ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease) Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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PRACTICE GUIDELINE: EXECUTIVE SUMMARY

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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Preamble

It is important that the medical profession play a central role in critically evaluating the use of diagnostic procedures and therapies introduced and tested for detection, management, or prevention of disease. Rigorous, expert analysis of the available data documenting absolute and relative benefits and risks of these procedures and therapies can produce guidelines that improve the effectiveness of care, optimize patient outcomes, and favorably affect the cost of care by focusing resources on the most effective strategies.

The American College of Cardiology Foundation (ACCF) and the American Heart Association (AHA) have jointly engaged in the production of guidelines in the area of cardiovascular disease since 1980. The American College of Cardiology (ACC)/AHA Task Force on Practice Guidelines is charged with developing, updating, and revising practice guidelines for cardiovascular diseases and procedures and directs this effort. Writing committees are charged with assessing the evidence as an independent group of authors to develop, update, or revise recommendations for clinical practice.

Experts in the subject under consideration have been selected from both organizations to examine subject-specific data and write guidelines in partnership with representatives from other medical practitioner and specialty groups. Writing committees are specifically charged to perform a formal literature review, weigh the strength of evidence for or against particular treatments or procedures, and include estimates of expected health outcomes where data exist. Patient-specific modifiers, comorbidities, and issues of patient preference that might influence the choice of tests or therapies are considered, as well as the frequency of follow-up and cost-effectiveness. When available, information from studies on cost is considered, but data on efficacy and clinical outcomes constitute the primary basis for recommendations in these guidelines.

The ACC/AHA Task Force on Practice Guidelines makes every effort to avoid actual, potential, or perceived conflicts of interest that might arise as a result of industry relationships or personal interests among the writing committee. Specifically, all members of the writing committee, as well as peer reviewers of the document, are asked to disclose all such relationships that might be perceived as real or potential conflicts of interest. Writing committee members are also strongly encouraged to declare previous relationships with industry that might be perceived as relevant to guideline development. If a writing committee member develops a new relationship with industry during their tenure, they are required to notify guideline staff in writing. These statements are reviewed by the parent task force, reported orally to all members at each meeting of the writing committee, and updated and reviewed by the writing committee as changes occur. Please refer to the methodology manual for ACC/AHA guideline writing committees for further description of the relationships with industry policy (1). See Appendix 1 for author relationships with industry and Appendix 2 for peer reviewer relationships with industry pertinent to this guideline.

These practice guidelines are intended to assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions. Clinicians should consider the quality and availability of expertise in the area where care is provided. These guidelines attempt to define practices that meet the needs of most patients in most circumstances. The recommendations reflect a consensus of expert opinion after a thorough review of the available current scientific evidence and are intended to improve patient care.

Patient adherence to prescribed and agreed upon medical regimens and lifestyles is an important aspect of treatment. Prescribed courses of treatment in accordance with these recommendations are only effective if they are followed. Because lack of patient understanding and adherence may adversely affect outcomes, physicians and other healthcare providers should make every effort to engage the patient's active participation in prescribed medical regimens and lifestyles.

If these guidelines are used as the basis for regulatory or payer decisions, the goal is quality of care and serving the patient's best interest. The ultimate judgment regarding care of a particular patient must be made by the healthcare provider and the patient in light of all of the circumstances presented by that patient. There are circumstances in which deviations from these guidelines are appropriate.

The guidelines will be reviewed annually by the ACC/AHA Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution. The Executive Summary and Recommendations are published in the December 2, 2008, issue of the *Journal of the American College of Cardiology* and the December 2, 2008, issue of *Circulation.* The full-text guidelines are e-published in the same issue of these journals and posted on the ACC (www.acc.org) and AHA (http://my.americanheart.org) World Wide Web sites.

Sidney C. Smith, Jr, MD, FACC, FAHA
Chair, ACC/AHA Task Force on Practice Guidelines

1. Introduction

1.1. Methodology and Evidence Review

The recommendations listed in this document are, whenever possible, evidence-based. Unlike other ACC/AHA practice guidelines, there is not a large body of peer-reviewed published evidence to support most recommendations, which will be clearly indicated in the text. An extensive literature survey was conducted that led to the incorporation of 647 references. Searches were limited to studies, reviews, and other evidence conducted in human subjects and published in English. Key search words included but were not limited to adult congenital heart disease (ACHD), atrial septal defect, arterial switch operation, bradycardia, cardiac catheterization, cardiac reoperation, coarctation, coronary artery abnormalities, cyanotic congenital heart disease, Doppler-echocardiography, d-transposition of the great arteries, Ebstein's anomaly, Eisenmenger physiology, familial, heart defect, medical therapy, patent ductus arteriosus, physical activity, pregnancy, psychosocial, pulmonary arterial hypertension, right heart obstruction, supravalvular pulmonary stenosis, surgical therapy, tachyarrhythmia, tachycardia, tetralogy of Fallot, transplantation, tricuspid atresia, and Wolff-Parkinson-White. Additionally, the writing committee reviewed documents related to the subject matter previously published by the ACC and AHA. References selected and published in this document are representative and not all-inclusive.

The committee reviewed and ranked evidence supporting current recommendations with the weight of evidence ranked as Level A if the data were derived from multiple randomized clinical trials involving a large number of individuals. The committee ranked available evidence as Level B when data were derived from a limited number of trials involving a comparatively small number of patients or from well-designed data analyses of nonrandomized studies or observational data registries. Evidence was ranked as Level C when the consensus of experts was the primary source of the recommendation. In the narrative portions of these guidelines, evidence is generally presented in chronological order of development. Studies are identified as observational, randomized, prospective, or retrospective. The committee emphasizes that for certain conditions for which no other therapy is available, the indications are based on expert consensus and years of clinical experience and are thus well supported, even though the evidence was ranked as Level C. An analogous example is the use of penicillin in pneumococcal pneumonia where there are no randomized trials and only clinical experience. When indications at Level C are supported by historical clinical data, appropriate references (eg, case reports and clinical reviews) are cited if available. When Level C indications are based strictly on committee consensus, no references are cited. The final recommendations for indications for a diagnostic procedure, a particular therapy, or an intervention in ACHD patients summarize both clinical evidence and expert opinion. The schema for classification of recommendations and level of evidence is summarized in Table 1, which also illustrates how the grading system provides an estimate of the size of treatment effect and an estimate of the certainty of the treatment effect.

1.2. Organization of Committee and Relationships With Industry

The ACC/AHA Task Force on Practice Guidelines was formed to create clinical practice guidelines for select cardiovascular conditions with important implications for public health. This guideline writing committee was assembled to adjudicate the evidence and construct recommendations regarding the diagnosis and treatment of ACHD. Writing committee members were selected with attention to ACHD subspecialties, broad geographic representation, and involvement in academic medicine and clinical practice. The writing committee included representatives of the American Society of Echocardiography, Canadian Cardiovascular Society, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

All members of the writing committee were required to disclose all relationships with industry relevant to the data under consideration (1).

1.3. Document Review and Approval

This document was reviewed by 3 external reviewers nominated from both the ACC and the AHA, as well as from the the American Society of Echocardiography, Canadian Cardiovascular Society, Heart Rhythm Society, International Society for Adult Congenital Cardiac Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons, and 20 individual content reviewers, which included reviewers from the ACC Congenital Heart Disease and Pediatric Cardiology Committee and the AHA Congenital Cardiac Defects Committee. All reviewer relationships with industry information were collected and distributed to the writing committee and are published in this document (see Appendix 2 for details). The committee thanks all reviewers for their comments. Many of their suggestions were incorporated into the final document.

This document was approved for publication by the governing bodies of the ACCF and the AHA and endorsed by the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

1.4. Epidemiology and Scope of the Problem

Remarkable improvement in survival of patients with congenital heart disease (CHD) has occurred over the past half century since reparative surgery has become commonplace. Since the advent of neonatal repair of complex lesions in the 1970s, an estimated 85% of patients survive into adult life. The 32nd Bethesda Conference report in 2000 estimated that there were approximately 800 000 adults with CHD in the United States (2,3). Given modern surgical mortality rates of less than 5%, one would expect that in the next decade, almost 1 in 150 young adults will have some form of CHD. In particular, there are a substantial number of young adults with single-ventricle physiology, systemic right ventricles, or

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Table 1. Applying Classification of Recommendations and Level of Evidence

ESTIMATE OF CERTAINTY (PRECISION) OF TREATMENT EFFECT ↓ / SIZE OF TREATMENT EFFECT →	CLASS I *Benefit >>> Risk* Procedure/Treatment **SHOULD** be performed/administered	CLASS IIa *Benefit >> Risk* *Additional studies with focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	CLASS IIb *Benefit ≥ Risk* *Additional studies with broad objectives needed; additional registry data would be helpful* Procedure/Treatment **MAY BE CONSIDERED**	CLASS III *Risk ≥ Benefit* Procedure/Treatment should **NOT** be performed/administered **SINCE IT IS NOT HELPFUL AND MAY BE HARMFUL**
LEVEL A Multiple populations evaluated* Data derived from multiple randomized clinical trials or meta-analyses	■ Recommendation that procedure or treatment is useful/effective ■ Sufficient evidence from multiple randomized trials or meta-analyses	■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from multiple randomized trials or meta-analyses	■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from multiple randomized trials or meta-analyses	■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Sufficient evidence from multiple randomized trials or meta-analyses
LEVEL B Limited populations evaluated* Data derived from a single randomized trial or nonrandomized studies	■ Recommendation that procedure or treatment is useful/effective ■ Evidence from single randomized trial or nonrandomized studies	■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from single randomized trial or nonrandomized studies	■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from single randomized trial or nonrandomized studies	■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Evidence from single randomized trial or nonrandomized studies
LEVEL C Very limited populations evaluated* Only consensus opinion of experts, case studies, or standard of care	■ Recommendation that procedure or treatment is useful/effective ■ Only expert opinion, case studies, or standard of care	■ Recommendation in favor of treatment or procedure being useful/effective ■ Only diverging expert opinion, case studies, or standard of care	■ Recommendation's usefulness/efficacy less well established ■ Only diverging expert opinion, case studies, or standard of care	■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Only expert opinion, case studies, or standard of care
Suggested phrases for writing recommendations†	should is recommended is indicated is useful/effective/beneficial	is reasonable can be useful/effective/beneficial is probably recommended or indicated	may/might be considered may/might be reasonable usefulness/effectiveness is unknown/unclear/uncertain or not well established	is not recommended is not indicated should not is not useful/effective/beneficial may be harmful

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

†In 2003, the ACC/AHA Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline 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 guidelines and will allow queries at the individual recommendation level.

complex intracardiac baffles who are now entering adult life and starting families. Young adults have many psychological, social, and financial issues that present barriers to proactive health management. The infrastructure that is provided to most pediatric cardiology centers, namely, case management by advanced practice nurses and social workers, is largely lacking within the adult healthcare system. Recognizing the compound effects of a complex and unfamiliar disease with an unprepared patient and healthcare system, the ACC/AHA ACHD Guideline Writing Committee has determined that the most immediate step it can take to support the practicing cardiologist in the care of ACHD patients is to provide a consensus document that outlines the most important diagnostic and management strategies and indicates when referral to a highly specialized center is appropriate. To provide ease of use, the writing committee constructed this document by lesion type and in each section included recommendations on topics common to all lesions (eg, infective endocarditis [IE] prophylaxis, pregnancy, physical activity, and medical therapy).

1.5. Recommendations for Delivery of Care and Ensuring Access

CLASS I

1. **The focus of current healthcare access goals for ACHD patients should include the following:**
   a. **Strengthening organization of and access to transition clinics for adolescents and young adults with CHD, including funding of allied healthcare providers to provide infrastructure compa-**

rable to that provided for children with CHD. (*Level of Evidence: C*)

b. Organization of outreach and education programs for patients, their families, and caregivers to recapture patients leaving pediatric supervisory care or who are lost to follow-up. Such programs can determine when and where further intervention is required. (*Level of Evidence: C*)

c. Enhanced education of adult cardiovascular specialists and pediatric cardiologists in the pathophysiology and management of ACHD patients. (*Level of Evidence: C*)

d. A liaison with regulatory agencies at the local, regional, state, and federal levels to create programs commensurate with the needs of this large cardiovascular population. (*Level of Evidence: C*)

2. Health care for ACHD patients should be coordinated by regional ACHD centers of excellence that would serve as a resource for the surrounding medical community, affected individuals, and their families. (Table 2)

a. Every academic adult cardiology/cardiac surgery center should have access to a regional ACHD center for consultation and referral. (*Level of Evidence: C*)

b. Each pediatric cardiology program should identify the ACHD center to which the transfer of patients can be made. (*Level of Evidence: C*)

c. All emergency care facilities should have an affiliation with a regional ACHD center. (*Level of Evidence: C*)

3. ACHD patients should carry a complete medical "passport" that outlines specifics of their past and current medical history, as well as contact information for immediate access to data and counsel from local and regional centers of excellence. (*Level of Evidence: C*)

4. Care of some ACHD patients is complicated by additional special needs, including but not restricted to intellectual incapacities or psychosocial limitations, and designated healthcare guardians should be included in all medical decision making. (*Level of Evidence: C*)

5. Every ACHD patient should have a primary care physician. To ensure and improve communication, current clinical records should be on file with the primary care physician and local cardiovascular specialist, as well as at a regional ACHD center; patients should also have copies of relevant records. (*Level of Evidence: C*)

6. Every cardiovascular family caregiver should have a referral relationship with a regional ACHD center so that all patients have geographically accessible care. (*Level of Evidence: C*)

Table 2. Personnel and Services Recommended for Regional ACHD Centers

Type of Service	Personnel/Resources
Cardiologist specializing in ACHD	One or several 24/7
Congenital cardiac surgeon	Two or several 24/7
Nurse/physician assistant/nurse practitioner	One or several
Cardiac anesthesiologist	Several 24/7
Echocardiography*	Two or several 24/7
• Includes TEE, intraoperative TEE	
Diagnostic catheterization*	Yes, 24/7
Noncoronary interventional catheterization*	Yes, 24/7
Electrophysiology/pacing/AICD implantation*	One or several
Exercise testing	• Echocardiography
	• Radionuclide
	• Cardiopulmonary
	• Metabolic
Cardiac imaging/radiology*	• Cardiac MRI
	• CT scanning
	• Nuclear medicine
Multidisciplinary teams	• High-risk obstetrics
	• Pulmonary hypertension
	• Heart failure/transplant
	• Genetics
	• Neurology
	• Nephrology
	• Cardiac pathology
	• Rehabilitation services
	• Social services
	• Vocational services
	• Financial counselors
Information technology	• Data collection
	• Database support
	• Quality assessment review/protocols

*These modalities must be supervised/performed and interpreted by physicians with expertise and training in CHD.

ACHD indicates adult congenital heart disease; 24/7, availability 24 hours per day, 7 days per week; TEE, transesophageal echocardiography; AICD, automatic implantable cardioverter defibrillator; MRI, magnetic resonance imaging; and CT, computed tomography.

The need for delivery of appropriate health care to ACHD patients largely remains unmet. The 32nd Bethesda Conference report in 2000 recommended organizing ACHD care within a regional and national system of specialized ACHD centers of excellence that would disseminate care, provide education, orchestrate research and innovation, and serve as a general resource for the region within this model (3) (Table 2). Such a system has been demonstrated to improve care for adults with similar chronic severe illness such as severe heart failure, for which measures of improvement surrounding uniformity of care within a guidelines framework, medical and surgical outcomes, decreased visits, improved patient quality of life, cost containment, data collection and knowledge dissemination, trials of new therapeutics, and enhanced insurability have been achieved.

The pediatric cardiology team should be paired with adult cardiologists to facilitate transition of care for affected individuals. It is recommended that all ACHD patients have a provider who constitutes the medical "home," as well as at least 1 overreaching visit with a cardiologist with advanced training and experience in caring for ACHD patients (4).

These models of care delivery for ACHD patients fall directly into concordance with the overarching goals of the guidelines currently established by this ACC/AHA Guideline

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Table 3. Types of Adult Congenital Heart Disease of Great Complexity*

Conduits, valved or nonvalved
Cyanotic congenital heart (all forms)
Double-outlet ventricle
Eisenmenger syndrome
Fontan procedure
Mitral atresia
Single ventricle (also called double inlet or outlet, common, or primitive)
Pulmonary atresia (all forms)
Pulmonary vascular obstructive disease
Transposition of the great arteries
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of atrioventricular or ventriculoarterial connection not included above (ie, crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

*These patients should be seen regularly at adult congenital heart disease centers.

Modified from Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170–5 (3).

Committee. A national ACHD database has been proposed to facilitate the establishment of medical and surgical outcomes and quality-of-life measures.

1.5.1. Recommendations for Access to Care

CLASS I

1. **An individual primary caregiver or cardiologist without specific training and expertise in ACHD should manage the care of adults with complex and moderate CHD (Tables 3 and 4) only in collaboration with level 2 or 3 ACHD specialists (4). (*Level of Evidence: C*)**
2. **For ACHD patients in the lowest-risk group (simple CHD; Table 5), cardiac follow-up at a regional ACHD center is recommended at least once to formulate future needs for follow-up. (*Level of Evidence: C*)**
3. **Frequent follow-up (generally every 12 to 24 months) at a regional ACHD center is recommended for the larger group of adults with complex and moderate CHD. A smaller group of adults with very complex CHD will require follow-up at a regional ACHD center at a minimum of every 6 to 12 months. (*Level of Evidence: C*)**
4. **Stabilized adult patients with CHD who require admission for urgent or acute care should be transferred to a regional ACHD center, except in some circumstances after consultation with the patient's primary level 2 or level 3 ACHD specialist (4). (*Level of Evidence: C*)**
5. **Diagnostic and interventional procedures including imaging (ie, echocardiography, magnetic resonance imaging [MRI], or computed tomography [CT]), advanced cardiac catheterization, and electrophysiology procedures for adults with complex and moderate CHD should be performed in a regional ACHD center with appropriate experience in CHD and in a laboratory with appropriate personnel and equipment. Personnel performing such procedures should work as part of a team with expertise in the surgical and transcatheter management of patients with CHD. (*Level of Evidence: C*)**
6. **Surgical procedures that require general anesthesia or conscious sedation in adults with moderate or complex CHD should be performed in a regional ACHD center with an anesthesiologist familiar with ACHD patients. (*Level of Evidence: C*)**
7. **ACHD patients should be transferred to an ACHD center for urgent or acute care of cardiac problems. (*Level of Evidence: C*)**
8. **Adult patients with complex or high-risk CHD should be transferred to an ACHD center for urgent or acute noncardiac problems. (*Level of Evidence: C*)**
9. **An ACHD specialist should be notified or consulted when a patient with simple or low-risk CHD is admitted to a non-ACHD center. (*Level of Evidence: C*)**

The features of an ACHD center, outlined in Table 2, describe a team that includes level 2 and 3 ACHD specialists. The 32nd Bethesda Conference described 3 levels of training of adult cardiovascular specialists in terms of experience in ACHD (6). Task Force 9 covered training in the care of adult patients with CHD and differentiated 3 levels of training and expected expertise. Level 1 training consists of basic exposure to CHD patients and organized educational material on CHD. To enable proper recognition of the problems of adults with CHD and to be cognizant of when specialized referral is needed, all medical cardiology fellows should achieve level 1 training in CHD. Level 1 trainees should be instructed by a

Table 4. Diagnoses in Adult Patients With Congenital Heart Disease of Moderate Complexity*

Aorto–left ventricular fistulas
Anomalous pulmonary venous drainage, partial or total
Atrioventricular septal defects (partial or complete)
Coarctation of the aorta
Ebstein's anomaly
Infundibular right ventricular outflow obstruction of significance
Ostium primum atrial septal defect
Patent ductus arteriosus (not closed)
Pulmonary valve regurgitation (moderate to severe)
Pulmonary valve stenosis (moderate to severe)
Sinus of Valsalva fistula/aneurysm
Sinus venosus atrial septal defect
Subvalvular AS or SupraAS (except HOCM)
Tetralogy of Fallot
Ventricular septal defect with:
Absent valve or valves
Aortic regurgitation
Coarctation of the aorta
Mitral disease
Right ventricular outflow tract obstruction
Straddling tricuspid/mitral valve
Subaortic stenosis

*These patients should be seen periodically at regional adult congenital heart disease centers.

Modified from Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170–5 (3).

AS indicates aortic stenosis; HOCM, hypertrophic obstructive cardiomyopathy; and SupraAS, supravalvular aortic stenosis.

Table 5. Diagnoses in Adult Patients With Simple Congenital Heart Disease*

Native disease
Isolated congenital aortic valve disease
Isolated congenital mitral valve disease (eg, except parachute valve, cleft leaflet)
Small atrial septal defect
Isolated small ventricular septal defect (no associated lesions)
Mild pulmonary stenosis
Small patent ductus arteriosus
Repaired conditions
Previously ligated or occluded ductus arteriosus
Repaired secundum or sinus venosus atrial septal defect without residua
Repaired ventricular septal defect without residua

*These patients can usually be cared for in the general medical community.
Modified from Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170–5 (3).

faculty member with level 2 or 3 training or its equivalent. A pediatric cardiologist should also be involved in these training programs.

Level 2 training represents additional training for fellows who plan to care for adult patients with CHD so that they may acquire expertise in the clinical evaluation and management of such patients. Level 2 training generally requires 1 year of training in ACHD: either a 1-year formal program at a regional or tertiary care ACHD center or cumulative experience of 12 months through repetitive rotations or electives as a cardiology fellow with experienced ACHD cardiologists. This training should prepare the individual to be well equipped for the routine care of even moderate to complex ACHD and to recognize when more advanced consultation or referral is advisable.

Level 3 training represents the level of knowledge needed by those graduates who wish to make a clinical and academic/research commitment to this field and not only become competent in the care of the entire spectrum of adult patients with CHD but also participate in the teaching and research of ACHD. Level 3 trainees generally require 2 years of training. These 24 months may either be consecutive or cumulative experience, and some recognition can be given to overall experience in CHD, be it pediatric, adolescent, or adult (eg, prior pediatric cardiology training or rotations). Such level 3 training would be sufficient to clinically manage the most complex ACHD patient in a regional or tertiary care center, to pursue an academic career, to train others in the field, or to direct an ACHD center program (7).

1.5.2. Recommendations for Psychosocial Issues

CLASS I

1. **Individual and family psychosocial screening (including knowledge assessment of cardiac disease and management; perceptions about health and the impact of CHD; social functioning with family, friends, and significant others; employment and insurability status; and screening for cognitive, mood, and psychiatric disorders) should be part of the care of ACHD patients. Advanced practice nurses, physician assistants, psychologists, and social workers should play an integral role in assessing and providing for the psychosocial needs of ACHD patients. (*Level of Evidence: C*)**
2. **Informational tools should be developed before transfer from adolescent to adult care and used for patient/family education regarding CHD, including the following elements, to be provided in electronic format:**
   a. **Demographic data, including physician contact. (*Level of Evidence: C*)**
   b. **Description of CHD, surgeries, interventional procedures, and most recent diagnostic studies. (*Level of Evidence: C*)**
   c. **Medications. (*Level of Evidence: C*)**
3. **Additional health maintenance screening and information should be offered to ACHD patients as indicated during each visit to their ACHD healthcare provider, including the following:**
   a. **Endocarditis prophylaxis measures (refer to Section 1.6, Recommendations for Infective Endocarditis). (*Level of Evidence: C*)**
   b. **Exercise prescription, guidelines for exercise, and athletic participation for patients with CHD should reflect the published recommendations of the 36th Bethesda Conference report (5). (*Level of Evidence: C*)**
   c. **Contraception and pregnancy information, including education regarding risk of CHD in offspring (for men and women). (*Level of Evidence: C*)**
   d. **General medical/dental preventive care (eg, smoking cessation, weight loss/maintenance, hypertension/lipid screening, oral care, and substance abuse counseling). (*Level of Evidence: A*)**
   e. **Recommended follow-up with cardiology. (*Level of Evidence: C*)**
4. **Vocational referral and health insurance information should be offered to ACHD patients during the transition period and refreshed at the time of their initial consultation in a tertiary referral center and intermittently as indicated by their social situation. (*Level of Evidence: C*)**
5. **A formal transition process should be used to provide optimal transfer of patients into ACHD care. This process should begin by 12 years of age and should be individualized on the basis of the patient's maturity level, with the goal being to transition and ultimately transfer the patient into adult care settings depending on the stability of the disease and psychosocial status. (*Level of Evidence: C*)**
6. **A psychological evaluation should be obtained if an adult's mental competency is in question and no appointed adult surrogate is available. (*Level of Evidence: C*)**
7. **All ACHD patients should be encouraged to complete an advance directive, ideally at a time during which they are not extremely ill or hospitalized, so that they can express their wishes thoughtfully in a less stressful setting and communicate these wishes to their families and caregivers. (*Level of Evidence: C*)**

New information is emerging about cognitive functioning in adolescents who underwent surgical repair in infancy with cardiopulmonary bypass that indicates some deficits in planning and self-management (8–12). Long-term behavioral outcome studies after the neonatal arterial switch operation (ASO) for transposition of the great arteries (TGA) have

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demonstrated highly specific disabilities that might impact the quality of self-care (13). Longer survival and decreasing morbidity among ACHD patients have made quality-of-life issues much more central to the understanding and management of this population (14–24). Some quality-of life-issues pertinent to ACHD patients, regardless of severity of disease, include independent living arrangements, education, employment, sports, health and life insurance acquisition, contraception, genetic counseling, and pregnancy concerns (25).

Circumstantial depression and anxiety are understandable in older adolescents and young adults with chronic health problems. One pilot study suggests that up to one third of ACHD patients may have a psychiatric disorder, with depression and anxiety being most prominent (26), whereas only 20% of the general population are afflicted with psychiatric illness (27). Accordingly, a careful assessment of depressive symptoms and their possible overlap with symptoms of medical illness or side effects of medications must be part of the clinical evaluation of ACHD patients (17,28).

1.5.3. Transition of Care

Physical and emotional maturity is the primary requirement for transfer of adolescent or young adult patients into adult care environments. The age at which this occurs varies and may range from the mid-teens to the mid-20s, depending on the patient. However, the process of transitioning, that is, preparing young patients for successful transfer to an adult healthcare provider at a later time, should begin by the age of 12 years (29). Strategies for transfer of patients with CHD into adult care settings are well described (30,31) and use a stepwise approach to establishing autonomy and understanding one's cardiac problem and lifestyle issues important to long-term stability of CHD.

Pertinent medical records, including diagrams of cardiac defects and operations, operative and procedural reports, recent physical examination, electrocardiograms (ECGs), and echocardiograms, should be provided to all cardiologists involved in the care of a patient with CHD. In addition, once patients are properly educated and aware of basic terminology pertaining to their own cardiac status, they should be offered copies of their medical reports, which implies and imparts responsibility and autonomy regarding their condition.

1.6. Recommendations for Infective Endocarditis

CLASS I

1. **ACHD patients must be informed of their potential risk for IE and should be provided with the AHA information card with instructions for prophylaxis. (*Level of Evidence: B*)**
2. **When patients with ACHD present with an unexplained febrile illness and potential IE, blood cultures should be drawn before antibiotic treatment is initiated to avoid delay in diagnosis due to "culture-negative" IE. (*Level of Evidence: B*)**
3. **Transthoracic echocardiography (TTE) should be performed when the diagnosis of native-valve IE is suspected. (*Level of Evidence: B*)**
4. **Transesophageal echocardiography (TEE) is indicated if TTE windows are inadequate or equivocal, in the presence of a prosthetic valve or material or surgically constructed shunt, in the presence of complex congenital cardiovascular anatomy, or to define possible complications of endocarditis (eg, sepsis, abscess, valvular destruction or dehiscence, embolism, or hemodynamic instability) (32). (*Level of Evidence: B*)**
5. **ACHD patients with evidence of IE should have early consultation with a surgeon with experience in ACHD because of the potential for rapid deterioration and concern about possible infection of prosthetic material. (*Level of Evidence: B*)**

CLASS IIa

1. **Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa is reasonable in patients with CHD with the highest risk for adverse outcome from IE, including those with the following indications:**
   a. **Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: B*)**
   b. **Previous IE. (*Level of Evidence: B*)**
   c. **Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: B*)**
   d. **Completely repaired CHD with prosthetic materials, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure. (*Level of Evidence: B*)**
   e. **Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization. (*Level of Evidence: B*)**
2. **It is reasonable to consider antibiotic prophylaxis against IE before vaginal delivery at the time of membrane rupture in select patients with the highest risk of adverse outcomes. This includes patients with the following indications:**
   a. **Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: C*)**
   b. **Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: C*)**

CLASS III

1. **Prophylaxis against IE is not recommended for nondental procedures (such as esophagogastroduodenoscopy or colonoscopy) in the absence of active infection. (*Level of Evidence: C*)**

The clinical setting and presentation of endocarditis have changed over the last 50 years, in part owing to technical advances (eg, cardiac surgery, hemodialysis), the use of prosthetic devices and indwelling lines, the increasing prevalence of intravenous drug abuse, the emergence of resistant organisms, and the continued development of increasingly potent antibiotics (33–38). True surgical cures of congenital cardiovascular disorders are infrequent, and almost all patients who have undergone surgery are left with some form of residua or sequelae, many of which predispose to IE (33,34,37–42).

A delay in diagnosis of IE carries the risk of significant morbidity and mortality. A high index of suspicion for IE in any patient with operated or unoperated CHD is a key to early diagnosis (34,38,39,41,43–46). Cardiac lesions and their relative risks of developing IE are listed in Table 6.

Table 6. Cardiac Conditions Associated With the Highest Risk of Adverse Outcome From Endocarditis for Which Prophylaxis With Dental Procedures Is Reasonable

Condition	Congenital Specific Condition*
• Previous infective endocarditis	• Unrepaired cyanotic CHD, including palliative shunts and conduits
• Prosthetic cardiac valve or prosthetic material used for cardiac valve repair	• 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†
	• Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization
	• Cardiac transplant recipients who develop cardiac valvulopathy

*Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD.

†Prophylaxis is reasonable because endothelialization of prosthetic material occurs within 6 months after the procedure.

Modified with permission to include footnotes from Wilson et al. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation.* 2007;116:1736–54 (32).

CHD indicates congenital heart disease.

The 2007 AHA guidelines for the prevention of endocarditis have substantially changed the recommendations for antibiotic prophylaxis on the basis of a consensus of expert opinions (32). The new, simplified recommendations are based on the proposition that most bacteremia occurs during activities of daily living, that IE is more likely to result from long-term cumulative exposure to these daily random bacteremias than from procedural bacteremias, and that proof is lacking that prophylaxis prevents any (or at most a very small number) cases of IE. They posit that the risks of antibiotic adverse events in the patient (allergic reactions) and the emergence of resistant organisms exceed any proven benefit of antibiotic prophylaxis against IE.

The new AHA guidelines appropriately emphasize maintenance of oral health and hygiene to reduce daily bacteremia and underscore that this is more important than any dental antibiotic prophylaxis. Accordingly, the 2007 writing committee for the updated guidelines on prevention of endocarditis concluded that antibiotic prophylaxis for dental procedures likely to induce procedural bacteremia (those that involve manipulation of gingival tissue or the periapical region of the teeth or perforation of the gingival mucosa) should be confined to cardiac conditions associated with the most significant adverse outcomes should IE develop (32). They included in this group those with previous IE, those with prosthetic cardiac valves or surgically constructed conduits or shunts, those with unrepaired cyanotic CHD or CHD repaired with prosthetic material or devices (until 6 months after the procedure), those with repaired CHD with residual defects at or adjacent to the site of a prosthetic patch or device, and cardiac transplant patients who develop valvulopathy. They specifically recommend no IE prophylaxis before gastrointestinal or genitourinary procedures, a major departure from previous guidelines. The present ACHD Guideline Committee understands that there may be reluctance to deviate from prior recommendations for patients with some forms of CHD. Accordingly, this committee recommends that healthcare providers discuss the rationale for these new changes with their patients, including the lack of scientific evidence demonstrating proven benefit for IE prophylaxis. In those settings, the clinician should determine that the risks associated with antibiotics are low before continuing a prophylaxis regimen.

The present writing committee proposes that the "high-risk" group in whom it is reasonable to give antibiotic prophylaxis before dental procedures would include the following: 1) those with a prosthetic cardiac valve; 2) those with prior IE; 3) those with unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits; 4) those with repaired CHD with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure; and 5) those with repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization.

We emphasize that nonchemotherapeutic methods are particularly important in the adolescent or young adult patient with CHD, among whom nail biting, acne, and problems with dental health are common. Oral prevention starts with meticulous oral care and routine preventive care by a dentist or oral hygienist. A patient with cyanotic heart disease often has spongy, friable gums, and a soft-bristle toothbrush must be used.

Female contraception should be planned with the risks and benefits of intrauterine devices kept in mind.

1.7. Recommendations for Noncardiac Surgery

CLASS I

1. **Basic preoperative assessment for ACHD patients should include systemic arterial oximetry, an ECG, chest x-ray, TTE, and blood tests for full blood count and coagulation screen. (*Level of Evidence: C*)**
2. **It is recommended that when possible, the preoperative evaluation and surgery for ACHD patients be performed in a regional center specializing in congenital cardiology, with experienced surgeons and cardiac anesthesiologists. (*Level of Evidence: C*)**
3. **Certain high-risk patient populations should be managed at centers for the care of ACHD patients under all circumstances, unless the operative intervention is an absolute emergency. High-risk categories include patients with the following:**
   a. **Prior Fontan procedure. (*Level of Evidence: C*)**
   b. **Severe pulmonary arterial hypertension (PAH). (*Level of Evidence: C*)**
   c. **Cyanotic CHD. (*Level of Evidence: C*)**
   d. **Complex CHD with residua such as heart failure, valve disease, or the need for anticoagulation. (*Level of Evidence: C*)**
   e. **Patients with CHD and malignant arrhythmias. (*Level of Evidence: C*)**

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Table 7. Congenital Cardiac Lesions and Perioperative Risk for Noncardiac Surgery

High risk
Pulmonary hypertension, primary or secondary
Cyanotic congenital heart disease
New York Heart Association class III or IV
Severe systemic ventricular dysfunction (ejection fraction less than 35%)
Severe left-sided heart obstructive lesions
Moderate risk
Prosthetic valve or conduit
Intracardiac shunt
Moderate left-sided heart obstruction
Moderate systemic ventricular dysfunction

4. **Consultation with ACHD experts regarding the assessment of risk is recommended for patients with CHD who will undergo noncardiac surgery. (*Level of Evidence: C*)**
5. **Consultation with a cardiac anesthesiologist is recommended for moderate- and high-risk patients. (*Level of Evidence: C*)**

Performance of any surgical procedure in ACHD patients carries a greater risk than in the normal population. Certain surgical procedures are frequently required in cyanotic patients, such as intervention for gallstones, scoliosis, and, less commonly, cerebral abscess. The risk for noncardiac surgery depends on the nature of the underlying CHD, the extent of the procedure, and the urgency of intervention. Table 7 lists lesions at moderate and high risk for noncardiac surgery.

A thorough evaluation of the patient with CHD should be undertaken before anticipated noncardiac surgery. Basic preoperative assessment includes an ECG, chest x-ray, TTE, and blood tests for full blood count and coagulation screen. It is recommended, when possible, that the preoperative evaluation and surgery be performed in an ACHD center with experienced surgeons and cardiac anesthesiologists. This allows close perioperative follow-up by an ACHD specialist.

Select high-risk patient populations should be managed at centers for the care of ACHD patients under all circumstances, unless the operative intervention is an absolute emergency. These patients include those with prior Fontan procedure, severe PAH, cyanotic CHD, or complex CHD with residua such as heart failure, valve disease, or the need for anticoagulation. Patients with cyanotic CHD, especially when associated with PAH, are at highest risk from noncardiac surgery (47).

Postoperatively, patients with CHD may need intensive care unit monitoring facilities even for relatively minor procedures. Nursing staff should be informed about the specific issues related to the CHD.

1.8. Recommendations for Pregnancy and Contraception

CLASS I

1. **Patients with CHD should have consultation with an ACHD expert before they plan to become pregnant to develop a plan for management of labor and the postpartum period that includes consideration of the appropriate response to potential complications. This care plan should be made available to all providers. (*Level of Evidence: C*)**
2. **Patients with intracardiac right-to-left shunting should have fastidious care taken of intravenous lines to avoid paradoxical air embolus. (*Level of Evidence: C*)**
3. **Prepregnancy counseling is recommended for women receiving chronic anticoagulation with warfarin to enable them to make an informed decision about maternal and fetal risks (48–50). (*Level of Evidence: B*)**

CLASS IIa

1. **Meticulous prophylaxis for deep venous thrombosis, including early ambulation and compression stockings, can be useful for all patients with an intracardiac right-to-left shunt. Subcutaneous heparin or low-molecular-weight heparin is reasonable for prolonged bed rest. Full anticoagulation can be useful for the high-risk patient. (*Level of Evidence: C*)**

CLASS III

1. **The estrogen-containing oral contraceptive pill is not recommended in ACHD patients at risk of thromboembolism, such as those with cyanosis related to an intracardiac shunt, severe PAH, or Fontan repair. (*Level of Evidence: C*)**

Congenital malformations now represent the most common cause of maternal morbidity and mortality from heart disease in North America. Both men and women with ACHD should have a thorough understanding of the risks of transmitting CHD to their offspring. Counseling by an ACHD expert before pregnancy is important and should include genetic evaluation and, specifically for women, assessment of potential fetal risk, risk of prematurity or low birth weight in the offspring, review of medications that may be deleterious to the fetus, appropriate management of anticoagulation, and discussion of potential maternal complications (51). If pregnancy occurs, fetal echocardiography should be obtained and its consequences discussed (51).

The outcome of pregnancy is favorable in most women with CHD provided that functional class and systemic ventricular function are good. PAH presents a serious risk during pregnancy, particularly when the pulmonary pressure exceeds 70% of systemic pressure, irrespective of functional class. Events often occur after delivery (52). The need for full anticoagulation during pregnancy, although not a contraindication, poses an increased risk to both mother and fetus (53). The relative risks and benefits of the different anticoagulant approaches need to be discussed fully with the prospective mother. There is a small group of patients with complex CHD or high-risk disorders in whom pregnancy is either dangerous or contraindicated because of risk to mother or fetus. If pregnancy occurs and continues with any of these disorders, these high-risk patients should be managed and delivered in specialized centers with multidisciplinary expertise and experience in CHD, obstetrics, anesthesiology, and neonatology. In patients in whom pregnancy termination is considered, the risks of termination versus continuation of pregnancy should be evaluated and discussed.

Although endocarditis is a recognized risk for maternal morbidity and mortality, endocarditis prophylaxis around the

time of delivery is not universally recommended for patients with structural heart disease, because some believe that the risk of bacteremia is low. Others routinely administer antibiotics because it is not known in advance whether or not instrumentation will be required. Thus, there is no consensus on this point (54). Antibiotics should be considered for those at highest risk of an adverse outcome and, when appropriate, given as the membranes rupture. Intravenous amoxicillin and gentamicin should be considered for women with high-risk anatomy or previous history of endocarditis (see Section 1.6, Recommendations for Infective Endocarditis).

1.8.1. Contraception

There are limited data on the safety of various contraceptive techniques in ACHD patients. The estrogen-containing oral contraceptive pill is generally not recommended in ACHD patients at risk of thromboembolism, such as those with cyanosis, prior Fontan procedure, atrial fibrillation, or PAH. In addition, this form of contraceptive therapy may upset anticoagulation control. However, medroxyprogesterone, the progesterone-only pills, and levonorgestrel may also cause fluid retention and should be used with caution in patients with heart failure. Depression and breakthrough bleeding may prevent the use of the progesterone-only pills, and there is a higher failure rate than with combined oral contraceptives.

Levonorgestrel, barrier methods, and tubal ligation are the recommended contraceptive methods for women with cyanotic CHD and PAH. The potential complications of the "morning after pill" (levonorgestrel "plan B") should be explained to those at risk of acute fluid retention. Tubal ligation, although the most secure method of contraception, can be a high-risk procedure in patients with complex CHD or those with PAH. Hysteroscopic sterilization (Essure) may be reasonable for high-risk patients (55). Sterilization of a male partner of a woman with CHD should only occur after full explanation of the prognosis to the patient. The specialist in the ACHD clinic needs to interact with both the general practitioner and the gynecologist to provide optimal advice regarding contraception. The risk of endocarditis with intrauterine devices in women with CHD is controversial, and recommendations should be individualized on the basis of discussions between the cardiologist and gynecologist.

Breast feeding is safe in women with CHD. Women requiring cardiovascular medications should be aware that many of the medications will cross into breast milk and should clarify the potential effect of medications on the infant with a pediatrician.

1.9. Recommendations for Arrhythmia Diagnosis and Management

CLASS I

1. **Complete and appropriate noninvasive testing, as well as clear knowledge of the specific anatomy and review of all surgical and procedural records, is recommended before electrophysiological testing or device placement is attempted in ACHD patients. (*Level of Evidence: C*)**
2. **Decisions regarding tachycardia management in ACHD patients should take into account the broad cardiovascular picture, particularly repairable hemodynamic issues that might favor a surgical or catheter-based approach to treatment. (*Level of Evidence: B*)**
3. **Catheter ablation procedures for ACHD patients should be performed at centers where the staff is experienced with the complex anatomy and distinctive arrhythmia substrates encountered in congenital heart defects. (*Level of Evidence: B*)**
4. **Pacemaker and device lead placement (or replacement) in ACHD patients should be performed at centers where the staff is familiar with the unusual anatomy of congenital heart defects and their surgical repair. (*Level of Evidence: B*)**
5. **Epicardial pacemaker and device lead placement should be performed in all cyanotic patients with intracardiac shunts who require devices. (*Level of Evidence: B*)**

CLASS IIa

1. **It is reasonable to recommend the use of an implantable cardioverter defibrillator for any patient who has had a cardiac arrest or experienced an episode of hemodynamically significant or sustained ventricular tachycardia. (*Level of Evidence: C*)**
2. **Pacemaker implantation can be beneficial in ACHD patients with bradyarrhythmias and may be helpful in overdrive pacing in patients with difficult-to-control tachyarrhythmias (see ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities) (56). (*Level of Evidence: B*)**

CLASS IIb

1. **Pacemaker implantation may be beneficial for asymptomatic adult patients with resting heart rates of less than 40 beats per minute or abrupt pauses in excess of 3 seconds. (*Level of Evidence: C*)**

Cardiac arrhythmias are a major source of morbidity and mortality for ACHD patients (Table 8). Although rhythm disorders can often be observed in adults with unrepaired or palliated defects, the most difficult cases usually involve patients who have undergone prior intracardiac repairs, especially when this reparative surgery was performed relatively late in life (57,58). In this setting, the electrical pathology stems from the unique and complex myocardial substrates created by septal patches and suture lines in combination with cyanosis and abnormal pressure/volume status of variable duration. Virtually the entire spectrum of rhythm disturbances is manifested in these patients, including some disorders that are specific to the anatomic defect or the surgical technique used for repair.

1.10. Cyanotic Congenital Heart Disease

Right-to-left intracardiac or extracardiac shunts result in hypoxemia, erythrocytosis, and cyanosis.

1.10.1. Recommendations for Hematologic Problems

CLASS I

1. **Indications for therapeutic phlebotomy are hemoglobin greater than 20 g per dL and hematocrit greater than 65%, associated with headache, increasing fatigue, or other symptoms of hyperviscosity in the absence of dehydration or anemia. (*Level of Evidence: C*)**

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Table 8. Rhythm Disturbances in Adults With Congenital Heart Disease

Rhythm Disturbance	Associated Lesions
Tachycardias	
Wolff-Parkinson-White syndrome	Ebstein's anomaly
	Congenitally corrected transposition
Intra-atrial reentrant tachycardia (atrial flutter)	Postoperative Mustard
	Postoperative Senning
	Postoperative Fontan
	Tetralogy of Fallot
	Other
Atrial fibrillation	Mitral valve disease
	Aortic stenosis
	Tetralogy of Fallot
	Palliated single ventricle
Ventricular tachycardia	Tetralogy of Fallot
	Aortic stenosis
	Other
Bradycardias	
Sinus node dysfunction	Postoperative Mustard
	Postoperative Senning
	Postoperative Fontan
	Sinus venosus ASD
	Heterotaxy syndrome
Spontaneous AV block	AV septal defects
	Congenitally corrected transposition
Surgically induced AV block	VSD closure
	Subaortic stenosis relief
	AV valve replacement

AV indicates atrioventricular; ASD, atrial septal defect; and VSD, ventricular septal defect.

CLASS III

1. Repeated routine phlebotomies are not recommended because of the risk of iron depletion, decreased oxygen-carrying capacity, and stroke. (*Level of Evidence: C*)

Cyanosis in patients with CHD has profound hematologic consequences that may affect many organ systems and need to be recognized and managed appropriately. The hematologic complications of chronic hypoxemia are erythrocytosis, iron deficiency, and bleeding diathesis (59). The increase in red blood cell mass that accompanies cyanosis is a compensatory response to improve oxygen transport. The white blood cell count is usually normal, and the platelet count may be normal or reduced.

The increased red blood cell mass may result in an increase in blood viscosity. However, the most likely cause of complications in adults with cyanotic CHD is aggressive phlebotomy or blood loss (60). Most cyanotic patients have compensated erythrocytosis with stable hemoglobin that requires no intervention.

The treatment for iron deficiency in a patient with destabilized erythropoiesis is challenging. Oral administration of iron frequently results in a rapid and dramatic increase in red cell mass; therefore, caution should be exercised and hemoglobin monitored. Once the serum ferritin and/or transferrin saturation is within the normal range, iron supplementation may be discontinued.

1.10.1.1. Hemostasis

Hemostatic abnormalities have been documented in up to 20% of cyanotic patients. Platelet dysfunction and clotting factor deficiencies combine to produce a bleeding tendency in these patients. Epistaxis, gingival bleeding, menorrhagia, and pulmonary hemorrhage are the most common causes of bleeding. The use of anticoagulants and antiplatelet agents, therefore, is controversial and confined to well-defined indications with careful monitoring of the degree of anticoagulation.

1.10.1.2. Renal Function

In chronic cyanosis, the renal glomeruli are abnormal, frequently hypercellular, and congested and eventually become sclerotic (61). This results in a reduction of the glomerular filtration rate, increased creatinine levels, and proteinuria. This may cause problems with radiopaque contrast material and dehydration, leading to uremia, oliguria, and even anuria. Thus, patients should be hydrated before procedures that involve contrast media.

1.10.1.3. Gallstones

The increased breakdown of red blood cells in chronic cyanosis results in an increased risk of calcium bilirubinate gallstones.

1.10.1.4. Orthopedic and Rheumatologic Complications

Hypertrophic osteoarthropathy with thickened, irregular periosteum occurs in the setting of cyanotic CHD. Scoliosis occurs in a high percentage of patients with cyanotic CHD and is occasionally severe enough to compromise pulmonary function and require surgical intervention. Preoperative evaluation by an ACHD cardiologist and cardiac anesthesiologist is recommended before the operation for scoliosis is undertaken because of the recognized increased risk of surgery in cyanotic patients, especially those with PAH, for whom this procedure may be contraindicated.

1.10.1.5. Neurological Complications

Neurological complications include an increased risk for paradoxical cerebral emboli. Brain abscess in cyanotic patients and thromboembolic events in patients with atrial tachycardia or atrial stasis associated with transvenous pacing leads can result in new neurological symptoms. These complications should be suspected in a cyanotic patient with headache, fever, and new neurological symptoms. Substantial cognitive and psychosocial issues are prevalent in this population, as discussed in Section 1.5.2, Recommendations for Psychosocial Issues.

1.11. Recommendations for General Health Issues for Cyanotic Patients

CLASS I

1. Cyanotic patients should drink nonalcoholic and noncaffeinated fluids frequently on long-distance flights to avoid dehydration. (*Level of Evidence: C*)

CLASS IIb

1. **Supplemental oxygenation may be considered for cyanotic patients during long-distance flights. (*Level of Evidence: C*)**

Cyanotic patients should use only pressurized commercial airplanes. Oxygen therapy, although often unnecessary, may be suggested for prolonged travel. Similarly, residence at high altitude is detrimental for patients with cyanosis. Dehydration should be avoided by frequent fluid intake on long flights and during sports activities.

Competitive sports should be avoided in cyanotic patients (62). Cyanosis is a recognized handicap to fetal growth and development, and pregnancy outcome is impacted, with an increased risk of congestive heart failure, preterm delivery, intrauterine growth retardation, and miscarriage. Increased maternal and fetal mortality are also noted and correlate with the degree of cyanosis, ventricular dysfunction, and pulmonary pressures (54).

1.11.1. Hospitalization and Operation

Cyanotic patients are at high risk during any hospitalization or operation. Management strategies that should be applied include those likely to reduce the risk of paradoxical emboli related to air in the intravenous lines. Medication adjustment may be needed, with cyanosis taken into account.

1.12. Recommendations for Heart and Heart/Lung Transplantation

CLASS I

1. **Patients with CHD and heart failure who may require heart transplantation should be evaluated and managed in tertiary care centers with medical and surgical personnel with experience and expertise in the management of both CHD and heart transplantation. (*Level of Evidence: C*)**
2. **Patients with CHD and heart or respiratory failure who may require lung or heart/lung transplantation should be evaluated and managed in tertiary care centers with medical and surgical personnel with experience and expertise in the management of CHD and lung or heart/lung transplantation. (*Level of Evidence: C*)**

The pretransplantation evaluation involves a multidisciplinary approach that addresses assessment of cardiopulmonary, renal, neurological, hepatic, infectious disease, socioeconomic, and psychological issues. In addition to history and physical examination, diagnostic studies include ECG, echocardiography, chest x-ray, and Holter monitoring. Cardiac catheterization is required to assess pulmonary vascular resistance (PVR) and transpulmonary gradient (63). In addition to cardiac catheterization, MRI or CT angiography is often performed to delineate the anatomy in patients with complex CHD (eg, patients with malpositioning of the great arteries and/or substernal position of an extracardiac conduit, abnormalities of systemic venous return, and situs abnormalities).

Many patients with long-standing heart failure may have elevated PVR. Consequently, donor right-sided heart failure may result when the heart is abruptly placed proximal to such a high-resistance pulmonary vascular bed.

Heart/lung transplantation is usually reserved for patients with uncorrectable or previously repaired or palliated CHD associated with significant pulmonary vascular obstructive disease, such as single-ventricle physiology with pulmonary vascular disease or left ventricular (LV) dysfunction with associated pulmonary vascular disease. When a simple cardiac defect is present, such as atrial septal defect (ASD), ventricular septal defect (VSD), or patent ductus arteriosus (PDA), the cardiac defect can often be repaired at lung transplantation (64). In the presence of more complex intracardiac abnormalities, combined heart/lung transplantation is usually most appropriate.

2. Atrial Septal Defect

One of the most common adult congenital heart defects, an ASD is a persistent communication between the atria. There are several different types of ASD: the secundum ASD in the region of the fossa ovalis; the primum ASD, positioned inferiorly near the crux of the heart; the sinus venosus ASD, located superiorly near the superior vena caval entry or inferiorly near the inferior vena caval entry; and the uncommon coronary sinus septal defect, which causes shunting through the ostium of the coronary sinus.

2.1. Unrepaired Atrial Septal Defect

The consequence of a left-to-right shunt across an ASD is right ventricular (RV) volume overload and pulmonary overcirculation. Large atrial shunts lead to symptoms from excess pulmonary blood flow and right-sided heart failure, including frequent pulmonary infections, fatigue, exercise intolerance, and palpitations. Atrial arrhythmias—atrial flutter, atrial fibrillation, and sick sinus syndrome—are a common result of long-standing right-sided heart volume and pressure overload. Flow-related PAH accompanies large left-to-right shunts, and pulmonary vascular obstructive disease may develop in adult years but occurs much later with ASD than with high-pressure left-to-right shunts such as VSD or PDA.

2.2. Recommendations for Evaluation of the Unoperated Patient

CLASS I

1. **ASD should be diagnosed by imaging techniques with demonstration of shunting across the defect and evidence of RV volume overload and any associated anomalies. (*Level of Evidence: C*)**
2. **Patients with unexplained RV volume overload should be referred to an ACHD center for further diagnostic studies to rule out obscure ASD, partial anomalous venous connection, or coronary sinoseptal defect. (*Level of Evidence: C*)**

CLASS IIa

1. **Maximal exercise testing can be useful to document exercise capacity in patients with symptoms that are discrepant with clinical findings or to document changes in oxygen saturation in patients with mild or moderate PAH. (*Level of Evidence: C*)**
2. **Cardiac catheterization can be useful to rule out concomitant coronary artery disease in patients at risk because of age or other factors. (*Level of Evidence: B*)**

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CLASS III

1. In younger patients with uncomplicated ASD for whom noninvasive imaging results are adequate, diagnostic cardiac catheterization is not indicated. (*Level of Evidence: B*)
2. Maximal exercise testing is not recommended in ASD with severe PAH. (*Level of Evidence: B*)

The diagnostic workup for a patient with a suspected ASD is directed at defining the presence, size, and location of the ASD; the functional effect of the shunt on the right and left ventricles and the pulmonary circulation; and any associated lesions.

2.3. Management Strategies

2.3.1. Recommendations for Medical Therapy

CLASS I

1. Cardioversion after appropriate anticoagulation is recommended to attempt restoration of the sinus rhythm if atrial fibrillation occurs. (*Level of Evidence: A*)
2. Rate control and anticoagulation are recommended if sinus rhythm cannot be maintained by medical or interventional means. (*Level of Evidence: A*)

Patients with small shunts and normal RV size are generally asymptomatic and require no medical therapy. Routine follow-up of the patient with a small ASD without evidence of RV enlargement or PAH should include assessment of symptoms, especially arrhythmias and possible paradoxical embolic events. A repeat echocardiogram should be obtained every 2 to 3 years to assess RV size and function and pulmonary pressure. Reductions in LV compliance related to hypertension, coronary artery disease, or acquired valvular disease increase the degree of left-to-right shunt across an existing ASD.

Atrial arrhythmias should be treated to restore and maintain sinus rhythm if possible (65). If atrial fibrillation occurs, both antiarrhythmic therapy and anticoagulation should be recommended.

2.3.2. Recommendations for Interventional and Surgical Therapy

CLASS I

1. Closure of an ASD either percutaneously or surgically is indicated for right atrial and RV enlargement with or without symptoms. (*Level of Evidence: B*)
2. A sinus venosus, coronary sinus, or primum ASD should be repaired surgically rather than by percutaneous closure. (*Level of Evidence: B*)
3. Surgeons with training and expertise in CHD should perform operations for various ASD closures. (*Level of Evidence: C*)

CLASS IIa

1. Surgical closure of secundum ASD is reasonable when concomitant surgical repair/replacement of a tricuspid valve is considered or when the anatomy of the defect precludes the use of a percutaneous device. (*Level of Evidence: C*)
2. Closure of an ASD, either percutaneously or surgically, is reasonable in the presence of:
   a. Paradoxical embolism. (*Level of Evidence: C*)
   b. Documented orthodeoxia-platypnea. (*Level of Evidence: B*)

CLASS IIb

1. Closure of an ASD, either percutaneously or surgically, may be considered in the presence of net left-to-right shunting, pulmonary artery pressure less than two thirds systemic levels, PVR less than two thirds systemic vascular resistance, or when responsive to either pulmonary vasodilator therapy or test occlusion of the defect (patients should be treated in conjunction with providers who have expertise in the management of pulmonary hypertensive syndromes). (*Level of Evidence: C*)
2. Concomitant Maze procedure may be considered for intermittent or chronic atrial tachyarrhythmias in adults with ASDs. (*Level of Evidence: C*)

CLASS III

1. Patients with severe irreversible PAH and no evidence of a left-to-right shunt should not undergo ASD closure. (*Level of Evidence: B*)

2.3.3. Indications for Closure of Atrial Septal Defect

Small ASDs with a diameter of less than 5 mm and no evidence of RV volume overload do not impact the natural history of the individual and thus may not require closure unless associated with paradoxical embolism. Larger defects with evidence of RV volume overload on echocardiography usually only cause symptoms in the third decade of life, and closure is usually indicated to prevent long-term complications such as atrial arrhythmias, reduced exercise tolerance, hemodynamically significant tricuspid regurgitation (TR), right-to-left shunting and embolism during pregnancy, overt congestive cardiac failure, or pulmonary vascular disease that may develop in up to 5% to 10% of affected (mainly female) individuals.

The majority of secundum ASDs can be closed with a percutaneous catheter technique. When this is not feasible or is not appropriate, surgical closure is recommended.

Sinus venosus, coronary sinus, and primum defects are not amenable to device closure. An ASD with a large septal aneurysm or a multifenestrated atrial septum requires careful evaluation by and consultation with interventional cardiologists before device closure is selected as the method of repair.

2.4. Recommendations for Postintervention Follow-Up

CLASS I

1. Early postoperative symptoms of undue fever, fatigue, vomiting, chest pain, or abdominal pain may represent postpericardiotomy syndrome with tamponade and should prompt immediate evaluation with echocardiography. (*Level of Evidence: C*)
2. Annual clinical follow-up is recommended for patients postoperatively if their ASD was repaired as an adult and the following conditions persist or develop:
   a. PAH. (*Level of Evidence: C*)
   b. Atrial arrhythmias. (*Level of Evidence: C*)
   c. RV or LV dysfunction. (*Level of Evidence: C*)
   d. Coexisting valvular or other cardiac lesions. (*Level of Evidence: C*)
3. Evaluation for possible device migration, erosion, or other complications is recommended for patients 3 months to 1 year after device closure and periodically thereafter. (*Level of Evidence: C*)

4. **Device erosion, which may present with chest pain or syncope, should warrant urgent evaluation. (*Level of Evidence: C*)**

Follow-up for patients after device closure requires clinical assessment of symptoms of arrhythmia, chest pain, or embolic events and echocardiographic surveillance for device position, residual shunting, and complications such as thrombus formation or pericardial effusion. Pericardial effusions and cardiac tamponade may occur up to several weeks after surgical repair of ASDs and should be evaluated by clinical examination and echocardiography before hospital discharge and at the early postoperative visits. Assessment of pulmonary pressure, RV function, and residual atrial shunting should also be made during follow-up echocardiography. Clinical and ECG surveillance for recurrent or new-onset arrhythmia is an important feature of postoperative evaluation.

2.4.1. Recommendation for Reproduction

CLASS III

1. **Pregnancy in patients with ASD and severe PAH (Eisenmenger syndrome) is not recommended owing to excessive maternal and fetal mortality and should be strongly discouraged. (*Level of Evidence: A*)**

Women with large shunts and PAH may experience arrhythmias, ventricular dysfunction, and progression of PAH. Pregnancy in patients with ASD and severe PAH (Eisenmenger syndrome) is contraindicated owing to excessive maternal and fetal mortality and should be strongly discouraged (66,67). Paradoxical embolism may occasionally be encountered in small and large ASDs (53,68).

3. Ventricular Septal Defect

VSD is the most common congenital heart defect at birth (69) and presents in approximately 3.0 to 3.5 infants per 1000 live births. Although VSD is most often an isolated lesion, it is a common component of complex abnormalities such as conotruncal defects (eg, tetralogy of Fallot and TGA). VSD can also be associated with left-sided obstructive lesions such as subaortic stenosis (SubAS) and coarctation of the aorta. A subpulmonary (supracristal) VSD is often associated with progressive aortic valve regurgitation caused by prolapse of the aortic cusp (usually right) through the defect.

It is unlikely for an adult with an isolated VSD to present with no prior workup/diagnosis. Possible scenarios include the following:

- An asymptomatic patient with a systolic murmur previously thought to be an innocent murmur
- Fever and bacteremia secondary to IE
- A new diastolic murmur of aortic regurgitation (AR) secondary to aortic valve prolapse
- Cyanosis and exercise intolerance secondary to the progressive development of pulmonary vascular disease.

3.1. Recommendations for Cardiac Catheterization

CLASS I

1. **Cardiac catheterization to assess the operability of adults with VSD and PAH should be performed in an ACHD regional center in collaboration with experts. (*Level of Evidence: C*)**

CLASS IIa

1. **Cardiac catheterization can be useful for adults with VSD in whom noninvasive data are inconclusive and further information is needed for management. Data to be obtained include the following:**
   a. **Quantification of shunting. (*Level of Evidence: B*)**
   b. **Assessment of pulmonary pressure and resistance in patients with suspected PAH. Reversibility of PAH should be tested with various vasodilators. (*Level of Evidence: B*)**
   c. **Evaluation of other lesions such as AR and double-chambered right ventricle. (*Level of Evidence: C*)**
   d. **Determination of whether multiple VSDs are present before surgery. (*Level of Evidence: C*)**
   e. **Performance of coronary arteriography is indicated in patients at risk for coronary artery disease. (*Level of Evidence: C*)**
   f. **VSD anatomy, especially if device closure is contemplated. (*Level of Evidence: C*)**

3.2. Management Strategies

3.2.1. Recommendation for Medical Therapy

CLASS IIb

1. **Pulmonary vasodilator therapy may be considered for adults with VSDs with progressive/severe pulmonary vascular disease (refer to Section 9, Pulmonary Hypertension/Eisenmenger Physiology). (*Level of Evidence: B*)**

3.2.2. Recommendations for Surgical Ventricular Septal Defect Closure

CLASS I

1. **Surgeons with training and expertise in CHD should perform VSD closure operations. (*Level of Evidence: C*)**
2. **Closure of a VSD is indicated when there is a Qp/Qs (pulmonary–to–systemic blood flow ratio) of 2.0 or more and clinical evidence of LV volume overload. (*Level of Evidence: B*)**
3. **Closure of a VSD is indicated when the patient has a history of IE. (*Level of Evidence: C*)**

CLASS IIa

1. **Closure of a VSD is reasonable when net left-to-right shunting is present at a Qp/Qs greater than 1.5 with pulmonary artery pressure less than two thirds of systemic pressure and PVR less than two thirds of systemic vascular resistance. (*Level of Evidence: B*)**
2. **Closure of a VSD is reasonable when net left-to-right shunting is present at a Qp/Qs greater than 1.5 in the presence of LV systolic or diastolic failure. (*Level of Evidence: B*)**

CLASS III

1. **VSD closure is not recommended in patients with severe irreversible PAH. (*Level of Evidence: B*)**

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3.2.3. Recommendation for Interventional Catheterization

CLASS IIb

1. **Device closure of a muscular VSD may be considered, especially if the VSD is remote from the tricuspid valve and the aorta, if the VSD is associated with severe left-sided heart chamber enlargement, or if there is PAH. (*Level of Evidence: C*)**

Indications for catheter device closure of VSD include residual defects after prior attempts at surgical closure, restrictive VSDs with a significant left-to-right shunt, trauma, or iatrogenic artifacts after surgical replacement of the aortic valve. Indications for closure of restrictive VSDs in the adult population include a history of bacterial endocarditis or a hemodynamically significant left-to-right shunt (Qp/Qs greater than 1.5:1).

3.3. Key Issues to Evaluate and Follow-Up

3.3.1. Recommendations for Surgical and Catheter Intervention Follow-Up

CLASS I

1. **Adults with VSD with residual heart failure, shunts, PAH, AR, or RV outflow tract (RVOT) or LV outflow tract (LVOT) obstruction should be seen at least annually at an ACHD regional center. (*Level of Evidence: C*)**
2. **Adults with a small residual VSD and no other lesions should be seen every 3 to 5 years at an ACHD regional center. (*Level of Evidence: C*)**
3. **Adults with device closure of a VSD should be followed up every 1 to 2 years at an ACHD center depending on the location of the VSD and other factors. (*Level of Evidence: C*)**

Adults with no residual VSD, no associated lesions, and normal pulmonary artery pressure do not require continued follow-up at a regional ACHD center except on referral from the patient's cardiologist or physician. Patients who develop bifascicular block or transient trifascicular block after VSD closure are at risk in later years for the development of complete heart block and should be followed up yearly by history and ECG and have periodic ambulatory monitoring and/or exercise testing.

3.3.2. Recommendation for Reproduction

CLASS III

1. **Pregnancy in patients with VSD and severe PAH (Eisenmenger syndrome) is not recommended owing to excessive maternal and fetal mortality and should be strongly discouraged. (*Level of Evidence: A*)**

Women with small VSDs, no PAH, and no associated lesions have no increased cardiovascular risk for pregnancy. Women with PAH should be counseled against pregnancy (refer to Section 9, Pulmonary Hypertension/Eisenmenger Physiology).

Pregnancy is generally well tolerated, with no maternal mortality and no significant maternal or fetal morbidity. Although the left-to-right shunt may increase with the increase in cardiac output during pregnancy, this is counterbalanced by the decrease in peripheral resistance. Women with large shunts and PAH may experience arrhythmias, ventricular dysfunction, and progression of PAH.

4. Atrioventricular Septal Defect

The terms atrioventricular septal defect (AVSD), atrioventricular (AV) canal defect, and endocardial cushion defect can be used interchangeably to describe this group of defects. Tetralogy of Fallot and other conotruncal anomalies and heterotaxy syndromes also occur in association with AVSD.

4.1. Recommendation for Heart Catheterization

CLASS IIa

1. **Cardiac catheterization is reasonable to assess PAH and test vasoreactivity in patients with repaired or unrepaired AVSD. (*Level of Evidence: B*)**

Heart catheterization has a limited role in the assessment of these patients unless noninvasive findings are equivocal. Evaluation of PAH and coronary anatomy may be needed when reoperation is being considered. Hemodynamic data may also be needed when noninvasive studies have not been able to provide this information.

4.2. Recommendations for Surgical Therapy

CLASS I

1. **Surgeons with training and expertise in CHD should perform operations for AVSD. (*Level of Evidence: C*)**
2. **Surgical reoperation is recommended in adults with previously repaired AVSD with the following indications:**
   a. **Left AV valve repair or replacement for regurgitation or stenosis that causes symptoms, atrial or ventricular arrhythmias, a progressive increase in LV dimensions, or deterioration of LV function. (*Level of Evidence: B*)**
   b. **LVOT obstruction with a mean gradient greater than 50 mm Hg or peak instantaneous gradient greater than 70 mm Hg, or a gradient less than 50 mm Hg in association with significant mitral regurgitation or AR. (*Level of Evidence: B*)**
   c. **Residual/recurrent ASD or VSD with significant left-to-right shunting (refer to Section 2.0, Atrial Septal Defect, and Section 3.0, Ventricular Septal Defect). (*Level of Evidence: B*)**

Primary operation is rarely recommended for complete AVSD in adults because of pulmonary vascular obstructive disease. Unoperated partial or transitional AVSD, also known as partial or transitional AV canal, may not be identified until adulthood. Primary repair is generally recommended provided there is no fixed PAH.

4.3. Recommendations for Endocarditis Prophylaxis

CLASS IIa

1. **Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of teeth**

or perforation of the oral mucosa is reasonable in patients with CHD with the highest risk for adverse outcome from IE, including those with the following indications:
   a. Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: B*)
   b. Previous IE. (*Level of Evidence: B*)
   c. Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: B*)
   d. Completely repaired CHD with prosthetic materials, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure. (*Level of Evidence: B*)
   e. Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization. (*Level of Evidence: B*)
2. It is reasonable to consider antibiotic prophylaxis against IE before vaginal delivery at the time of membrane rupture in select patients with the highest risk of adverse outcomes. This includes patients with the following indications:
   a. Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: C*)
   b. Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: C*)

CLASS III

1. Prophylaxis against IE is not recommended for nondental procedures (such as esophagogastroduodenoscopy or colonoscopy) in the absence of active infection. (*Level of Evidence: C*)

4.4. Recommendations for Pregnancy

CLASS I

1. All women with a history of AVSD should be evaluated before conception to ensure that there are no significant residual hemodynamic lesions that might complicate the management of pregnancy. (*Level of Evidence: C*)
2. The issue of pregnancy risk and preventive measures should be discussed with women with Down syndrome and their caregivers. (*Level of Evidence: C*)

Pregnancy is usually well tolerated by women who have had repair and who have no major residua, as well as by women with a primum defect who are functionally well. Pregnancy is not advised for women with severe PAH.

5. Patent Ductus Arteriosus

5.1. Recommendations for Evaluation of the Unoperated Patient

CLASS I

1. Definitive diagnosis of PDA should be based on visualization by imaging techniques and demonstrations of the shunting across the defect (with or without evidence of clinically significant LV volume overload). (*Level of Evidence: C*)

CLASS III

1. Diagnostic cardiac catheterization is not indicated for uncomplicated PDA with adequate noninvasive imaging. (*Level of Evidence: B*)
2. Maximal exercise testing is not recommended in PDA with significant PAH. (*Level of Evidence: B*)

The diagnostic workup for a patient with a suspected PDA is directed at defining the presence and size of the PDA, the functional effect of the shunt on the left atrium and left ventricle, the pulmonary circulation, and any associated lesions.

5.2. Management Strategies

5.2.1. Recommendations for Medical Therapy

CLASS I

1. Routine follow-up is recommended for patients with a small PDA without evidence of left-sided heart volume overload. Follow-up is recommended every 3 to 5 years for patients with a small PDA without evidence of left-heart volume overload. (*Level of Evidence: C*)

CLASS III

1. Endocarditis prophylaxis is not recommended for those with a repaired PDA without residual shunt. (*Level of Evidence: C*)

5.2.2. Recommendations for Closure of Patent Ductus Arteriosus

CLASS I

1. Closure of a PDA either percutaneously or surgically is indicated for the following:
   a. Left atrial and/or LV enlargement or if PAH is present, or in the presence of net left-to-right shunting. (*Level of Evidence: C*)
   b. Prior endarteritis. (*Level of Evidence: C*)
2. Careful evaluation and consultation with ACHD interventional cardiologists is recommended before surgical closure is selected as the method of repair for patients with a calcified PDA. (*Level of Evidence: C*)
3. Surgical repair, by a surgeon experienced in CHD surgery, is recommended when:
   a. The PDA is too large for device closure. (*Level of Evidence: C*)
   b. Distorted ductal anatomy precludes device closure (eg, aneurysm or endarteritis) (42). (*Level of Evidence: B*)

CLASS IIa

1. It is reasonable to close an asymptomatic small PDA by catheter device. (*Level of Evidence: C*)
2. PDA closure is reasonable for patients with PAH with a net left-to-right shunt. (*Level of Evidence: C*)

CLASS III

1. PDA closure is not indicated for patients with PAH and net right-to-left shunt. (*Level of Evidence: C*)

5.2.3. Surgical/Interventional Therapy

Surgical closure of PDA in the adult may pose some problems due to the friability and/or calcification of the ductus, atherosclerosis, and aneurysm formation, as well as the presence of other unrelated comorbid conditions, such as coronary atherosclerosis or renal disease, that may adversely affect the perioperative risk. Adults with PDA are better suited for percutaneous closure with either the occlusion device or coils because of its high success and few compli-

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cations (70). If the PDA is associated with other conditions that require surgical correction, the ductus may be closed during the same operation, although percutaneous closure of the PDA before other cardiac surgery may decrease the risk of cardiopulmonary bypass.

5.3. Key Issues to Evaluate and Follow-Up

Adults with large PDAs are likely to have Eisenmenger physiology. Such patients require frequent follow-up to monitor their progress/deterioration. Problems associated with Eisenmenger physiology are discussed in Section 9, Pulmonary Hypertension/Eisenmenger Physiology.

Patients who have undergone surgical/PDA closure can be discharged safely from follow-up once complete closure of the ductus is documented by TTE. Antibiotic prophylaxis is discontinued 6 months after PDA closure. Follow-up approximately every 5 years for patients who received a device is recommended because of the lack of long-term data on device closure with the occlusion device.

6. Left-Sided Heart Obstructive Lesions: Aortic Valve Disease, Subvalvular and Supravalvular Aortic Stenosis, Associated Disorders of the Ascending Aorta, and Coarctation

LVOT obstruction syndromes include SubAS, valvular AS, SupraAS, and aortic coarctation (71). Obstruction can occur singly or at multiple levels, as an isolated lesion, or in combination with septal defects or conotruncal anomalies. Bicuspid aortic valve (BAV) is one of the most common congenital cardiovascular malformations, with an estimated incidence of 1% to 2% of the population.

6.1. Associated Lesions

Abnormalities associated with BAV disease include SubAS, parachute mitral valve, VSD, PDA, or coarctation of the aorta with varying degrees of arch hypoplasia. A left-dominant coronary artery system is more frequent with BAV (72). Turner syndrome may be associated with AS in addition to aortic coarctation. The presence of multiple levels of left-sided heart obstructions (eg, SubAS, BAV, AS, coarctation, parachute mitral valve, or supramitral ring) is termed Shones syndrome. Patients presenting in childhood with LVOT obstruction generally have more complex or severe disease than those found to have BAV in adult life. BAV disease can be associated with progressive dilation of the aortic root, aortic aneurysm, and even rupture or dissection; intrinsic abnormalities of aortic wall elastin may result in ascending aortic dilation even with a normally functioning aortic valve.

6.2. Recommendations for Evaluation of the Unoperated Patient

Recommendations and guidelines concerning AS, BAV, and AR in the adult patient are also discussed in the 2006 valvular heart disease guidelines (73).

CLASS I

1. **Primary imaging and hemodynamic assessment of AS and aortic valve disease are recommended by echocardiography-Doppler to evaluate the presence and severity of AS or AR; LV size, function, and mass; and dimensions and anatomy of the ascending aorta and associated lesions. (*Level of Evidence: B*)**
2. **Echocardiography is recommended for reevaluation of patients with AS who experience a change in signs or symptoms and for assessment of changes in AS hemodynamics during pregnancy. (*Level of Evidence: B*)**
3. **In asymptomatic adolescents and young adults, echocardiography-Doppler is recommended yearly for AS with a mean Doppler gradient greater than 30 mm Hg or peak instantaneous gradient greater than 50 mm Hg and every 2 years for patients with lesser gradients. (*Level of Evidence: C*)**
4. **Cardiac catheterization is recommended when noninvasive measurements are inconclusive or discordant with clinical signs. (*Level of Evidence: C*)**
5. **Coronary angiography is recommended before aortic valve surgery for coronary angiography in adults at risk for coronary artery disease. (*Level of Evidence: B*)**
6. **Coronary angiography is recommended before a Ross procedure if noninvasive imaging of the coronary arteries is inadequate. (*Level of Evidence: C*)**
7. **A yearly ECG is recommended in young adults less than 30 years of age with mean Doppler gradients greater than 30 mm Hg or peak Doppler gradients greater than 50 mm Hg. (*Level of Evidence: C*)**
8. **An ECG is recommended every other year in young adults less than 30 years of age with mean Doppler gradients less than 30 mm Hg or peak Doppler gradients less than 50 mm Hg. (*Level of Evidence: C*)**

CLASS IIa

1. **In asymptomatic young adults less than 30 years of age, exercise stress testing is reasonable to determine exercise capability, symptoms, and blood pressure response. (*Level of Evidence: C*)**
2. **Exercise stress testing is reasonable for patients with a mean Doppler gradient greater than 30 mm Hg or peak Doppler gradient greater than 50 mm Hg if the patient is interested in athletic participation or if clinical findings differ from noninvasive measurements. (*Level of Evidence: C*)**
3. **Exercise stress testing is reasonable for the evaluation of an asymptomatic young adult with a mean Doppler gradient greater than 40 mm Hg or a peak Doppler gradient greater than 64 mm Hg or when the patient anticipates athletic participation or pregnancy. (*Level of Evidence: C*)**
4. **Dobutamine stress testing can be beneficial in the evaluation of a mild aortic valve gradient in the face of low LV ejection fraction and reduced cardiac output. (*Level of Evidence: B*)**
5. **MRI/CT can be beneficial to add important information about the anatomy of the thoracic aorta. (*Level of Evidence: C*)**
6. **Exercise stress testing can be useful to evaluate blood pressure response or elicit exercise-induced symptoms in asymptomatic older adults with AS. (*Level of Evidence: B*)**

CLASS IIb

1. **Magnetic resonance angiography may be beneficial in quantifying AR when other data are ambiguous or borderline. (*Level of Evidence: C*)**

CLASS III

1. **Exercise stress testing should not be performed in symptomatic patients with AS or those with repolarization abnormality on ECG or systolic dysfunction on echocardiography. (*Level of Evidence: C*)**

6.3. Problems and Pitfalls

Problems and pitfalls regarding BAV stenosis include the following:

- The click murmur of a BAV may be misdiagnosed as mitral valve prolapse.
- A systolic murmur may be thought to be "benign" because an ejection click is not recognized.
- To quantify the severity of valvular AS by echocardiography-Doppler, mean gradient and aortic valve area should be used rather than relying only on peak systolic gradient, which may overestimate the severity of stenosis. The aortic valve area should be indexed to body surface area to correct for different body sizes and habitus.
- Progressive aortic dilatation may occur in patients with BAV even in the absence of significant AS or AR.
- In the presence of increased LV dimensions and normal wall thickness, an increased LV mass is present. LV mass calculations are needed and should be indexed to body surface area (74).

6.4. Management Strategies for Left Ventricular Outflow Tract Obstruction and Associated Lesions

6.4.1. Recommendations for Medical Therapy

CLASS IIa

1. **It is reasonable to treat systemic hypertension in patients with AS while monitoring diastolic blood pressure to avoid reducing coronary perfusion. (*Level of Evidence: C*)**
2. **It is reasonable to administer beta blockers in patients with BAV and aortic root dilatation. (*Level of Evidence: C*)**
3. **It is reasonable to use long-term vasodilator therapy in patients with AR and systemic hypertension while carefully monitoring diastolic blood pressure to avoid reducing coronary perfusion. (*Level of Evidence: C*)**

CLASS IIb

1. **It may be reasonable to treat patients with BAV and risk factors for atherosclerosis with statins with the aim of "slowing down degenerative changes in the aortic valve and preventing atherosclerosis. (*Level of Evidence: C*)**

CLASS III

1. **Vasodilator therapy is not indicated for long-term therapy in AR for the following:**
   a. **The asymptomatic patient with only mild to moderate AR and normal LV function. (*Level of Evidence: B*)**
   b. **The asymptomatic patient with LV systolic dysfunction who is otherwise a candidate for aortic valve replacement (AVR). (*Level of Evidence: B*)**
   c. **The asymptomatic patient with either LV systolic function or mild to moderate LV diastolic dysfunction who is otherwise a candidate for AVR. (*Level of Evidence: C*)**

There are currently no established medical treatments proven to alter the natural history or halt the progression of stenosis in BAV disease (refer to Section 1.6, Recommendations for Infective Endocarditis, for additional information). Beta-blockers may be administered to delay or prevent aortic root dilatation or progression, but benefit has only been validated in patients with Marfan syndrome or acute aortic dissections. Judicious afterload reduction in patients with hypertension to reduce systolic blood pressure and lower LV wall tension may delay onset of LV dilatation or dysfunction but should be balanced against the risk of reducing diastolic coronary perfusion. There is no clear evidence that afterload reduction decreases the volume of AR or reduces the need for AVR (75). Multimodality molecular imaging has identified proteolytic and osteogenic activity in early aortic valve disease, a precursor to atherosclerotic and calcific degenerative AS (76). Thus, statins may slow the progression of acquired or calcific degenerative AS and probably have a role in the treatment of BAV disease, early in the process, before significant calcification and AS or AR have developed (77). Although no clinical trials have confirmed the benefits of statins in BAV disease, it appears reasonable to treat those patients who have risk factors for atherosclerosis.

6.4.2. Catheter and Surgical Intervention

6.4.2.1. Recommendations for Catheter Interventions for Adults With Valvular Aortic Stenosis

CLASS I

1. **In young adults and others without significantly calcified aortic valves and no AR, aortic balloon valvotomy is indicated in the following patients:**
   a. **Those with symptoms of angina, syncope, dyspnea on exertion, and peak-to-peak gradients at catheterization greater than 50 mm Hg. (*Level of Evidence: C*)**
   b. **Asymptomatic adolescents or young adults who demonstrate ST or T-wave abnormalities in the left precordial leads on ECG at rest or with exercise and a peak-to-peak catheter gradient greater than 60 mm Hg. (*Level of Evidence: C*)**

CLASS IIa

1. **Aortic balloon valvotomy is reasonable in the asymptomatic adolescent or young adult with AS and a peak-to-peak gradient on catheterization greater than 50 mm Hg when the patient is interested in playing competitive sports or becoming pregnant. (*Level of Evidence: C*)**

CLASS IIb

1. **Aortic balloon valvotomy may be considered as a bridge to surgery in hemodynamically unstable adults with AS, adults at high risk for AVR, or when AVR cannot be performed secondary to significant comorbidities. (*Level of Evidence: C*)**

CLASS III

1. **In older adults, aortic balloon valvotomy is not recommended as an alternative to AVR, although certain younger patients may be an exception and should be referred to a center with experience in aortic balloon valvuloplasties. (*Level of Evidence: B*)**
2. **In asymptomatic adolescents and young adults, aortic balloon valvotomy should not be performed with a peak-to-peak gradi-**

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ent less than 40 mm Hg without symptoms or ECG changes. (*Level of Evidence: B*)

6.4.2.2. Recommendations for Aortic Valve Repair/Replacement and Aortic Root Replacement

CLASS I

1. Aortic valvuloplasty, AVR, or Ross repair is indicated in patients with severe AS or chronic severe AR while they undergo coronary artery bypass grafting surgery on the aorta, or surgery on other heart valves. (*Level of Evidence: C*)
2. AVR is indicated for patients with severe AS and LV dysfunction (LV ejection fraction less than 50%). (*Level of Evidence: C*)
3. AVR is indicated in adolescents or young adults with severe AR who have:
   a. Development of symptoms. (*Level of Evidence: C*)
   b. Development of persistent LV dysfunction (LV ejection fraction less than 50%) or progressive LV dilatation (LV end-diastolic diameter 4 standard deviations above normal). (*Level of Evidence: C*)
4. Surgery to repair or replace the ascending aorta in a patient with a BAV is recommended when the ascending aorta diameter is 5.0 cm or more or when there is progressive dilation at a rate greater than or equal to 5 mm per year (73). (*Level of Evidence: B*)

CLASS IIa

1. AVR is reasonable for asymptomatic patients with severe AR and normal systolic function (ejection fraction greater than 50%) but with severe LV dilatation (LV end-diastolic diameter greater than 75 mm or end-systolic dimension greater than 55 mm*). (*Level of Evidence: B*)
2. Surgical aortic valve repair or replacement is reasonable in patients with moderate AS undergoing coronary artery bypass grafting or other cardiac or aortic root surgery. (*Level of Evidence: B*)

CLASS IIb

1. AVR may be considered for asymptomatic patients with any of the following indications:
   a. Severe AS and abnormal response to exercise. (*Level of Evidence: C*)
   b. Evidence of rapid progression of AS or AR. (*Level of Evidence: C*)
   c. Mild AS while undergoing coronary artery bypass grafting or other cardiac surgery and evidence of a calcific aortic valve. (*Level of Evidence: C*)
   d. Extremely severe AS (aortic valve area less than 0.6 cm and/or mean Doppler systolic AV gradient greater than 60 mm Hg) in an otherwise good operative candidate. (*Level of Evidence: C*)
   e. Moderate AR undergoing coronary artery bypass grafting or other cardiac surgery. (*Level of Evidence: C*)
   f. Severe AR with rapidly progressive LV dilation, when the degree of LV dilation exceeds an end-diastolic dimension of 70 mm or end-systolic dimension of 50 mm, with declining exercise tolerance, or with abnormal hemodynamic response to exercise. (*Level of Evidence: C*)
2. Surgical repair may be considered in adults with AS or AR and concomitant ascending aortic dilatation (ascending aorta diameter greater than 4.5 cm) coexisting with AS or AR. (*Level of Evidence: B*)
3. Early surgical repair may be considered in adults with the following indications:
   a. AS and a progressive increase in ascending aortic size. (*Level of Evidence: C*)
   b. Mild AR if valve-sparing aortic root replacement is being considered. (*Level of Evidence: C*)

CLASS III

1. AVR is not useful for prevention of sudden death in asymptomatic adults with AS who have none of the findings listed under the Class IIa/IIb indications. (*Level of Evidence: B*)
2. AVR is not indicated in asymptomatic patients with AR who have normal LV size and function. (*Level of Evidence: B*)

When valvular AS is secondary to bicuspid commissural fusion, especially in young adults, the potential exists for successful balloon dilation with gradient reduction and extended freedom from reintervention (78). Increasing calcification, with concomitantly increasing transvalvular gradient with increasing patient age, limits results in older adults, in whom AVR is the intervention of choice (78). Criteria for intervention vary, with typical indications including a valve area less than or equal to 0.45 cm^2 per m^2 (if not indexed, 0.8 cm^2 for an average-sized adult with a height of 1.7 m^2), especially in the setting of the symptoms of dyspnea, angina, or syncope or with worsening ventricular function. Balloon valvuloplasty may be considered in younger patients in whom there is a need to have augmented cardiac output, such as those with a desire to become pregnant or to participate in vigorous sports. When balloon valvuloplasty is indicated, patients should be referred to a center experienced in the procedure.

In BAV disease, there is no consensus regarding the specific diameter of the ascending aorta for which replacement is indicated, but greater than or equal to 5.0 cm has been suggested by some (73). Whether aortic root replacement or wrapping is optimal in such patients is a matter of debate; results of AVR in CHD have an acceptable medium-term result (79).

6.5. Recommendations for Key Issues to Evaluate and Follow-Up

CLASS I

1. Lifelong cardiology follow-up is recommended for all patients with aortic valve disease (AS or AR) (operated or unoperated; refer to Section 6.2, Recommendations for Evaluation of the Unoperated Patient). (*Level of Evidence: A*)
2. Serial imaging assessment of aortic root anatomy is recommended for all patients with BAV, regardless of severity. The frequency of imaging would depend on the size of the aorta at initial assessment: if less than 40 mm, it should be reimaged approximately every 2 years; if greater than or equal to 40 mm, it should be reimaged yearly or more often as progression of root dilation warrants or whenever there is a change in clinical symptoms or findings. (*Level of Evidence: B*)
3. Prepregnancy counseling is recommended for women with AS who are contemplating pregnancy. (*Level of Evidence: B*)
4. Patient referral to a pediatric cardiologist experienced in fetal echocardiography is indicated in the second trimester of preg-

*Consider lower threshold values for patients of small stature of either gender.

nancy to search for cardiac defects in the fetus. (*Level of Evidence: C*)

5. **Women with BAV and ascending aorta diameter greater than 4.5 cm should be counseled about the high risks of pregnancy. (*Level of Evidence: C*)**
6. **Patients with moderate to severe AS should be counseled against competitive athletics and strenuous isometric exercise. (*Level of Evidence: B*)**
7. **Echocardiographic screening for the presence of BAV is recommended for first-degree relatives of patients with BAV. (*Level of Evidence: B*)**

Progressive or recurrent AS, AR, or aortic enlargement may occur in the presence of a BAV. Patients with or without intervention should be followed up at least yearly for symptoms and findings of progressive AS/AR ventricular dysfunction and arrhythmia. This includes resting and stress ECGs to look for ischemic changes or arrhythmia; echocardiography-Doppler to monitor LV size/volume and systolic and diastolic function, aortic valve function, and aortic root size and anatomy; and 24-hour ambulatory ECG monitoring.

6.6. Isolated Subaortic Stenosis

SubAS refers to a discrete fibrous ring or fibromuscular narrowing and is distinct from genetic hypertrophic cardiomyopathy with dynamic LVOT obstruction. SubAS may occur as an associated defect with VSDs, AVSD, or conotruncal anomalies and may develop after patch closure of a perimembranous or misaligned VSD or AVSD (80).

6.6.1. Clinical Course With/Without Previous Intervention

The course of SubAS is often progressive. The unrepaired history includes progressive aortic valve damage, ventricular dysfunction, IE, and sudden cardiac death. The dominant feature may be obstruction or AR (81–83). AR occurs in more than 50% of those with SubAS. Once the peak Doppler gradient across the SubAS is more than 30 mm Hg, and if the membrane is immediately adjacent to the aortic valve or there is extension of the membrane onto the mitral valve, LVOT obstruction is likely to be progressive (82). Once the peak instantaneous Doppler LVOT gradient reaches 50 mm Hg or more, there is increased risk for moderate or severe AR (82). Patients are at risk for endocarditis, which will contribute to worsening AR (84).

6.7. Recommendations for Surgical Intervention

CLASS I

1. **Surgical intervention is recommended for patients with SubAS and a peak instantaneous gradient of 50 mm Hg or a mean gradient of 30 mm Hg on echocardiography-Doppler. (*Level of Evidence: C*)**
2. **Surgical intervention is recommended for SubAS with less than a 50 mm Hg peak or less than a 30 mm Hg mean gradient and progressive AR and an LV dimension at end-systolic diameter of 50 mm or LV ejection fraction less than 55%. (*Level of Evidence: C*)**

CLASS IIb

1. **Surgical resection may be considered in patients with a mean gradient of 30 mm Hg, but careful follow-up is required to detect progression of stenosis or AR. (*Level of Evidence: C*)**
2. **Surgical resection may be considered for patients with less than a 50 mm Hg peak gradient or less than a 30 mm Hg mean gradient in the following situations:**
   a. **When LV hypertrophy is present. (*Level of Evidence: C*)**
   b. **When pregnancy is being planned. (*Level of Evidence: C*)**
   c. **When the patient plans to engage in strenuous/competitive sports. (*Level of Evidence: C*)**

CLASS III

1. **Surgical intervention is not recommended to prevent AR for patients with SubAS if the patient has trivial LVOT obstruction or trivial to mild AR. (*Level of Evidence: C*)**

Surgical intervention should be recommended for patients with SubAS when the peak instantaneous echocardiographic gradient is greater than 50 mm Hg, the mean gradient is greater than 30 mm Hg, or catheter measurement of the resting peak-to-peak gradient is greater than 50 mm Hg. Patients with lesser degrees of obstruction may be considered for surgery in the presence of LV systolic dysfunction or significant aortic valve regurgitation, or if the patient desires to become pregnant or to participate in active sports.

Postoperative complications may include damage to the aortic or mitral valve, heart block, iatrogenic VSD, and IE. SubAS may recur after surgical repair; repair of SubAS in children does not necessarily prevent AR development in adults (81,85). However, data exist to suggest that surgical resection of fixed SubAS before the development of a more than 40 mm Hg LVOT gradient may prevent reoperation and secondary progressive aortic valve disease (86). Although catheter palliation has been performed in some centers on an experimental basis, its efficacy has not been demonstrated (87).

6.8. Recommendations for Key Issues to Evaluate and Follow-Up

CLASS I

1. **Lifelong cardiology follow-up, including evaluation by and/or consultation with a cardiologist with expertise in ACHD, is recommended for all patients with SubAS, repaired or not. (*Level of Evidence: C*)**
2. **The unoperated asymptomatic adult with stable LVOT obstruction due to SubAS and a mean gradient less than 30 mm Hg without LV hypertrophy or significant AR should be monitored at yearly intervals for increasing obstruction, the development or progression of AR, and the evaluation of systolic and diastolic LV function. (*Level of Evidence: B*)**

CLASS IIa

1. **Stress testing to determine exercise capability, symptoms, ECG changes or arrhythmias, or increase in LVOT gradient is reasonable in the presence of otherwise equivocal indications for intervention. (*Level of Evidence: C*)**

Progressive and/or recurrent obstruction and progressive AR may occur in patients with or without intervention. Recurrent obstruction is frequent after resection of SubAS

and occurs at a rate of approximately 20% over 10 years. In addition, AR may occur despite resection of the subaortic membrane.

6.9. Supravalvular Aortic Stenosis

Supravalvular aortic stenosis (SupraAS) is a fixed obstruction that arises from just above the sinus of Valsalva and extends a variable distance along the aorta. The origin of the coronary arteries is usually proximal to the obstruction, which subjects them to high systolic pressure and limited diastolic flow. There may be partial or complete ostial obstruction of the coronary arteries, ectasia, or aneurysm of the coronary arteries (88).

SupraAS is commonly seen in Williams syndrome and can be associated with hypoplasia of the entire aorta, renal artery stenosis, stenoses of other major aortic branches, and long-segment peripheral pulmonary artery stenosis. Williams syndrome, an autosomal dominant disorder due to an elastin gene mutation, is associated with abnormal (elfin) facies, cognitive and behavioral disorders, and joint abnormalities. Familial non-Williams SubAS is also associated with branch pulmonary artery stenosis and hypoplasia, as well as hypoplastic descending aorta and renal artery stenosis.

6.9.1. Clinical Course (Unrepaired)

Most patients with SubAS will be followed up from childhood and may present in adult life with symptoms due to significant outflow obstruction, systemic hypertension, or ischemia. Clinical presentation with ischemic symptomatology referable to insufficient coronary artery flow has been reported due to either anatomic obstruction or myocardial hypertrophy that limits nonepicardial coronary flow (89).

6.10. Recommendations for Evaluation of the Unoperated Patient

CLASS I

1. **TTE and/or TEE with Doppler and either MRI or CT should be performed to assess the anatomy of the LVOT, the ascending aorta, coronary artery anatomy and flow, and main and branch pulmonary artery anatomy and flow. (*Level of Evidence: C*)**
2. **Assessment of anatomy and flow in the proximal renal arteries is recommended in ACHD patients with SupraAS. (*Level of Evidence: C*)**
3. **Assessment of systolic and diastolic ventricular function is recommended in ACHD patients with SupraAS. (*Level of Evidence: C*)**
4. **Assessment of aortic and mitral valve anatomy and function is recommended in ACHD patients with SupraAS. (*Level of Evidence: C*)**
5. **Adults with a history or presence of SupraAS should be screened periodically for myocardial ischemia. (*Level of Evidence: C*)**

CLASS IIa

1. **Exercise testing, dobutamine stress testing, positron emission tomography, or stress sestamibi with adenosine studies can be useful to evaluate the adequacy of myocardial perfusion. (*Level of Evidence: C*)**

6.10.1. Imaging

TTE and TEE demonstrate the diameter and anatomy of the aortic sinus, sinotubular ridge, and proximal ascending aorta; the origins of the coronary arteries; the systolic gradient across the SupraAS obstruction; and the degree of left ventricle hypertrophy. MRI/CT is required to more precisely define the anatomy of the aorta and branches, as well as the pulmonary arteries. As with any long-segment obstruction, assessment of the gradient can be challenging and may require cardiac catheterization for complete assessment of hemodynamic severity of the stenosis. Patients with Williams syndrome should have imaging of the entire aorta, including the renal arteries, because of the association with arterial stenosis at any level.

6.10.2. Myocardial Perfusion Imaging

Patients with an inability to perform maximal stress testing secondary to limited cognitive function or physical capacity may undergo perfusion imaging with pharmacologic stress (adenosine or dobutamine) nuclear imaging with positron emission tomography, single photon emission computed tomography, or MRI.

6.10.3. Cardiac Catheterization

Diagnostic catheterization may help to delineate anatomy and accurately measure gradients. Selective coronary angiography should be approached with caution after thorough noninvasive and angiographic examination of the aortic root, because coronary ostial stenosis is a frequent occurrence in this population. Intravascular ultrasonography may provide definition of coronary artery anatomy and define the nature and extent of the diseased vessel before consideration of repair.

6.11. Management Strategies for Supravalvular Left Ventricular Outflow Tract

6.11.1. Recommendations for Interventional and Surgical Therapy

CLASS I

1. **Operative intervention should be performed for patients with supravalvular LVOT obstruction (discrete or diffuse) with symptoms (ie, angina, dyspnea, or syncope) and/or mean gradient greater than 50 mm Hg or peak instantaneous gradient by Doppler echocardiography greater than 70 mm Hg. (*Level of Evidence: B*)**
2. **Surgical repair is recommended for adults with lesser degrees of supravalvular LVOT obstruction and the following indications:**
   a. **Symptoms (ie, angina, dyspnea, or syncope). (*Level of Evidence: B*)**
   b. **LV hypertrophy. (*Level of Evidence: C*)**
   c. **Desire for greater degrees of exercise or a planned pregnancy. (*Level of Evidence: C*)**
   d. **LV systolic dysfunction. (*Level of Evidence: C*)**
3. **Interventions for coronary artery obstruction in patients with SupraAS should be performed in ACHD centers with demonstrated expertise in the interventional management of such patients. (*Level of Evidence: C*)**

Surgical relief of SupraAS is accomplished with the use of complex patching of the aorta, with reconstruction of the coronary ostia or bypass grafting, depending on the anatomy of the lesion. Surgical results with reconstruction of the coronary ostium or bypass grafting, depending on anatomy of the lesions noted, have been described without long-term follow-up (90). Branch pulmonary artery stenosis may be addressed during the same surgical procedure. There are no long-term follow-up data on adults after surgery for SupraAS. Catheter-based techniques have not been described for this lesion.

6.11.2. Recommendations for Key Issues to Evaluate and Follow-Up

CLASS I

1. **Both operated and unoperated patients with SupraAS should be followed up annually at a regional ACHD center. (*Level of Evidence: C*)**
2. **Long-term psychosocial assessment and oversight, including the need for legal guardianship, are recommended for patients with Williams syndrome. (*Level of Evidence: C*)**

Repair of SupraAS results in low early and late mortality and a low incidence of recurrent obstruction. The durability of patch material requires long-term observation for assessment of aneurysm formation. Both operated and unoperated patients with SupraAS require lifelong annual follow-up to evaluate the degree of obstruction and LV compensation, the development of coronary insufficiency or systemic hypertension, and the development of mitral regurgitation.

Patients with Williams syndrome require long-term psychosocial follow-up to assess competency for self-care and recommend appropriate measures. This is particularly important because these patients have verbal and social skills that result in an overestimation of their executive functioning.

6.11.3. Recommendations for Reproduction

CLASS I

1. **SupraAS, whether associated with Williams syndrome or nonsyndromic, has a strong likelihood of being an inherited disorder. Undetected family members may be at risk for hypertension, coronary disease, or stroke; therefore, all available relatives should be screened. (*Level of Evidence: C*)**
2. **Patients with SupraAS and significant obstruction, coronary involvement, or aortic disease should be counseled against pregnancy. (*Level of Evidence: C*)**

6.12. Aortic Coarctation

Discrete coarctation of the aorta consists of short-segment narrowing in the region of the ligamentum arteriosum adjacent to the origin of the left subclavian artery. In some cases, there is also narrowing of the aortic arch or isthmus. Extensive collateral vessels may arise proximal to the obstruction, masking the severity of obstruction. An associated intrinsic abnormality in the aortic wall predisposes to dissection or rupture in the ascending aorta or the area of the coarctation. Associated lesions include BAV, SubAS, mitral valve abnormalities such as parachute mitral stenosis, VSD, and circle of Willis cerebral artery aneurysm.

6.12.1. Recommendations for Clinical Evaluation and Follow-Up

CLASS I

1. **Every patient with systemic arterial hypertension should have the brachial and femoral pulses palpated simultaneously to assess timing and amplitude evaluation to search for the "brachial-femoral delay" of significant aortic coarctation. Supine bilateral arm (brachial artery) blood pressures and prone right or left supine leg (popliteal artery) blood pressures should be measured to search for differential pressure. (*Level of Evidence: C*)**
2. **Initial imaging and hemodynamic evaluation by TTE, including suprasternal notch acoustic windows, is useful in suspected aortic coarctation. (*Level of Evidence: B*)**
3. **Every patient with coarctation (repaired or not) should have at least 1 cardiovascular MRI or CT scan for complete evaluation of the thoracic aorta and intracranial vessels. (*Level of Evidence: B*)**

Aortic coarctation may be recognized in the adult, usually because of systemic arterial hypertension and discrepant upper- and lower-extremity pulses. Patients may complain of exertional headaches, leg fatigue, or claudication. Occasionally, the patient may come to medical attention because of a murmur due to BAV or VSD.

6.13. Management Strategies for Coarctation of the Aorta

6.13.1. Medical Therapy

Hypertension should be controlled by beta blockers, ACE inhibitors, or angiotensin-receptor blockers as first-line medications. The choice of beta blockers or vasodilators may be influenced in part by the aortic root size, the presence of AR, or both.

6.13.2. Recommendations for Interventional and Surgical Treatment of Coarctation of the Aorta in Adults

CLASS I

1. **Intervention for coarctation is recommended in the following circumstances:**
   a. **Peak-to-peak coarctation gradient greater than or equal to 20 mm Hg. (*Level of Evidence: C*)**
   b. **Peak-to-peak coarctation gradient less than 20 mm Hg in the presence of anatomic imaging evidence of significant coarctation with radiological evidence of significant collateral flow. (*Level of Evidence: C*)**
2. **Choice of percutaneous catheter intervention versus surgical repair of native discrete coarctation should be determined by consultation with a team of ACHD cardiologists, interventionalists, and surgeons at an ACHD center. (*Level of Evidence: C*)**
3. **Percutaneous catheter intervention is indicated for recurrent, discrete coarctation and a peak-to-peak gradient of at least 20 mm Hg. (*Level of Evidence: B*)**
4. **Surgeons with training and expertise in CHD should perform operations for previously repaired coarctation and the following indications:**
   a. **Long recoarctation segment. (*Level of Evidence: B*)**
   b. **Concomitant hypoplasia of the aortic arch. (*Level of Evidence: B*)**

CLASS IIb

1. **Stent placement for long-segment coarctation may be considered, but the usefulness is not well established, and the long-term efficacy and safety are unknown. (*Level of Evidence: C*)**

The appropriate type of treatment for native coarctation of the aorta in adults remains somewhat controversial. In particular, for women who are or will be of childbearing age after repair, there is a concern about the tissue integrity of the paracoarctation region, particularly during pregnancy. As such, one may select direct surgical repair with excision of the paracoarctation tissue for those individuals. For recurrent aortic coarctation (coarctation after surgical repair), the prevailing opinion now is that catheter-based intervention (balloon or stent) is generally safe and the preferred alternative to surgery in the absence of confounding features (eg, aneurysm or pseudoaneurysm formation, or significant coarctation that affects the adjoining arch arterial branches). For localized discrete narrowing, balloon angioplasty is an acceptable alternative to surgical repair as a primary intervention but is still considered less suitable for long-segment or tortuous forms of coarctation.

In the majority of circumstances, discrete recoarctation is managed with balloon dilation with or without stent placement. In many ACHD centers, surgery is reserved for patients who are unsuitable for percutaneous treatment or who have undergone unsuccessful percutaneous treatment.

The use of partial or full cardiopulmonary bypass may be required to prevent paralytic complications. Rebound hypertension can occur early after repair and may be prevented or blunted by preoperative administration of a beta blocker. Morbidity in adults with reoperation for coarctation can be considerable and may include significant early postoperative bleeding, pleural effusion, lung contusion, recurrent laryngeal nerve palsy, or phrenic nerve injury (with hemidiaphragmatic paresis or paralysis). Other postoperative complications include recoarctation and hypertension. Aneurysm formation at the repair site can occur after patch aortoplasty (particularly with the use of a Dacron patch) or resection of the coarctation shelf. False aneurysms may also occur at the repair site. Late dissection proximal or distal to the repair site can occur. Paraplegia secondary to spinal cord ischemia is rare but is more common with poor collateral circulation. Arm claudication or subclavian steal syndrome is rare but in particular may occur after use of the subclavian flap technique.

6.13.3. Recommendations for Key Issues to Evaluate and Follow-Up

CLASS I

1. **Lifelong cardiology follow-up is recommended for all patients with aortic coarctation (repaired or not), including an evaluation by or consultation with a cardiologist with expertise in ACHD. (*Level of Evidence: C*)**
2. **Patients who have had surgical repair of coarctation at the aorta or percutaneous intervention for coarctation of the aorta should have at least yearly follow-up. (*Level of Evidence: C*)**
3. **Even if the coarctation repair appears to be satisfactory, late postoperative thoracic aortic imaging should be performed to assess for aortic dilatation or aneurysm formation. (*Level of Evidence: B*)**
4. **Patients should be observed closely for the appearance or reappearance of resting or exercise-induced systemic arterial hypertension, which should be treated aggressively after recoarctation is excluded. (*Level of Evidence: B*)**
5. **Evaluation of the coarctation repair site by MRI/CT should be performed at intervals of 5 years or less, depending on the specific anatomic findings before and after repair. (*Level of Evidence: C*)**

CLASS IIb

1. **Routine exercise testing may be performed at intervals determined by consultation with the regional ACHD center. (*Level of Evidence: C*)**

All patients with either interventional catheterization or surgical repair of coarctation of the aorta should have close follow-up and aggressive management of blood pressure and other risk factors for cardiovascular disease. This should include at least yearly cardiology evaluations. Consultation with a cardiologist with special expertise in ACHD should be obtained on initial contact to determine risk factors specific for the patient's anatomy and the presence of associated lesions. Evaluation of the repair site by MRI/CT should be repeated at intervals of 5 years or less, depending on the specific anatomic findings before and after repair.

7. Right Ventricular Outflow Tract Obstruction

Obstruction to the RVOT in the adult patient can be either congenital or acquired. Congenital obstruction can be at the pulmonary valve, below the pulmonary valve, or above the pulmonary valve. Below the pulmonary valve, obstruction can be either at the infundibular or the subinfundibular level. Infundibular stenosis is a crucial component of tetralogy of Fallot.

7.1. Valvular Pulmonary Stenosis

Valvular PS is usually an isolated lesion. In long-standing, severe PS, there may be an element of infundibular hypertrophy and potential obstruction.

7.1.1. Recommendations for Evaluation of the Unoperated Patient

CLASS I

1. **Two-dimensional echocardiography-Doppler, chest x-ray, and ECG are recommended for the initial evaluation of patients with valvular PS. (*Level of Evidence: C*)**
2. **A follow-up physical examination, echocardiography-Doppler, and ECG are recommended at 5-year intervals in the asymptomatic patient with a peak instantaneous valvular gradient by Doppler less than 30 mm Hg. (*Level of Evidence: C*)**
3. **A follow-up echocardiography-Doppler is recommended every 2 to 5 years in the asymptomatic patient with a peak instantaneous valvular gradient by Doppler greater than 30 mm Hg. (*Level of Evidence: C*)**

CLASS III

1. **Cardiac catheterization is unnecessary for diagnosis of valvular PS and should be used only when percutaneous catheter intervention is contemplated. (*Level of Evidence: C*)**

There is little progression in PS severity when the gradient is less than 30 mm Hg; such patients can be followed up at least every 5 years with a clinical examination and Doppler-echocardiography. Those with more significant stenosis should be followed up on an annual basis. Most patients with PS who reach adulthood are asymptomatic and require no specific therapy.

Patients with valvular PS do not require cardiac catheterization for diagnosis, and the relationship between the peak-to-peak invasive hemodynamic gradient and the Doppler peak instantaneous gradient becomes relevant in deciding appropriateness for invasive evaluation and intervention. There are recent data that suggest the peak-to-peak gradient by cardiac catheterization correlates best with the mean Doppler (and not peak instantaneous Doppler) gradient in this situation (91) and that the peak instantaneous gradient systematically overestimates the peak-to-peak cardiac catheterization gradient by slightly more than 20 mm Hg. Correlation of the echocardiography-Doppler gradient with other clinical findings is important.

7.1.2. Management Strategies

7.1.2.1. Recommendations for Intervention in Patients With Valvular Pulmonary Stenosis

CLASS I

1. **Balloon valvotomy is recommended for asymptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg (in association with less than moderate pulmonary valve regurgitation). (*Level of Evidence: B*)**
2. **Balloon valvotomy is recommended for symptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg (in association with less than moderate pulmonary regurgitation). (*Level of Evidence: C*)**
3. **Surgical therapy is recommended for patients with severe PS and an associated hypoplastic pulmonary annulus, severe pulmonary regurgitation, subvalvular PS, or supravalvular PS. Surgery is also preferred for most dysplastic pulmonary valves and when there is associated severe TR or the need for a surgical Maze procedure. (*Level of Evidence: C*)**
4. **Surgeons with training and expertise in CHD should perform operations for the RVOT and pulmonary valve. (*Level of Evidence: B*)**

CLASS IIb

1. **Balloon valvotomy may be reasonable in asymptomatic patients with a dysplastic pulmonary valve and a peak instantaneous gradient by Doppler greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg. (*Level of Evidence: C*)**
2. **Balloon valvotomy may be reasonable in selected symptomatic patients with a dysplastic pulmonary valve and peak instantaneous gradient by Doppler greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg. (*Level of Evidence: C*)**

CLASS III

1. **Balloon valvotomy is not recommended for asymptomatic patients with a peak instantaneous gradient by Doppler less than 50 mm Hg in the presence of normal cardiac output. (*Level of Evidence: C*)**
2. **Balloon valvotomy is not recommended for symptomatic patients with PS and severe pulmonary regurgitation. (*Level of Evidence: C*)**
3. **Balloon valvotomy is not recommended for symptomatic patients with a peak instantaneous gradient by Doppler less than 30 mm Hg. (*Level of Evidence: C*)**

Since the initial successful report of percutaneous balloon valvotomy for pulmonary valve stenosis in 1982 (92), the procedure has evolved to be the treatment of choice for patients with classic domed valvular PS. Balloon valvotomy produces relief of the gradient by commissural splitting. As might be expected from the morphology, results in patients with a dysplastic pulmonary valve are less impressive.

Because of the elasticity of the pulmonary annulus, it has been found that oversizing the balloons up to 1.4 times the measured pulmonary annulus is more effective in achieving a successful result (usually defined by a final valvular gradient of less than 20 mm Hg). To accomplish this oversizing in adults, a double-balloon procedure is frequently used. In general, acute complications from the procedure have been minimal. During the acute performance of the valvotomy, vagal symptoms predominate, along with catheter-induced ventricular ectopy and occasionally right bundle-branch block.

Other complications include pulmonary valve regurgitation, pulmonary edema (presumably from increasing pulmonary blood flow to previously underperfused lungs), cardiac perforation and tamponade, high-grade AV nodal block, and transient RVOT obstruction. The latter is sometimes referred to as a “suicidal right ventricle” and is due to abrupt infundibular obstruction once the pulmonary valve obstruction has been relieved (93). This may be alleviated by volume expansion and beta blockade. This postprocedural infundibular obstruction tends to regress over time.

In patients with PS and significant valvular regurgitation, valve replacement may be required. Mechanical valve replacement (94) is rarely used because of concerns regarding thrombosis. Bioprosthetic valves (95) can be effectively implanted with good durability in patients of all ages, although valvular degeneration eventually ensues in all.

7.1.3. Recommendation for Clinical Evaluation and Follow-Up After Intervention

CLASS I

1. **Periodic clinical follow-up is recommended for all patients after surgical or balloon pulmonary valvotomy, with specific attention given to the degree of pulmonary regurgitation; RV pressure, size, and function; and TR. The frequency of follow-up should be determined by the severity of hemodynamic abnormalities but should be at least every 5 years. (*Level of Evidence: C*)**

Long-term follow-up after balloon valvuloplasty in patients without valve dysplasia suggests a low (0% to 4.8%) incidence of restenosis (96,97) and a moderate (39%) incidence of pulmonary regurgitation. In adult patients without valve dysplasia, excellent results were also observed, with a residual gradient primarily found only in those patients who had an inadequate initial result (98). In the Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) registry (99), follow-up data were available on 533 patients a mean of 8.7 years after valvotomy. A suboptimal result (defined as gradient greater than 35 mm Hg at the end of the procedure) was present in 23%. Valve morphology and annulus size were the most significant predictors of long-term results. Pulmonary regurgitation was more commonly seen when the balloon-to-annulus ratio exceeded 1.4, which suggests an optimal ratio of 1.2 to 1.4. When restenosis does occur after percutaneous balloon pulmonary valvotomy, it appears that a repeat procedure is effective in patients without dysplastic pulmonary valves (100).

Percutaneous balloon valvotomy thus appears to be an excellent alternative to surgical valvuloplasty or valve replacement in most patients with classic, doming, valvular PS. Its use in patients with a dysplastic valve is much less established. After surgical valvotomy, pulmonary regurgitation is common, and after 3 to 4 decades, RV dysfunction and secondary TR may ensue, necessitating pulmonary valve replacement in some patients. This should be undertaken before there is severe RV enlargement and any more than mild RV dysfunction. Deteriorating exercise capacity or the onset of atrial or ventricular arrhythmias is also a sign of the need for pulmonary valve replacement. This emphasizes the need for lifelong follow-up in such patients (101).

7.2. Right-Sided Obstruction Due to Supravalvular, Branch, and Peripheral Pulmonary Artery Stenosis

The pulmonary arterial segments distal to patent stenotic lesions often exhibit poststenotic dilation. Central and peripheral pulmonary artery stenosis may be a major cardiovascular feature in the Alagille and Keutel syndromes (102–106). Pulmonary artery stenoses are also sequelae of the congenital Rubella syndrome, Williams syndrome, or scarring at the site of a previous pulmonary artery band or aorticopulmonary shunt.

7.2.1. Clinical Course

Peripheral pulmonary artery stenoses tend to occur in multiple tertiary branches of the pulmonary tree and are progressive, and by the time patients are seen as adults, there may be considerable loss of lung parenchyma due to totally occluded segmental pulmonary arteries. With PAH, pulmonary valve regurgitation may be expected.

7.2.2. Recommendations for Evaluation of Patients With Supravalvular, Branch, and Peripheral Pulmonary Stenosis

CLASS I

1. **Patients with suspected supravalvular, branch, or peripheral PS should have baseline imaging with echocardiography-Doppler plus 1 of the following: MRI angiography, CT angiography, or contrast angiography. (*Level of Evidence: C*)**
2. **Once the diagnosis is established, follow-up echocardiography-Doppler to assess RV systolic pressure should be performed periodically, depending on severity. (*Level of Evidence: C*)**

TTE-Doppler helps confirm the presence of RV systolic hypertension and any pulmonary valve regurgitation. Echocardiography may also be able to define proximal pulmonary branch stenosis. It is of much less value in the identification of peripheral PS. TEE is likewise useful only when there are proximal pulmonary artery lesions. Radionuclide studies reveal the severity of peripheral PS in different lung segments. Cardiac MRI with pulmonary angiography and CT are much superior to echocardiography-Doppler for imaging these lesions, and both can help confirm the diagnosis.

7.2.3. Recommendations for Interventional Therapy in the Management of Branch and Peripheral Pulmonary Stenosis

CLASS I

1. **Percutaneous interventional therapy is recommended as the treatment of choice in the management of appropriate focal branch and/or peripheral pulmonary artery stenosis with greater than 50% diameter narrowing, an elevated RV systolic pressure greater than 50 mm Hg, and/or symptoms. (*Level of Evidence: B*)**
2. **In patients with the above indications for intervention, surgeons with training and expertise in CHD should perform operations for management of branch pulmonary artery stenosis not anatomically amenable to percutaneous interventional therapy. (*Level of Evidence: B*)**

Branch pulmonary artery stenosis and/or hypoplasia may be associated with a variety of cardiac malformations or may be a residual from prior surgical intervention, such as an anastomotic lesion at the distal site of a prior Blalock-Taussig or Potts shunt procedure. Surgical exposure to these areas is often difficult, which favors attempts at percutaneous approaches.

The highly elastic pulmonary arteries have proven to be resilient to balloon procedures, and angioplasty methods have generally given way to stent procedures in which there appears to be a higher initial success rate and a lower intermediate-term incidence of restenosis (107). Stenting of branch PS has also been used in the operating room as adjunctive therapy. The use of balloon angioplasty and stenting may also be applied to more distal peripheral PS, although the results have generally been less impressive than with branch stenosis (108).

7.2.4. Recommendations for Evaluation and Follow-Up

CLASS I

1. **Patients with peripheral PS should be followed up every 1 to 2 years, on the basis of severity, with a clinical evaluation and echocardiography-Doppler to evaluate RV systolic pressure and RV function. (*Level of Evidence: C*)**

2. Discussion with a cardiac surgeon with expertise in CHD should take place before percutaneous peripheral pulmonary artery interventions are undertaken. (*Level of Evidence: C*)

The lesions in peripheral PS may be progressive, so patients should be followed up every 1 to 2 years with echocardiography-Doppler to assess RV peak systolic pressure and function. Restenosis of these lesions is common, and repeat percutaneous angioplasty, stenting, and/or surgical intervention may be required when this occurs.

7.3. Right-Sided Heart Obstruction Due to Stenotic Right Ventricular–Pulmonary Artery Conduits or Bioprosthetic Valves

Some gradient is to be expected across any RV–pulmonary artery conduit or any bioprosthetic valve placed in the RVOT, depending on the valve size and flow across the valve.

7.3.1. Recommendation for Evaluation and Follow-Up After Right Ventricular–Pulmonary Artery Conduit or Prosthetic Valve

CLASS I

1. After surgical relief of RVOT obstruction with a conduit or prosthetic valve, patients should be followed up on a 1- to 2-year basis with echocardiography-Doppler assessment of RV systolic pressure and function, as well as a measurement of the gradient across the RVOT. (*Level of Evidence: C*)

7.3.2. Echocardiography

TTE and Doppler are particularly helpful in delineating hemodynamics and facilitate measurement of RV pressure, RV size and function, and gradient across the conduit and prosthetic valve. However, tubular narrowing in a conduit is often associated with underestimation of the gradient.

7.3.3. Magnetic Resonance Imaging/ Computed Tomography

CT and MRI can be used to help define lesion severity and may demonstrate conduit adherence to the sternum, something of interest to the surgeon if a reoperation is contemplated.

7.3.4. Cardiac Catheterization

Because distal conduit stenosis is frequent, MRI and CT, as well as cardiac catheterization and angiography, can define the level and severity of stenosis.

7.3.5. Recommendations for Reintervention in Patients With Right Ventricular–Pulmonary Artery Conduit or Bioprosthetic Pulmonary Valve Stenosis

CLASS I

1. Surgeons with training and expertise in CHD should perform operations for patients with severe pulmonary prosthetic valve stenosis (peak gradient greater than 50 mm Hg) or conduit regurgitation and any of the following:
   a. Decreased exercise capacity. (*Level of Evidence: C*)
   b. Depressed RV function. (*Level of Evidence: C*)
   c. At least moderately enlarged RV end-diastolic size. (*Level of Evidence: C*)
   d. At least moderate TR. (*Level of Evidence: C*)

CLASS IIa

1. Either surgical or percutaneous therapy can be useful in symptomatic patients with discrete RV–pulmonary artery conduit obstructive lesions with greater than 50% diameter narrowing or when a bioprosthetic pulmonary valve has a peak gradient by Doppler greater than 50 mm Hg or a mean gradient greater than 30 mm Hg. (*Level of Evidence: C*)
2. Either surgical or percutaneous therapy can be useful in asymptomatic patients when a pulmonary bioprosthetic valve has a peak Doppler gradient greater than 50 mm Hg. (*Level of Evidence: C*)

CLASS IIb

1. Surgical intervention may be considered preferable to percutaneous catheter intervention when an associated Maze procedure is being considered for the treatment of atrial arrhythmia. (*Level of Evidence: C*)

Both angioplasty and stenting have been applied to obstruction in an RV–to–pulmonary artery conduit. Such cases can present difficult issues, and the decision to proceed with a percutaneous intervention should be made in association with an ACHD surgeon or an ACHD interventionalist. Surgical intervention is generally required once there is evidence of important RV enlargement or the development of significant TR.

7.3.6. Key Issues to Evaluate and Follow-Up

Most patients are not limited physically unless the gradient across the conduits or prosthetic valves is greater than 50 mm Hg.

7.4. Double-Chambered Right Ventricle

In patients with a double-chambered right ventricle, the right ventricle is divided into a high-pressure proximal and lower-pressure distal chamber by anomalous myocardial muscle bundles.

7.4.1. Recommendations for Intervention in Patients With Double-Chambered Right Ventricle

CLASS I

1. Surgery is recommended for patients with a peak midventricular gradient by Doppler greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg, regardless of symptoms. (*Level of Evidence: B*)

CLASS IIb

1. Symptomatic patients with a peak midventricular gradient by Doppler greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg may be considered for surgical resection if no other cause of symptoms can be discerned. (*Level of Evidence: C*)

Peak RV systolic pressure, as estimated by echocardiography-Doppler via the TR jet, may be the result of more than 1 level of obstruction, and it is important to investigate this possibility thoroughly before surgical intervention is considered. This is particularly important in the adult, in whom prior surgical procedures and other causes of PAH may complicate the clinical picture.

In patients with a double-chambered right ventricle, resection and outflow-enlarging procedures have been very effective, with excellent long-term results (109). Many such patients also require repair of an associated VSD.

8. Coronary Artery Abnormalities

8.1. General Recommendations for Evaluation and Surgical Intervention

CLASS I

1. **Any patient with CHD who has had coronary artery manipulation should be evaluated for coronary artery patency, function, and anatomic integrity at least once in adulthood. (*Level of Evidence: C*)**
2. **Surgeons with training and expertise in CHD should perform operations for the treatment of coronary artery anomalies. (*Level of Evidence: C*)**

Because there is no long-term follow-up information about the sequelae of manipulation of the coronary arteries in the various forms of CHD, it is prudent to evaluate these patients at least once during adult life for late development of coronary artery disease.

8.2. Recommendations for Coronary Anomalies Associated With Supravalvular Aortic Stenosis

CLASS I

1. **Adults with a history or presence of SupraAS should be screened every 1 or 2 years for myocardial ischemia. (*Level of Evidence: C*)**
2. **Interventions for coronary artery obstruction in patients with SupraAS should be performed in ACHD centers with demonstrated expertise in the interventional management of these patients. (*Level of Evidence: C*)**

SupraAS may be associated with coronary obstruction from partial to complete ostial obliteration, and these patients are also at risk for ectasia and aneurysm of the coronary arteries (88). Pathological specimens with diffuse or focal intimal and medial fibrosis, hyperplasia, dysplasia, adventitial fibroelastosis, and occasional intramedial dissection have been reported in children and more commonly in adults (110–112).

8.3. Recommendation for Coronary Anomalies Associated With Tetralogy of Fallot

CLASS I

1. **Coronary artery anatomy should be determined before any intervention for RV outflow. (*Level of Evidence: C*)**

The most common and important abnormality is the left anterior descending coronary artery arising from the right coronary artery and crossing the RV outflow, which occurs in approximately 3% to 7% of persons with tetralogy of Fallot. The occurrence is more common when the aortic root is more anterior, rightward, or lateral (113).

8.3.1. Preintervention Evaluation

Coronary artery origin and course should be delineated before any surgical or interventional procedure, because the potential exists for damage to anomalous coronary arteries to occur during cardiac exposure, surgery on the RVOT, and stenting of RV outflow.

8.4. Recommendation for Coronary Anomalies Associated With Dextro-Transposition of the Great Arteries After Arterial Switch Operation

CLASS I

1. **Adult survivors with dextro-TGA (d-TGA) after arterial switch operation (ASO) should have noninvasive ischemia testing every 3 to 5 years. (*Level of Evidence: C*)**

The coronary artery course plays an important role in the surgical repair of d-TGA. The most common anatomic arrangement occurs in nearly two thirds of patients, with the left coronary artery arising from the anterior facing sinus and the right coronary artery from the posterior facing sinus. Sixteen percent of patients with d-TGA have a circumflex that arises from the right coronary artery, and the remaining patients have inverted coronary artery variants, single coronary arteries, or intramural coronary arteries (114). Damage to the sinus node coronary artery, whether during surgery or during balloon septostomy, has been implicated in the occurrence of atrial arrhythmias and sinus node dysfunction after repair.

8.4.1. Clinical Course

After great artery translocation and transfer of coronary arteries, early and late postoperative loss of coronary perfusion may occur due to causes such as anatomic torsion, extrinsic compression, focal or diffuse fibrocellular intimal thickening, and small-caliber distal coronary arteries with functional decrease in coronary flow reserve (115–117). Survival free of coronary events has been reported as 93% and 88% at 1 and 15 years, respectively, with many reports associating coronary events with increased mortality (117).

8.4.2. Clinical Features and Evaluation After Arterial Switch Operation

No single ischemia provocation test has been shown to be both sufficiently sensitive and specific to screen for coronary flow abnormalities after a switch repair of d-TGA. Combinations of testing, including echocardiography, nuclear scintigraphy, and exercise testing, have been suggested to improve sensitivity and specificity (117).

Given the emergence of an adult population of survivors with d-TGA after ASO, with undefined future course and morbidity, the present writing committee recommends episodic noninvasive ischemia provocation testing every 3 to 5 years. Positive results should be pursued by invasive catheterization with measurement of coronary flow reserve and intravascular ultrasound when appropriate.

8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

CLASS I

1. The evaluation of individuals who have survived unexplained aborted sudden cardiac death or with unexplained life-threatening arrhythmia, coronary ischemic symptoms, or LV dysfunction should include assessment of coronary artery origins and course. (*Level of Evidence: B*)
2. CT or magnetic resonance angiography is useful as the initial screening method in centers with expertise in such imaging. (*Level of Evidence: B*)
3. Surgical coronary revascularization should be performed in patients with any of the following:
   a. Anomalous left main coronary artery coursing between the aorta and pulmonary artery. (*Level of Evidence: B*)
   b. Documented coronary ischemia due to coronary compression (when coursing between the great arteries or in intramural fashion). (*Level of Evidence: B*)
   c. Anomalous origin of the right coronary artery between aorta and pulmonary artery with evidence of ischemia. (*Level of Evidence: B*)

CLASS IIa

1. Surgical coronary revascularization can be beneficial in the setting of documented vascular wall hypoplasia, coronary compression, or documented obstruction to coronary flow, regardless of inability to document coronary ischemia. (*Level of Evidence: C*)
2. Delineation of potential mechanisms of flow restriction via intravascular ultrasound can be beneficial in patients with documented anomalous coronary artery origin from the opposite sinus. (*Level of Evidence: C*)

CLASS IIb

1. Surgical coronary revascularization may be reasonable in patients with anomalous left anterior descending coronary artery coursing between the aorta and pulmonary artery. (*Level of Evidence: C*)

8.5.1. Definition, Associated Lesions, and Clinical Course

Congenital anomalous origin of the coronary arteries may occur in 1% to 1.2% of all coronary angiograms performed, with 0.5% of them having the highest-risk lesions of the left main or left anterior descending branch artery arising from the opposite sinus of Valsalva (118). Coronary anomalies account for approximately 15% of sudden cardiac deaths in athletes (potentially due to torsion or slitlike compression of the proximal coronary artery, exercise-induced compression, vasospasm, or ischemic or scar-induced ventricular arrhythmia) (119,120). In 80% of autopsies in athletes with sudden cardiac death and anomalous coronary artery origins, the affected coronary artery coursed between the aorta and the pulmonary artery (120,121).

8.6. Recommendations for Anomalous Left Coronary Artery From the Pulmonary Artery

CLASS I

1. In patients with an anomalous left coronary artery from the pulmonary artery (ALCAPA), reconstruction of a dual coronary artery supply should be performed. The surgery should be performed by surgeons with training and expertise in CHD at centers with expertise in the management of anomalous coronary artery origins. (*Level of Evidence: C*)
2. For adult survivors of ALCAPA repair, clinical evaluation with echocardiography and noninvasive stress testing is indicated every 3 to 5 years. (*Level of Evidence: C*)

ALCAPA is relatively rare, occurring in 1 in 300 000 live births.

8.7. Management Strategies

8.7.1. Surgical Intervention

If patients present in adulthood with decreased systolic function and previously unrecognized ALCAPA, the present writing committee suggests surgical myocardial revascularization to achieve a dual coronary supply, regardless of myocardial viability testing, given the lack of current data to correlate such testing with outcomes. Given the increasing awareness of residual coronary artery, myocardial, and valvular abnormalities, the present writing committee suggests surveillance with echocardiography and noninvasive ischemia provocation testing every 3 to 5 years for patients after repair of ALCAPA.

8.7.2. Surgical and Catheterization-Based Intervention

Surgical repair by either arterial bypass or, more commonly, reimplantation of the anomalous coronary into the aorta is indicated because of the risk of sudden cardiac death (122,123). If ischemia is demonstrated in patients after repair of ALCAPA with either concomitant symptomatology or echocardiographic changes, the present writing committee recommends invasive catheterization with planned intervention determined by clinical findings.

8.8. Recommendations for Coronary Arteriovenous Fistula

CLASS I

1. If a continuous murmur is present, its origin should be defined either by echocardiography, MRI, CT angiography, or cardiac catheterization. (*Level of Evidence: C*)
2. A large coronary arteriovenous fistula (CAVF), regardless of symptomatology, should be closed via either a transcatheter or surgical route after delineation of its course and its potential to fully obliterate the fistula. (*Level of Evidence: C*)
3. A small to moderate CAVF in the presence of documented myocardial ischemia, arrhythmia, otherwise unexplained ventricular systolic or diastolic dysfunction or enlargement, or endarteritis should be closed via either a transcatheter or surgical approach after delineation of its course and its potential to fully obliterate the fistula. (*Level of Evidence: C*)

CLASS IIa

1. Clinical follow-up with echocardiography every 3 to 5 years can be useful for patients with small, asymptomatic CAVF to exclude development of symptoms or arrhythmias or progression of size or chamber enlargement that might alter management. (*Level of Evidence: C*)

CLASS III

1. **Patients with small, asymptomatic CAVF should not undergo closure of CAVF. (*Level of Evidence: C*)**

Fistulas arise from either or both coronary arteries, with drainage more typically to the right atrium, right ventricle, or right atrium–superior vena cava junction, and occasionally to the coronary sinus or left side of the heart. Although the potential for associated myocardial ischemia and infarction, endarteritis, dissection, and rupture has been documented, there are few data associating occurrence, shunt properties, anatomic features, and outcomes. Small fistulas may slowly increase in size with advancing age and changes in systemic blood pressure and aortic compliance. Periodic clinical evaluation with imaging such as echocardiography to assess both the size of the fistula and ventricular function is reasonable. Sometimes small fistulas are detected as an incidental finding on echocardiography.

8.9. Recommendations for Management Strategies

CLASS I

1. **Surgeons with training and expertise in CHD should perform operations for management of patients with CAVF. (*Level of Evidence: C*)**
2. **Transcatheter closure of CAVF should be performed only in centers with expertise in such procedures. (*Level of Evidence: C*)**
3. **Transcatheter delineation of CAVF course and access to distal drainage should be performed in all patients with audible continuous murmur and recognition of CAVF. (*Level of Evidence: C*)**

Surgical closure of audible CAVF with appropriate anatomy is recommended in all large CAVFs and in small to moderate CAVFs in the presence of symptoms of myocardial ischemia, threatening arrhythmia, unexplained ventricular dysfunction, or left atrial hypertension. Numerous reports of transcatheter closure with coils or detachable devices describe near or complete CAVF occlusion in attempted closure procedures (124). Criteria for transcatheter closure of CAVF are similar to those used for surgical closure of CAVF. Transcatheter closure of CAVF should be performed only in centers with particular expertise in such intervention.

8.9.1. Preintervention Evaluation After Surgical or Catheterization-Based Repair

Patients with CAVF, even after repair, may still have large, patulous epicardial conduits. Intermediate- and longer-term follow-up of these thin-walled, ectatic coronary arteries after either surgical or transcatheter repair appears mandated.

9. Pulmonary Hypertension/Eisenmenger Physiology

PAH, a progressive increase in PVR, can lead to subpulmonary ventricular failure and death. PAH can frequently be related to pulmonary venous hypertension (most commonly due to left AV valve disorders, volume excess, or SV end-diastolic pressure elevation) and can be classified as World Health Organization PAH class II (due to "left heart disease") with therapies guided toward improving these causes. Within this section, however, the present writing committee will primarily focus on disorders in which PAH is due to other abnormalities and is generally hemodynamically defined as a mean pulmonary artery pressure greater than 25 mm Hg at rest or greater than 30 mm Hg with exercise, pulmonary capillary wedge pressure less than or equal to 15 mm Hg, and PVR greater than 3 mm Hg per L per min per m^2. Particular CHD-related PAH (CHD-PAH) occurs in a number of different scenarios, including the following:

a. "Dynamic" PAH related to high shunt flow that responds to reduction of the shunt
b. Immediate postoperative or "reactive" PAH
c. Late, postoperative PAH
d. Secondary to lesions that cause pulmonary venous hypertension
e. Shunt reversal (eg, Eisenmenger physiology).

These guidelines will largely focus on management of dynamic PAH and Eisenmenger physiology. Recently, CHD-PAH has been recognized to have potentially differing pathogenetic mechanisms, therapeutic goals, treatment plans, and outcomes than idiopathic PAH.

9.1. Clinical Course

9.1.1. Dynamic Congenital Heart Disease–Pulmonary Arterial Hypertension

The development of CHD-PAH associated with systemic–to–pulmonary artery shunts is dependent on both the type and size of the underlying anatomic defect, as well as the magnitude of shunt flow (shear stress and structural changes lead to intravascular and matrix-dependent inflammatory mediator release and changes).

9.2. Recommendations for Evaluation of the Patient With Congenital Heart Disease–Pulmonary Arterial Hypertension

CLASS I

1. **Care of adult patients with CHD-related PAH should be performed in centers that have shared expertise and training in both ACHD and PAH. (*Level of Evidence: C*)**
2. **The evaluation of all ACHD patients with suspected PAH should include noninvasive assessment of cardiovascular anatomy and potential shunting, as detailed below:**
   a. **Pulse oximetry, with and without administration of supplemental oxygen, as appropriate. (*Level of Evidence: C*)**
   b. **Chest x-ray. (*Level of Evidence: C*)**
   c. **ECG. (*Level of Evidence: C*)**
   d. **Diagnostic cardiovascular imaging via TTE, TEE, MRI, or CT as appropriate. (*Level of Evidence: C*)**
   e. **Complete blood count and nuclear lung scintigraphy. (*Level of Evidence: C*)**
3. **If PAH is identified but its causes are not fully recognized, additional testing should include the following:**
   a. **Pulmonary function tests with volumes and diffusion capacity (diffusing capacity of the lung for carbon monoxide). (*Level of Evidence: C*)**

b. Pulmonary embolism–protocol CT with parenchymal lung windows. (*Level of Evidence: C*)
c. Additional testing as appropriate to rule out contributing causes of PAH. (*Level of Evidence: C*)
d. Cardiac catheterization at least once, with potential for vasodilator testing or anatomic intervention, at a center with expertise in catheterization, PAH, and management of CHD-PAH. (*Level of Evidence: C*)

CLASS IIa

1. It is reasonable to include a 6-minute walk test or similar nonmaximal cardiopulmonary exercise test as part of the functional assessment of patients with CHD-PAH. (*Level of Evidence: C*)

9.2.1. Dynamic Congenital Heart Disease–Pulmonary Arterial Hypertension

Surgical experience has suggested that the changes that occur with shunt-mediated PAH are reversible, provided the surgery is performed before pulmonary vascular changes are "fixed." Catheterization-based calculations of pulmonary blood flow (Qp) with isolation of all sources of Qp, individualized measurements of resistance in isolated lung segments, and direct measurement of pulmonary venous pressure are typically used to assess PAH reversibility and the likelihood of surgical success. Acute administration of inhaled (nitric oxide) or intravenously administered (prostacyclin) pulmonary vascular agents is frequently used in such investigations to assess for acute reactivity.

9.2.2. Eisenmenger Physiology

Diagnosis and evaluation of Eisenmenger physiology require a detailed history to look for all possible PAH triggers and a thorough understanding of current and past anatomy, as well as knowledge of all past surgical and medical interventions. Documentation of the size and direction of intracardiac or intravascular shunts present at the atrial, ventricular, or great arterial level is required, as is a precise documentation of pulmonary arteriolar resistance. A suggested basic evaluation of adults with presumed Eisenmenger physiology includes assessment of anatomy, degree of PAH, ventricular function, and both the presence and magnitude of secondary complications.

9.3. Management Strategies

9.3.1. Recommendations for Medical Therapy of Eisenmenger Physiology

CLASS I

1. It is recommended that patients with Eisenmenger syndrome avoid the following activities or exposures, which carry increased risks:
   a. Pregnancy. (*Level of Evidence: B*)
   b. Dehydration. (*Level of Evidence: C*)
   c. Moderate and severe strenuous exercise, particularly isometric exercise. (*Level of Evidence: C*)
   d. Acute exposure to excessive heat (eg, hot tub or sauna). (*Level of Evidence: C*)
   e. Chronic high-altitude exposure, because this causes further reduction in oxygen saturation and increased risk of altitude-related cardiopulmonary complications (particularly at an elevation greater than 5000 feet above sea level). (*Level of Evidence: C*)
   f. Iron deficiency. (*Level of Evidence: B*)
2. Patients with Eisenmenger syndrome should seek prompt therapy for arrhythmias and infections. (*Level of Evidence: C*)
3. Patients with Eisenmenger syndrome should have hemoglobin, platelet count, iron stores, creatinine, and uric acid assessed at least yearly. (*Level of Evidence: C*)
4. Patients with Eisenmenger syndrome should have assessment of digital oximetry, both with and without supplemental oxygen therapy, at least yearly. The presence of oxygen-responsive hypoxemia should be investigated further. (*Level of Evidence: C*)
5. Exclusion of air bubbles in intravenous tubing is recommended as essential during treatment of adults with Eisenmenger syndrome. (*Level of Evidence: C*)
6. Patients with Eisenmenger syndrome should undergo noncardiac surgery and cardiac catheterization only in centers with expertise in the care of such patients. In emergent or urgent situations in which transportation is not feasible, consultation with designated caregivers in centers with expertise in the care of patients with Eisenmenger syndrome should be performed and sustained throughout care. (*Level of Evidence: C*)

CLASS IIa

1. All medications given to patients with Eisenmenger physiology should undergo rigorous review for the potential to change systemic blood pressure, loading conditions, intravascular shunting, and renal or hepatic flow or function. (*Level of Evidence: C*)
2. Pulmonary vasodilator therapy can be beneficial for patients with Eisenmenger physiology because of the potential for improved quality of life. (*Level of Evidence: C*)

An emphasis on patient education and avoidance of destabilizing situations and volume shifts that result in alteration of catecholamines, extreme fatigue, high-altitude exposure, contact with cigarette smoke, changes in renal or hepatic function, or use of medications that may modulate flow to or function of these organs is advocated. Avoidance of pregnancy and iron deficiency and prompt therapy for arrhythmia or infection are recommended. A concept of team planning for all procedures is mandated because of the potential for morbid and mortal outcomes of even the simplest of interventions for any ailment. The optimal type and mode of anesthetic administration should be individualized by experts in the care of persons with Eisenmenger physiology. Risk of right-to-left embolization warrants avoidance of bubbles, and consideration of the use of air filters on all venous catheters still tends to be advocated, although controversy exists regarding the relative benefit obtained compared with meticulous guarding of all intravenous administration systems.

Therapies for adults with CHD-PAH have been limited and have included oxygen, warfarin, diuretics, calcium channel blockers, long-term continuous intravenous epoprostenol, oral prostacyclin analogues, oral endothelin antagonists, oral phosphodiesterase inhibition, and lung or lung/heart transplantation. The benefit of supplemental oxygen administration is a matter of debate, given the conflict between recognized concomitant oxygen-responsive and -unresponsive components

to hypoxemia in many patients and the lack of sufficient trial data to assess benefit (125,126).

In adults with Eisenmenger physiology, recognition of in vivo pulmonary thrombus (127) contrasted with reports of in vitro abnormalities of coagulation in persons with cyanosis (128) has led to debate over the potential benefit of oral anticoagulant therapy, particularly with the concomitant bleeding diathesis inherent in the condition. In patients with active or chronic hemoptysis, anticoagulation is contraindicated.

The theoretical possibility of worsening of right-to-left shunting raises questions about the safety of using pulmonary artery modulating therapies that also have systemic vasodilator potential. Nevertheless, some of these agents (eg, intravenous prostacyclin and oral sildenafil) have yielded improvements in hemodynamics, exercise tolerance, and/or systemic arterial oxygen saturation in limited case studies (128–134). The potential for significant adverse reaction due to these agents has been recognized.

9.4. Key Issues to Evaluate and Follow-Up

9.4.1. Recommendations for Reproduction

CLASS I

1. **Women with severe CHD-PAH, especially those with Eisenmenger physiology, and their partners should be counseled about the absolute avoidance of pregnancy in view of the high risk of maternal death, and they should be educated regarding safe and appropriate methods of contraception. (*Level of Evidence: B*)**
2. **Women with CHD-PAH who become pregnant should:**
   a. **Receive individualized counseling from cardiovascular and obstetric caregivers collaborating in care and with expertise in management of CHD-PAH. (*Level of Evidence: C*)**
   b. **Undergo the earliest possible pregnancy termination after such counseling. (*Level of Evidence: C*)**
3. **Surgical sterilization carries some operative risk for women with CHD-PAH but is a safer option than pregnancy. In view of advances in minimally invasive techniques, the risks and benefits of sterilization modalities should be discussed with an obstetrician experienced in management of high-risk patients, as well as with a cardiac anesthesiologist. (*Level of Evidence: C*)**

CLASS IIb

1. **Pregnancy termination in the last 2 trimesters of pregnancy poses a high risk to the mother. It may be reasonable, however, after the risks of termination are balanced against the risks of continuation of the pregnancy. (*Level of Evidence: C*)**

CLASS III

1. **Pregnancy in women with CHD-PAH, especially those with Eisenmenger physiology, is not recommended and should be absolutely avoided in view of the high risk of maternal mortality. (*Level of Evidence: B*)**
2. **The use of single-barrier contraception alone in women with CHD-PAH is not recommended owing to the frequency of failure. (*Level of Evidence: C*)**
3. **Estrogen-containing contraceptives should be avoided. (*Level of Evidence: C*)**

Pregnancy carries particular risks for individual with CHD-PAH, especially those with Eisenmenger physiology, with mostly older case series suggesting maternal mortality in the latter group of up to 50% and similarly high levels of fetal loss. Even after a successful pregnancy, maternal mortality may be particularly increased in the first several days after delivery (135). Termination of pregnancy, particularly in its mid and later phases, with its concomitant volume and hormonal fluctuations, also carries a high maternal risk. Termination in the first trimester is the safer option. Counseled contraception is strongly advised, although the particular method of such is a matter of debate. Maternal sterilization carries a defined operative risk of mortality, and endoscopic sterilization may be the safer option. Hormonal therapies increase the preexisting potential for thrombosis, although progesterone-only preparations may be considered. Barrier methods have an increased rate of failure, and intrauterine device implantation carries anecdotally increased infection risk, although the highest risk is for local infection in multipartner couples. There is no consensus on comparative contraceptive risks; therefore, the patient should discuss the options with a high-risk obstetrician (maternal fetal medicine specialist).

9.4.2. Recommendations for Follow-Up

CLASS I

1. **Patients with CHD-related PAH should:**
   a. **Have coordinated care under the supervision of a trained CHD and PAH provider and be seen by such individuals at least yearly. (*Level of Evidence: C*)**
   b. **Have yearly comprehensive evaluation of functional capacity and assessment of secondary complications. (*Level of Evidence: C*)**
   c. **Discuss all medication changes or planned interventions with their CHD-related PAH caregiver. (*Level of Evidence: C*)**

CLASS III

1. **Endocardial pacing is not recommended in patients with CHD-PAH with persistent intravascular shunting, and alternative access for pacing leads should be sought (the risks should be individualized) (136). (*Level of Evidence: B*)**

10. Tetralogy of Fallot

Tetralogy of Fallot has 4 components: subpulmonary infundibular stenosis, a VSD, an aorta that overrides the VSD by less than 50% of its diameter, and RV hypertrophy. The single and large VSD is usually in the subaortic position. The pulmonary valve is often small and stenotic. Pulmonary artery anomalies are frequent and include hypoplasia and stenosis. Pulmonary artery hypoplasia may involve the pulmonary trunk or the branch pulmonary arteries. Pulmonary artery stenosis at any of these levels is common.

10.1. Clinical Course (Unrepaired)

10.1.1. Presentation as an Unoperated Patient

An occasional patient is seen with relatively mild pulmonary obstruction and mild cyanosis (the so-called pink tetralogy), in which the diagnosis may not be made until adult life.

10.2. Recommendations for Evaluation and Follow-Up of the Repaired Patient

CLASS I

1. **Patients with repaired tetralogy of Fallot should have at least annual follow-up with a cardiologist who has expertise in ACHD. (*Level of Evidence: C*)**
2. **Patients with tetralogy of Fallot should have echocardiographic examinations and/or MRIs performed by staff with expertise in ACHD. (*Level of Evidence: C*)**
3. **Screening for heritable causes of their condition (eg, 22q11 deletion) should be offered to all patients with tetralogy of Fallot. (*Level of Evidence: C*)**
4. **Before pregnancy or if a genetic syndrome is identified, consultation with a geneticist should be arranged for patients with tetralogy of Fallot. (*Level of Evidence: B*)**
5. **Patients with unrepaired or palliated forms of tetralogy should have a formal evaluation at an ACHD center regarding suitability for repair. (*Level of Evidence: B*)**

All patients should have regular follow-up with a cardiologist who has expertise in ACHD (3,4,29,42,137–139). The frequency, although typically annual, may be determined by the extent and degree of residual abnormalities. Key postoperative issues are summarized below:

- Residual pulmonary regurgitation
- RV dilation and dysfunction due to pulmonary regurgitation, possibly with associated TR
- Residual RVOT obstruction
- Branch pulmonary artery stenosis or hypoplasia
- Sustained ventricular tachycardia
- Sudden cardiac death
- AV block, atrial flutter, and/or atrial fibrillation
- Progressive AR
- Syndromal associations.

The most common problem encountered in the adult patient after repair is that of pulmonary regurgitation.

10.2.1. Recommendation for Imaging

CLASS 1

1. **Comprehensive echocardiographic imaging should be performed in a regional ACHD center to evaluate the anatomy and hemodynamics in patients with repaired tetralogy of Fallot. (*Level of Evidence: B*)**

Echocardiography is usually very helpful in assessing a patient after repair of tetralogy. The presence and severity of residual RVOT obstruction and pulmonary regurgitation can usually be assessed along with the presence or absence of TR. The tricuspid regurgitant velocity facilitates measurement of the RV pressure. A residual VSD may be seen. RV volume and wall motion are not reliably quantified by standard techniques, although size and function can be determined qualitatively. Doppler measurement of the RV myocardial performance index may be a useful adjunct to serial assessment of RV systolic function. Atrial size can be assessed. Aortic root dilation and AR should be sought and evaluated at regular intervals.

MRI is now seen as the reference standard (140,141) for assessment of RV volume and systolic function. It can be helpful in assessing the severity of pulmonary regurgitation and in evaluating important associated pathology, especially involving the pulmonary arteries and the ascending aorta. Left-sided heart disease can also be evaluated. Recently, CT scanning has become available (142–144) to make similar measurements of RV volume and systolic function and is potentially helpful in patients who cannot have an MRI.

10.3. Recommendations for Diagnostic and Interventional Catheterization for Adults With Tetralogy of Fallot

CLASS I

1. **Catheterization of adults with tetralogy of Fallot should be performed in regional centers with expertise in ACHD. (*Level of Evidence: C*)**
2. **Coronary artery delineation should be performed before any intervention for the RVOT. (*Level of Evidence: C*)**
3. **Interventional catheterization in an ACHD center is indicated for patients with previously repaired tetralogy of Fallot with the following indications:**
   a. **To eliminate residual native or palliative systemic–pulmonary artery shunts. (*Level of Evidence: B*)**
   b. **To manage coronary artery disease. (*Level of Evidence: B*)**

CLASS IIa

1. **Interventional catheterization in an ACHD center is reasonable in patients with repaired tetralogy of Fallot to eliminate a residual ASD or VSD with a left-to-right shunt greater than 1.5:1 if it is in an appropriate anatomic location. (*Level of Evidence: C*)**

CLASS IIb

1. **In adults with repaired tetralogy of Fallot, catheterization may be considered to better define potentially treatable causes of otherwise unexplained LV or RV dysfunction, fluid retention, chest pain, or cyanosis. In these circumstances, transcatheter interventions may include:**
   a. **Elimination of residual shunts or aortopulmonary collateral vessels. (*Level of Evidence: C*)**
   b. **Dilation (with or without stent implantation) of RVOT obstruction. (*Level of Evidence: B*)**
   c. **Elimination of additional muscular or patch-margin VSD. (*Level of Evidence: C*)**
   d. **Elimination of residual ASD. (*Level of Evidence: B*)**

Interventional catheterization in previously repaired tetralogy of Fallot should be planned carefully with the medical and surgical team in an ACHD center. Although there is experience in the use of catheter devices to close residual shunts, experience with the use of percutaneous stent-valve implants in the RV outflow for patients with pulmonary regurgitation and right-sided heart failure is recent, and efficacy/safety remains undefined, but this technique appears promising.

For the unusual case of a patient with tetralogy of Fallot who has undergone palliation with a surgical shunt, catheterization should be performed to assess the potential for repair. The presence or absence of additional muscular VSDs may be

determined, as well as the course and anatomy of the epicardial coronary arteries. The pulmonary architecture and vascular pressure and resistance should be delineated, because pulmonary artery distortion and PAH are frequent sequelae of palliative surgical shunts. Potential catheter interventions include the elimination of collateral vessels or systemic–pulmonary artery shunts, dilation/stent implantation of obstructed pulmonary arteries, and, more recently, the possibility of percutaneous pulmonary valve implantation. Heart catheterization is not used routinely in the assessment of patients who have undergone repair, except when surgery or other therapy is being considered or for evaluation of the pulmonary and coronary arteries.

10.3.1. Branch Pulmonary Artery Angioplasty

Balloon angioplasty of a branch pulmonary artery may be considered when the RV pressure is greater than 50% of the systemic level or at lower pressure when there is RV dysfunction. Balloon pulmonary artery angioplasty may also be considered when there is unbalanced pulmonary blood flow greater than 75%, 25%, or otherwise unexplained dyspnea with severe vascular stenosis (145,146). Pulmonary artery balloon angioplasty may be an effective way to reduce obstruction to pulmonary blood flow, thereby increasing pulmonary vascular capacitance and decreasing PVR (147). A transcatheter approach to the management of residual muscular or patch-margin VSDs (indications for which typically include a Qp/Qs greater than 1.5 to 2.0, or less in the setting of PAH, left atrial hypertension, or LV failure) remains an effective alternative to reoperative surgical closure (148,149).

10.4. Recommendations for Surgery for Adults With Previous Repair of Tetralogy of Fallot

CLASS I

1. **Surgeons with training and expertise in CHD should perform operations in adults with previous repair of tetralogy of Fallot. (*Level of Evidence: C*)**
2. **Pulmonary valve replacement is indicated for severe pulmonary regurgitation and symptoms or decreased exercise tolerance. (*Level of Evidence: B*)**
3. **Coronary artery anatomy, specifically the possibility of an anomalous anterior descending coronary artery across the RVOT, should be ascertained before operative intervention. (*Level of Evidence: C*)**

CLASS IIa

1. **Pulmonary valve replacement is reasonable in adults with previous tetralogy of Fallot, severe pulmonary regurgitation, and any of the following:**
   a. **Moderate to severe RV dysfunction. (*Level of Evidence: B*)**
   b. **Moderate to severe RV enlargement. (*Level of Evidence: B*)**
   c. **Development of symptomatic or sustained atrial and/or ventricular arrhythmias. (*Level of Evidence: C*)**
   d. **Moderate to severe TR. (*Level of Evidence: C*)**
2. **Collaboration between ACHD surgeons and ACHD interventional cardiologists, which may include preoperative stenting, intraoperative stenting, or intraoperative patch angioplasty, is reasonable to determine the most feasible treatment for pulmonary artery stenosis. (*Level of Evidence: C*)**
3. **Surgery is reasonable in adults with prior repair of tetralogy of Fallot and residual RVOT obstruction (valvular or subvalvular) and any of the following indications:**
   a. **Residual RVOT obstruction (valvular or subvalvular) with peak instantaneous echocardiography gradient greater than 50 mm Hg. (*Level of Evidence: C*)**
   b. **Residual RVOT obstruction (valvular or subvalvular) with RV/LV pressure ratio greater than 0.7. (*Level of Evidence: C*)**
   c. **Residual RVOT obstruction (valvular or subvalvular) with progressive and/or severe dilatation of the right ventricle with dysfunction. (*Level of Evidence: C*)**
   d. **Residual VSD with a left-to-right shunt greater than 1.5:1. (*Level of Evidence: B*)**
   e. **Severe AR with associated symptoms or more than mild LV dysfunction. (*Level of Evidence: C*)**
   f. **A combination of multiple residual lesions (eg, VSD and RVOT obstruction) leading to RV enlargement or reduced RV function. (*Level of Evidence: C*)**

Late survival after tetralogy repair is excellent; 35-year survival is approximately 85%. The need for reintervention, usually for pulmonary valve insertion, increases after the second decade of life. Surgical intervention is indicated for symptomatic patients with severe pulmonary regurgitation or asymptomatic patients with severe PS or pulmonary regurgitation in association with signs of progressive or severe RV enlargement or dysfunction. Patients with RV–to–pulmonary artery conduit repairs often require further intervention for conduit stenosis or regurgitation. Any intervention that involves the RVOT requires careful preoperative assessment of the coronary anatomy to avoid interruption of an important coronary vessel. Some patients experience increasing AR, which requires surgical intervention.

10.5. Key Issues to Evaluate and Follow-Up

10.5.1. Recommendations for Arrhythmias: Pacemaker/Electrophysiology Testing

CLASS I

1. **Annual surveillance with history, ECG, assessment of RV function, and periodic exercise testing is recommended for patients with pacemakers/automatic implantable cardioverter defibrillators. (*Level of Evidence: C*)**

CLASS IIa

1. **Periodic Holter monitoring can be beneficial as part of routine follow-up. The frequency should be individualized depending on the hemodynamics and clinical suspicion of arrhythmia. (*Level of Evidence: C*)**

CLASS IIb

1. **Electrophysiology testing in an ACHD center may be reasonable to define suspected arrhythmias in adults with tetralogy of Fallot. (*Level of Evidence: C*)**

Despite overall excellent hemodynamic outcomes after surgery for tetralogy of Fallot, there remains a concerning incidence of unexpected sudden death during long-term

follow-up. Ventricular tachycardia appears to be the mechanism for most of these events, although rapidly conducted intra-atrial reentrant tachycardia (IART; atrial flutter) or AV block may be responsible in some cases. The incidence of sudden death for the adult tetralogy population can be estimated from several large series to be on the order of 2.5% per decade of follow-up (150–154).

Worrisome symptoms (ie, palpitations, dizziness, or an episode of syncope) should obviously heighten the index of suspicion for serious arrhythmias in tetralogy patients and trigger a prompt evaluation, including hemodynamic catheterization and electrophysiology study. Repairable hemodynamic issues may also be identified by echocardiography or cardiac catheterization that could possibly shift therapy toward a surgical approach, such as closure of a residual septal defect or relief of valve regurgitation, combined with intraoperative ventricular tachycardia mapping and ablation. Serious symptoms in adult patients with tetralogy of Fallot (ie, documented sustained ventricular tachycardia or cardiac arrest) are now managed with implantable cardioverter defibrillators at almost all centers.

11. Dextro-Transposition of the Great Arteries

TGA implies that each great artery arises from the wrong ventricle. TGA is AV concordance with ventriculoarterial discordance. As such, d-TGA implies that the aorta arises rightward and anterior to the pulmonary artery and arises from the systemic right ventricle.

Patients with d-TGA by definition have abnormal origins of the aorta and pulmonary artery. Anomalies of the coronary ostia are also common, and clear delineation is required. Additional congenital cardiac lesions include VSD, which occurs in up to 45% of cases, LVOT obstruction in approximately 25% of cases, and coarctation of the aorta in approximately 5%.

11.1. Recommendation for Evaluation of the Operated Patient With Dextro-Transposition of the Great Arteries

CLASS I

1. **Patients with repaired d-TGA should have annual follow-up with a cardiologist who has expertise in the management of ACHD patients. (*Level of Evidence: C*)**

Most adults born with d-TGA will have had 1 or more operations in childhood. All patients should have regular follow-up with a cardiologist who has expertise in ACHD. The frequency may be determined by the degree of residual hemodynamic abnormalities, and these become more common, along with the occurrence of arrhythmias, with advancing age.

All operated d-TGA patients should be seen at least annually by a specialist in an ACHD regional center, with attention given to rhythm disorders, as well as ventricular and valvular function. Stress testing, including cardiopulmonary stress testing, should be applied selectively. If specialized testing is performed, it is best done at a regional center. If significant abnormalities are uncovered by these examinations, or if the patient is symptomatic, more frequent follow-up visits are indicated.

11.1.1. Clinical Features and Evaluation of Dextro-Transposition of the Great Arteries After Atrial Baffle Procedure

Because the ASO only gained acceptance in the 1980s, many adults with d-TGA will have had a Mustard or Senning procedure. These procedures involve an atrial baffle that redirects the systemic venous blood to the mitral valve and left ventricle, which remains committed to the pulmonary artery. The pulmonary venous blood is redirected to the tricuspid valve and right ventricle, which remains committed to the aorta.

The atrial baffle (Mustard or Senning) procedure for d-TGA has characteristic long-term problems. The most common early structural complications include baffle obstruction, which most commonly affects the superior limb rather than the inferior vena cava. Facial suffusion and "superior vena cava syndrome" may result. Inferior vena cava obstruction may cause hepatic congestion or even cirrhosis. Baffle leaks occur in up to 25% of patients. Most are small but many pose a risk of paradoxical embolus, particularly in the setting of atrial arrythmias and an endocardial pacemaker. Pulmonary venous obstruction may also occur but less common. Subpulmonary stenosis and PS may occur, in part related to the abnormal geometry of the left ventricle, which becomes distorted and compressed by the enlarged systemic right ventricle. Long-term, the most important complication after atrial baffle is failure of the systemic right ventricle and systemic TR. These complications have a major impact on morbidity and mortality. Important but less common complications include PAH, residual VSD, dynamic subpulmonic stenosis, and a host of conduction and arrhythmia disturbances with the potential for implantation of permanent pacemakers or sudden death (23,155–157).

11.1.2. Imaging for Dextro-Transposition of the Great Arteries After Atrial Baffle Procedure

CLASS I

1. **In patients with d-TGA repaired by atrial baffle procedure, comprehensive echocardiographic imaging should be performed in a regional ACHD center to evaluate the anatomy and hemodynamics. (*Level of Evidence: B*)**
2. **Additional imaging with TEE, CT, or MRI, as appropriate, should be performed in a regional ACHD center to evaluate the great arteries and veins, as well as ventricular function, in patients with prior atrial baffle repair of d-TGA. (*Level of Evidence: B*)**

CLASS IIa

1. **Echocardiography contrast injection with agitated saline can be useful to evaluate baffle anatomy and shunting in patients with previously repaired d-TGA after atrial baffle. (*Level of Evidence: B*)**
2. **TEE can be effective for more detailed baffle evaluation for patients with d-TGA. (*Level of Evidence: B*)**

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Evaluation for intra-atrial baffle anatomy and shunting or obstruction may warrant echocardiography contrast injection. Assessment of systemic RV function is challenging by echocardiography. In addition to routine evaluation of ventricular size and function, measurement of the dP/dt of the AV regurgitant jet, Doppler tissue indices of annular motion, and the myocardial performance index may provide further insight (155–160). Tissue Doppler evaluation of myocardial acceleration during isovolumic contraction has been validated as a sensitive, noninvasive method to assess RV contractility (161,162). The myocardial performance index has the advantage of representing indices of both systolic and diastolic function without geometric constraints and has shown a relationship to brain natriuretic peptide levels in ACHD patients (163). The coronary anatomy may be difficult to evaluate by echocardiography in the adult patient.

TEE is used to provide complementary information, including imaging of atrial anatomy, the presence of baffle leak or obstruction, and intracardiac thrombus. Radiological imaging with MRI or CT can be used to further assess atrial baffle patency, systemic ventricular function, and coronary anatomy. MRI or magnetic resonance angiography is usually superior for evaluation of the extracardiac great arteries and veins.

11.1.3. Cardiac Catheterization

Cardiac catheterization is used to assess hemodynamics, baffle leak, superior vena cava or inferior vena cava pathway obstruction, pulmonary venous pathway obstruction, myocardial ischemia, unexplained systemic RV dysfunction, significant LV dysfunction (subpulmonary [LVOT] obstruction), or PAH, with potential for vasodilator testing. Cardiac catheterization in patients after atrial baffle also provides the opportunity for intervention. For adults after palliative atrial baffle repair for d-TGA, VSD, and pulmonary vascular disease, catheterization may be indicated to assess the potential for pulmonary artery vasomodulator therapy.

11.2. Clinical Features and Evaluation of Dextro-Transposition of the Great Arteries After Arterial Switch Operation

Long-term concerns after the ASO include coronary insufficiency, myocardial ischemia, ventricular dysfunction and arrhythmias, and issues regarding stenosis at the great arterial anastomotic sites, as well as development of aortic or pulmonary regurgitation. Significant neoaortic root dilatation and neoaortic valve regurgitation may develop over time, in part related to older age at the time of ASO or to an associated VSD with previous pulmonary artery banding (164).

11.2.1. Recommendations for Imaging for Dextro-Transposition of the Great Arteries After Arterial Switch Operation

CLASS I

1. **Comprehensive echocardiographic imaging to evaluate the anatomy and hemodynamics in patients with d-TGA and prior ASO repair should be performed at least every 2 years at a center with expertise in ACHD. (*Level of Evidence: C*)**
2. **After prior ASO repair for d-TGA, all adults should have at least 1 evaluation of coronary artery patency. Coronary angiography should be performed if this cannot be established noninvasively. (*Level of Evidence: C*)**

CLASS IIa

1. **Periodic MRI or CT can be considered appropriate to evaluate the anatomy and hemodynamics in more detail. (*Level of Evidence: C*)**

Echocardiography after ASO may demonstrate minimal findings or 1 or more of the recognized complications after ASO, which include the following: 1) stenosis at the arterial anastomotic sites, most commonly PS (165); 2) aortic root dilatation; and 3) neoaortic valve regurgitation (native pulmonary valve) (166). Coronary complications cannot be assessed adequately by echocardiography, but stress echocardiography may facilitate detection of ischemia. CT angiography has been used recently. Patients with intramural or single coronary arteries have increased mortality compared with those with the typical coronary pattern (167).

11.2.2. Recommendation for Cardiac Catheterization After Arterial Switch Operation

CLASS IIa

1. **Coronary angiography is reasonable in all adults with d-TGA after ASO to rule out significant coronary artery obstruction. (*Level of Evidence: C*)**

Coronary ischemia is a recognized late complication after ASO, with concern about ischemia or infarction reported in up to 8% of patients after ASO. These complications are due to reimplantation of the coronary arteries during surgery (165). Noninvasive testing for coronary ischemia may not be sufficiently sensitive, and coronary arteriography has been recommended 5, 10, and 15 years after ASO to detect significant late coronary artery stenosis. Aortic root angiography is recommended to detect ostial coronary artery disease.

Hemodynamic cardiac catheterization is used to assess pulmonary and aortic anastomosis obstruction when incompletely evaluated by other imaging modalities. Cardiac catheterization in patients after ASO also provides the opportunity for intervention.

11.3. Clinical Features and Evaluation of Dextro-Transposition of the Great Arteries After Rastelli Operation

The Rastelli operation for a combination of d-TGA, PS, and VSD has recognized complications that include RVOT or pulmonary conduit obstruction, superimposed RV failure, and TR. LVOT obstruction may also occur from the intraventricular baffle, arrhythmias from atriotomy and/or ventriculotomy incisions, residual VSD, myocardial hypertrophy, chamber enlargement, aortic root dilatation, and aortic valve regurgitation. The 3 most common late causes of postoperative death are sudden cardiac death, heart failure, and reoperation.

11.4. Recommendations for Diagnostic Catheterization for Adults With Repaired Dextro-Transposition of the Great Arteries

CLASS I

1. Diagnostic catheterization of the adult with d-TGA should be performed in centers with expertise in the catheterization and management of ACHD patients. (*Level of Evidence: C*)

CLASS IIa

1. For adults with d-TGA after atrial baffle procedure (Mustard or Senning), diagnostic catheterization can be beneficial to assist in the following:
   a. Hemodynamic assessment. (*Level of Evidence: C*)
   b. Assessment of baffle leak. (*Level of Evidence: B*)
   c. Assessment of superior vena cava or inferior vena cava pathway obstruction. (*Level of Evidence: B*)
   d. Assessment of pulmonary venous pathway obstruction. (*Level of Evidence: B*)
   e. Suspected myocardial ischemia or unexplained systemic RV dysfunction. (*Level of Evidence: B*)
   f. Significant LV outflow obstruction at any level (LV pressure greater than 50% of systemic levels, or less in the setting of RV dysfunction). (*Level of Evidence: B*)
   g. Assessment of PAH, with potential for vasodilator testing. (*Level of Evidence: C*)
2. For adults with d-TGA, VSD, and PS, after Rastelli-type repair, diagnostic catheterization can be beneficial to assist in the following:
   a. Coronary artery delineation before any intervention for RVOT obstruction. (*Level of Evidence: C*)
   b. Assessment of residual VSD. (*Level of Evidence: C*)
   c. Assessment of PAH, with potential for vasodilator testing. (*Level of Evidence: C*)
   d. Assessment of subaortic obstruction across the left ventricle–to–aorta tunnel. (*Level of Evidence: C*)

11.5. Recommendations for Interventional Catheterization for Adults With Dextro-Transposition of the Great Arteries

CLASS IIa

1. Interventional catheterization of the adult with d-TGA can be performed in centers with expertise in the catheterization and management of ACHD patients. (*Level of Evidence: C*)
2. For adults with d-TGA after atrial baffle procedure (Mustard or Senning), interventional catheterization can be beneficial to assist in the following:
   a. Occlusion of baffle leak. (*Level of Evidence: B*)
   b. Dilation or stenting of superior vena cava or inferior vena cava pathway obstruction. (*Level of Evidence: B*)
   c. Dilation or stenting of pulmonary venous pathway obstruction. (*Level of Evidence: B*)
3. For adults with d-TGA after ASO, interventional catheterization can be beneficial to assist in dilation or stenting of supravalvular and branch pulmonary artery stenosis. (*Level of Evidence: B*)
4. For adults with d-TGA, VSD, and PS, after Rastelli-type repair, interventional catheterization can be beneficial to assist in the following:
   a. Dilation with or without stent implantation of conduit obstruction (RV pressure greater than 50% of systemic levels, or peak-to-peak gradient greater than 30 mm Hg; these indications may be lessened in the setting of RV dysfunction). (*Level of Evidence: C*)
   b. Device closure of residual VSD. (*Level of Evidence: C*)

11.6. Recommendations for Surgical Interventions

11.6.1. After Atrial Baffle Procedure (Mustard, Senning)

CLASS I

1. Surgeons with training and expertise in CHD should perform operations in patients with d-TGA and the following indications:
   a. Moderate to severe systemic (morphological tricuspid) AV valve regurgitation without significant ventricular dysfunction (168). (*Level of Evidence: B*)
   b. Baffle leak with left-to-right shunt greater than 1.5:1, right-to-left shunt with arterial desaturation at rest or with exercise, symptoms, and progressive ventricular enlargement that is not amenable to device intervention. (*Level of Evidence: B*)
   c. Superior vena cava or inferior vena cava obstruction not amenable to percutaneous treatment. (*Level of Evidence: B*)
   d. Pulmonary venous pathway obstruction not amenable to percutaneous intervention. (*Level of Evidence: B*)
   e. Symptomatic severe subpulmonary stenosis. (*Level of Evidence: B*)

11.6.2. After Arterial Switch Operation

CLASS I

1. It is recommended that surgery be performed in patients after the ASO with the following indications:
   a. RVOT obstruction peak-to-peak gradient greater than 50 mm Hg or right ventricle/left ventricle pressure ratio greater than 0.7, not amenable or responsive to percutaneous treatment; lesser degrees of obstruction if pregnancy is planned, greater degrees of exercise are desired, or concomitant severe pulmonary regurgitation is present. (*Level of Evidence: C*)
   b. Coronary artery abnormality with myocardial ischemia not amenable to percutaneous intervention. (*Level of Evidence: C*)
   c. Severe neoaortic valve regurgitation. (*Level of Evidence: C*)
   d. Severe neoaortic root dilatation (greater than 55 mm) after ASO (169). (This recommendation is based on data for other forms of degenerative aortic root aneurysms.) (*Level of Evidence: C*)

11.6.3. After Rastelli Procedure

CLASS I

1. Reoperation for conduit and/or valve replacement after Rastelli repair of d-TGA is recommended in patients with the following indications:
   a. Conduit obstruction peak-to-peak gradient greater than 50 mm Hg. (*Level of Evidence: C*)
   b. RV/LV pressure ratio greater than 0.7. (*Level of Evidence: C*)
   c. Lesser degrees of conduit obstruction if pregnancy is being planned or greater degrees of exercise are desired. (*Level of Evidence: C*)
   d. Subaortic (baffle) obstruction (mean gradient greater than 50 mm Hg). (*Level of Evidence: C*)

e. Lesser degrees of subaortic baffle obstruction if LV hypertrophy is present, pregnancy is being planned, or greater degrees of exercise are desired. (*Level of Evidence: C*)
f. Presence of concomitant severe AR. (*Level of Evidence: C*)

2. Reoperation for conduit regurgitation after Rastelli repair of d-TGA is recommended in patients with severe conduit regurgitation and the following indications:
   a. Symptoms or declining exercise tolerance. (*Level of Evidence: C*)
   b. Severely depressed RV function. (*Level of Evidence: C*)
   c. Severe RV enlargement. (*Level of Evidence: C*)
   d. Development/progression of atrial or ventricular arrhythmias. (*Level of Evidence: C*)
   e. More than moderate TR. (*Level of Evidence: C*)
3. Collaboration between surgeons and interventional cardiologists, which may include preoperative stenting, intraoperative stenting, or intraoperative patch angioplasty with or without conduit replacements, is recommended to determine the most feasible treatment for pulmonary artery stenosis. (*Level of Evidence: C*)
4. Surgical closure of residual VSD in adults after Rastelli repair of d-TGA is recommended with the following indicators:
   a. Qp/Qs greater than 1.5:1. (*Level of Evidence: B*)
   b. Systolic pulmonary artery pressure greater than 50 mm Hg. (*Level of Evidence: B*)
   c. Increasing LV size from volume overload. (*Level of Evidence: C*)
   d. Decreasing RV function from pressure overload. (*Level of Evidence: C*)
   e. RVOT obstruction (peak instantaneous gradient greater than 50 mm Hg). (*Level of Evidence: B*)
   f. Pulmonary artery pressure less than two thirds of systemic pressure, or PVR less than two thirds of systemic vascular resistance, with a net left-to-right shunt of 1.5:1, or a decrease in pulmonary artery pressure with pulmonary vasodilators (oxygen, nitric oxide, or prostaglandins). (*Level of Evidence: B*)
5. Surgery is recommended after Rastelli repair of d-TGA in adults with branch pulmonary artery stenosis not amenable to percutaneous treatment. (*Level of Evidence: C*)
6. In the presence of a residual intracardiac shunt or significant systemic venous obstruction, permanent pacing, if indicated, should be performed with epicardial leads (168). (*Level of Evidence: B*)

CLASS IIa

1. A concomitant Maze procedure can be effective for the treatment of intermittent or chronic atrial tachyarrhythmias in adults with d-TGA requiring reoperation for any reason. (*Level of Evidence: C*)

11.6.4. Reoperation After Atrial Baffle Procedure

Reoperation after atrial baffle procedure in adults is recommended for patients with a baffle leak that is not amenable to device intervention, demonstrates a left-to-right shunt greater than 1.5:1 or a right-to-left shunt with arterial desaturation at rest or with exercise, symptoms, or progressive ventricular enlargement. Although late conversion to an ASO has been attempted in some centers, it has not proved successful and is not generally considered a reasonable option for the management of systemic ventricular failure in patients with TGA.

Patients with severe symptomatic superior or inferior vena cava obstruction or pulmonary venous pathway obstruction not amenable to percutaneous treatment should be referred for operative intervention. Patients with severe symptomatic subpulmonary stenosis should also be considered for operative intervention.

Severe symptomatic systemic AV (morphological tricuspid) valve regurgitation may prompt surgical referral when the problem relates to intrinsic tricuspid valve disease and is not secondary to systemic ventricular dysfunction. This is a rare occurrence, because most TR after atrial baffle procedure is secondary to systemic ventricular dysfunction. Alternative techniques include tricuspid valve replacement, pulmonary artery band placement, and transplantation.

11.6.5. Reoperation After Arterial Switch Operation

Reoperation after ASO should be considered for adults with the following: severe RVOT obstruction peak-to-peak gradient greater than 50 mm Hg or RV/LV pressure ratio greater than 0.7, not amenable or responsive to percutaneous treatment, or lesser degrees of obstruction that are dynamic if pregnancy is planned or greater degrees of exercise are desired. Pulmonary valve replacement or repair should be considered when severe pulmonary regurgitation is present and there is significant RV dilatation or RV dysfunction.

Coronary ostial stenosis late after the ASO may be repaired by coronary bypass grafting or ostial arterioplasty techniques. Patients who have developed neoaortic root dilation without severe AR may be treated with valve-sparing root-replacement techniques when the aortic root diameter is greater than 55 mm.

11.6.6. Other Reoperation Options

A concomitant Maze procedure can be effective for the treatment of intermittent or chronic atrial tachyarrhythmias in adults with d-TGA who are undergoing reoperation. This option for arrhythmia management should be considered preoperatively.

Cardiac transplantation may be required in failing systemic ventricular circulations; given that there are frequently anomalous venous or arterial connections, cardiac malpositioning, or both, technical anastomotic issues are common (170). In addition, many patients have had multiple surgeries and have more adhesions, which makes postoperative bleeding more of a concern, with the need for more blood transfusions and consequently more antigenic exposure, which leads to accelerated rejection.

11.7. Recommendations for Electrophysiology Testing/Pacing Issues in Dextro-Transposition of the Great Arteries

CLASS I

1. Clinicians should be mindful of the risk of sudden arrhythmic death among adults after atrial baffle repair of d-TGA. These events usually relate to ventricular tachycardia but may be caused in some cases by rapidly conducted IART or progressive AV block. (*Level of Evidence: B*)

2. **Consultation with an electrophysiologist who is experienced with CHD is recommended to assist with treatment decisions. (*Level of Evidence: B*)**
3. **Pacemaker implantation is recommended for patients with d-TGA with either symptomatic sinus bradycardia or sick sinus syndrome. (*Level of Evidence: B*)**

CLASS IIa

1. **Routine surveillance with history, ECG, assessment of RV function, and periodic Holter monitoring can be beneficial as part of routine follow-up. (*Level of Evidence: B*)**

The most significant arrhythmia issue facing adults with d-TGA is the high incidence of tachy-brady syndrome that occurs in those who have undergone the Mustard or Senning operations (171). There is little doubt that these arrhythmias relate directly to the extensive suture lines created during atrial baffling. Some degree of sinus node dysfunction will be observed in more than half of the Mustard and Senning populations by the time they reach adulthood, probably due to surgical trauma in the vicinity of the sinus node or its arterial supply during creation of the superior vena cava limb of the atrial baffle (171). In addition, up to 30% of these patients will develop episodic IART or atrial flutter, which typically involves a macroreentry circuit around the atrial border of the tricuspid valve that is supported by the narrow conduction corridor between the inferior vena cava limb of the baffle and the valve ring (172). Patients can become highly symptomatic from either tachycardia or bradycardia, including the possibility of sudden death due to an episode of rapidly conducted IART (173). In patients who have advanced dysfunction of their systemic right ventricle, ventricular arrhythmias may also develop.

11.8. Key Issues to Evaluate and Follow-Up

11.8.1. Recommendations for Endocarditis Prophylaxis

CLASS IIa

1. **Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa is reasonable in those with the following indications:**
   a. **Prosthetic cardiac valve. (*Level of Evidence: B*)**
   b. **Previous IE. (*Level of Evidence: B*)**
   c. **Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: B*)**
   d. **Completely repaired CHD with prosthetic materials, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure. (*Level of Evidence: B*)**
   e. **Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization. (*Level of Evidence: B*)**
2. **It is reasonable to consider antibiotic prophylaxis against IE before vaginal delivery at the time of membrane rupture in select patients with the highest risk of adverse outcomes. This includes patients with the following indications:**
   a. **Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: C*)**
   b. **Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: C*)**

CLASS III

1. **Prophylaxis against IE is not recommended for nondental procedures (such as esophagogastroduodenoscopy or colonoscopy) in the absence of active infection. (*Level of Evidence: C*)**

11.8.2. Recommendation for Reproduction

CLASS I

1. **Before women with d-TGA contemplate pregnancy, a comprehensive clinical, functional, and echocardiographic evaluation should be performed at a center with expertise in ACHD. (*Level of Evidence: C*)**

Comprehensive evaluation is recommended before pregnancy in all patients with d-TGA and prior repair. For patients after atrial baffle, major prepregnancy concerns include ventricular function assessment, systemic AV regurgitation, and atrial arrhythmias. There is a small but recognized risk of cardiovascular complications during pregnancy after the atrial baffle procedure. The physiological stresses of pregnancy, although clinically well tolerated late after a Mustard procedure, carry an increased risk of RV dysfunction that may be irreversible (174).

After a Rastelli operation, pregnancy should be well tolerated, assuming the absence of LV or RV obstruction and preservation of ventricular function. Isolated reports are available on the outcome of pregnancy after ASO. In the absence of important cardiovascular residua, pregnancy is well tolerated. A comprehensive anatomic and functional assessment, including assessment of coronary artery anatomy, is recommended before a patient proceeds with pregnancy.

12. Congenitally Corrected Transposition of the Great Arteries

Congenitally corrected TGA (CCTGA) is a complex congenital anomaly with a wide spectrum of morphological features and clinical profiles. The term “corrected” refers to the physiologically normal direction of blood flow caused by this “double discordance,” which makes the term “corrected” misleading (175). The term “l-transposition” is synonymous with CCTGA and indicates that the morphological RV is to the left of the morphological LV. In addition, the aorta is usually anterior to and to the left of the pulmonary artery.

12.1. Associated Lesions

Only 1% of cases are uncomplicated, that is, they do not have associated anomalies. Frequently associated structural anomalies include the following:

- VSD occurs in 70% of patients and is usually perimembranous.
- PS occurs in 40% of patients and is often subvalvular.
- Some abnormality of the systemic AV valve occurs in 90% of patients. Most commonly, this is an Ebstein-like malformation in which the valve is displaced inferiorly toward the cardiac apex (176).

The AV node and His bundle are often in an unusual position, and an accessory AV node is present in many patients (177). Conduction abnormalities are also common, with spontaneous complete heart block occurring at a rate of approximately 2% per year, and these are related to the abnormal position of the AV node (177–179).

12.2. Presentation in Adulthood: Unoperated

Some patients were diagnosed in childhood but did not require operation. In some adults, the diagnosis is made for the first time because of a heart murmur or incidentally when an ECG, chest x-ray, or echocardiogram is performed for other reasons (180). The diagnosis is often missed in cardiology practice because of the failure to recognize the abnormal position of the ventricles and the associated AV valves (181).

12.3. Recommendations for Evaluation and Follow-Up of Patients With Congenitally Corrected Transposition of the Great Arteries

CLASS I

1. **All patients with CCTGA should have a regular follow-up with a cardiologist who has expertise in ACHD. (*Level of Evidence: C*)**
2. **Echocardiography-Doppler study and/or MRI should be performed yearly or at least every other year by staff trained in imaging complex CHD. (*Level of Evidence: C*)**
3. **The following diagnostic evaluations are recommended for patients with CCTGA:**
   a. **ECG. (*Level of Evidence: C*)**
   b. **Chest x-ray. (*Level of Evidence: C*)**
   c. **Echocardiography-Doppler study. (*Level of Evidence: C*)**
   d. **MRI. (*Level of Evidence: C*)**
   e. **Exercise testing. (*Level of Evidence: C*)**

The frequency of follow-up visits may be determined by the presence or absence of associated lesions but is often annual. More frequent visits may be necessary for those with ventricular dysfunction and systemic AV valve regurgitation, regardless of whether they are symptomatic. Clinical examination, ECG, chest x-ray, and cardiopulmonary exercise testing will usually be performed. If progression of heart block is suspected by history or ECG, ambulatory ECG monitoring for 24 hours should be considered. Patients who have implantation of an endocardial pacemaker warrant more frequent observation, because "septal shift" may cause deterioration in systemic ventricle (SV) dysfunction.

12.4. Interventional Therapy

12.4.1. Recommendations for Catheter Interventions

CLASS IIa

1. **For patients with unrepaired CCTGA, cardiac catheterization can be effective to assess the following:**
   a. **Hemodynamic status in the setting of arrhythmia. (*Level of Evidence: C*)**
   b. **Unexplained SV dysfunction, to define the degree of systemic AV valve regurgitation, degree of intracardiac shunting, and coronary artery anatomy. (*Level of Evidence: C*)**
   c. **Unexplained volume retention or cyanosis, especially when noninvasive assessment of pulmonary outflow obstruction is limited. (*Level of Evidence: C*)**

Combined with noninvasive imaging techniques, diagnostic and interventional cardiopulmonary catheterization play important roles in the management of many adults with CCTGA, both in the unoperated native state and after surgical repair with VSD patch or Rastelli-type LV–pulmonary artery connections.

12.4.2. Recommendations for Surgical Intervention

CLASS I

1. **Surgeons with training and expertise in CHD should perform operations for patients with CCTGA for the following indications:**
   a. **Unrepaired CCTGA and severe AV valve regurgitation. (*Level of Evidence: B*)**
   b. **Anatomic repair with atrial and arterial level switch/Rastelli repair in cases in which the left ventricle is functioning at systemic pressures. (*Level of Evidence: B*)**
   c. **Simple VSD closure when the VSD is not favorable for LV-to-aorta baffling or is restrictive. (*Level of Evidence: B*)**
   d. **LV–to–pulmonary artery conduit in rare cases with LV dysfunction and severe LV outflow obstruction. (*Level of Evidence: B*)**
   e. **Evidence of moderate or progressive systemic AV valve regurgitation. (*Level of Evidence: B*)**
   f. **Conduit obstruction with systemic or nearly systemic RV pressures and/or RV dysfunction after anatomic repair. (*Level of Evidence: B*)**
   g. **Conduit obstruction and systemic or suprasystemic LV pressures in a patient with nonanatomic correction. (*Level of Evidence: B*)**
   h. **Moderate or severe AR/neo-AR and onset of ventricular dysfunction or progressive ventricular dilatation. (*Level of Evidence: B*)**

Indications for surgery in patients who have undergone previous operations include repair or replacement of the systemic AV valve when a nonanatomic repair has been done previously, conduit replacement in patients who had a Rastelli-type anatomic repair and resection of LV outflow obstruction in the same group. Aortic valve and mitral valve repair/replacement are occasionally required in patients who have undergone anatomic repair. AR is seen more commonly in patients who underwent pulmonary arterial banding before ASO as part of staged anatomic repair.

12.5. Recommendations for Postoperative Care

CLASS I

1. **Patients with prior repair of CCTGA should have regular follow-up with a cardiologist with expertise in ACHD. (*Level of Evidence: C*)**
2. **Echocardiography-Doppler study and/or MRI should be performed yearly or at least every other year by staff trained in imaging complex CHD. (*Level of Evidence: C*)**

Regular follow-up (usually annually) is necessary (182), with particular emphasis on the following:

- Function of the SV
- Maintenance of sinus rhythm when possible
- Function of the systemic AV valve or systemic AV valve prosthesis if present
- Function of the pulmonary conduit or prosthesis
- Residual septal defects
- Development or progression of AR
- Degree of PAH, if any

12.5.1. Recommendations for Endocarditis Prophylaxis

CLASS IIa

1. **Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa is reasonable in those with the following indications:**
   a. **Prosthetic cardiac valve. (*Level of Evidence: B*)**
   b. **Previous IE. (*Level of Evidence: B*)**
   c. **Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: B*)**
   d. **Completely repaired CHD with prosthetic materials, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure. (*Level of Evidence: B*)**
   e. **Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization. (*Level of Evidence: B*)**
2. **It is reasonable to consider antibiotic prophylaxis against IE before vaginal delivery at the time of membrane rupture in select patients with the highest risk of adverse outcomes. This includes patients with the following indications:**
   a. **Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: C*)**
   b. **Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: C*)**

CLASS III

1. **Prophylaxis against IE is not recommended for nondental procedures (such as esophagogastroduodenoscopy or colonoscopy) in the absence of active infection. (*Level of Evidence: C*)**

12.5.2. Recommendation for Reproduction

CLASS I

1. **All women with CCTGA (whether repaired or not) should seek counseling from a cardiologist with expertise in ACHD before proceeding with a pregnancy. (*Level of Evidence: C*)**

The volume load of pregnancy may pose too great a burden for a compromised SV, particularly with associated systemic AV valve regurgitation. A careful and comprehensive clinical evaluation should be performed when pregnancy is contemplated. This should include a careful history, clinical examination, ECG, chest x-ray, and an assessment of the hemodynamics, presence or absence of valvular lesions, and ejection fraction. This should be evaluated with echocardiography and/or MRI study. An exercise test is helpful in determining the functional capacity of patients, and in general, it is unlikely pregnancy will be well tolerated if the functional aerobic capacity is less than 75% of predicted.

13. Ebstein's Anomaly

13.1. Initial Adult Presentation

Patients with mild Ebstein's anomaly may be asymptomatic with no functional limitation. Survival to the ninth decade has been reported (183). Electrophysiological rather than hemodynamic symptoms are more common in patients over the age of 10 years at presentation. Patients with Ebstein's anomaly who reach late adolescence and adulthood often have an excellent outcome (184).

13.2. Clinical Features and Evaluation of the Unoperated Patient

The disorder has the following features in common:

- Adherence of the tricuspid valve leaflets to the underlying myocardium (failure of delamination)
- Apical displacement of the septal and posterior leaflets of the tricuspid valve below the AV junction in the right ventricle
- Atrialization and dilation of the inflow of the right ventricle to varying degrees
- Redundancy, tethering, and fenestrations of the anterior tricuspid valve leaflet
- Varying degrees of TR
- Enlargement of the right atrium
- Varying degrees of cyanosis

Associated lesions include the following:

- More than 50% of patients have a shunt at the atrial level with either a patent foramen ovale or secundum ASD, which results in varying degrees of cyanosis
- One or more accessory conduction pathways, increasing the risk of atrial tachycardias (approximately 25%)
- VSD
- Varying degrees of anatomic and physiological RVOT obstruction
- Occasionally, other anomalies such as mitral valve prolapse
- Abnormalities of LV morphology and function

13.3. Recommendation for Evaluation of Patients With Ebstein's Anomaly

CLASS I

1. **All patients with Ebstein's anomaly should have periodic evaluation in a center with expertise in ACHD. (*Level of Evidence: C*)**

All patients with Ebstein's anomaly should have regular follow-up in a center for congenital cardiology. Unoperated patients need serial monitoring for features that suggest that surgical intervention is required or medical therapy is indicated. An assessment of functional limitation should also be performed.

13.4. Recommendations for Diagnostic Tests

CLASS I

1. **ECG, chest x-ray, and echocardiography-Doppler are recommended for the diagnostic evaluation of Ebstein's anomaly in adult patients. (*Level of Evidence: C*)**

CLASS IIa

1. **Pulse oximetry at rest and/or during exercise can be useful in the diagnostic evaluation of Ebstein's anomaly in adult patients. (*Level of Evidence: C*)**
2. **An electrophysiological study can be useful in the diagnostic evaluation of Ebstein's anomaly in adult patients if a supraventricular arrhythmia is documented or suspected (subsequent radiofrequency catheter ablation should be considered if clinically feasible). (*Level of Evidence: C*)**
3. **The following additional diagnostic tests can be useful for the comprehensive evaluation of Ebstein's anomaly in adult patients:**
   a. **Doppler TEE examination if the anatomic information is not provided by transthoracic imaging. (*Level of Evidence: B*)**
   b. **Holter monitoring. (*Level of Evidence: B*)**
   c. **Electrophysiological study for history or ECG evidence of accessory pathway(s). (*Level of Evidence: B*)**
   d. **Coronary angiography, when surgical repair is planned, if there is a suspicion of coronary artery disease, and in men 35 years or older, premenopausal women 35 years or older who have coronary risk factors, and postmenopausal women. (*Level of Evidence: B*)**

The ECG is valuable in the diagnosis of Ebstein's anomaly. Preexcitation may be present, usually via a right bypass tract. Multiple bypass tracts may also occur. The P waves are often very tall and peaked (so-called Himalayan P waves). A QR pattern is often seen in lead V_1 and may extend to V_4. The QRS duration is usually prolonged, with a right bundle-branch pattern, but is often "splintered," followed by inverted T waves.

The chest x-ray may be nearly normal in mild cases and in more severe cases shows severe enlargement. Right atrial enlargement is prominent, with a "globular" cardiac contour and clear lung fields. The great arteries are usually small, and the aortic root is inconspicuous or absent.

The diagnosis of Ebstein's anomaly is most commonly confirmed by TTE Doppler evaluation by a skilled echocardiographer, preferably with expertise in CHD. Echocardiography is the diagnostic test of choice and should document the severity of the degree of right-sided cardiac enlargement, RV dysfunction, and TR. TTE supplemented with intraoperative TEE usually provides sufficient data to permit operative intervention without the need to obtain additional preoperative diagnostic structural information in patients with Ebstein's anomaly (185–187). The diagnostic workup may require a TEE to assess the presence of an ASD or to delineate intracardiac anatomy in patients with suboptimal TTE images.

Hemodynamic cardiac catheterization is rarely required in patients with Ebstein's anomaly before surgical intervention is considered. In select high-risk patients, hemodynamic assessment by cardiac catheterization may be helpful for risk stratification. Coronary angiography should be performed before surgical intervention if there is a concern about coronary artery disease.

13.5. Management Strategies

13.5.1. Recommendation for Medical Therapy

CLASS I

1. **Anticoagulation with warfarin is recommended for patients with Ebstein's anomaly with a history of paradoxical embolus or atrial fibrillation. (*Level of Evidence: C*)**

The large right atrium predisposes to thrombus formation, particularly in association with atrial fibrillation. The potential for right-to-left shunting at the atrial level raises the risk of paradoxical embolus.

13.6. Recommendation for Catheter Interventions for Adults With Ebstein's Anomaly

CLASS I

1. **Adults with Ebstein's anomaly should have catheterization performed at centers with expertise in catheterization and management of such patients. (*Level of Evidence: C*)**

Catheterization is not usually required for the evaluation and management of Ebstein's anomaly; therefore, it should only be performed at regional centers for very specific indications, after thorough noninvasive evaluation.

13.6.1. Recommendation for Electrophysiology Testing/Pacing Issues in Ebstein's Anomaly

CLASS IIa

1. **Catheter ablation can be beneficial for treatment of recurrent supraventricular tachycardia in some patients with Ebstein's anomaly. (*Level of Evidence: B*)**

Supraventricular tachycardia related to accessory pathways is a frequent accompaniment of Ebstein's anomaly (188). Catheter ablation has become the most attractive treatment for this condition, although the procedure can be quite challenging. Overall, success rates are lower and recurrence rates are higher than those reported for ablation in a structurally normal heart (189,190), in part because multiple accessory pathways are present in nearly 50% of these patients (191). Any patient suspected of having an accessory pathway should undergo electrophysiology study before surgical repair, so that the pathway(s) may be localized and catheter ablation attempted. If catheter ablation is unsuccessful or deemed inappropriate for any reason, surgical interruption can be performed in the operating room. For any patients with history of atrial flutter, a right atrial Maze procedure can be incorporated into the surgery, and for those with atrial fibrillation, a biatrial Maze can be performed.

13.6.2. Recommendations for Surgical Interventions

CLASS I

1. **Surgeons with training and expertise in CHD should perform tricuspid valve repair or replacement with concomitant closure**

of an ASD, when present, for patients with Ebstein's anomaly with the following indications:
   a. Symptoms or deteriorating exercise capacity. (*Level of Evidence: B*)
   b. Cyanosis (oxygen saturation less than 90%). (*Level of Evidence: B*)
   c. Paradoxical embolism. (*Level of Evidence: B*)
   d. Progressive cardiomegaly on chest x-ray. (*Level of Evidence: B*)
   e. Progressive RV dilation or reduction of RV systolic function. (*Level of Evidence: B*)
2. Surgeons with training and expertise in CHD should perform concomitant arrhythmia surgery in patients with Ebstein's anomaly and the following indications:
   a. Appearance/progression of atrial and/or ventricular arrhythmias not amenable to percutaneous treatment. (*Level of Evidence: B*)
   b. Ventricular preexcitation not successfully treated in the electrophysiology laboratory. (*Level of Evidence: B*)
3. Surgical rerepair or replacement of the tricuspid valve is recommended in adults with Ebstein's anomaly with the following indications:
   a. Symptoms, deteriorating exercise capacity, or New York Heart Association functional class III or IV. (*Level of Evidence: B*)
   b. Severe TR after repair with progressive RV dilation, reduction of RV systolic function, or appearance/progression of atrial and/or ventricular arrhythmias. (*Level of Evidence: B*)
   c. Bioprosthetic tricuspid valve dysfunction with significant mixed regurgitation and stenosis. (*Level of Evidence: B*)
   d. Predominant bioprosthetic valve stenosis (mean gradient greater than 12 to 15 mm Hg). (*Level of Evidence: B*)
   e. Operation can be considered earlier with lesser degrees of bioprosthetic stenosis with symptoms or decreased exercise tolerance. (*Level of Evidence: B*)

The primary operation generally consists of closure of any interatrial communications; antiarrhythmia procedures such as surgical division of accessory conduction pathways, cryoablation of AV node reentry tachycardia, or Maze procedure; and tricuspid valve surgery. The tricuspid valve is repaired when feasible, and tricuspid valve replacement is performed with a mechanical or heterograft bioprosthesis when repair is not feasible or the repair result is not satisfactory. A right reduction atrioplasty is often performed.

Reoperation usually requires tricuspid valve replacement or rereplacement (tissue or mechanical). Rerepair of the tricuspid valve is rarely successful. Other procedures are performed as with the primary operation. A concomitant Maze procedure may be performed for intermittent or chronic atrial fibrillation/flutter.

13.7. Problems and Pitfalls

The problems and pitfalls associated with the management of adults with Ebstein's anomaly are as follows:

- Patients with Ebstein's anomaly may be referred for percutaneous or surgical ASD closure; however, the presence of Ebstein's anomaly may alter the recommendation for intervention.
- Percutaneous ablation of an accessory pathway should be performed with caution in patients with Ebstein's anomaly and an interatrial communication with right-to-left shunt because of the risk of paradoxical embolus.
- The presence of multiple accessory pathways should raise the suspicion for Ebstein's anomaly.
- Patients with Ebstein's anomaly and marked cardiomegaly may complain of few symptoms despite marked limitation. Exercise testing will demonstrate functional limitation and should be included as part of the regular assessment of these patients. Exercise testing should include monitoring of oxygen saturation, because exercise-induced cyanosis may occur.
- Patients with newly diagnosed Ebstein's anomaly may be told they have concomitant PAH, particularly when cyanosis and right-sided heart enlargement are present. This is usually a misdiagnosis, because PAH is very rare among Ebstein patients.
- Other tricuspid valve disorders may be misdiagnosed as Ebstein's anomaly (refer to Section 13.4, Recommendations for Diagnostic Tests).

13.8. Recommendation for Reproduction

CLASS I

1. Women with Ebstein's anomaly should undertake prepregnancy counseling with a physician with expertise in ACHD. (*Level of Evidence: C*)

Most women with Ebstein's anomaly can have a successful pregnancy with proper care, but there is an increased risk of low birth weight and fetal loss if significant cyanosis is present. The risk of CHD in the offspring (in the absence of a family history) is approximately 6% (192).

13.9. Recommendation for Endocarditis Prophylaxis

CLASS IIa

1. Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of the teeth or perforation of the oral mucosa is reasonable in cyanotic patients with Ebstein's anomaly and postoperative patients with a prosthetic cardiac valve. (*Level of Evidence: C*)

Antibiotic prophylaxis is usually unnecessary in the acyanotic, unoperated patient (refer to Section 1.6, Recommendations for Infective Endocarditis, for additional information).

14. Tricuspid Atresia/Single Ventricle

This section focuses on conditions that are not amenable to biventricular repair and will include the various types of so-called univentricular hearts, such as tricuspid atresia, mitral atresia, double-inlet left ventricle, single ventricle, hypoplastic right ventricle or left ventricle, and heterotaxia syndromes.

14.1. Clinical Course (Unoperated and Palliated)

Patients usually fall into 2 general categories. The first category includes those patients with no anatomic restrictions

to pulmonary blood flow with early postnatal development of a large left-to-right shunt and symptoms of congestive heart failure. The second major clinical presentation is severe cyanosis due to obstruction to pulmonary flow, frequently caused by valvular/subvalvular PS or atresia.

A small number of patients will present with mild cyanosis and no congestive heart failure. These patients have sufficient PS to limit pulmonary flow to levels that do not cause heart failure symptoms but is adequate to prevent severe hypoxemia. The vast majority of adults with these conditions will have undergone previous palliation with some type of systemic–to–pulmonary artery shunt, cavopulmonary connection (bidirectional Glenn), or 1 of the modifications of the Fontan operation (193,194).

14.2. Recommendation for Catheterization Before Fontan Procedure

CLASS I

1. **In the evaluation of hemodynamics to assess the potential for definitive palliation of unoperated or shunt-palliated adults with univentricular hearts, catheterization is indicated to:**
   a. **Assess the nature of pulmonary artery obstruction, with potential to restore maximal continuous, effective, unimpeded systemic venous flow to the maximal number of pulmonary artery segments. (*Level of Evidence: C*)**
   b. **Assess and eliminate systemic–to–pulmonary vein collaterals. (*Level of Evidence: C*)**
   c. **Assess and eliminate systemic–to–pulmonary artery connections. (*Level of Evidence: C*)**
   d. **For adults with systemic–to–pulmonary shunts, the potential for perioperative transcatheter shunt exclusion should be examined. (*Level of Evidence: C*)**

Data to be obtained include intracardiac, pulmonary artery, and aortic pressures; oxygen saturations; and estimations of pulmonary and systemic blood flow and resistances. Imaging data would include angiograms of systemic venous anatomy, great vessel anatomy (specifically, anatomy of the pulmonary arteries and estimations of ventricular volume), hypertrophy, and ejection fraction. Coronary angiography is needed for older patients and those with questionable ischemia or coronary anomalies. Assessment of venous and arteriopulmonary collaterals is also important, because these may be amenable to coil occlusion.

14.3. Recommendation for Surgical Options for Patients With Single Ventricle

CLASS I

1. **Surgeons with training and expertise in CHD should perform operations for single-ventricle anatomy or physiology. (*Level of Evidence: C*)**

Surgical options for the treatment of adults with tricuspid atresia/single ventricle are outlined below.
Systemic–to–pulmonary artery shunt:

- Often from the ascending aorta to the main or right pulmonary artery; rarely performed as an isolated procedure, and only if a cavopulmonary connection is contraindicated.

Bidirectional Glenn (bidirectional cavopulmonary anastomosis [BDCPA]):

- Most commonly performed in infancy or early childhood as a staged procedure toward the Fontan completion; this provides a stable source of pulmonary blood flow without volume loading the SV; it generally should not be the sole source of pulmonary blood flow (except as the stage II procedure for hypoplastic left-sided heart syndrome).

BDCPA plus additional pulmonary blood flow:

- The most reliable source of additional pulmonary blood flow is via the native RVOT with native PS or with a pulmonary artery band. A concomitant systemic–to–pulmonary artery shunt may be added if an increase in systemic oxygenation is required, but this is at the expense of an increase in volume load on the SV and often elevated superior vena cava pressure.

Single-ventricle repair:

- When the rudimentary pulmonary ventricle is less than 30% of its normal volume, a Fontan-type of operation is performed. The operation has gone through many modifications; each allows systemic venous return to enter the pulmonary circulation directly.

Modified Fontan Procedures:

- Extracardiac conduit–BDCPA plus conduit from inferior vena cava to right pulmonary artery/main pulmonary artery
- Intra-atrial conduit–BDCPA plus intra-atrial conduit from inferior vena cava to right pulmonary artery/main pulmonary artery; preferred when the ventricular mass would lie on top of an extracardiac conduit, eg, isolated dextrocardia or isolated levocardia with situs inversus
- Intracardiac lateral tunnel plus BDCPA
- Intracardiac lateral tunnel
- Atriopulmonary connection (rarely used in the current era)
- Fenestration between systemic venous pathway and left atrium

1.5-Ventricle Repair:

A term used to describe a procedure for cyanotic CHD performed when the pulmonary ventricle is insufficiently developed to accept the entire systemic venous return. A BDCPA is constructed to direct superior vena cava blood directly into the pulmonary arteries while the inferior caval blood is directed to the lungs via the small pulmonary ventricle.

2-Ventricle Repair:

A term used to describe a procedure for cyanotic CHD with a common ventricle or adequately sized pulmonary ventricles and SVs that communicate via a VSD. The pulmonary and systemic circulations are surgically septated by placement of an interventricular patch (for common ventricle) or VSD patch (for separate pulmonary and SV cavities).

Transplantation:

Heart and/or heart/lung transplantation are reserved for severe SV failure with or without PAH when there is no conventional surgical option.

14.4. Recommendation for Evaluation and Follow-Up After Fontan Procedure

CLASS I

1. **Lifelong follow-up is recommended for patients after a Fontan-type operation; this should include a yearly evaluation by a cardiologist with expertise in the care of ACHD patients. (*Level of Evidence: C*)**

All patients should have follow-up with a cardiologist who has expertise in ACHD. The frequency, although typically annual, may be determined by the extent and degree of residual abnormalities. Long-term problems include atrial arrhythmias and right atrial thrombus, especially common in direct atrium–to–pulmonary artery connections, ventricular dysfunction and edema, need for reoperation, hepatic congestion and dysfunction, and protein-losing enteropathy.

14.5. Recommendation for Imaging

CLASS I

1. **All patients with prior Fontan-type repair should have periodic echocardiographic and/or magnetic resonance examinations performed by staff with expertise in ACHD. (*Level of Evidence: C*)**

Echocardiography is the cornerstone of the postoperative evaluation. Spontaneous contrast is often seen in the Fontan circuit and represents slow flow in the pathway. It is important to image the Fontan pathway in its entirety, and TEE is often necessary to accomplish this. In addition, TEE is needed to rule out right atrial thrombus. The presence or absence of a Fontan fenestration should be sought, and if present, the gradient across the fenestration should be measured.

14.6. Recommendation for Diagnostic and Interventional Catheterization for Adults After Fontan Procedure

CLASS I

1. **Catheterization of adults with a Fontan-type repair of single-ventricle physiology should be performed in regional centers with expertise in ACHD. (*Level of Evidence: C*)**

For adults after Fontan palliation, cardiac catheterization, often assisted by contrast echocardiography, is indicated to investigate and potentially treat unexplained volume retention, fatigue or exercise limitation, atrial arrhythmia, or cyanosis and hemoptysis. To further investigate oxygen-unresponsive hypoxemia in the adult Fontan survivor, catheterization is directed at assessing and potentially relieving (when applicable) the following: persistent Fontan fenestration; systemic–to–pulmonary venous collaterals; pulmonary arteriovenous malformations; and the cause of volume retention, increasing Fontan pathway pressure and resistance, and thereby worsening right-to-left shunting.

Evaluation of postoperative Fontan patients with worsening cyanosis (oxygen saturation usually 90% or less at rest and decreasing with exercise):

In addition to pressure and resistance data, an angiographic search for atrial right-to-left shunts and shunts from inferior cava, superior cava, and innominate vein to the left atrium should be performed, as well as a search for pulmonary arteriovenous malformations. Interventional closure of residual shunts by coils or ASD devices is often possible.

14.7. Recommendations for Management Strategies for the Patient With Prior Fontan Repair

CLASS I

1. **Management of patients with prior Fontan repair should be coordinated with a regional ACHD center. Local cardiologists, internists, and family care physicians should develop ongoing relationships with such a center with continuous availability of specialists. (*Level of Evidence: C*)**
2. **At least yearly follow-up is recommended for patients after Fontan repair. (*Level of Evidence: C*)**
3. **Arrhythmia management is frequently an issue, and consultation with an electrophysiologist is recommended as a vital part of care. (*Level of Evidence: C*)**
4. **New-onset atrial tachyarrhythmia should prompt a comprehensive noninvasive imaging evaluation to identify associated atrial/baffle thrombus, anatomic abnormalities of the Fontan pathway, or ventricular dysfunction. (*Level of Evidence: C*)**

Patients with single-ventricle physiology should be considered to have a chronic disease that requires active management to prevent secondary disability and preserve the function of the single ventricle for the maximum possible time. This requires close follow-up and coordination between all providers. Ventricular dysfunction, congestive heart failure, symptomatic arrhythmias, thromboembolism, and edema are all possible findings on long-term follow-up and require management directed by an ACHD center as defined in these guidelines.

14.7.1. Recommendations for Medical Therapy

CLASS I

1. **Warfarin should be given for patients who have a documented atrial shunt, atrial thrombus, atrial arrhythmias, or a thromboembolic event. (*Level of Evidence: C*)**

CLASS IIa

1. **It is reasonable to treat SV dysfunction with ACE inhibitors and diuretics. (*Level of Evidence: C*)**

Anticoagulants should be given to all patients with atrial arrhythmias even if atrial thrombus has not been documented. Warfarin should also be given to those with a residual ASD, especially those with dual atrial pulmonary connections, spontaneous right atrial contrast on echocardiography, and an ejection fraction less than 40%.

Ventricular dysfunction, congestive heart failure, symptomatic arrhythmias, thromboembolism, and edema are all possible findings on long-term follow-up and require management directed by an ACHD center as defined in these guidelines. Many patients require afterload reduction with

ACE inhibitors. Many adult survivors also require diuretic therapy.

14.8. Recommendations for Surgery for Adults With Prior Fontan Repair

CLASS I

1. Surgeons with training and expertise in CHD should perform operations on patients with prior Fontan repair for single-ventricle physiology. (*Level of Evidence: C*)
2. Reoperation after Fontan is indicated for the following:
   a. Unintended residual ASD that results in right-to-left shunt with symptoms and/or cyanosis not amenable to transcatheter closure. (*Level of Evidence: C*)
   b. Hemodynamically significant residual systemic artery–to–pulmonary artery shunt, residual surgical shunt, or residual ventricle–to–pulmonary artery connection not amenable to transcatheter closure. (*Level of Evidence: C*)
   c. Moderate to severe systemic AV valve regurgitation. (*Level of Evidence: C*)
   d. Significant (greater than 30 mm Hg peak-to-peak) subaortic obstruction. (*Level of Evidence: C*)
   e. Fontan pathway obstruction. (*Level of Evidence: C*)
   f. Development of venous collateral channels or pulmonary arteriovenous malformation not amenable to transcatheter management. (*Level of Evidence: C*)
   g. Pulmonary venous obstruction. (*Level of Evidence: C*)
   h. Rhythm abnormalities, such as complete AV block or sick sinus syndrome, that require epicardial pacemaker insertion. (*Level of Evidence: C*)
   i. Creation or closure of a fenestration not amenable to transcatheter intervention. (*Level of Evidence: C*)

CLASS IIa

1. Reoperation for Fontan conversion (ie, revision of an atriopulmonary connection to an intracardiac lateral tunnel, intra-atrial conduit, or extracardiac conduit) can be useful for recurrent atrial fibrillation or flutter without hemodynamically significant anatomic abnormalities. A concomitant Maze procedure should also be performed. (*Level of Evidence: C*)

CLASS IIb

1. Heart transplantation may be beneficial for severe SV dysfunction or protein-losing enteropathy. (*Level of Evidence: C*)

Reoperation includes valve repair or replacement for systemic AV valve regurgitation, resection of subaortic obstruction, closure of an unintended residual shunt, revision of Fontan pathway obstruction, or Fontan conversion for atrial tachyarrhythmias with or without anatomic abnormalities (195). Venous collateral channels or arteriovenous malformations in the right lung in the presence of a classic Glenn shunt may be ameliorated by conversion to a modified Fontan procedure. This enables hepatic venous blood to perfuse the right-sided pulmonary vascular bed (195). Arteriovenous malformations often regress, provided they are not large and have not been long-standing. Clinically significant persistent venous collateral channels or systemic aortopulmonary collaterals are usually treated with transcatheter occlusion.

Atrial tachycardias can be treated by catheter ablation versus Fontan conversion with Maze procedure (196). Complete AV block or sick sinus syndrome commonly requires permanent pacing, usually epicardial.

Protein-losing enteropathy not amenable to medical or catheter therapy may be treated by creation of an atrial septal fenestration or Fontan conversion. The Fontan revision carries an operative mortality rate of 5% to 25% in reported series (197,198). If protein-losing enteropathy is due to Fontan pathway obstruction, successful revision of the Fontan communication may be curative. Protein-losing enteropathy often requires heart transplantation (197). although the protein-losing enteropathy does not always resolve. Severe SV dysfunction often requires heart transplantation (199).

14.9. Key Issues to Evaluate and Follow-Up

14.9.1. Recommendations for Electrophysiology Testing/Pacing Issues in Single-Ventricle Physiology and After Fontan Procedure

CLASS I

1. Arrhythmia management is frequently an issue in patients after the Fontan procedure, and consultation with an electrophysiologist with expertise in CHD is recommended as a vital part of care. (*Level of Evidence: C*)
2. New-onset atrial tachyarrhythmias should prompt a comprehensive noninvasive imaging evaluation to identify associated atrial/baffle thrombus, anatomic abnormalities of the Fontan pathway, or ventricular dysfunction. (*Level of Evidence: C*)
3. Electrophysiological studies in adults with Fontan physiology should be performed at centers with expertise in the management of such patients. (*Level of Evidence: C*)
4. Clinicians must be mindful of the high risk for symptomatic IART in adult patients who have undergone the Fontan operation. This arrhythmia can cause serious hemodynamic compromise and contribute to atrial thrombus formation. Treatment is often difficult, and consultation with an electrophysiologist who is experienced with CHD is recommended whenever recurrent IART is detected. (*Level of Evidence: C*)

The most significant rhythm issue facing adults who have undergone the Fontan operation is recurrent IART. This arrhythmia is a major source of morbidity in the post-Fontan population, especially for patients who have undergone an atriopulmonary connection and subsequently developed advanced degrees of dilation, thickening, and scarring of their right atrial chamber (200). Beyond the surgical technique, other risk factors for development of IART include concomitant sinus node dysfunction and older age at time of Fontan operation (57). Tachycardia episodes can result in significant hemodynamic compromise and, if long in duration, clot formation within the dilated right artery.

14.9.2. Recommendations for Endocarditis Prophylaxis

CLASS IIa

1. Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa is reasonable in those patients with the following indications:
   a. Prosthetic cardiac valve. (*Level of Evidence: B*)

b. Previous IE. (*Level of Evidence: B*)
c. Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: B*)
d. Completely repaired CHD with prosthetic materials, whether placed by surgery or catheter intervention, during the first 6 months after the procedure. (*Level of Evidence: B*)
e. Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization. (*Level of Evidence: B*)

2. It is reasonable to consider antibiotic prophylaxis against IE before vaginal delivery at the time of membrane rupture in select patients with the highest risk of adverse outcomes. This includes patients with the following indications:
   a. Prosthetic cardiac valve or prosthetic material used for cardiac valve repair. (*Level of Evidence: C*)
   b. Unrepaired and palliated cyanotic CHD, including surgically constructed palliative shunts and conduits. (*Level of Evidence: C*)

CLASS III

1. Prophylaxis against IE is not recommended for nondental procedures (such as esophagogastroduodenoscopy or colonoscopy) in the absence of active infection. (*Level of Evidence: C*)

14.9.3. Recommendations for Reproduction

CLASS I

1. All women with a Fontan operation should have a comprehensive evaluation by a physician with expertise in ACHD before proceeding with a pregnancy. (*Level of Evidence: C*)

CLASS III

1. Pregnancy should not be planned without consultation and evaluation at a comprehensive ACHD center with experience and expertise in maternal and prenatal management of complex CHD. (*Level of Evidence: C*)

Successful pregnancy has been reported in postoperative Fontan patients, but atrial arrhythmias, ventricular dysfunction, edema, and ascites have been reported as maternal complications (201,202). In addition, there is an increased risk for spontaneous abortion and premature birth. For those patients undergoing warfarin anticoagulation, this poses the additional risk of fetal exposure in the first trimester and resulting fetal embryopathy. In each case, management must be individualized.

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Appendixes

Appendix 1. Author Relationships With Industry and Other Entities—ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

Committee Member	Research Grant	Speakers' Bureau	Stock Ownership	Board of Directors	Consultant/Advisory Member
Dr. Carole A. Warnes *(Co-Chair)*	None	None	None	None	None
Dr. Roberta G. Williams *(Co-Chair)*	None	None	None	None	None
Dr. Thomas M. Bashore	None	None	None	None	None
Dr. John S. Child	None	None	None	None	None
Dr. Heidi M. Connolly	None	None	None	None	None
Dr. Joseph A. Dearani	None	None	None	None	None
Dr. Pedro del Nido	None	None	None	None	None
Dr. James W. Fasules	None	None	None	None	None
Dr. Thomas P. Graham, Jr	None	None	None	None	None
Dr. Ziyad M. Hijazi	None	None	None	None	• AGA Medical
Dr. Sharon A. Hunt	None	None	None	None	None
Dr. Mary Etta King	None	None	None	None	None
Dr. Michael J. Landzberg	• Actelion • AGA Medical • Myogen • NMT Medical • Pfizer	None	None	None	None
Dr. Pamela D. Miner	None	None	None	None	None
Dr. Martha J. Radford	None	None	None	None	None
Dr. Edward P. Walsh	None	None	None	None	None
Dr. Gary D. Webb	None	None	None	None	None

This table represents the relevant relationships of committee members with industry and other entities that were reported orally at the initial writing committee meeting and updated in conjunction with all meetings and conference calls of the writing committee during the document development process. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of $10 000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships in this table are modest unless otherwise noted.

Appendix 2. Peer Reviewer Relationships With Industry and Other Entities—ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

Reviewer	Representation	Consultant	Speakers' Bureau	Ownership/Partnership/ Principal	Research	Other
Dr. Ann G. Bolger	Official—American Heart Association	None	None	None	None	None
Dr. James W. Fasules	Official—American College of Cardiology Foundation Board of Trustees	None	None	None	None	None
Dr. Sharon A. Hunt	Official—American College of Cardiology/American Heart Association Lead Task Force Reviewer	None	None	None	None	None
Dr. William Mahle	Official—American Heart Association	None	None	None	• Sanofi-Aventis	None
Dr. Brian McCrindle	Official—American Heart Association	None	None	None	None	None
Dr. Kevin Mulhern	Official—American College of Cardiology Foundation Board of Governors	None	None	None	None	None
Dr. Emily Bacha	Organizational—Society of Thoracic Surgeons	None	None	None	• Cryolife* • Medtronic	None
Dr. Jack Colman	Organizational—International Society for Adult Congenital Heart Disease	None	None	None	None	• 2007 Plaintiff–Pregnancy and Congenital Heart Disease • 2007 Defense–Pregnancy and Congenital Heart Disease
Dr. Elyse Foster	Organizational—International Society for Adult Congenital Heart Disease	None	None	None	• Evalve* • Guidant/ Boston Scientific*	None
Dr. Robert Hamilton	Organizational—Heart Rhythm Society	None	None	None	None	None
Dr. Thomas K. Jones	Organizational—Society for Cardiovascular Angiography and Interventions	• AGA Medical* • Copatus Medical* • WL Gore & Associates	None	None	None	None
Dr. Thomas R. Kimball	Organizational—American Society of Echocardiography	None	None	None	None	None
Dr. Rachel Lampert	Organizational—Heart Rhythm Society	None	None	None	• Guidant/ Boston Scientific* • Medtronic* • St. Jude Medical*	None
Dr. Louis Bezold	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	None	None	None
Dr. Frank Cetta	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	None	None	None
Dr. Barbara Deal	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	None	None	None
Dr. John Deanfield	Content—Individual	None	None	None	None	None
Dr. Christopher C. Erickson	Content—American Heart Association Congenital Cardiac Defects Committee	• Medtronic	None	None	None	• Expert witness for defense regarding ablation case, 2005

(*Continued*)

Appendix 2. Continued

Reviewer	Representation	Consultant	Speakers' Bureau	Ownership/Partnership/ Principal	Research	Other
Dr. Michael Gatzoulis	Content—American Heart Association	• Actelion • Pfizer	• Boston Scientific* • Medtronic • St. Jude	• Hansen Medical*	• Actelion* • Pfizer*	None
Dr. Welton Gersony	Content—Individual	None	None	None	None	None
Dr. Michael Gewitz	Content—Individual	None	None	None	None	None
Dr. David Gregg	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	• Johnson & Johnson* • Schering-Plough	None	None
Dr. Daphne Hsu	Content—American Heart Association Congenital Cardiac Defects Committee	None	None	None	None	None
Dr. Walter H. Johnson	Content—American Heart Association Congenital Cardiac Defects Committee	None	None	None	None	None
Dr. Karen S. Kuehl	Content—Individual	None	None	None	None	None
Dr. Gerard R. Martin	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	None	None	None
Dr. G. Paul Matherne	Content—American Heart Association Congenital Cardiac Defects Committee	None	None	None	None	None
Dr. Geoffrey L. Rosenthal	Content—Individual	None	None	None	None	None
Dr. Craig Sable	Content—American Heart Association Congenital Cardiac Defects Committee	None	None	None	None	None
Dr David J. Sahn	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	None	None	None
Dr. Jane Somerville	Content—Individual	None	None	None	None	None
Dr. Kathryn Taubert	Content—American Heart Association	None	None	None	None	None
Dr. Judith Therrien	Content—Canadian Congenital Heart Alliance Committee	None	None	None	None	None
Dr. Elizabeth Tong	Content—Individual	None	None	None	None	None
Dr. Jeffery Towbin	Content—American College of Cardiology Foundation Congenital Heart Disease/Pediatric Cardiology Committee	None	None	None	None	None
Dr. Catherine Webb	Content—American Heart Association Congenital Cardiac Defects Committee	None	None	• Johnson & Johnson* • Tyco* • Wyeth*	None	None

This table represents the relevant relationships with industry and other entities that were disclosed at the time of peer review. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of $10 000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships in this table are modest unless otherwise noted. Names are listed in alphabetical order within each category of review.

*Significant (greater than $10 000) relationship.

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KEY WORDS: ACC/AHA Practice Guidelines ■ congenital heart disease ■ cardiac defects ■ congenital heart surgery ■ unoperated/repaired heart defects ■ medical therapy ■ cardiac catheterization.

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