been induced.

Balloon dilation provides excellent palliation for most children with congenital valvar AS. It cannot be considered curative (as is the case with all current treatment options), because important valve regurgitation will occur in some patients, and valve restenosis will eventually occur in the majority of patients late after the dilation procedure.

Our recommendations reflect the understanding that balloon dilation is a palliative intervention and are consistent with the published natural history studies.\textsuperscript{127,128}

Recommendations for Aortic Valvuloplasty

Class I
1. Aortic valvuloplasty is indicated regardless of valve gradient in the newborn with isolated critical valvar AS who is ductal dependent or in children with isolated valvar AS who have depressed left ventricular systolic function (Level of Evidence: B).
2. Aortic valvuloplasty is indicated in children with isolated valvar AS who have a resting peak systolic valve gradient (by catheter) of \(\geq 50\) mm Hg\textsuperscript{†} (Level of Evidence: B).
3. Aortic valvuloplasty is indicated in children with isolated valvar AS who have a resting peak systolic valve gradient (by catheter) of \(\geq 40\) mm Hg\textsuperscript{†} if there are symptoms of angina or syncope or ischemic ST-T-wave changes on electrocardiography at rest or with exercise (Level of Evidence: C).

Class IIb
1. Aortic valvuloplasty may be considered in a child or adolescent with a resting peak systolic valve gradient (by catheter) of \(\geq 40\) mm Hg\textsuperscript{†} but without symptoms or ST–T-wave changes if the patient desires to become pregnant or to participate in strenuous competitive sports (Level of Evidence: C).
2. Aortic valvuloplasty may be considered for asymptomatic patients with a catheter-obtained peak systolic gradient of \(<50\) mm Hg when the patient is heavily sedated or anesthetized if a nonsedated Doppler study finds the mean valve gradient to be \(\geq 50\) mm Hg (Level of Evidence: C).

Class III
1. Aortic valvuloplasty is not indicated in children with isolated valvar AS who have a resting peak systolic valve gradient (by catheter) of \(<40\) mm Hg\textsuperscript{†} and who have no symptoms or ST–T-wave changes on electrocardiography (Level of Evidence: C).
2. Aortic valve balloon dilation is not indicated in children with isolated valvar AS who also have a degree of aortic regurgitation that warrants surgical aortic valve replacement or repair (Level of Evidence: C).

5.3. Mitral Valvuloplasty
Mitral valve stenosis in children can be separated into 2 broad categories: rheumatic and congenital. Although rheumatic mitral valve stenosis is commonly due to the development of thickened mitral leaflets and fused commissures, congenital mitral stenosis encompasses a broad spectrum of anatomic variants, including the “typical” variant with thickened leaflets, shortened chordae, and decreased interchordal spaces;\textsuperscript{†}

\textsuperscript{†}These refer to pressure gradients measured with a patient sedated during cardiac catheterization. Catheter gradients obtained with a patient under general anesthesia are likely to be somewhat lower.
valve stenosis. The double-transseptal technique was developed to minimize vascular trauma in infants and smaller children. The Inoue balloon (Toray International America) was first developed for mitral valve dilations in adults and later used in older patients with congenital mitral stenosis and double-orifice mitral stenosis. The Inoue balloon occluded the mitral valve as an alternative to surgery. One large multicenter adult study suggested that balloon mitral valvuloplasty is more favorable in patients with moderate to severe mitral stenosis who have a calcified mitral valve, as an alternative to surgery. Class IIb indications are for asymptomatic adult patients with moderate to severe mitral valve stenosis who have associated cardiac defects. Balloon dilation to treat rheumatic mitral stenosis, the majority of reports for congenital mitral valve stenosis are limited to small series. The largest report is from Children’s Hospital–Boston (Boston, MA), comprising 64 patients. Because surgical repair of mitral valve stenosis in infants and young children has high mortality and morbidity, the less invasive transcatheter technique remains a reasonable alternative, although the results remain variable, in part because of the varied morphology of the mitral valve stenosis. Balloon dilation procedures can have good immediate and even midterm relief of gradients, but progressive mitral regurgitation and restenosis remain problematic. Torn mitral leaflet or chordal attachments and papillary muscle rupture are the most common causes of regurgitation after dilation. In selected cases, transcatheter dilation may delay the need for surgical mitral valve replacement. Successful transcatheter dilation to treat mitral restenosis after surgical repair of rheumatic mitral stenosis has also been reported. Success is based on accurate preintervention diagnosis of the anatomic substrate of the mitral stenosis, and associated left-sided heart obstructive lesions are important factors to consider when deciding whether a patient should undergo balloon dilation or surgical intervention. In general, balloon dilation is more favorable in those variants of mitral stenosis with commissural fusion and more balanced chordal attachments, and worse outcome is seen with parachute mitral valves, supramitral rings, or small mitral annulus; in younger patients; and in those who develop significant mitral regurgitation.

In the adult literature, the severity of mitral stenosis is based on a constellation of data that include symptomatology (at rest and at exercise testing), mitral valve gradient, mitral valve area, and pulmonary artery systolic pressure. Class I indications are for symptomatic adult patients with moderate to severe mitral stenosis and asymptomatic patients with moderate to severe mitral stenosis and pulmonary hypertension (>50 mm Hg at rest or >60 mm Hg with exercise). Class IIa indications are for symptomatic adult patients with moderate to severe mitral stenosis who have a calcified valve and are high-risk candidates for surgery. Class IIb indications are for asymptomatic adult patients with moderate to severe mitral stenosis who have new onset of atrial fibrillation in the absence of left atrial thrombus or moderate to severe mitral regurgitation; symptomatic patients with mitral valve area >1.5 cm² but with significant mitral valve stenosis based on pulmonary artery systolic pressure >60 mm Hg, pulmonary artery wedge pressure of >25 mm Hg, or a mean mitral gradient of >15 mm Hg with exercise; or symptomatic patients with moderate to severe mitral valve stenosis who have a calcified mitral valve, as an alternative to surgery. One large multicenter adult study involved 738 patients (most of whom had rheumatic mitral stenosis) indicating the predominant symptom before the procedure was heart failure (94%), with 64% of patients in New York Heart Association class III or IV. The analysis was broken down into 2 groups based on single- versus double-balloon technique. For the single-balloon group, echocardiographic data showed the mitral valve area increased from 1.1 to 1.7 cm². Catheterization data showed the mitral valve area increased from 14 to 37 mm Hg. Mitral valve area increased from 0.9±0.4 to 1.7±0.7 cm², left atrial pressure decreased from 26±7 to 19±6 mm Hg, and pulmonary artery pressure decreased from 40±14 to 34±12 mm Hg. For the double-balloon group, the mitral valve area increased from 10±4 to 5±3 mm Hg and mitral valves area increased from 1.1±0.3 to 1.7±0.5 cm². Catheterization data showed the mitral valve area increased from 1.0±0.3 to 2.0±0.8 cm², left atrial pressure decreased from 25±7 to 16±7 mm Hg, and pulmonary artery pressure decreased from 35±13 to 29±11 mm Hg.

Unfortunately, there are few comparative data for infants and children with rheumatic or congenital mitral valve stenosis in either the surgical or interventional literature. Although the hemodynamic classifications of severity of mitral stenosis may be similar, symptoms in infants and young children may be limited to tachypnea, dyspnea, and/or failure to thrive. One surgical series for congenital mitral valve stenosis reported the average age at operation was 5.1±3.2 years, with 54% in functional class III and 24% in class IV. The average preoperative pulmonary arterial pressure was 13 to 34 mm Hg, with a mean capillary wedge pressure of 21 mm Hg. Reported a series of 30 children with rheumatic mitral stenosis who underwent balloon valvuloplasty. Although indications
were not discussed, the cohort that was included had mean left atrial pressure of 25±5 mm Hg, mean mitral gradient of 16±4 mm Hg, and mitral area of 0.7 cm² by catheterization and 0.8 cm² by echocardiography. Another small series of 8 patients with congenital mitral stenosis who underwent balloon valvuloplasty had a mean mitral gradient of 18 mm Hg and left atrial pressure of 25 mm Hg. McElhinney et al reported the combined outcomes of transcatheter and surgical therapy of 108 infants and children with congenital mitral stenosis. The hemodynamic profile of the 64 patients who underwent balloon dilation were as follows: Echocardiography peak and mean gradients were 21.8±7.1 and 15.1±4.5 mm Hg, respectively; mean left atrial pressure was 24.0±5.8 mm Hg; the calculated mitral valve area was 0.9±0.3 cm²; and mean pulmonary arterial pressures were in the mid-40s. Given the lack of data on indications, current publications would suggest that the hemodynamic profile of an infant or child with at least moderate mitral stenosis who met indications for a transcatheter intervention would include peak transmural gradients of 20 mm Hg or higher, mean transmural gradient of 15 mm Hg or higher, near-systemic pulmonary artery pressure, and calculated mitral valve area of <1 cm/m² with respiratory symptoms or failure to thrive.

Risks/Complications
The most important risk and complication for this procedure is iatrogenic mitral regurgitation. Although rheumatic mitral stenosis is often due to fusion of the commissures, which can be safely widened with a balloon, the anatomic causes of congenital mitral stenosis are often more complicated, and dilation may result in torn chordae or even a flailed leaflet with significant mitral regurgitation. Other major complications reported include cardiac arrest, atrial or ventricular perforation, transient rhythm abnormalities, stroke, and femoral artery or vein trauma. On the other hand, surgical repair for congenital mitral stenosis also carries significant mortality and morbidity, especially in infants and small children <5 years of age. When mitral valve repair is not possible, replacement becomes the only option, which also carries significant risks in the young patient. The decision to proceed with transcatheter balloon dilation must be weighed against the alternative risks and benefits of surgery.

Recommendations for Mitral Valvuloplasty

Class I
1. Transcatheter balloon valvuloplasty is indicated for symptomatic patients with moderate to severe isolated rheumatic mitral stenosis or asymptomatic patients with moderate to severe rheumatic mitral valve stenosis/restenosis associated with pulmonary hypertension (Level of Evidence: B).

Class IIa
1. Transcatheter balloon valvuloplasty is reasonable for the treatment of symptomatic children ≥5 years of age with congenital mitral stenosis or restenosis who have mitral valve morphology that is favorable for balloon valvuloplasty (ie, thickened leaflets and fused commissure) (Level of Evidence: B).

Class IIb
1. Transcatheter balloon valvuloplasty may rarely be considered at centers of expertise for patients who are <5 years of age with congenital mitral valve stenosis with moderate to severe residual stenosis or with valve restenosis who have already undergone surgical valvuloplasty at centers with expertise in mitral valve intervention in young children (Level of Evidence: C).
2. Transcatheter balloon valvuloplasty at centers with surgical and interventional expertise in mitral valve procedures may be considered in the management of infants and young children <5 years of age with moderate to severe congenital mitral valve stenosis who would otherwise undergo mitral valve replacement or when mitral valve replacement is thought to be problematic (Level of Evidence: C).
3. Transcatheter balloon valvuloplasty may be considered as a palliative measure in the management of patients with moderate to severe congenital mitral valve stenosis (ie, thickened leaflets, shortened chordae, and decreased interchordal spaces) (Level of Evidence: B).

Class III
1. Transcatheter balloon valvuloplasty is not indicated for patients with congenital mitral valve stenosis due to supramitral valve ring or associated with hypoplastic left ventricle (Level of Evidence: B).

5.4. Tricuspid Valvuloplasty
Given the rarity of tricuspid valve stenosis as an isolated congenital lesion and the lack of literature on tricuspid valve interventions in the pediatric age group, no specific recommendations are offered by the authors.

6. Transcatheter Balloon Angioplasty and/or Stent Placement for Obstructive Lesions
There is controversy and uncertainty with regard to transcatheter treatment in coarctation of the aorta. The reader is reminded, as stated in the Preamble, that transcatheter treatment does not necessarily replace surgical management. In several instances, both transcatheter and surgical intervention are viable treatment options.

6.1. Native Coarctation and Recoarctation
Coarctation of the aorta is a common form of CHD, accounting for ~6% to 8% of all cardiac defects. The prevalence of coarctation is increased in certain disorders, such as Turner syndrome. The most common associated cardiac anomaly is bicuspid aortic valve, which is present in >30% to 40% of all cases. The most important noncardiac associated anomaly is intracerebral aneurysm (berry aneurysm), present in ~10% of all cases. The usual location of coarctation is juxtaaudical, just distal to the left subclavian artery; less often, the coarctation is proximal to the origin of the left subclavian artery. Clinical manifestations of coarctation are age dependent. Neonates with coarctation of the aorta may present with signs and symptoms of low cardiac output and shock once the ductus arteriosus closes. Older infants and children may present with signs and symptoms of failure to thrive, and
older children and adults may present with hypertension, headaches, and claudication. The diagnosis of coarctation is suggested by the presence of a blood pressure gradient between an arm and leg and the presence of weak femoral pulses. Transthoracic echocardiography confirms the diagnosis. Other imaging modalities (CT/MRI angiograms, cardiac catheterization) may not be obtained routinely unless the diagnosis is in doubt or additional information is needed to plan a surgical or catheter intervention; however, the information obtained by these studies will likely impact our understanding of coarctation repair outcome. Noninvasive MRI/MRA and CT angiography provide a comprehensive view of the thoracic aorta, including the arch, coarctation site, and associated collateral vessels. MRI/MRA is particularly valuable in the posttreatment follow-up of patients and permits monitoring of coarctation outcome.

Treatment of coarctation of the aorta has evolved over the years. For native coarctation of the aorta, initially, surgical repair (extended resection with an end-to-end anastomosis) was the primary treatment at most centers and remains the “gold standard” therapeutic option. However, in recent years, balloon angioplasty with or without stent implantation has emerged as an alternative, less invasive option. For most patients with discrete recurrent/residual coarctation after surgical repair, balloon angioplasty has been shown to be the best therapeutic option. Balloon angioplasty for native coarctation can also be performed safely and successfully beyond the neonatal period. Young patients (>1 month but <6 months of age) with discrete narrowing and no evidence of arch hypoplasia may benefit from balloon angioplasty. This criterion applies to relatively few patients in this age group, because arch hypoplasia commonly accompanies the coarctation of the aorta. However, the recurrence rate is higher for younger patients (<6 months of age), and there is a small but important incidence of aneurysm formation after balloon dilation of native coarctation at any age. Stent implantation for native coarctation or recoarctation of the aorta has also emerged as a beneficial therapeutic option for patients who can receive a stent that can be expanded to an adult size (minimum of 2 cm in diameter). Many interventionalists perform primary stent implantation in patients with suitable anatomy in place of balloon angioplasty because of the perceived superior beneficial short-term results of stent implantation; however, the downside to this procedure is the need for future procedures to expand the stent as the patient grows in size. Newer stent technology may mitigate these stent size issues. In theory, stent implantation provides the potential for long-term repair with less chance of coarctation recurrence or aneurysm formation, but of course, the long-term benefit has yet to be proven. The reader is referred to excellent review articles for full technical details of balloon angioplasty and stent implantation.

Risks/Complications
Complications encountered during or after balloon angioplasty for both native and recurrent coarctation include femoral artery injury, dissection, and aneurysm formation at the site of angioplasty. The potential need for stent reexpansion in growing children limits the use of stent implantation to patients in whom stents can be implanted that can reach an eventual adult size. Complications encountered during or after stent implantation are similar to those encountered after balloon angioplasty, with the added potential risk of stent malposition. The incidence of aneurysm formation after stent implantation is less than for balloon angioplasty alone. Although short- and medium-term data are available in humans and animals, the long-term implications of stent placement in the aorta are unknown.

Recommendations for Transcatheter Balloon Angioplasty of Coarctation/Recoarctation of the Aorta

Class I
1. Balloon angioplasty of recoarctation is indicated when associated with a transcatheter systolic coarctation gradient of >20 mm Hg and suitable anatomy, irrespective of patient age (Level of Evidence: C).
2. Balloon angioplasty of recoarctation is indicated when associated with a transcatheter systolic coarctation gradient of <20 mm Hg and in the presence of significant collateral vessels and suitable angiographic anatomy, irrespective of patient age, as well as in patients with univentricular heart or with significant ventricular dysfunction (Level of Evidence: C).

Class IIa
1. It is reasonable to consider balloon angioplasty of native coarctation as a palliative measure to stabilize a patient irrespective of age when extenuating circumstances are present such as severely depressed ventricular function, severe mitral regurgitation, low cardiac output, or systemic disease affected by the cardiac condition (Level of Evidence: C).

Class IIb
1. Balloon angioplasty of native coarctation may be reasonable in patients beyond 4 to 6 months of age when associated with a transcatheter systolic coarctation gradient >20 mm Hg and suitable anatomy (Level of Evidence: C).
2. Balloon angioplasty of native or recurrent coarctation of the aorta might be considered in patients with complex coarctation anatomy or systemic conditions such as connective tissue disease or Turner syndrome but should be scrutinized on a case-by-case basis (Level of Evidence: C).

Recommendations for Stent Placement in Native Coarctation and Recoarctation of the Aorta

Class I
1. Stent placement is indicated in patients with recurrent coarctation who are of sufficient size for safe stent placement, in whom the stent can be expanded to an adult size, and who have a transcatheter systolic coarctation gradient >20 mm Hg (Level of Evidence: B).

Class IIa
1. It is reasonable to consider placement of a stent that can be expanded to an adult size for the initial
treatment of native or recurrent coarctation of the aorta in patients with:

- a transcatheter systolic coarctation gradient of >20 mm Hg (Level of Evidence: B).
- a transcatheter systolic coarctation gradient of <20 mm Hg but with systemic hypertension associated with an anatomic narrowing that explains the hypertension (Level of Evidence: C).
- a long-segment coarctation with a transcatheter systolic coarctation gradient >20 mm Hg (Level of Evidence: B).

2. Stent implantation for the treatment of coarctation (native or recurrent) is reasonable in patients in whom balloon angioplasty has failed, as long as a stent that can be expanded to an adult size can be implanted (Level of Evidence: B).

Class IIb

1. It may be reasonable to consider stent implantation for the treatment of coarctation in infants and neonates when complex aortic arch obstruction exists despite surgical or catheter-mediated attempts to relieve this obstruction and when further surgery is regarded as high risk. Implantation of a stent with less than adult-sized potential implies a commitment on the part of the surgical team to remove or enlarge this stent at a later date when the final diameter of this device is no longer adequate to maintain unobstructed aortic flow (Level of Evidence: C).

2. It may be reasonable to consider placement of a stent that can be expanded to an adult size for the initial treatment of native or recurrent coarctation of the aorta in patients with:

- a transcoarctation gradient of <20 mm Hg but with an elevated left ventricular end-diastolic pressure and an anatomic narrowing (Level of Evidence: C).
- a transcoarctation gradient of <20 mm Hg but in whom significant aortic collaterals exist, which results in an underestimation of the coarctation (Level of Evidence: C).

6.2. Pulmonary Artery Angioplasty and Stent Placement

Pulmonary Artery Angioplasty

Balloon angioplasty alone is indicated for both severe main pulmonary artery and severe branch pulmonary artery stenosis, particularly in very small patients or in those with pulmonary arteries with very complicated anatomy, in whom, in either case, primary stent implantation is not a viable option. Significant stenosis is obvious when there are measureable gradients of >20 to 30 mm Hg across the stenosis area, when there is elevation of the RV or proximal main pulmonary artery pressure to greater than one half to two thirds of systemic pressure secondary to the more distal obstruction, or when there is relative flow discrepancy between the 2 lungs of 35%/65% or worse. However, the significance of the stenosis is often more subtle and is determined by the subjective anatomic (angiographic) appearance of narrowing or by the discrepant blood flow away from specific areas rather than by a specific gradient across a segment. In low-pulmonary-flow situations such as with Glenn shunts and Fontan circulations, gradients in the pulmonary bed are notoriously unreliable determinants of the degree of stenosis. Likewise, underestimation of obstruction in the pulmonary circuit, even in the setting of normal pulmonary blood flow, can exist because of decompression runoff. In the case of isolated pulmonary arterial obstruction runoff to the opposite lung or adjacent segments of the lung, very significant (>90%) unilateral or isolated segments of branch stenosis may result in minimal or even no pressure drop across the obstruction.

Balloon angioplasty is applicable for both congenital and acquired pulmonary artery stenosis; however, balloon dilation alone has rarely been effective in the long term for such lesions. Balloon angioplasty of these lesions frequently does produce up to 50% improvement in the diameter of the involved vessels and a 50% decrease in the gradient across the lesion, as well as comparable increases in the blood flow in the involved vessel, but dilation alone seldom produces complete or permanent resolution of the obstruction or normalization of flow to the area, and the improvement achieved is often transient. There is little evidence that balloon dilation alone produces persistence of significant improvement over the long term, with no published long-term data. More recent studies on dilation of pulmonary artery stenosis have concentrated on either balloon angioplasty augmented by cutting balloons or primary stent implants, both of which are covered in the specific indications for cutting balloons and for pulmonary artery stent implants.

Pulmonary artery stents are indicated in main or branch pulmonary artery stenosis that is not expected to have, or has not had, an adequate or persistent response to primary pulmonary artery balloon dilation. Intravascular stents are effective for congenital stenosis, surgically created stenosis of vessels, or stenosis due to compression from an adjacent structure. Single stents, multiple tandem stents, and bifurcating stents all are effective and can be used when indicated by the underlying pulmonary artery anatomy.

Risks/Complications

The inherent risks associated with any complex catheter manipulations are present in those required to position wires and balloons for a balloon angioplasty. The risks for pulmonary angioplasty particularly include vessel perforation and arrhythmia. The major additional risks associated with angioplasty of the main or branch pulmonary arteries are injuries to the pulmonary vessels. To achieve a successful dilation, it is necessary to significantly overdilate both the area of stenosis and the adjacent vessel. To achieve a more effective dilation, at least a tear in the intima of the wall of the vessel is considered necessary. Generally, the pulmonary arteries are very compliant, which allows overdistension to even 2 to 3 times their normal diameter. However, the stenotic areas are not normal tissue and generally have lower compliance, which makes the vessels more susceptible to tears and can lead to vessel rupture, which can result in massive bleeding with hemothorax and even death. Although not considered a “complication,” most pulmonary arteries and branches are only dilated to ~50% of the vessel’s nominal diameter, and
many dilated pulmonary arteries and pulmonary artery branches experience restenosis within a relatively short time and often recontract to their original degree of stenosis.169

**Pulmonary Artery Stent Placement**

Stent implantation for central and proximal pulmonary arteries in growing patients should only be accomplished with balloon-dilatable stents. The stents must have adequate wall strength to support the vessel, have some flexibility, and be larger-diameter stents that are capable of eventual dilation to the largest potential adult diameters of the pulmonary arteries in that location of that particular patient.29,170 It is typically in the best interest of patients to have stents with adult-size potential implanted into the central branch pulmonary arteries, and this should always be considered as the first and ideal option. In certain instances, such as small patient size, small vessel size, or proximity in time to recent heart surgery, smaller stents without adult-size potential have been used successfully to treat branch pulmonary artery stenosis. Use of these smaller stents must be recognized as a palliative procedure, and a commitment must be made before implantation that surgical removal or enlargement of these stents will be needed at a future date. This can often be accomplished at the time of a planned future operation, such as RV–pulmonary artery conduit replacement or Fontan completion. It is recommended that use of such smaller stents be considered only with consultation with a pediatric cardiac surgeon.

Routine predilation of the stenosis in a pulmonary artery is not recommended because of the risk of vascular rupture; however, there are 2 circumstances in which predilation of a significant stenosis in a pulmonary artery is definitely recommended before stent implantation. The first circumstance is a stenosis of a pulmonary artery that could not be expanded to the desired diameter with the available balloons during a previous catheterization. The second indication for predilation is when the diameter of the area of stenosis is so tight that the delivery sheath/dilator set cannot be advanced through the stenosis without predilation of the area. This degree of stenosis would be <2 to 3 mm in diameter. There may be other circumstances in lesions <8 mm in size in which the operator believes that dilation alone or cutting balloon dilation may relieve significant stenosis without a stent, and the dilation may be considered before a stent is implanted.

**Risks/Complications**

Larger and stiffer wires and delivery systems are required for implantation of stents into the pulmonary arteries. This slightly increases the general risks of the wire/sheath manipulation; however, the risk of tearing a pulmonary artery during implantation of stents into pulmonary arteries is less than the risk of balloon dilation alone because overdilation of the individual vessels is not necessary.

When the balloon/stent is not sized properly or is not positioned exactly in the stenotic lesion during implantation, the stent can be misplaced or migrate after implantation. This can result in abnormal positioning in the vessel or in “jailing” of branch or side vessels. Abnormal positioning of the stent can also lead to late abnormal intimal proliferation and restenosis of the vessel. Gross undersizing or malpositioning of the stent can lead to retrograde embolization to the main pulmonary artery or even the RV, which may require surgical removal.

**Recommendations for Pulmonary Angioplasty**

**Class I**

1. Pulmonary angioplasty is indicated for the treatment of significant peripheral branch pulmonary artery stenosis (see text for definition of “significant” stenosis) or for pulmonary artery stenosis in very small patients in whom primary stent implantation is not an option (Level of Evidence: B).

**Class IIa**

1. Pulmonary angioplasty is reasonable to consider for treatment of significant distal arterial stenosis (as defined in the introduction to “Pulmonary Artery Angioplasty and Stent Placement”) or for stenosis in larger, more proximal branch pulmonary arteries that do not appear to be amenable to primary stent implantation (Level of Evidence: B).

**Class IIb**

1. Pulmonary angioplasty may be considered for treatment of significant main pulmonary artery stenosis that results in an elevation of pressure to more than two thirds of systemic pressure in the proximal pulmonary artery segment or in the RV (in the absence of pulmonary valve stenosis). This stenosis is usually a form of supravalvular pulmonary stenosis, which is not particularly responsive to balloon dilation alone (Level of Evidence: C).

**Recommendations for Pulmonary Artery Stent Placement**

**Class I**

1. Primary intravascular stent implantation is indicated for the treatment of significant proximal or distal branch pulmonary artery stenosis when the vessel/patient is large enough to accommodate a stent that is capable of being dilated to the adult diameter of that vessel (Level of Evidence: B).

**Class IIa**

1. It is reasonable to consider pulmonary artery stent implantation in critically ill postoperative cardiac patients when it has been determined that significant branch pulmonary artery stenosis is resulting in a definite hemodynamic compromise in a patient/vessel of any size, particularly if balloon dilation is unsuccessful (Level of Evidence: B).

2. Primary intravascular stent implantation is reasonable in the treatment of significant stenosis of the main pulmonary artery segment that results in elevation of the RV pressure, provided that the stent definitely will not compromise a functioning pulmonary valve and will not impinge on the pulmonary artery bifurcation (Level of Evidence: B).

**Class IIb**

1. It may be reasonable to implant small pulmonary artery stents that lack the potential to achieve adult size in small children as part of a cooperative
surgical strategy to palliate severe branch pulmonary artery stenosis. These stents may need to be enlarged surgically or removed during a future planned operation (eg, conduit replacement, Fontan completion) (Level of Evidence: C).

6.3. Systemic Venous Balloon Angioplasty and Stenting

Systemic Veins
Systemic venous stenosis occurs as a congenital defect, subsequent to surgery on or about the venous system, from indwelling lines in the veins or from external compression. Because of the low-pressure venous circulation, “significant” gradients usually are not present, and even in the presence of complete obstruction of a venous channel, only a 1- to 2-mm Hg gradient may be present. The areas of significant obstruction are easily visible. There often will be distension of the veins proximal to the obstruction, as well as multiple collateral venous channels around the obstruction. The venous obstruction may demonstrate congestion and swelling of tissues proximal to the area of significant obstruction.

Isolated balloon angioplasty is seldom indicated as the definitive treatment of systemic venous stenosis/occlusions, with most of these lesions currently being treated with intravascular stents. As is the case with balloon angioplasty of pulmonary artery stenosis, some short-term improvement usually is achieved, with both an increase in the diameter of the involved vessel and an increase in flow through the area; however, significant long-term improvement of either diameter or flow seldom is maintained.¹⁷¹,¹⁷² This applies to congenital venous stenosis and the much more common stenosis of peripheral veins that develops in particular with the presence of indwelling lines, as well as the stenosis of central venous channels after surgical venous reconstructions (Mustard, Senning, and Fontan-type procedures). Similar to balloon angioplasty alone for pulmonary artery stenosis, isolated balloon angioplasty may be appropriate in very small veins in small patients or in complex areas of stenosis such as the more peripheral systemic veins, where the obstructed veins are in areas of the body that are subject to bending or flexing (eg, peripheral veins in the neck, axilla, or groin) and, as a consequence, not applicable for implantation of the available intravascular stents. Initial balloon angioplasty of systemic veins also is often necessary to allow access through a very tight stenosis in the veins to deliver the larger sheaths needed for the implantation of intravascular stents into the area. In very small (peripheral) vein occlusions, previously dilated veins frequently reocclude totally, which makes access to the vessel even more difficult.

Risks/Complications
The dilation of systemic veins is associated with very few complications. Most stenotic veins have fairly straightforward access and do not require the more dangerous complex catheter manipulations to access. The risk of vascular perforation is possible when it is necessary to access a totally occluded vein by purposeful perforation.

The normal and perhaps the stenotic systemic vein may be very compliant, allowing dilation to 3 to 4 times its nominal diameter without obvious damage to the vein; however, restenosis of dilated veins is common, probably even more so than in the pulmonary arteries. The risk of venous rupture does exist, but in practice, this has been extremely rare. Thrombosis of dilated systemic veins is common if adequate flow is not established through the dilated vein.

Systemic Venous Stenting
Intravascular stents are indicated in the treatment of all types of systemic vein stenosis and total occlusions, including lesions in the iliofemoral vein, innominate veins, central subclavian vein, superior vena cava, and inferior vena cava, and in central venous baffle stenosis, such as that found in postoperative Mustard/Senning and Fontan types of procedures. Because of the historical lack of persistent success of balloon dilation alone and the frequent difficulty in accessing or traversing these systemic venous lesions, primary stent implantation is indicated as the treatment at the time these lesions are encountered.

The majority of the favorable experience in the use of stents in systemic vein stenosis/obstruction has come with the use of balloon-expandable stents. As with all stents in growing patients, the stents implanted should be capable of eventual dilation to the adult diameters of the vessels involved. The stents used currently in systemic veins should be positioned to avoid areas of repeated flexion of the vessel to prevent bending or breaking of the stent. Persistent patency of stents placed in systemic veins depends on the establishment of good flow through the stented vessel at the end of the procedure.¹⁷⁰

There have been no controlled or randomized trials in the use of stents in the systemic veins. Like stents in the pulmonary arteries, stents were adapted for use in the systemic veins after repeated failure of dilation alone to provide sustained relief of systemic vein stenosis, and they have shown favorable immediate and midterm results.¹⁷³

Risks/Complications
As with the dilation of systemic veins, access to stenotic systemic veins for the placement of stents is usually more direct, with less complex manipulations and less risk incurred during the implantation. Because of the compliance of the systemic veins, displacement of stents during implantation, including remote embolization of the stent, is more likely than in other areas. There is a very low risk of venous rupture with stent implantation. The embolized stent in a systemic vein has the likelihood of passing centrally into the right atrium or RV, which makes surgical removal potentially more difficult and dangerous. Systemic venous stents can collapse if exposed to any flexing or bending and then are very prone to reobstruction. If adequate flow is not established through systemic venous stents at the time of implantation, they will often become thrombosed. The risk of venous stent thrombosis appears to be greater during the perioperative period.

Recommendations for Systemic Venous Balloon Angioplasty

Class IIa
1. Venous balloon angioplasty is reasonable to consider for the management of peripheral venous obstruction
or “complex” venous stenosis in cases in which no viable alternative therapy exists (Level of Evidence: C).

**Class Ib**

1. Venous balloon angioplasty may be reasonable in the management of central venous obstruction, but its usefulness as a definitive therapy has not been established (Level of Evidence: C).

**Recommendations for Systemic Venous Stenting**

**Class I**

1. Stenting is indicated for the relief of significant systemic venous obstruction inferior to the clavicles and above the inguinal ligaments (Level of Evidence: B).

6.4. Pulmonary Veins

Pulmonary vein stenosis can present as an isolated congenital lesion or in association with other cardiac defects. It can also be acquired after corrective surgery for various anomalous pulmonary venous connections or after lung transplantation. Adult patients can acquire pulmonary vein stenosis after radiofrequency ablation for atrial fibrillation. More rare causes include external compression from tumors, sarcoidosis, and mediastinitis. This lesion is often progressive and fatal if it occurs bilaterally. The use of stents for treatment of pulmonary vein stenosis was first reported in 1991. Since then, the literature on pulmonary vein stenting has been limited to small series and tends to be divided into 2 major categories: Adults who acquire pulmonary vein stenosis after radiofrequency ablation therapy for atrial arrhythmias and children who either develop pulmonary vein stenosis after surgical repair of associated CHD or those with congenital pulmonary vein stenosis. Other treatment modalities include surgery, balloon angioplasty, and use of cutting balloons. But to date, there is not a single procedure that produces a high rate of patency consistently and does not require reintervention.

Comparison data of balloon angioplasty versus stent dilatation suggest that stents achieve better results and have longer patency rates. Furthermore, final stent diameter is a factor in delay of restenosis. This is particularly true when the stents are dilated to >8 to 10 mm in the case of adults with acquired pulmonary vein stenosis after pulmonary vein ablation for atrial fibrillation. Prieto et al. found a 42% success rate and 72% restenosis rate for angioplasty compared with a 95% success rate and 33% restenosis rate for stents. There was freedom from restenosis if the stent diameter was ≥10 mm. Similarly, Neumann et al. found a restenosis rate as low as 23% when the stents were dilated to ≥10 mm. Kluge et al. showed that 6 mm is the “critical diameter” for a low-flow state in the pulmonary veins in adults. Even so, other studies have shown a restenosis rate of up to 60% whether angioplasty or stents were used. In the pediatric literature, the surgical data indicate the “sutureless marsupialization” technique to be most effective, but freedom from reoperation or death still hovers around 50%. Restenosis rates after angioplasty and stent data are similarly high and particularly worse for congenital pulmonary vein stenosis. Because surgery after stent implantation is very difficult, it is important to implant a stent that can be serially dilated because of restenosis or to accommodate growth of the child to an adult. Often, reinterventions are performed as a bridge to lung transplantation as the disease progresses into the intraparenchymal veins. It is in the context of poor therapy options that our recommendations are made.

CT imaging can overestimate the rate of complete occlusion. Pulmonary artery wedge angiograms are more sensitive to diagnose patency, which permits at least an attempt to salvage the pulmonary vein. The transcatheter technique is straightforward and well described in the literature. A transseptal puncture is required if the atrial septum is intact. The course from the femoral veins to the pulmonary veins is not tortuous in general, and the delivery systems are well tolerated by patients.

**Risks/Complications**

Complications are similar to stenting in other vascular structures and include vessel dissection, stent malposition, and embolization. Because the procedure is performed in the left atrium, an embolized stent can have devastating consequences. Alternatively, the stent implantation can be performed intraoperatively under direct vision. The advantages of this technique are the ability for precise positioning of the stent within the left atrium, which permits future easy access for reinterventions, and its suitability in infants and small children. Although immediate results are excellent, midterm and long-term results are less optimal, with restenosis being the major issue.

**Recommendations for Pulmonary Venous Angioplasty and Stenting**

**Class I**

1. Pulmonary venous angioplasty and stenting are indicated for the management of acquired significant pulmonary vein stenosis after radiofrequency ablation procedures (Level of Evidence: C).

2. Pulmonary venous angioplasty and stenting are indicated for the management of acquired pulmonary vein stenosis after lung transplantation or for external compression due to tumors in older children and adolescents (Level of Evidence: B).

**Class IIa**

1. Pulmonary venous angioplasty is reasonable to consider in the management of pulmonary vein stenosis after surgical repair for anomalous pulmonary vein connections (Level of Evidence: C).

**Class IIb**

1. Pulmonary venous angioplasty and stenting may be considered in the management of isolated congenital pulmonary vein stenosis (Level of Evidence: C).

2. Pulmonary venous angioplasty and stenting may be considered for the management of acquired pulmonary vein stenosis after lung transplantation or for external compression due to tumors in infants and young children (Level of Evidence: C).
3. Pulmonary venous stenting may be considered for the management of postoperative pulmonary vein stenosis after surgical repair for anomalous pulmonary vein connections (Level of Evidence: C).

Class III

1. Pulmonary venous angioplasty and stenting should not be considered in the management of pulmonary vein stenosis associated with other CHD that requires surgical intervention (Level of Evidence: C).

6.5. Patent Ductus Arteriosus Stenting

Stenting of the ductus arteriosus as a means to establish a reliable source of pulmonary blood flow for palliation of cyanotic heart disease is a relatively new transcatheter intervention. Stenting of the ductus arteriosus for the purpose of establishing systemic blood flow will be addressed later in the present statement. (The reader is referred to Section 9 for stenting of the patent ductus arteriosus [PDA]) to augment systemic blood flow.) Compared with surgical alternatives, ductal stenting is attractive because it can avoid complications such as chylothorax, phrenic, or recurrent laryngeal nerve injury or pulmonary artery distortion, which are well described after placement of a surgical aortopulmonary shunt. However, data describing the outcomes of ductal stenting in newborns with cyanotic CHD are currently available from relatively few single-institution case series. The advantages and disadvantages of this procedure, therefore, are not yet fully understood.

The initial results of ductal stenting were not favorable because of technical failures and early ductal restenosis.224,225 During the past decade, however, results have improved for ductal stenting in select infants with cyanotic CHD.226–232

The improved outcomes described in more recent reports are due in part to the availability of flexible, low-profile stents designed for coronary artery use that can be delivered safely through a 4F to 5F sheath. Patient selection has also improved with the identification of those newborns who are more likely to benefit from ductal stenting. Currently, the literature suggests that the balance of benefits to risks with ductal stenting is most favorable for newborns with a relatively straight ductus (no more than 1 to 2 bends) who require reliable palliation for no more than 3 to 6 months. These criteria are typically met in infants with critical pulmonary stenosis or pulmonary atresia with intact ventricular septum that may require an additional source of pulmonary blood flow after a pulmonary valve dilation procedure. Newborns with a more tortuous ductus (>2 turns) present a technical challenge to ductal stenting, and therefore, although successful stenting of such ducts has been reported, a higher rate of procedural failure or early restenosis may be expected.

Early restenosis is common in the stented ductus, and consequently, the palliation provided by this procedure is less reliable and often of shorter duration than can be expected from a surgical aortopulmonary shunt. Alwi and colleagues,227 for example, reported the outcome of ductal stenting in 51 infants with cyanotic CHD. Freedom from reintervention was 89% at 6 months and decreased to 55% at 12 months. Restenosis most commonly is caused by in-stent tissue ingrowth but also occurs predictably at the aortic or pulmonary artery end of the ductus if even a 1- to 2-mm length is left unstented. In fact, most reports have stressed the critical importance of stenting the entire length of the duct, from its pulmonary artery to aortic end, to avoid early restenosis.

Ductal stenting can be performed from an antegrade or a retrograde approach. In newborns with critical pulmonary stenosis or pulmonary atresia after pulmonary valve dilation, who typically have a relatively short and straight ductus, the transvenous antegrade approach is preferable; this approach can enable an arterial catheter to be used for angiography to assist in stent positioning. A retrograde arterial approach is used in newborns without a patent RV outflow tract and has been described from the femoral, umbilical, or axillary artery. It is important that the ductus be somewhat restrictive, to provide a stable site for anchoring the stent. For this reason, prostaglandin E1 infusion may need to be stopped several hours before the procedure in some patients.

Flexible premounted stents designed for coronary artery use are used for ductal stenting. These can be inserted through 4F to 5F sheaths over a 0.014-inch wire. The necessary stent length will vary depending on ductal anatomy, but to avoid early restenosis, it is important that the stent cover the entire ductus from pulmonary artery to aortic end. If a single stent does not provide sufficient length, then 2 overlapping stents can be implanted. In full-term newborns, a stent diameter of 3.5 to 4 mm generally provides adequate palliation of pulmonary blood flow without leading to excessive pulmonary blood flow. In small newborns, a diameter of 3.0 mm may be adequate. To help diminish in-stent restenosis, long-term antiplatelet therapy is generally recommended after the procedure.

Ductal stenting may provide acceptable short-term palliation in cyanotic newborns who have another source of pulmonary blood flow, such as patients with critical pulmonary stenosis or pulmonary atresia with intact ventricular septum after RV outflow tract dilation. Such patients often require only short-term additional pulmonary blood flow from the stented duct, and they also typically have a relatively straight ductus, which is most amenable to stenting. Patients with a more tortuous ductus (>2 turns) may also benefit from ductal stenting, but the literature suggests that stenting of such ducts is technically challenging, and the procedural risk-benefit ratio is currently not as favorable. Because restenosis is relatively common, ductal stenting may not be as appropriate in patients who are absolutely dependent on ductal patency for pulmonary blood flow and who therefore require a secure and reliable palliation. Certainly, if such a newborn is deemed to be at too high a risk for a surgical shunt, then ductal stenting may be reasonable. However, very careful follow-up is mandatory, because early and unpredictable restenosis of the stented duct makes such palliation less reliable and potentially hazardous in such a patient.

Risks/Complications

Complications of the procedure include those associated with cardiac catheterization in newborns, particularly femoral vessel injury or occlusion. In patients who are absolutely “ductal dependent,” the interventional team should be prepared for the possible need for extracorporeal membrane oxygenation or bypass should the ductus unexpectedly spasm...
or be injured during manipulation. The PDA stenting procedure itself can traumatize the ductus during wire/catheter positioning and can lead to ductus perforation or occlusion before stenting. Stent malposition or embolization can occur and may require a difficult retrieval procedure. As noted above, if any portion of the ductus remains uncovered by the stent, then early ductal stenosis is common and may lead to important arterial hypoxemia in a cyanotic infant. Shunting through a stented PDA may be generous, which may lead to complications of excessive pulmonary blood flow. The stenting of the PDA does add to the surgical complexity at the time of the second-stage palliation (eg, control of pulmonary blood flow, need for pulmonary angioplasty). Close collaboration with the surgical and interventional teams in adopting this strategy is necessary.

**Recommendations for PDA Stenting for the Purpose of Augmenting Pulmonary Blood Flow**

The recommendations that follow reflect the understanding that despite avoiding surgical morbidities, palliation obtained from ductal stenting is less reliable and of shorter duration than that expected from a surgical aortopulmonary shunt. The reader is referred to section 9 on PDA stenting to augment systemic blood flow.

**Class IIa**

1. It is reasonable to stent an anatomically suitable ductus arteriosus in an infant with cyanotic CHD who has >1 source of pulmonary blood flow (eg, antegrade pulmonary blood flow or collateral blood flow) but who requires additional pulmonary blood flow from the stented ductus for a relatively short period of time (3 to 6 months) (**Level of Evidence: B**).

**Class IIb**

1. It might be reasonable to stent an anatomically suitable PDA in an infant with cyanotic CHD whose sole source of pulmonary blood flow is the ductus (**Level of Evidence: C**).

**Class III**

1. Ductal stenting should not be performed in an infant with cyanotic CHD who has obvious proximal pulmonary artery stenosis in the vicinity of the ductal insertion (**Level of Evidence: C**).

### 6.6. Conduit Intervention

Surgically implanted conduits commonly are used to establish RV-to-pulmonary artery continuity and for the extracardiac completion of Fontan repairs. Obstruction of these conduits occurs secondary to multiple factors. With a conduit implanted in smaller patients, the conduit becomes obstructed with growth of the patient. Additionally, kinking or external compression of the conduit, stenosis of the bioprosthetic valve within the conduit, and intimal proliferation and calcification within the conduit all can cause significant obstruction. The definitive treatment for conduit obstruction is surgical replacement of the conduit.

However, significant obstructions can be palliated by dilation or stenting of the conduit, and this represents an alternative treatment in patients with conduit obstruction and RV pressures greater than two thirds of the systemic pressure. This palliation will create or aggravate pulmonary regurgitation, which must be considered. However, successful enlargement of the obstructive diameter often will suffice for many months or even years until a more invasive and definitive surgical replacement must be performed. Balloon dilation alone may be successful short-term but seldom produces sustained results, so that most interventional therapy of conduit obstruction now includes the implantation of a stent to maintain the dilated diameter. The reader is referred to section 7 for a discussion of intravascular stent implantation in association with a transcatheter pulmonary valve implantation.

**Risks/Complications**

The risks of balloon dilation/stent implantation in conduits include not only the creation of pulmonary regurgitation but the risk of a tear or even rupture of the conduit. To reduce this risk, the dilation/stent should not exceed the original diameter of the conduit. Coronary artery compression may occur as a result of RV-to-pulmonary artery conduit stenting; it is best avoided by careful assessment of coronary artery proximity to the conduit and the use of coronary angiography during conduit test dilation.

**Recommendation for Balloon Dilation of Conduit Obstruction**

**Class IIb**

1. As a palliative measure, balloon dilation of significant conduit obstruction may be indicated to relieve obstruction or to test the compliance of the obstruction before stent implantation (**Level of Evidence: C**).

**Recommendation for Intravascular Stent Implantation for Conduit Obstruction**

**Class I**

1. Primary intravascular stent implantation is indicated for the treatment of significant stenosis of an RV-to-main pulmonary artery/pulmonary artery conduit when there is a predominant and significant stenosis of the conduit and it is believed that (a) the stent will significantly prolong the life of the conduit before further intervention becomes necessary, (b) the pulmonary regurgitation created will be tolerated better than the stenosis, and (c) the stent definitely will not impinge on the pulmonary artery bifurcation or compromise the coronary circulation by compression (**Level of Evidence: B**).

### 7. Transcatheter Vascular Occlusion

#### 7.1. Patent Ductus Arteriosus

Occlusion of the PDA was first described in 1971 with an Ivalon plug.233 This was followed by Rashkind et al, who used a hooked single-disk device in 1979 and a double-disk device in 1987.234,235 These systems used rather large delivery systems.235 In 1992, Cambier et al236 reported a small series using Gianturco coils to occlude small PDAs. The major advantage of this technique was the use of small (5F) delivery systems. Since then, transcatheter occlusion of PDAs with a variety of devices and innovative techniques has been re-
ported.237–295 Both safety and efficacy have been established in many series, and this treatment modality has become a standard of care at many centers except in very low birthweight patients or those with unsuitable anatomy, such as the type B (also known as the AP window type) PDA.296 In the mid-1990s, the PDA Coil Registry, which represented 46 institutions, reported a 95% success rate in a series of 535 patients with a median minimal PDA diameter of 2.0 mm, with complete occlusion achieved in 75% within 24 hours.240 The European Pediatric Cardiology Registry reported a large series of 1291 attempted PDA coil occlusions in 1258 patients with an immediate occlusion rate of 59%, which increased to 95% at 1-year follow-up.241 Complete closure has approached 100% in late-term follow-up in later series. Subsequently, moderate-sized and large PDAs were closed successfully with multiple coils or other devices by use of a variety of techniques.237,244,253,261,262,264,270,280,283,284,286,288–290,295 One study reported a series of 104 patients with moderate to large PDAs who underwent coil occlusion with a success rate of 100% and 98% complete occlusion at 2- to 16-month follow-up, respectively.237 With the recent US Food and Drug Administration approval of the AMPLATZER Duct Occluder in 2003, PDAs as large as 16 mm can be closed rather easily with no long-term residual shunting. Although original recommendations from the manufacturer for ductal occlusion exclude patients who weigh <6 kg, successful use in infants as small as 2.5 kg has been reported, although such patients are more challenging technically.

The most common devices used currently are various kinds of coils and the AMPLATZER ductal occluder device. Many prograde and antegrade techniques have been developed to deliver coils to maximize occlusion and to minimize the risk of coil embolization.237,242,297–302 The AMPLATZER ductal occluder device is generally implanted by the antegrade approach. Standard delivery techniques are well described in the literature.

Indications for PDA occlusion are elimination of pulmonary overcirculation and subsequent development of obstructive pulmonary vascular disease, as well as prevention of endocarditis/endarteritis. There is controversy related to occlusion of so-called silent ductus. Endocarditis in the silent ductus has been found only in single-case reports.303 In general, there are few data on the benefits of occluding the silent ductus because of its small size and presumably lack of significant flow turbulence and endothelial damage.

In those patients with a large PDA and bidirectional flow due to pulmonary vascular disease, occlusion may be beneficial only if the pulmonary lung bed shows some reactivity to pulmonary vasodilator therapy.304,305 These patients should undergo hemodynamic assessment and pulmonary vasoactivity testing before consideration for ductal occlusion. However, data on this group of patients are scant, and long-term follow-up data are unknown. Should pulmonary vascular disease continue to progress, the ductus will no longer be available to prevent the RV pressures from becoming suprasystemic.

Finally, in older patients who have developed Eisenmenger syndrome due to an unrestrictive ductus, occlusion of the ductus is contraindicated. At the other end of the spectrum, small infants (<2.4 kg) would benefit from elimination of their ductus, but risks of the transcatheter approach render this option less desirable than surgical ligation and division.

**Risks/Complications**

The PDA occlusion procedure is relatively straightforward. Rare complications have been reported, including inadvertent device embolization into the pulmonary and systemic circulation; device obstruction to aortic (creating an iatrogenic coarctation) or pulmonary flow, especially in small infants; transient left ventricular systolic dysfunction; hemolysis; and recanalization.237,241,252,278,279,285,289,306–312 Careful ductal and ampulla measurements for device selection and postimplantation evaluation before device release are of the utmost importance to minimize these risks. Residual shunting after coil occlusion may require additional coils. Although it is common to see initial residual shunting through an AMPLATZER PDA occluder, a multicenter trial indicated 99.7% complete occlusion at 1-year follow-up.352 More challenging anatomy includes the type B PDA and the calcified ductus in the elderly, whereas small infants and patients with pulmonary vascular disease pose another set of issues related to PDA occlusion.

**Recommendations for Transcatheter PDA Occlusion**

**Class I**

1. Transcatheter PDA occlusion is indicated for the treatment of a moderate-sized or large PDA with a left-to-right shunt that results in any of the following: Congestive heart failure, failure to thrive, pulmonary overcirculation (with or without pulmonary hypertension), or an enlarged left atrium or left ventricle, provided the anatomy and patient size are suitable (Level of Evidence: B).

**Class IIa**

1. Transcatheter PDA occlusion is reasonable in the presence of a small left-to-right shunt with normal-sized heart chambers when the PDA is audible by standard auscultation techniques (Level of Evidence: C).

**Class IIb**

1. In rare instances, transcatheter PDA occlusion may be considered in the presence of a bidirectional PDA shunt due to pulmonary hypertension and obstructive pulmonary vascular disease but reversible to a pure left-to-right shunting with pulmonary vasodilator therapy (Level of Evidence: C).

**Class III**

1. Transcatheter PDA occlusion should not be attempted in a patient with a PDA with severe pulmonary hypertension associated with bidirectional or right-to-left shunting that is unresponsive to pulmonary vasodilator therapy (Level of Evidence: C).
7.2. Aortopulmonary Collateral Vessels

Aortopulmonary collaterals can be found in association with various CHDs, from simple lesions such as the PDA to very complex malformed hearts, resulting in varying degrees of left-to-right shunting. There are 3 major categories of patients in whom occlusion of these vessels may be considered when found.

Aortopulmonary collaterals are often found in single-ventricle patients who have undergone a Glenn shunt procedure or Fontan repair. The resultant left-to-right shunting may be detrimental to the single-ventricle physiology, for example, by causing increased volume load and filling pressure to the single ventricle, and it may act as competitive flow in the pulmonary circulation and increase pulmonary pressures in the Glenn or Fontan circulation. In the early postoperative period, the increased left-to-right shunting can increase ventilator time and prolong the patient’s stay in the intensive care unit. These collaterals may contribute to the development of pleural effusions or protein-losing enteropathy, but the nature and extent of the contribution are not clear because many other hemodynamic, anatomic, and neurohumoral factors are involved. There are many reports in the literature that document the clinical improvement of ill single-ventricle patients after occlusion of these vessels, but the benefits of routine occlusion of aortopulmonary collaterals during pre-Glenn or pre-Fontan cardiac catheterization in the otherwise asymptomatic patient are not well defined. Certain benefits of collateral occlusion for the surgical team include reducing the pulmonary venous blood return during cardiopulmonary bypass, thus improving visibility and effective tissue perfusion during cardiopulmonary bypass.

Patients with tetralogy of Fallot and pulmonary atresia with VSD commonly have associated aortopulmonary collaterals. Interestingly, in the presence of cyanosis and decreased pulmonary flow, the left-to-right shunt of the aortopulmonary collateral may be beneficial in augmenting pulmonary flow. This is especially true when aortopulmonary collaterals are associated with hypoplastic native pulmonary arteries. The anatomy of aortopulmonary collaterals in this group of patients varies widely. They can be the major source of pulmonary flow and require surgical unifocalization procedures to be incorporated into the pulmonary arterial circulation, or they may contribute to only a few segments of the pulmonary circulation. They may or may not have dual supply with the native pulmonary arterial tree. The origins of the aortopulmonary collaterals can arise anywhere along the aorta or its major side branches, including the coronary arteries. The decision to occlude these aortopulmonary collaterals depend on several factors: Degree of left-to-right shunting, degree of cyanosis and effective pulmonary flow, and degree of dual supply between the aortopulmonary collateral and the native pulmonary artery to the segmental branches and pulmonary vascular bed. Collaboration and careful planning with the surgeons are important, with the ultimate goal of maximizing and evenly distributing true pulmonary flow and minimizing pulmonary pressure.

Aortopulmonary collaterals can be found in patients with other types of CHD that result in excessive left-to-right shunting. Often, they are masked by another predominant cardiac lesion and are not discovered until after surgical repair of the major lesion. The typical situation is a postoperative patient who remains in congestive heart failure with pulmonary overcirculation of unclear origin. Echocardiography may or may not discover the presence of the aortopulmonary collateral. Further workup with cardiac catheterization is required when the aortopulmonary collateral is discovered and occluded. Further workup with MRI or CT may be helpful with subsequent catheterization and occlusion of the aortopulmonary collateral vessels at the time of cardiac catheterization.

Transcatheter occlusion of vascular structures has been reported since the 1970s with a variety of devices and materials, which fall into 4 categories: Particulate agents, liquids, detachable balloons or plugs, and coils. At the present time, transcatheter occlusion of aortopulmonary collaterals is performed most commonly with coils and AMPLATZER vascular plug occluders. Techniques of occlusion are well described in the literature. It is important to recognize that aortopulmonary collaterals may have multiple sources of arterial supply, and occlusion devices should be delivered as selectively and as deeply into the lesion as possible to block all potential arterial supply to the final pulmonary exit point.

### Risks/Complications

The risks of occluding aortopulmonary collaterals are related to inadvertent device embolization into an important systemic artery. Because many of these collaterals arise from the head vessels off the aortic arch, such as the carotid or subclavian artery, it is important to select a device that can be accommodated by the collateral vessel such that it does not inadvertently fall out of the collateral during implantation. Rarely, extravasation of contrast material is reported with manipulation of catheters into small collaterals.

### Recommendations for Aortopulmonary Collateral Occlusion

#### Class I

1. Transcatheter occlusion of the aortopulmonary collateral vessels is indicated for treatment of aortopulmonary collateral vessels with documented large left-to-right shunting in biventricular or single-ventricle physiology that results in congestive heart failure, pulmonary overcirculation, and respiratory compromise, or development of pleural effusion or protein-losing enteropathy (Level of Evidence: B).

#### Class IIb

1. Transcatheter occlusion of aortopulmonary collateral vessels may be considered in the presence of moderate-sized collaterals found in asymptomatic single-ventricle patients undergoing routine pre-Glenn or pre-Fontan cardiac catheterization (Level of Evidence: B).

2. Transcatheter occlusion of aortopulmonary collateral vessels may be considered in association with surgical consultation in patients with pulmonary atresia and aortopulmonary collaterals that have adequate dual supply from native pulmonary arteries (Level of Evidence: B).
Class III
1. Transcatheter occlusion is not recommended for the presence of aortopulmonary collaterals of any size in biventricular or single-ventricle patients who have significant cyanosis due to decreased pulmonary flow (Level of Evidence: C).
2. Transcatheter occlusion is not recommended for patients with pulmonary atresia with aortopulmonary collaterals that can be unifocalized into native pulmonary arteries (Level of Evidence: C).

7.3. Surgically Created Systemic-to–Pulmonary Artery Shunts

The use of surgical aortopulmonary shunts such as the Blalock-Taussig shunt (BTS) for palliation of cyanotic CHD has waned in favor of more definitive surgical repair. Use of so-called Potts and Waterston shunts was discontinued because of pulmonary artery distortion and a high risk of pulmonary hypertension but is still occasionally seen in adults with palliated CHD. As patients progress through more advanced surgical correction, alternate flow may become unnecessary, and with prolonged patency, shunts may compromise more anatomically and physiologically (surgically) corrected blood flow. Further surgical intervention to ligate or occlude these shunts is associated with risk of morbidity, which include injury to the phrenic nerve, recurrent laryngeal nerve, and thoracic duct. Surgical occlusion of the shunt requires an incision, usually requires a longer hospital stay, and may be associated with morbidity because of prolonged dissection and a long operation. With the development of methods and available devices, several literature studies and reports have shown that percutaneous transcatheter interventions provide an effective, reproducible, and safe route of shunt closure. The use of the percutaneous route is advantageous overall for shunt closure when it is the sole step remaining for elimination of extraneous sources of pulmonary blood flow, which may put the patient at long-term risk of pulmonary hypertension, ventricular volume overload, and infective endocarditis.

The literature consists of small series of patients or case reports. The largest series is from Sivakumar et al., of 22 patients who had occlusion of BTS before surgical correction of tetralogy of Fallot. Earlier, Moore et al. had reported percutaneous closure of BTS in 18 patients from 1993 to 1998 using both coils and vascular occlusion devices; of the 19 closures, all were complete and without complications, embolism, or the need for further corrective surgical interventions. These studies support the overall feasibility and high rate of success associated with the percutaneous route for transcatheter closure of surgically created shunts.

Shunt occlusion is performed during cardiac catheterization either under general anesthesia or with sedation. Antibiotics are given; patients are fully heparinized, and continuous heparinized fluid infusion is flushed through large sheaths to prevent thrombus formation. Larger sheaths may kink because of acute angles created by sharp angles at the takeoff and insertion of surgically created shunts.

Various devices have been used for shunt occlusions. Most commonly, coils are chosen for BTS occlusion because of the smaller catheter and sheath requirements for deployment from the arterial end of the shunt. Coils may be either the standard Gianturco coil (0.038-inch or the sturdier 0.052-inch), the sturdier 0.052-inch pushable coils (with controlled-release mechanisms that use a bioptome), the MReye Flipper or Jackson coil (Cook Medical), or the “DuctOcclud” coil (PFM Medical, Cologne, Germany). Controlled-release coils are preferable when there is no stenosis within the shunt to ensure safe deployment before release. In older patients or if venous access to the shunt can be achieved, a larger sheath can be used to deliver an AMPLATZER vascular plug or AMPLATZER duct occluder. Other devices that have been used for BTS occlusion in the past include the Gianturco-Grifka vascular occlusion device (Cook), the Rashkind double-umbrella occluding device, and detachable balloons.

The risk of embolization of a device placed in a BTS (especially where there is no stenosis in the shunt) can be minimized either by stent placement in the pulmonary artery at the shunt anastomosis site or by temporary occlusion of the distal end of the shunt with a balloon catheter or temporary balloon occlusion of the pulmonary artery across the area of the shunt. Techniques of BTS occlusion are sometimes easier when an exchange length wire introduced into the shunt from the arterial end is snared and exteriorized out of the femoral vein to form an arteriovenous loop.

The Potts shunt is probably best closed with a covered aortic endoluminal stent graft. Use of an AMPLATZER MVSD occluder (AGA) has been reported as well as other devices.

For direct ascending aorta-to–pulmonary artery connections (Waterston anastomosis), double-umbrella devices (Rashkind PDA, Lock Clamshell, and CardioSEAL ASD devices [NMT Medical]) and AMPLATZER ASD devices have all been used. A through-and-through arteriovenous exchange length wire helps prevent kinking of the long delivery sheath in the unfavorable angles on this shunt.

Risks/Complications

The high rate of flow through shunts from the arterial system (eg, BTS) or directly from the aorta to the pulmonary system (Potts and Waterston shunts) increases the risk of pulmonary embolism. However, technical modifications to control either flow into the shunt or distal flow exiting the shunt, by catheter, balloon, or stent, are effective in minimizing such complications.

Nineteen shunt occlusions reported in 2000 by Moore et al. from 2 institutions resulted in no procedural complications, no embolization of any device or coil, and no protrusion into pulmonary or systemic arteries. In a more recent series, transcatheter closure with various coils (bioptome released) or detachable devices was successful in 21 of 22 patients, with 1 coil embolization and 1 procedural failure due to acute angle takeoff of the shunt. In this latter series, reported procedural time, fluoroscopic time, and radiation dose were reasonable (respective means: 42 minutes, 8.5 minutes, and 14.2 Gy-cm²).
Heparinization is used at 100 U/kg to prevent thrombus in the sheaths and femoral artery occlusion. However, it has been shown to have no measurable effect on occlusion rate when coils are used.373

An important consideration in BTS transcatheter device occlusion is the long-term consideration of leaving behind a fixed length of Gore-Tex tube, which has the potential to cause “tenting” upward of the pulmonary artery in a growing child. Long-term follow-up of this potential complication is therefore required. Tenting of the pulmonary artery was not reported in the 2 larger series of transcatheter occlusion of BTS or from institutions that surgically clip or ligate BTS rather than dividing the shunt.357,358

**Recommendations for Transcatheter Device Occlusion of Surgically Created Shunts**

**Class I**

1. Transcatheter occlusion of BTS is indicated for patients with pulmonary atresia with intact ventricular septum or critical pulmonary stenosis who have previously undergone a decompressive surgical or interventional procedure (eg, pulmonary valvuloplasty) but demonstrate evidence of no longer being dependent on the systemic-to-pulmonary shunt for maintenance of adequate oxygenation (Level of Evidence: C).

2. Transcatheter coil or device occlusion of BTS (or Potts or Waterston shunts) is indicated after surgical correction of a congenital heart defect in which a previous palliative surgical shunt remains patent with a residual significant left-to-right shunt (Level of Evidence: C).

**Class IIa**

1. Transcatheter coil or device occlusion of a palliative BTS (or Potts or Waterston shunt) is reasonable just before corrective surgery if the surgeon predicts difficult or increased risk with surgical closure of the shunt and the patient does not become significantly desaturated during test occlusion (Level of Evidence: C).

**Class III**

1. Transcatheter coil or device occlusion of a BTS (or Potts or Waterston shunt) is not recommended before the cardiac defect has been corrected if the patient develops unsatisfactory hypoxemia with balloon occlusion of the shunt (Level of Evidence: C).

### 7.4. Transcatheter Occlusion of Other Vascular Abnormalities

**Coronary Fistula**

Coronary fistulae may arise from the right or left coronary artery and drain most commonly into the right atrium, ventricle, or pulmonary artery. With the advent of Doppler echocardiography, these previously rare connections are being detected with increasing frequency. Most patients are asymptomatic, and spontaneous regression has been reported.374 In the adult population, the frequency of symptoms increases. Thrombosis within the fistula is rare but may cause acute myocardial infarction, paroxysmal atrial fibrillation, and ventricular arrhythmias. Embolization can be performed with coils or other occluding devices.375–377 Complications may include incomplete occlusion with residual shunting, myocardial ischemia if a more distal coronary artery is inadvertently occluded, and distal embolization of a coil to the right side of the heart or the pulmonary artery, which requires retrieval. Late recanalization or endarteritis has not been reported after coil embolization of coronary artery fistulae.

**Pulmonary Arteriovenous Malformations**

Pulmonary arteriovenous malformations (PAVMs) are abnormal direct connections between the pulmonary arteries and veins (bypassing the capillaries); although uncommon, they may be life-threatening. PAVMs may be congenital and isolated or associated with liver disease, palliated CHD (in which no hepatic flow travels through the lung), cancer, or trauma. There is a strong association between PAVMs and Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia); 50% to 80% of PAVMs are associated with this syndrome, and 5% to 15% of patients with hereditary hemorrhagic telangiectasia have PAVMs. Manifestations of PAVMs include cyanosis (most commonly), paradoxical embolization, brain abscess, and rupture. Lesions are generally classified on the basis of size (small versus large) and whether an aneurysm or anomalous drainage exists.378 Transcatheter occlusion is recommended for all symptomatic patients and for asymptomatic patients with discrete lesions feeding arteries ≥3 mm in diameter to prevent neurological complications and progressive cyanosis.379 It should be understood that generalized diffuse microscopic PAVMs are not typically amenable to transcatheter occlusion. Transcatheter techniques have proven to be safe and effective, with excellent long-term results.380 The development of new vascular occluding devices has expanded the number of options for closure and has allowed for successful closure of even large PAVMs.381–384

**Risks/Complications**

Complications, although infrequent, may include incomplete occlusion, thrombus formation in the venous channels or veins “collecting system,” and malposition/embolization of the closure devices. Care must be taken to avoid the occlusion of nearby nonfistulous branches.

**Recommendations for Transcatheter Occlusion of Coronary and Pulmonary Arteriovenous Fistulas**

**Class I**

1. Transcatheter occlusion is indicated for closure of discrete PAVMs in patients who have evidence of significant cyanosis or for patients at risk for or who have a documented history of a systemic embolic event (Level of Evidence: B).

2. Transcatheter occlusion is indicated for patients with symptomatic coronary artery fistulae (Level of Evidence: B).

**Class IIa**

1. Transcatheter occlusion is reasonable for the management of patients with moderate or large coronary artery fistulae without clinical symptoms (Level of Evidence: C).
Both surgery and transcatheter PVL occlusion have limitations in this unstable and ill group of patients. In the largest series most recently published, 27 patients underwent attempted percutaneous closure of mitral PVL with an AMPLATZER ductal occluder device. The device was implanted successfully in 63%, and the degree of regurgitation was reduced in 50%. There was significant comorbidity (>60%) in this group of patients. Another smaller series of transcatheter PVL occlusion has shown better procedural success, with mild or no residual regurgitation in 17 of 21 attempts. Five of 10 patients reported in 2006 had sustained symptomatic improvement after 1 year, but 4 of the 10 required a second procedure.

### Risks/Complications

There is currently no device specifically developed for closure of PVL through a transcatheter approach. With the high regurgitant flow and varying diameter of the PVL, there is a risk of device dislodgement. Large defects are associated with greater risk of embolization, especially after attempted closure with coil devices. Impingement of the device on prosthetic valvular leaflets or residual leak may remain after closure. The procedures tend to be difficult, with prolonged radiation and procedural times. These risks may be mitigated by the thorough use of advanced imaging techniques to assess the exact conformation of the leak and to confirm proper and thorough placement of the device or devices within the defect. Adequate closure is also dependent on choice of an appropriate device. Residual shunt is not uncommon and sometimes necessitates a second procedure. Mortality is usually related to the comorbidities of the patient.

### Recommendation for Transcatheter Device Occlusion of PVLs

#### Class I

1. Transcatheter device occlusion of PVL is indicated for patients with PVL-associated hemolysis, recurrent blood transfusions, or hemodynamically significant heart failure and is deemed at high risk for surgical intervention after consultation with surgical colleagues (Level of Evidence: C).

#### Class III

1. Transcatheter device occlusion of PVLs is not indicated for patients with clinically insignificant coronary arteriovenous fistulae (eg, normal-sized cardiac chambers) (Level of Evidence: C).

#### Class III

1. Transcatheter device occlusion of PVL is indicated when it is determined that there is inadequate space in which to seat the device without impairing valvar function (Level of Evidence: C).

### 7.6. Venovenous Channels

Venovenous collaterals are found most often in children with a univentricular heart after a bidirectional Glenn or modified Fontan procedure. The resulting increased systemic venous pressure after these palliations allows for flow through the pulmonary vasculature. The development of collaterals from the “high-pressure” systemic venous system to the low-pressure venous system serves as a right-to-left shunt and may lead to (significant) cyanosis. The cause is unclear but may represent either de novo angiogenesis or reopening of previously existing channels. Venovenous collaterals occur in approximately one third of patients after Glenn or Fontan procedures, and approximately one third of those require intervention. The only predictor of development of collaterals is a higher transpulmonary gradient or higher systemic venous pressure.

Collateral channels to the pulmonary venous atrium (eg, directly to the heart, pulmonary veins, or coronary sinus) should be embolized at the time of diagnosis. Collaterals found to drain from the upper-body systemic venous return to below the diaphragm should be embolized preoperatively (pre-Glenn). There is no need to embolize collateral channels...
that drain below the diaphragm in an asymptomatic or mildly cyanotic pre-Fontan patient if a Fontan completion is planned. Once the Fontan procedure is completed, venous collateral channels that drain below the heart are of no physiological significance.

Risks/Complications
As with other embolization procedures, the most common complications are incomplete occlusion and device migration/embolization with the need for retrieval. Late recanalization may occur if coils are placed too proximally in the vessel to allow other collaterals to reenter the occluded vessel distal to the occlusion point.

Recommendations for Transcatheter Device Occlusion of Venovenous Anomalous Channels

Class I

1. Transcatheter embolization of a venovenous collateral vessel that drains to the pulmonary venous atrium either directly or via the pulmonary veins or coronary sinus is indicated if the collateral vessels are clinically relevant (e.g., significant cyanosis, risk for or documentation of a systemic embolic event) (Level of Evidence: C).

2. Transcatheter embolization is indicated for the management of venovenous collaterals that have developed after the modified Glenn procedure and that drain below the diaphragm, leading to clinically important cyanosis, unless Fontan completion is imminent (Level of Evidence: C).

Class IIa

1. It is reasonable to consider transcatheter embolization of venovenous collateral vessels not associated with significant cyanosis when such vessels are found at the time of pre-Fontan catheterization (Level of Evidence: C).

Class IIb

1. Venovenous collaterals that drain below the diaphragm in a pre-Glenn patient may be embolized (Level of Evidence: B).

Class III

1.Venovenous collaterals that drain below the diaphragm in a patient scheduled to undergo Fontan completion need not be embolized (Level of Evidence: C).

8. Transcatheter Pulmonary Valve Replacement

Pulmonary regurgitation may result in progressive RV dilation and failure, arrhythmias, and death. Resurrection of the pulmonic valve at an appropriate age may restore RV function and improve symptoms. Surgical valve implantation requires cardiopulmonary bypass, which may aggravate an already failing RV.\(^{405,406}\) Currently, surgeons use one of several types of valved conduits to replace the pulmonic valve: Homografts from cadavers, valved conduits, and the Contegra bovine jugular vein graft or a bioprosthetic valve implanted directly in the RV outflow tract. The primary mode of failure in all 3 is stenosis; however, regurgitation can occur, and often, the reason for failure is a combination of stenosis and regurgitation. Tweddell and colleagues\(^{407}\) reported on factors that affect homograft longevity and discovered that by approximately 4 to 5 years after homograft replacement, \(\approx25\%\) of patients would require reintervention on these conduits.

Bonhoeffer et al\(^{408}\) were the first to report on a novel percutaneous pulmonary valve. Since the first report, hundreds of patients have received this valve with good results.\(^{409}\) The Bonhoeffer valve (Melody transcatheter pulmonary valve) is made of a bovine jugular vein sewn inside a Cheatham-Platinum stent (NuMED Inc). The valve is hand-cramped on a BIB (balloon-in-balloon) catheter and is delivered while protected by a retractable cover that requires a 22F delivery system. Garay and colleagues\(^{410}\) reported on the use of another percutaneous valve, the Edwards-Cribier transcatheter heart valve (Edwards Lifesciences), in a patient with a failed homograft. This valve is made of 3 bovine pericardial leaflets sewn inside a stainless steel stent. The valve is available in 2 sizes, 23 or 26 mm in diameter, and requires a 22F to 24F delivery sheath. The valve is crimped by use of a crimper. The delivery sheath is 35 cm long; therefore, it is delivered unprotected by the sheath. The valve and delivery system are being modified to allow deployment in a smaller delivery system (18F); furthermore, a 20- and 29-mm-diameter valve will be added to the trial in the future.

The Melody transcatheter pulmonary valve (Medtronic) received US Food and Drug Administration approval for humanitarian device exemption use in January 2010.\(^{411}\) The Edwards SAPIEN transcatheter heart valve is still in clinical trials in the United States, and patients are eligible to receive these valves if they meet certain inclusion and exclusion criteria. The current transcatheter valve systems are designed to treat conduit and bioprosthetic valve failure only at this time. They are not intended to be used to treat patients who have undergone RV outflow tract reconstruction by a transannular patch technique. There were some members of the writing committee who believed that recent publications and experience with the transcatheter pulmonary valve warranted a Class I recommendation for its use.\(^{412-419}\) The committee ultimately elected to assign this a Class IIa recommendation given the relatively recent release for its use in the United States and the still limited number of centers that provide the procedure.

Risks/Complications
There are potential risks associated with transfemoral access and potential risks in those patients who require general anesthesia. Risks uniquely associated with the use of the pulmonic valve include the following:

1. Device instability and/or dislodgement: This may lead to percutaneous device retrieval and redeployment or may necessitate urgent surgery to retrieve the device. Therefore, such procedures should be performed in centers where surgical backup is available.

2. Coronary compression due to stent placement: Coronary artery compression can be avoided by careful
CT/MRA before the procedure to assess the distance between the conduit (and site of the stent valve implantation) and the coronary artery. In addition, at the time of the procedure, one should perform balloon inflation in the RV outflow tract during aortic root angiography or selective coronary angiography.

3. Pulmonary artery obstruction: Right or left pulmonary artery obstruction can be avoided by careful preprocedural CT/MRI assessment and meticulous stent and valve placement with fluoroscopy and angiography at the time of implantation. The appropriate-length stent should be positioned away from the origin of either branch pulmonary artery.

4. Homograft rupture: Factors related to rupture include degree of deterioration of the conduit (eg, severe calcification) and aggressive oversizing of balloon dilation. The importance of appropriate sizing for multiple modalities (CT/MRI, echocardiography, balloon-sizing angiography) cannot be overemphasized. Furthermore, the availability of covered stents for bailout in such circumstances is of crucial importance.

5. Stent fracture: This complication can lead to an increase in RV outflow tract gradient and RV pressure. The incidence of this complication was as high as 21% in 1 series that used the Melody valve. In that series, repeat interventions after pulmonic valve implantation were performed predominantly because of stent fractures. Nordmeyer and colleagues have devised a risk-stratification, systemic classification and anticipatory management strategy to effectively manage stent fracture after percutaneous pulmonary valve implantation.

6. Local vascular complications due to the large size required for valve delivery: Currently, some centers advocate venous cut-down and repair of the vein to better manage the vessel after intervention. Other centers use femoral vessel closure devices to achieve hemostasis after valve delivery. Several brands are available.

Recommendations for Percutaneous Pulmonary Valve Replacement

Class IIa

1. It is reasonable to consider percutaneous pulmonary valve replacement in a patient with an RV-to-pulmonary artery conduit with associated moderate to severe pulmonary regurgitation or stenosis provided the patient meets inclusion/exclusion criteria for the available valve (Level of Evidence: B).

9. Hybrid Procedures

9.1. HLHS and Complex Single-Ventricle Physiology

The "hybrid" approach to CHD combines traditional surgical and interventional catheterization procedures into 1 strategy. Successful execution of a hybrid procedure necessitates a close and dynamic working relationship between the interventionalist and surgeon that begins before and continues throughout and after the hybrid procedures. Perhaps in no cardiac lesion is the hybrid approach more embraced than in HLHS.

HLHS is a fatal disease, with ~90% of patients dying within the first month of life without some sort of intervention. The objective of stage I palliation for HLHS or any single-ventricle physiology is to provide an unobstructed systemic outflow tract, unrestricted interatrial communication, a controlled source of pulmonary blood flow, and a reliable source of coronary blood flow. The Norwood procedure described in 1981 and the Sano modification of the Norwood procedure described in 2001 both meet the above objectives. Although individual centers worldwide have had success with 1 or more of the surgical strategies, the learning curve has been steep, and in recent years, survival through all 3 stages of palliation has appeared to plateau.

In 1992, Gibbs and colleagues proposed palliating a newborn with HLHS by percutaneous PDA stent implantation and bilateral pulmonary artery branch banding during off-pump surgery. The next year, Ruiz et al used the same palliation as a bridge to cardiac transplantation in infants with HLHS. However, similar to the Norwood operation, the initial results of off-pump transcatheter surgical palliation were poor. A more contemporary attempt at a similar approach (selective pulmonary artery bandings without cardiopulmonary bypass followed by percutaneous PDA stent implantation and balloon dilation of the atrial septum) was made by Akintuerk et al in 2002. This group in Giessen, Germany, has found an overall actuarial survival rate after transcatheter surgery of 83%, with a 21% combined mortality rate for patients with a stage I, interstage, and stage II repair.

Numerous reports have been published by Galantowicz and Cheatham related to the evolution of the hybrid approach to HLHS. Their hybrid approach consists of an initial placement of surgical pulmonary artery bands through a small median sternotomy off cardiopulmonary bypass, with PDA stent delivery through a sheath placed directly through a purse-string suture in the main pulmonary artery above the pulmonary valve. A balloon atrial septostomy is performed later as a percutaneous therapy before discharge of the patient, which allows the left atrium to enlarge and a larger balloon to be used to give long-lasting results. These authors advocate that this hybrid technique minimizes comorbidities of HLHS stage I palliation such as small patient size, severity of ventricular dysfunction or tricuspid regurgitation, size of the ascending aorta, and multisystem organ failure. For this reason, many centers regard the hybrid stage I as a "rescue" procedure for high-risk HLHS and single-ventricle patients or as a bridge to heart transplantation in infants with HLHS. Only retrograde aortic arch obstruction with the PDA fully open is considered a contraindication to the hybrid stage I palliation because of potential acute obstruction to coronary flow, with the cells of the stent covering the tiny retrograde orifice. To avoid hemodynamic issues related to retrograde obstruction, one center has advocated the use of the "reverse" BTS at the time of hybrid palliation to protect coronary blood flow, but this is not applied universally in hybrid palliation.

A focus for patients, parents, and healthcare professionals alike is the long-term neurodevelopmental outcome and quality of life for HLHS patients. Multiple studies have raised concern about abnormal IQ and neurodevelopmental testing...
after the Norwood procedure. The impact that the hybrid approach will have on this issue remains unknown.

**Risks/Complications**
The immediate risks of the hybrid palliation include stent embolization, development of retrograde coarctation, and development of proximal stenosis of uncovered ductus. Pulmonary artery banding may result in pulmonary arterial distortion and need for angioplasty at the time of the second palliation. Balloon atrial septostomy in these patients may result in inadequate decompression or need for repeat balloon atrial septostomy. As noted above, reinterventions are common among hybrid-palliated HLHS patients. Heralded by a deterioration in RV pressure, increasing tricuspid regurgitation, or both, in-stent stenosis of the PDA stent or development of a retrograde coarctation may be observed. Likewise, close observation for restenosis of the atrial septum is necessary.

**Recommendations for a Hybrid Approach to HLHS and Complex Single Ventricle**

**Class IIa**

1. It is reasonable to perform hybrid stage I palliation, consisting of right and left pulmonary artery banding, PDA stent implantation, and creation of an unrestrictive atrial communication, by combining transcatheter and surgical techniques without cardiopulmonary bypass as an alternative to conventional surgery in neonates with HLHS or complex single ventricle, in high-risk surgical candidates, and as a bridge to heart transplantation *(Level of Evidence: B).*

**Class IIb**

1. Hybrid stage I palliation may not be indicated in a patient who has significant retrograde aortic arch obstruction at the time of initial diagnosis that might be further compromised by placement of the PDA stent. This decision should be a collaborative decision between the interventional cardiologist and a congenital heart surgeon *(Level of Evidence: C).*

9.2. **Perventricular Device Closure of MVSDs**

The surgical and percutaneous device closure of some forms of MVSDs remains a challenging problem. In the US feasibility trial evaluating the AMPLATZER MVSD occluder for percutaneous closure, a weight of <5 kg was significantly correlated with procedure- or device-related complications (53.8% compared with 38.1% in infants ≥5 kg). The reasons for this increase in complication rate are attributed to the necessary arterial venous loop required for percutaneous delivery, the relatively stiff exchange guidewire needed, the stiff delivery cable, the less than ideal angle of device delivery to MVSD anatomy, and the necessity for delivery across the tricuspid valve, which causes tricuspid regurgitation. All of these obstacles are magnified in the small infant and result in hemodynamic instability during device delivery.

In 1999, Amin et al described an alternative method of device closure of MVSD and perimembranous VSD using a left thoracotomy incision and performing “perventricular” closure without the use of cardiopulmonary bypass in catar pigs, which raised the potential for a hybrid approach to MVSD. Previously, there were reports of intraoperative device closure of MVSD while patients were on cardiopulmonary bypass, but a perventricular approach through a median sternotomy could potentially avoid cardiopulmonary bypass, and such a hybrid approach could be used in babies and small infants. In 2003, Bach et al described the technique and results of perventricular MVSD closure in 6 consecutive infants without cardiopulmonary bypass. Either a small median sternotomy or a subxiphoid minimally invasive incision was performed. Using TEE, the free wall of the RV was punctured with a needle through a purse-string suture after a suitable location was established by the surgeon by pushing on the RV free wall with his or her finger under TEE guidance; this allowed a perpendicular approach to the MVSD and avoided the papillary muscles. Under continuous TEE guidance, a soft angled-tip guidewire was introduced through the needle and across the MVSD into the left ventricle and then directed into either the aorta or left atrium. An appropriately sized short sheath and dilator were then introduced over the guidewire through the small perventricular purse-string suture and placed in the left ventricle. An AMPLATZER MVSD occluder 1 to 2 mm larger than the TEE-measured diameter of the MVSD was chosen and attached to the delivery cable, then transferred from the loading sheath into the short delivery sheath. Using TEE, the left ventricular disk was deployed, avoiding the mitral valve chordal attachments, and pulled against the left ventricular side of the septum, which allowed the middle waist to expand and fill the defect, with eventual deployment of the RV disk. Once the device was believed to be secure, the release mechanism was engaged and the device released. After that, the sheath was withdrawn from the perventricular entry site. It is possible that multiple MVSDs may be closed by use of this technique. With this technique, there is no hemodynamic compromise because the atrioventricular and semilunar valves are not crossed; there is no tension on the delivery cable because the device is delivered perpendicular to the defect; and there is virtually no blood loss. On completion of the procedure, the purse-string suture is tightened to close the entry site in the RV free wall, and the small median sternotomy or subxiphoid incision is closed. There were no complications in the initial 6 patients, with no significant shunting across the MVSD on discharge transthoracic echocardiography. In fact, 4 of the 6 patients still required cardiopulmonary bypass for additional complex CHD repair, but this hybrid strategy shortened their time on cardiopulmonary bypass and resulted in excellent clinical outcomes in complicated small infants.

By 2005, a multicenter experience with perventricular device closure of MVSDs was reported in 12 infants, with complete closure in 10 of the 12 and small shunts in 2, which demonstrated that this technology could be easily transferred to other hybrid teams. In 2007, a report was published that described AMPLATZER MVSD occluder closure of MVSDs in infants <1 year of age.

That same year, successful perventricular closure of MVSDs using the CardioSEAL occluder was reported in infants who weighed <8 kg, with 80% complete closure.
All of these reports were from centers within the United States, but by 2008, perventricular device closure of MVSD was being performed worldwide with excellent results and a high percentage of complete closure or trivial residual flow. Challenges remain for teams that perform perventricular MVSD closure. Defects either in the posterior, anterior, or apical septum can be difficult to access from the free wall of the RV and may require an even more in-depth hybrid approach. With a combined percutaneous approach to cross these defects from the left ventricle and place the guidewire into the pulmonary artery or RA, a perventricular sheath can be placed through the RV to facilitate snare capture of the guidewire and exteriorization through the RV sheath. The AMPLATZER MVSD occluder can then be delivered by a perventricular approach to maximize the optimal delivery angle for successful implantations.

MVSDs that would be suitable for hybrid closure include those with a hemodynamically significant shunt; a low likelihood of spontaneous closure; location of the defect in an anatomic area accessible by this method that would allow device closure without impingement on other intracardiac structures, such as the atrioventricular valves; and of a size and location that would indicate that the device will remain in a stable position (ie, with acceptable rims surrounding the defect).

Risks/Complications
The perventricular delivery of an MVSD device carries the risk of device embolization and inability to adequately anchor the device (which could lead to an aborted attempt, residual interventricular shunt, or atrioventricular valve compromise), similar to that seen in transvascular device placement. Although these intraprocedural complications are minimized by use of a hybrid perventricular approach that avoids hemodynamic compromise, arrhythmias, device malposition, and blood transfusions, unusual late complications have been reported. Nearly 10 months after successful perventricular closure, a left ventricular pseudoaneurysm was identified during cardiac catheterization to close additional defects. The pseudoaneurysm was believed to be secondary to a stiff guidewire, sheath, or dilator injury of the left ventricular free wall at the time of implantation. The pseudoaneurysm was treated successfully by transcatheter embolization, and this complication could be avoided with careful placement of the guidewire, sheath, dilator, and device under expert TEE guidance.

Recommendations for Hybrid Perventricular Device Closure of MVSDs

**Class IIa**

1. It is reasonable to perform hybrid perventricular device closure of MVSDs in small infants, thereby avoiding the cardiopulmonary bypass required in surgical closure and minimizing the adverse events associated with percutaneous device delivery (Level of Evidence: B).

**Class IIb**

1. Hybrid perventricular device closure of an MVSD in patients may be considered if there are concomitant surgical procedures that have additional risks or if the defect location might be better approached by this technique, as determined by a joint decision between the interventional cardiologist and cardiac surgeon (Level of Evidence: C).

9.3. Stent Implantation
Endovascular stents have become a valuable tool in the treatment of a variety of vascular stenoses in patients with CHD. Stent placement has been used to successfully treat vascular narrowing in the systemic and pulmonary arterial and venous circulations, as well as a variety of prosthetic tubes such as systemic-pulmonary shunts and RV-to-pulmonary artery conduits. Although the effectiveness of stent therapy is well accepted, the technical demands of implantation have at times limited the applicability of these devices, particularly in small, critically ill children. Although improvements in delivery technique and balloon, stent, and sheath technology have made percutaneous stent implantation a more accomplishable procedure, there are numerous clinical scenarios in which intraoperative or hybrid stent implantation may be preferable.

Intraoperative stent implantation into the pulmonary arteries was first reported in 1992. The technique of implantation has evolved and varies between centers but typically involves either placement under direct vision, videoscopic guidance with cardiopulmonary bypass, or angiographic image guidance performed in a beating heart without bypass. When placed under direct vision or with the aid of videoscopic guidance, balloon-expandable stents of a predetermined size are delivered to the target area without the use of an introducer sheath and expanded to a predetermined diameter. The stent dimensions and implantation balloon size are determined by preprocedural imaging (angiography, MRI, or CT scan). Hybrid stent implantation with surgical sheath placement (eg, delivery sheath surgically placed into the RV outflow tract to deliver adult-sized pulmonary artery stents in a small infant) uses serial angiography through the side arm of the sheath to determine stent and implantation balloon size. This technique allows for postimplantation imaging and the performance of preintervention and postintervention hemodynamic measurements. Although the greatest experience with hybrid stent implantation has been with branch pulmonary artery stenosis, more recently, this technique has been used to treat recurrent aortic arch obstruction, systemic–pulmonary artery shunt obstruction, and central venous stenoses and for the maintenance of ductal patency. The major advantage of these techniques is freedom from sheath constraints, which allows stents with adult-sized growth potential to be placed in young infants and children. Additional benefits of hybrid stent placement include a simplified catheter course, which may result in improved speed and ease of implantation, decreased exposure to ionizing radiation, improved hemodynamic stability during implantation, and the ability to combine 2 procedures (surgery and interventional catheterization) under 1 anesthetic, thereby reducing cumulative morbidity. Because of these benefits, intraoperative imaging can be helpful on completion of the surgical procedure.
Risks/Complications
The risks of hybrid placement of endovascular stents are the same as those outlined for individual use of transcatheter stent placement in the text.

Recommendations for Hybrid Stent Implantation
Class IIa
1. It is reasonable to perform hybrid stent implantation when a patient is undergoing an open cardiac surgical procedure and a stenotic lesion has been identified, either preoperatively or intraoperatively, that could be treated concomitantly with stent implantation (Level of Evidence: C).

10. Unique Catheter/Interventional Care Considerations
10.1. Postoperative Patients, Including Those on Extracorporeal Membrane Oxygenation
Patients who experience difficulty recovering after congenital heart surgery have a high incidence of residual structural lesions that may be amenable to further therapy (either surgical, catheter-directed, or hybrid).473–475 This is particularly true for patients who cannot be separated from cardiopulmonary bypass or who require mechanical cardiopulmonary support in the early postoperative period. Not surprisingly, these patients have a hospital mortality rate significantly higher than that for similar patients with an uncomplicated postoperative course.476–482 Reported survival to discharge in patients who require mechanical cardiopulmonary support after congenital heart surgery ranges between 33% and 55%. Noninvasive diagnostic imaging modalities (eg, transthoracic echocardiography, TEE, CT scan, and MRI) may be useful in determining the presence or absence of hemodynamically significant residual anatomic lesions, but these techniques may be limited in this setting. Diagnostic cardiac catheterization can be performed safely in the early postoperative period and often results in the discovery of previously undiagnosed important anatomic lesions that require further surgical or catheter intervention.483–485

Although interventional catheterization has become a mainstay in the treatment of many late postoperative lesions, such as branch pulmonary artery stenosis, residual shunts, and recurrent coartation of the aorta, these interventions have historically been avoided in the early postoperative period. Reasons for this include concerns about difficulties transporting these critically ill patients, concerns about worsening clinical status as a result of the procedure, hemodynamic instability during the catheterization procedure, and fear of disruption of recently placed suture lines. It is commonly believed that 6 weeks must pass after cardiac surgery to allow for adequate formation of scar tissue around new anastomotic sites before a catheter intervention such as angioplasty or stent implantation can be performed safely. Recent data, however, suggest that angioplasty, stent implantation, and device occlusion of intracardiac and vascular shunts can all be performed safely, with acceptable results, in the early postoperative period (<6 weeks after surgery).159,474,486

Furthermore, performance of successful catheter intervention in the early postoperative period may improve survival to discharge for patients who require mechanical cardiopulmonary support.489 Prerequisites for the safe performance of these procedures include (1) the involvement of a multidisciplinary team that includes a dedicated interventional cardiologist, congenital heart surgeon, cardiac intensivist, cardiac anesthesiologist, and, when indicated, perfusion team, and (2) a catheterization laboratory with substantial experience in the treatment of critically ill patients and complex CHD.

Recommendations for Catheterization and Transcatheter Interventions on Postoperative Patients
Class I
1. Cardiac catheterization with the potential for intervention is indicated early in the postoperative period in critically ill patients when there is a high index of suspicion for the presence of a residual structural lesion(s) but noninvasive diagnostics fail to identify a cause for hemodynamic and/or clinical compromise (Level of Evidence: B).

2. Cardiac catheterization with potential for intervention is indicated early in the postoperative period in any patient who requires mechanical cardiopulmonary support without a clear cause (Level of Evidence: B).

Class IIa
1. It is reasonable to perform cardiac catheterization with potential for catheter intervention early in the postoperative period in patients who are not pursuing a typical postoperative course (eg, prolonged mechanical ventilation, systemic oxygen desaturation) and in whom noninvasive diagnostic testing does not determine a reason for this course (Level of Evidence: C).

10.2. Decompression of a Fontan Circuit
Although sparsely reported in the literature, atrial septostomy has also been used in patients with Fontan circuits, both soon after surgery to augment cardiac output and chronically for treatment of such complications as a failing Fontan physiology or protein-losing enteropathy.487–492 Preoperative catheterization of Fontan candidates usually identifies those patients liable to encounter early postoperative issues due to elevated pulmonary resistance. These patients often undergo a fenestrated Fontan. However, pulmonary resistance may increase in patients postoperatively for a number of unforeseen reasons that might jeopardize the Fontan physiology. Likewise, a fenestration may be inadequate in size or may even close soon after surgery in patients at risk. The creation or enlargement of a fenestration in the Fontan circuit (or in the case of an extracardiac Fontan, stenting of the fenestration) can lead to improved Fontan hemodynamics by relieving the systemic venous hypertension and providing added preload to the single ventricle. The concept works much the same way for patients with chronically failing Fontan physiology. In both scenarios, however, the benefits may be countered by the negative effects of increased hypoxemia or increased load on a failing ventricle. The increasing use
of the extracardiac conduit Fontan limits this as an option. Notably, radiofrequency perforation is not applicable for perforation through artificial materials used in many intracardiac baffles.

Recommendations for Decompression of a Fontan Circuit

Class IIa

1. As a means to augment cardiac output, atrial septostomy is indicated for the immediate postoperative patient with a nonfenestrated Fontan procedure (or fenestrated with a small or occluded fenestration) with a low cardiac output state secondary to elevated pulmonary vascular resistance that is unresponsive or minimally responsive to therapy (Level of Evidence: C).

2. It is reasonable to consider atrial septostomy in an effort to augment cardiac output in patients with a chronically failing Fontan physiology (Level of Evidence: C).

10.3. Endocarditis Prophylaxis Issues in Cardiac Catheterization and Intervention

Most children with uncorrected or incompletely corrected CHD are at increased risk for developing infective endocarditis. This increased risk goes away after successful repair of the cardiac defect, although in other children, the risk is lifelong. In the past, it was recommended that all individuals with an increased risk for infective endocarditis receive antimicrobial prophylaxis before certain dental or gastrointestinal/genitourinary tract procedures or surgeries. In 2007, the AHA made major changes to the recommendations for endocarditis prophylaxis. They concluded that only an extremely small number of cases of infective endocarditis might be prevented by antibiotic prophylaxis even if such prophylactic therapy were 100% effective. The recommendation for routine prophylaxis was deemed reasonable only for patients with underlying cardiac conditions associated with the highest risk of adverse outcome from infective endocarditis. These cardiac conditions associated with the highest risk of adverse outcome from infective endocarditis and for which prophylaxis before certain dental procedures include:

- Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
- Previous infective endocarditis
- CHD, including the following:
  - Unrepaired cyanotic CHD, including palliative shunts and conduits
  - Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure (endothelialization of prosthetic material occurs within 6 months after the procedure)
  - Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)

Table. Regimens for Infective Endocarditis Prophylaxis Before a Dental Procedure

<table>
<thead>
<tr>
<th>Situation</th>
<th>Agent</th>
<th>Pediatric Dose*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral</td>
<td>Amoxicillin</td>
<td>50 mg/kg</td>
</tr>
<tr>
<td>Unable to take oral medication</td>
<td>Amoxicillin OR Cefazolin or ceftriaxone</td>
<td>50 mg/kg IM or IV</td>
</tr>
<tr>
<td>Allergic to penicillins or ampicillin—oral</td>
<td>Cephalaxin OR Clindamycin OR Azithromycin or clarithromycin</td>
<td>50 mg/kg or 20 mg/kg or 15 mg/kg</td>
</tr>
<tr>
<td>Allergic to penicillins or ampicillin and unable to take oral medication</td>
<td>Cefazolin or ceftriaxone OR Clindamycin</td>
<td>50 mg/kg IM or IV or 20 mg/kg IM or IV</td>
</tr>
</tbody>
</table>

IM indicates intramuscular; IV, intravenous.
*Medication given as single dose 30 to 60 minutes before the procedure. Total pediatric dose should not exceed total adult dose (see reference 493 for adult doses).
†Cephalosporins should not be used in an individual with a history of anaphylaxis, angioedema, or urticaria with penicillins or ampicillin.
Adapted from Wilson et al. Copyright © 2007, American Heart Association, Inc.

- Cardiac transplantation recipients who develop cardiac valvulopathy. For children with these underlying cardiac conditions, prophylaxis is reasonable for all dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa. Specific antibiotic regimens are listed in the Table. Antimicrobial prophylaxis solely to prevent endocarditis is no longer recommended for any patients who undergo a gastrointestinal/genitourinary tract procedure.

- Infective endocarditis as a complication of cardiac catheterization is exceedingly rare, and routine antibiotic prophylaxis is not recommended. Bacteremia is rarely observed during diagnostic cardiac catheterization, and infective endocarditis as a complication after pediatric cardiac catheterization is rare. Trauma to the endothelium of a valve or to the endocardium during catheterization can induce deposition of platelets and fibrin, which leads to a nonbacterial thrombotic endocardial lesion that makes the site vulnerable to infection. In addition, a congenitally abnormal structure within the heart or great vessels provides a site that could become infected. Without associated bacteremia, however, endocarditis will not occur. This points to the need for strict attention to sterile technique at the wound entry site.

- With regard to placement of intracardiac and intravascular prosthetic devices such as stents, coils, and occluding devices, it remains the practice of most interventional cardiologists to provide antibiotic coverage (typically a ceftazidime) during and after the case.
## Disclosures

### Writing Group Disclosures

<table>
<thead>
<tr>
<th>Writing Group Member</th>
<th>Employment</th>
<th>Research Grant</th>
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<td>Timothy F. Feltes</td>
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<td>Robert H. Beekman III</td>
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<td>Jeffrey A. Feinstein</td>
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<td>Michael de Moor</td>
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<td>Kathryn A. Taubert‡</td>
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*This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all writing group members are required to complete and submit. A relationship is considered to be “significant” if (1) the person receives $10,000 or more during any 12-month period or 5% or more of the person’s gross income; or (2) the person owns 5% or more of the voting stock or share of the entity or owns $10,000 or more of the fair market value of the entity. A relationship is considered to be “modest” if it is less than “significant” under the preceding definition.

†Significant.
‡Dr. Taubert is now with the World Heart Federation, Geneva, Switzerland.

### Reviewer Disclosures

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<td>Jacqueline Kreutzer</td>
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*Modest.
References


**KEY WORDS:** AHA Scientific Statements • catheterization • pediatrics • heart defects, congenital • valvuloplasty, balloon • cardiovascular diseases