JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY
© 2019 BY THE AMERICAN HEART ASSOCIATION, INC., AND AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION.
PUBLISHED BY ELSEVIER

VOL. 73, NO. 12, 2019

CLINICAL PRACTICE GUIDELINE: EXECUTIVE SUMMARY

2018 AHA/ACC Guideline 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 Clinical Practice Guidelines

Developed in Collaboration with the American Association for Thoracic Surgery, 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

Writing Committee Members*

Karen K. Stout, MD, FACC, *Chair*†
Curt J. Daniels, MD, *Vice Chair**†‡
Jamil A. Aboulhosn, MD, FACC, FSCAI*§
Biykem Bozkurt, MD, PhD, FACC, FAHA‖
Craig S. Broberg, MD, FACC*†
Jack M. Colman, MD, FACC†
Stephen R. Crumb, DNP, AACC†
Joseph A. Dearani, MD, FACC¶
Stephanie Fuller, MD, MS, FACC#
Michelle Gurvitz, MD, FACC**
Paul Khairy, MD, PhD*†
Michael J. Landzberg, MD, FACC*†
Arwa Saidi, MB, BCH, FACC*†
Anne Marie Valente, MD, FACC, FAHA, FASE††
George F. Van Hare, MD, FACC‡‡

*Writing committee members are required to recuse themselves from voting on sections to which their specific relationships with industry may apply; see Appendix 1 for recusal information. †ACC/AHA Representative. ‡International Society for Adult Congenital Heart Disease Representative. §Society for Cardiovascular Angiography and Interventions Representative. ‖ACC/AHA Task Force on Clinical Practice Guidelines Liaison. ¶Society of Thoracic Surgeons Representative. #American Association for Thoracic Surgery Representative. **ACC/AHA Task Force on Performance Measures Liaison. ††American Society of Echocardiography Representative. ‡‡Heart Rhythm Society Representative.

This document was approved by the American College of Cardiology Clinical Policy Approval Committee in May 2018, the American Heart Association Science Advisory and Coordinating Committee in June 2018, and the American Heart Association Executive Committee in July 2018.

The American College of Cardiology requests that this document be cited as follows: Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, Crumb SR, Dearani JA, Fuller S, Gurvitz M, Khairy P, Landzberg MJ, Saidi A, Valente AM, Van Hare GF. 2018 AHA/ACC guideline 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 Clinical Practice Guidelines. J Am Coll Cardiol 2019;73:1494-563.

This article has been copublished in *Circulation*.

ISSN 0735-1097/$36.00

https://doi.org/10.1016/j.jacc.2018.08.1028

ACC/AHA Task Force Members

Glenn N. Levine, MD, FACC, FAHA, *Chair*
Patrick T. O'Gara, MD, MACC, FAHA, *Chair-Elect*
Jonathan L. Halperin, MD, FACC, FAHA, *Immediate Past Chair*

Nancy M. Albert, PhD, RN, FAHA§§
Sana M. Al-Khatib, MD, MHS, FACC, FAHA
Joshua A. Beckman, MD, MS, FAHA
Kim K. Birtcher, PharmD, MS, AACC
Biykem Bozkurt, MD, PhD, FACC, FAHA§§
Ralph G. Brindis, MD, MPH, MACC§§
Joaquin E. Cigarroa, MD, FACC
Lesley H. Curtis, PhD, FAHA§§
Anita Deswal, MD, MPH, FACC, FAHA
Lee A. Fleisher, MD, FACC, FAHA
Federico Gentile, MD, FACC
Samuel S. Gidding, MD, FAHA§§
Zachary D. Goldberger, MD, MS, FACC, FAHA
Mark A. Hlatky, MD, FACC
John Ikonomidis, MD, PhD, FAHA
José Joglar, MD, FACC, FAHA
Richard J. Kovacs, MD, FACC, FAHA§§
Laura Mauri, MD, MSc, FAHA
E. Magnus Ohman, MD, FACC§§
Mariann R. Piano, RN, PhD, FAHA, FAAN
Susan J. Pressler, PhD, RN, FAHA§§
Barbara Riegel, PhD, RN, FAHA
Frank W. Sellke, MD, FACC, FAHA§§
Win-Kuang Shen, MD, FACC, FAHA§§
Duminda N. Wijeysundera, MD, PhD

§§Former Task Force member; current member during the writing effort.

TABLE OF CONTENTS

PREAMBLE

Since 1980, the American College of Cardiology (ACC) and American Heart Association (AHA) have translated scientific evidence into clinical practice guidelines (guidelines) with recommendations to improve cardiovascular health. These guidelines, which are based on systematic methods to evaluate and classify evidence, provide a cornerstone for quality cardiovascular care. The ACC and AHA sponsor the development and publication of guidelines without commercial support, and members of each organization volunteer their time to the writing and review efforts. Guidelines are official policy of the ACC and AHA.

Intended Use

Practice guidelines provide recommendations applicable to patients with or at risk of developing cardiovascular disease. The focus is on medical practice in the United States, but guidelines developed in collaboration with other organizations can have a global impact. Although guidelines may be used to inform regulatory or payer decisions, they are intended to improve patients' quality of care and align with patients' interests. Guidelines are intended to define practices meeting the needs of patients in most, but not all, circumstances and should not replace clinical judgment.

Clinical Implementation

Management in accordance with guideline recommendations is effective only when followed by both practitioners and patients. Adherence to recommendations can be enhanced by shared decision making between clinicians and patients, with patient engagement in selecting interventions on the basis of individual values, preferences, and associated conditions and comorbidities.

Methodology and Modernization

The ACC/AHA Task Force on Clinical Practice Guidelines (Task Force) continuously reviews, updates, and modifies guideline methodology on the basis of published standards from organizations, including the Institute of Medicine (P-1, P-2), and on the basis of internal reevaluation. Similarly, the presentation and delivery of guidelines are reevaluated and modified on the basis of evolving technologies and other factors to facilitate optimal dissemination of information to healthcare professionals at the point of care.

Toward this goal, this guideline continues the introduction of an evolved format of presenting guideline recommendations and associated text called the "modular knowledge chunk format." Each modular "chunk" includes a table of related recommendations, a brief

synopsis, recommendation-specific supportive text, and when appropriate, flow diagrams or additional tables. References are provided at the end of the document in their respective sections. Additionally, this format will facilitate seamless updating of guidelines with focused updates as new evidence is published, as well as content tagging for rapid electronic retrieval of related recommendations on a topic of interest. This evolved approach format was instituted when this guideline was near completion; therefore, the present document represents a transitional format that best suits the text as written. Future guidelines will fully implement this format, including provisions for limiting the amount of text in a guideline.

Recognizing the importance of cost-value considerations in certain guidelines, when appropriate and feasible, an analysis of the value of a drug, device, or intervention may be performed in accordance with the ACC/AHA methodology (P-3).

To ensure that guideline recommendations remain current, new data are reviewed on an ongoing basis, with full guideline revisions commissioned in approximately 6-year cycles. Publication of new, potentially practice-changing study results that are relevant to an existing or new drug, device, or management strategy will prompt evaluation by the Task Force, in consultation with the relevant guideline writing committee, to determine whether a focused update should be commissioned. For additional information and policies regarding guideline development, we encourage readers to consult the ACC/AHA guideline methodology manual (P-4) and other methodology articles (P-5–P-8).

Selection of Writing Committee Members

The Task Force strives to avoid bias by selecting experts from a broad array of backgrounds. Writing committee members represent different geographic regions, sexes, ethnicities, races, intellectual perspectives/biases, and scopes of clinical practice. The Task Force may also invite organizations and professional societies with related interests and expertise to participate as partners, collaborators, or endorsers.

Relationships With Industry and Other Entities

The ACC and AHA have rigorous policies and methods to ensure that guidelines are developed without bias or improper influence. The complete relationships with industry and other entities (RWI) policy can be found online. Appendix 1 of the present document lists writing committee members' relevant RWI. For the purposes of full transparency, writing committee members' comprehensive disclosure information is available online. Comprehensive disclosure information for the Task Force is also available online.

Evidence Review and Evidence Review Committees

In developing recommendations, the writing committee uses evidence-based methodologies that are based on all available data (P-4–P-7). Literature searches focus on randomized controlled trials but also include registries, nonrandomized comparative and descriptive studies, case series, cohort studies, systematic reviews, and expert opinion. Only key references are cited.

An independent evidence review committee (ERC) is commissioned when there are 1 or more questions deemed of utmost clinical importance that merit formal systematic review. The systematic review will determine which patients are most likely to benefit from a drug, device, or treatment strategy and to what degree. Criteria for commissioning an ERC and formal systematic review include: a) the absence of a current authoritative systematic review, b) the feasibility of defining the benefit and risk in a time frame consistent with the writing of a guideline, c) the relevance to a substantial number of patients, and d) the likelihood that the findings can be translated into actionable recommendations. ERC members may include methodologists, epidemiologists, healthcare providers, and biostatisticians. The recommendations developed by the writing committee on the basis of the systematic review are marked with "SR".

Guideline-Directed Management and Therapy

The term guideline-directed management and therapy (GDMT) encompasses clinical evaluation, diagnostic testing, and pharmacological and procedural treatments. For these and all recommended drug treatment regimens, the reader should confirm the dosage by reviewing product insert material and evaluate the treatment regimen for contraindications and interactions. The recommendations are limited to drugs, devices, and treatments approved for clinical use in the United States.

Class of Recommendation and Level of Evidence

The Class of Recommendation (COR) indicates the strength of the recommendation, encompassing the estimated magnitude and certainty of benefit in proportion to risk. The Level of Evidence (LOE) rates the quality of scientific evidence that supports the intervention on the

TABLE 1 Applying Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care* (Updated August 2015)

CLASS (STRENGTH) OF RECOMMENDATION	
CLASS I (STRONG)	**Benefit >>> Risk**
Suggested phrases for writing recommendations: ▪ Is recommended ▪ Is indicated/useful/effective/beneficial ▪ Should be performed/administered/other ▪ Comparative-Effectiveness Phrases†: ○ Treatment/strategy A is recommended/indicated in preference to treatment B ○ Treatment A should be chosen over treatment B	
CLASS IIa (MODERATE)	**Benefit >> Risk**
Suggested phrases for writing recommendations: ▪ Is reasonable ▪ Can be useful/effective/beneficial ▪ Comparative-Effectiveness Phrases†: ○ Treatment/strategy A is probably recommended/indicated in preference to treatment B ○ It is reasonable to choose treatment A over treatment B	
CLASS IIb (WEAK)	**Benefit ≥ Risk**
Suggested phrases for writing recommendations: ▪ May/might be reasonable ▪ May/might be considered ▪ Usefulness/effectiveness is unknown/unclear/uncertain or not well established	
CLASS III: No Benefit (MODERATE) *(Generally, LOE A or B use only)*	**Benefit = Risk**
Suggested phrases for writing recommendations: ▪ Is not recommended ▪ Is not indicated/useful/effective/beneficial ▪ Should not be performed/administered/other	
CLASS III: Harm (STRONG)	**Risk > Benefit**
Suggested phrases for writing recommendations: ▪ Potentially harmful ▪ Causes harm ▪ Associated with excess morbidity/mortality ▪ Should not be performed/administered/other	

LEVEL (QUALITY) OF EVIDENCE‡	
LEVEL A	
▪ High-quality evidence‡ from more than 1 RCT ▪ Meta-analyses of high-quality RCTs ▪ One or more RCTs corroborated by high-quality registry studies	
LEVEL B-R	**(Randomized)**
▪ Moderate-quality evidence‡ from 1 or more RCTs ▪ Meta-analyses of moderate-quality RCTs	
LEVEL B-NR	**(Nonrandomized)**
▪ Moderate-quality evidence‡ from 1 or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies ▪ Meta-analyses of such studies	
LEVEL C-LD	**(Limited Data)**
▪ Randomized or nonrandomized observational or registry studies with limitations of design or execution ▪ Meta-analyses of such studies ▪ Physiological or mechanistic studies in human subjects	
LEVEL C-EO	**(Expert Opinion)**
Consensus of expert opinion based on clinical experience	

COR and LOE are determined independently (any COR may be paired with any LOE).

A recommendation with LOE C does not imply that the recommendation is weak. Many important clinical questions addressed in guidelines do not lend themselves to clinical trials. Although RCTs are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

* The outcome or result of the intervention should be specified (an improved clinical outcome or increased diagnostic accuracy or incremental prognostic information).

† For comparative-effectiveness recommendations (COR I and IIa; LOE A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

‡ The method of assessing quality is evolving, including the application of standardized, widely used, and preferably validated evidence grading tools; and for systematic reviews, the incorporation of an Evidence Review Committee.

COR indicates Class of Recommendation; EO, expert opinion; LD, limited data; LOE, Level of Evidence; NR, nonrandomized; R, randomized; and RCT, randomized controlled trial.

basis of the type, quantity, and consistency of data from clinical trials and other sources (**Table 1**) (P-4–P-6).

The reader is encouraged to consult the full-text guideline (P-9) for additional guidance and details about adult congenital heart disease because this executive summary contains mainly the recommendations.

Glenn N. Levine, MD, FACC, FAHA
Chair, ACC/AHA Task Force on Clinical Practice Guidelines

1. INTRODUCTION

1.1. Methodology and Evidence Review

The recommendations listed in this guideline are, whenever possible, evidence-based. An initial extensive evidence review, which included literature derived from research involving human subjects, published in English, and indexed in MEDLINE (through PubMed), EMBASE,

the Cochrane Library, the Agency for Healthcare Research and Quality, and other selected databases relevant to this guideline, was conducted from April 2014 to November 2014. Key search words included but were not limited to the following: *adult congenital heart disease, anesthesia, aortic aneurysm, aortic stenosis, atrial septal defect, arterial switch operation, bradycardia, bicuspid aortic valve, cardiac catheterization, cardiac imaging, cardiovascular magnetic resonance, cardiac reoperation, cardiovascular surgery, chest x-ray, cirrhosis, coarctation of the aorta, congenital heart defects, congenitally corrected transposition of the great arteries, contraception, coronary artery abnormalities, cyanotic congenital heart disease, dextro-transposition of the great arteries, double inlet left ventricle, Ebstein anomaly, echocardiography, Eisenmenger syndrome, electrocardiogram, endocarditis, exercise test, Fontan, heart catheterization, heart defect, heart failure, infertility, l-transposition of the great arteries, medical therapy, myocardial infarction, noncardiac surgery, patent ductus arteriosus, perioperative care, physical activity, postoperative complications, pregnancy, preoperative assessment, psychosocial, pulmonary arterial hypertension, hypoplastic left heart syndrome, pulmonary regurgitation, pulmonary stenosis, pulmonary valve replacement, right heart obstruction, right ventricle to pulmonary artery conduit, single ventricle, supravalvular pulmonary stenosis, surgical therapy, tachyarrhythmia, tachycardia, tetralogy of Fallot, transplantation, tricuspid atresia, Turner syndrome,* and *ventricular septal defect.* Additional relevant studies published through January 2018, during the guideline writing process, were also considered by the writing committee, and added to the evidence tables when appropriate. The final evidence tables, included in the Online Data Supplement, summarize the evidence used by the writing committee to formulate recommendations. References selected and published in this document are representative and not all-inclusive.

As noted in the preamble, an independent ERC was commissioned to perform a formal systematic review of critical clinical questions related to adult congenital heart disease (ACHD), the results of which were considered by the writing committee for incorporation into this guideline. Concurrent with this process, writing committee members evaluated study data relevant to the rest of the guideline. The findings of the ERC and the writing committee members were formally presented and discussed, and then recommendations were developed. The systematic review reports on "Medical Therapy for Systemic Right Ventricles: A Systematic Review (Part 1) for the 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease" (S1.1-1) and "Interventional Therapy Versus Medical Therapy for Secundum Atrial Septal Defect: A Systematic Review (Part 2) for the 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease" (S1.1-2) are published in conjunction with this guideline.

1.2. Organization of the Writing Committee

The writing committee consisted of pediatric and adult congenital cardiologists, interventional cardiologists, electrophysiologists, surgeons, and an advance practice nurse. The writing committee included representatives from the ACC, AHA, and American Association for Thoracic Surgery (AATS), American Society of Echocardiography (ASE), Heart Rhythm Society (HRS), International Society for Adult Congenital Heart Disease (ISACHD), Society for Cardiovascular Angiography and Interventions (SCAI), and the Society of Thoracic Surgeons (STS).

1.3. Document Review and Approval

This document was reviewed by 3 official reviewers each nominated by the ACC and AHA, and 1 to 2 reviewers each from the AATS, ASE, HRS, ISACHD, SCAI, STS; and 32 individual content reviewers. Reviewers' RWI information was distributed to the writing committee and is published in this document (Appendix 2).

This document was approved for publication by the governing bodies of the ACC and the AHA and endorsed by the AATS, ASE, HRS, ISACHD, SCAI, and STS.

1.4. Scope of the Guideline

The 2018 ACHD guideline is a full revision of the "2008 ACC/AHA Guidelines for the Management of Adults with Congenital Heart Disease" (S1.4-1), which was the first U.S. guideline to be published on the topic. This revision uses the 2008 ACHD guideline as a framework and incorporates new data and growing ACHD expertise to develop recommendations. Congenital heart disease (CHD) encompasses a range of structural cardiac abnormalities present before birth attributable to abnormal fetal cardiac development but does not include inherited disorders that may have cardiac manifestations such as Marfan syndrome or hypertrophic cardiomyopathy. Also not included are anatomic variants such as patent foramen ovale. Valvular heart disease (VHD) may be congenital, so management overlaps with the "2014 AHA/ACC Guidelines for the Management of Patients With Valvular Heart Disease" (S1.4-2), particularly for bicuspid aortic valve (BAV) disease. Where overlap exists, this document focuses on the diagnosis and treatment of congenital valve disease when it differs from acquired valve disease, whether because of anatomic differences, presence of concomitant lesions, or differences to consider given the relatively young age of patients with ACHD. This

guideline is not intended to apply to children (<18 years of age) with CHD or adults with acquired VHD, heart failure (HF), or other cardiovascular disease.

The prevalence of ACHD is growing because of the success of pediatric cardiology and congenital cardiac surgery in diagnosing and treating congenital heart defects in children. Improved survival to adulthood is most striking for those with the most severe disease, with survival to age 18 years now expected for 90% of children diagnosed with severe CHD (S1.4-3-S1.4-5). Patients with ACHD are a heterogeneous population, both in underlying anatomy and physiology, as well as surgical repair or palliation. Consequently, although the prevalence of ACHD is increasing, the population of patients with a given congenital abnormality or specific repair may be relatively small (S1.4-3, S1.4-6-S1.4-8).

Patients with CHD are not cured of their disease after successful treatment in childhood. Almost all patients with ACHD will have sequelae of either their native CHD or its surgical repair or palliation, although these sequelae can take decades to manifest. The heterogeneity of the population and the long symptom-free intervals constrain the ability to generate data applicable across the population of ACHD or to adults with specific lesions or repairs. Despite the difficulty in studying ACHD populations, there is a growing body of high-quality data in these patients to guide the care of this relatively "new" population, and whenever feasible, these data were used to develop recommendations. Recommendations are made based on the available data; however, when important clinical issues lacked data, first principles, extrapolation from data in other populations, and expert consensus are used to guide care. Patients with ACHD may have concomitant disease to which other existing guidelines apply, such as coronary artery disease, HF, and arrhythmias. The data from acquired heart disease populations may apply to some patients with ACHD, and those circumstances are acknowledged in these recommendations and referenced accordingly.

Patients with ACHD who are cared for in ACHD centers have better outcomes than those cared for in centers without ACHD expertise (S1.4-9), and this need for specialized care is noted throughout the guideline. These recommendations are intended to provide guidance to a wide variety of providers caring for patients with ACHD, including general, pediatric, and ACHD cardiologists, as well as surgeons, primary care providers, and other healthcare providers.

In developing the 2018 ACHD guideline, the writing committee reviewed previously published guidelines and related scientific statements. **Table 2** contains a list of publications and scientific statements deemed pertinent to this writing effort; it is intended for use as a resource and does not repeat existing guideline recommendations.

1.5. Abbreviations

Abbreviation	Meaning/Phrase
AAOCA	anomalous aortic origin of the coronary artery
ACHD	adult congenital heart disease
AP	anatomic and physiological
AR	aortic regurgitation
ASD	atrial septal defect
AVSD	atrioventricular septal defect
BAV	bicuspid aortic valve
CCT	cardiac computed tomography
CCTGA	congenitally corrected transposition of the great arteries
CHD	congenital heart disease
CMR	cardiovascular magnetic resonance
CoA	coarctation of the aorta
CPET	cardiopulmonary exercise test
CT	computed tomography
CTA	computed tomography angiography
d-TGA	dextro-transposition of the great arteries
ECG	electrocardiogram
ERC	Evidence Review Committee
GDMT	guideline-directed management and therapy
HF	heart failure
ICD	implantable cardioverter-defibrillator
IE	infective endocarditis
LV	left ventricular
LVOT	left ventricular outflow tract
PA	pulmonary artery
PAH	pulmonary arterial hypertension
PDA	patent ductus arteriosus
PR	pulmonary regurgitation
PS	pulmonary stenosis
Qp:Qs	pulmonary–systemic blood flow ratio
RV	right ventricular
RVOT	right ventricular outflow tract
SCD	sudden cardiac death
TEE	transesophageal echocardiography
TGA	transposition of the great arteries
TOF	tetralogy of Fallot
TR	tricuspid regurgitation
TTE	transthoracic echocardiography
VHD	valvular heart disease
VSD	ventricular septal defect

TABLE 2 Associated Guidelines and Statements

Title	Organization	Publication Year (Reference)
Guidelines		
Syncope	ACC/AHA/HRS	2017 (S1.4-10)
Supraventricular tachycardia	ACC/AHA/HRS	2015 (S1.4-11)
Cardiopulmonary resuscitation and emergency cardiovascular care—Part 8: postcardiac arrest care	AHA	2015 (S1.4-12)
Non-ST-elevation acute coronary syndromes	AHA/ACC	2014 (S1.4-13)
Perioperative cardiovascular evaluation and noncardiac surgery	ACC/AHA	2014 (S1.4-14)
Atrial fibrillation	AHA/ACC/HRS	2014 (S1.4-15)
Stable ischemic heart disease	ACC/AHA/ACP/AATS/PCNA/SCAI/STS	2014 (S1.4-16), 2012 (S1.4-17)
Assessment of cardiovascular risk	ACC/AHA	2014 (S1.4-18)
Blood cholesterol to reduce atherosclerotic cardiovascular risk in adults	ACC/AHA	2014 (S1.4-19)
Overweight and obesity in adults	AHA/ACC/TOS	2014 (S1.4-20)
Lifestyle management to reduce cardiovascular risk	AHA/ACC	2014 (S1.4-21)
Valvular heart disease	AHA/ACC	2017 (S1.4-22)
High blood pressure in adults	ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA	2017 (S1.4-23)
Aortic valve and ascending aorta	STS	2013 (S1.4-24)
ST-elevation myocardial infarction	ACC/AHA	2013 (S1.4-25)
Heart failure	ACC/AHA/HFSA	2017 (S1.4-26)
Device-based therapy for cardiac rhythm abnormalities	ACC/AHA/HRS	2012 (S1.4-27)
Coronary artery bypass graft surgery	ACC/AHA	2011 (S1.4-28)
Percutaneous coronary intervention	ACC/AHA/SCAI	2011 (S1.4-29)
Secondary prevention and risk reduction therapy	AHA/ACC	2011 (S1.4-30)
Cardiovascular disease in women	AHA/ACC	2011 (S1.4-31)
Grown-up congenital heart disease	ESC	2010 (S1.4-32)
Thoracic aortic disease	ACC/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM	2010 (S1.4-33)
Adult congenital heart disease	CCS	2010 (S1.4-34)
Infective endocarditis	ESC	2009 (S1.4-35)
Scientific statements		
Imaging for patients with transposition of the great arteries	ASE	2016 (S1.4-36)
Cardiac chamber quantification by echocardiography	ASE	2015 (S1.4-37)
Consensus on arrhythmia management in ACHD	PACES/HRS	2014 (S1.4-38)
Imaging for patients with repaired tetralogy of Fallot	ASE	2014 (S1.4-39)
Thoracic aortic disease	CCS	2014 (S1.4-40)
Promotion of physical activity for children and adults with CHD	AHA	2013 (S1.4-41)
Neurodevelopmental outcomes in children with CHD	AHA	2012 (S1.4-42)
Pregnancy in women with heart disease	ESC	2011 (S1.4-43)
Transition to adulthood for adolescents with CHD	AHA	2011 (S1.4-44)
Pulmonary hypertension	ACC/AHA	2009 (S1.4-45)
Prevention of infective endocarditis	AHA	2007 (S1.4-46)

AATS indicates American Association for Thoracic Surgery; ABC, Association of Black Cardiologists; ACC, American College of Cardiology; ACHD, adult congenital heart disease; ACP, American College of Physicians; ACPM, American College of Preventive Medicine; ACR, American College of Radiology; AGS, American Geriatrics Society; AHA, American Heart Association; APhA, American Pharmacists Association; ASA, American Stroke Association; ASE, American Society of Echocardiography; ASH, American Society of Hypertension; ASPC, American Society of Preventive Cardiology; CCS, Canadian Cardiovascular Society; CHD, congenital heart disease; ESC, European Society of Cardiology; HFSA, Heart Failure Society of America; HRS, Heart Rhythm Society; NMA, National Medical Association; PACES, Pediatric and Congenital Electrophysiology Society; PCNA, Preventive Cardiovascular Nurses Association; SCA, Society of Cardiovascular Anesthesiologists; SCAI, Society for Cardiovascular Angiography and Interventions; SIR, Society of Interventional Radiology; STS, Society of Thoracic Surgeons; SVM, Society for Vascular Medicine; and TOS, The Obesity Society.

2. BACKGROUND AND PATHOPHYSIOLOGY

2.1. Anatomic and Physiological Terms

The International Society for Nomenclature of Pediatric and Congenital Heart Disease (also known as the Nomenclature Working Group) defined, codified, mapped, and archived examples of nomenclatures and developed standards for terminology (S2.1-1–S2.1-5). The International Paediatric and Congenital Cardiac Code (IPCCC) nomenclature for anatomic lesions and repairs is used in this guideline (http://ipccc.net) (S2.1-6).

2.2. Severity of ACHD

In a patient with CHD, severity of disease is determined by native anatomy, surgical repair, and current physiology. Prior documents, including the 2008 ACHD guideline (S2.2-1), relied primarily on anatomic classifications to rank severity of disease. However, patients with the same underlying anatomy may have very different repairs and experienced variable physiological consequences of those repairs. For example, a patient with tetralogy of Fallot (TOF) after a valve-sparing primary repair may have excellent biventricular function with normal exercise capacity and no arrhythmias, whereas another patient of the same age with TOF may have had palliative shunting followed by a transannular patch repair resulting in severe pulmonary regurgitation (PR) with right ventricular (RV) enlargement, biventricular dysfunction, and ventricular tachycardia. To categorize disease severity in CHD in a more comprehensive way, the writing committee developed an ACHD Anatomic and Physiological (AP) classification system (**Tables 3 and 4**) that incorporates the previously described CHD anatomic variables as well as physiological variables, many of which have prognostic value in patients with ACHD.

2.3. The ACHD AP Classification

The ACHD AP classification (**Tables 3 and 4**), newly elaborated in this guideline, is intended to capture the complexity of ACHD anatomy and physiology, which are not always correlated. Certain anatomic abnormalities of clinical importance are shared across diagnoses (e.g., aortic enlargement), which may be found in patients with BAV, coarctation of the aorta (CoA), transposition of the great arteries, and TOF, amongst others. In every patient, anatomic and physiological variables should be considered. In using **Tables 3 and 4**, a patient should be classified based on the "highest" relevant anatomic or physiological feature. For example, a normotensive patient with repaired CoA, normal exercise capacity, and normal end-organ function would be ACHD AP classification IIA, whereas an otherwise similar patient with ascending aortic diameter of 4.0 cm would be ACHD AP classification IIB, and if moderate aortic stenosis were also present, the ACHD AP classification would be IIC.

Patients with ACHD may have baseline exercise limitations, cyanosis, end-organ dysfunction, or other clinically important comorbidities related to their CHD. They are also at risk of HF, arrhythmias, sudden cardiac death (SCD), and development or progression of cardiac symptoms such as dyspnea, chest pain, and exercise intolerance. Concomitant valvular disease or aortic pathology may be present. There are growing data regarding the prognostic implications of these variables in patients with ACHD, but not the abundance of data available for patients with acquired heart disease (S2.3-1–S2.3-16).

The variables forming part of the ACHD AP classification (**Table 3**) were selected because data exist suggesting their importance in prognosis, management, or quality of life. As new data become available, we expect changes in the relative weights attributed to the components of the ACHD AP classification and perhaps new components, resulting in a scheme that ever more precisely tracks overall severity of disease and need for more or less intensive follow-up and management.

Similar to the New York Heart Association (NYHA) classification of functional status, patients may move from one ACHD AP classification to another over time. If clinical status worsens, the classification will change to a higher severity group, but improvement in status, for example after an intervention such as valve replacement or control of arrhythmia, can result in change to a lower severity classification. Such movement among classes is unlike the AHA HF A to D classification (S2.3-17), in which patients move in only one direction. This ACHD AP classification is used throughout this document, particularly when considering follow-up visits and need for testing. As the ACHD AP classification worsens because of changes in physiology (e.g., development of arrhythmias, HF, end-organ disease), the nature and frequency of recommended follow-up visits and testing will also change, adapting to the patient's changing circumstance instead of depending solely on a description of anatomic disease, which may not adequately discriminate physiological changes that alter severity over time.

Some patients with ACHD may have substantial acquired comorbidities unrelated to CHD, and as a consequence, their follow-up strategies might be more appropriately be based on other existing guidelines for acquired heart disease. For example, an 80-year-old patient who has a small atrial septal defect (ASD), but whose symptoms are related to diastolic HF, chronic kidney disease caused by hypertension and diabetes mellitus, and moderate aortic stenosis is well-suited to be followed according to existing guidelines for those diseases, rather than according to the ACHD AP classification for the ASD.

TABLE 3 Physiological Variables as Used in ACHD AP Classification

Variable	Description
Aortopathy	Aortic enlargement is common in some types of CHD and after some repairs. Aortic enlargement may be progressive over a lifetime. There is no universally accepted threshold for repair, nor is the role of indexing to body size clearly defined in adults, as is done in pediatric populations. For purposes of categorization and timing of follow-up imaging (S2.2-2–S2.2-4): ■ Mild aortic enlargement is defined as maximum diameter 3.5–3.9 cm ■ Moderate aortic enlargement is defined as maximum diameter 4.0–4.9 cm ■ Severe aortic enlargement is defined as maximum diameter ≥5.0 cm

Arrhythmia	Arrhythmias are very common in patients with ACHD and may be both the cause and consequence of deteriorating hemodynamics, valvular dysfunction, or ventricular dysfunction. Arrhythmias are associated with symptoms, outcomes, and prognosis (S2.2-5–S2.2-8), thus are categorized based on presence and response to treatment.
	■ No arrhythmia–No documented clinically relevant atrial or ventricular tachyarrhythmias
	■ Arrhythmia not requiring treatment–Bradyarrhythmia, atrial or ventricular tachyarrhythmia not requiring antiarrhythmic therapy, cardioversion, or ablation
	■ Arrhythmia controlled with therapy:
	  ■ Bradyarrhythmia requiring pacemaker implantation
	  ■ Atrial or ventricular tachyarrhythmia requiring antiarrhythmic therapy, cardioversion, or ablation
	  ■ AF and controlled ventricular response
	  ■ Patients with an ICD
	■ Refractory arrhythmias:
	  ■ Atrial or ventricular tachyarrhythmia currently unresponsive to or refractory to antiarrhythmic therapy or ablation

Concomitant VHD	Severity defined according to the 2014 VHD guideline (S2.2-2). ■ Mild VHD ■ Moderate VHD ■ Severe VHD
End-organ dysfunction	Clinical and/or laboratory evidence of end-organ dysfunction (S2.2-9–S2.2-11) including ■ Renal (kidney) ■ Hepatic (liver) ■ Pulmonary (lung)
Exercise capacity	Patients with ACHD are often asymptomatic notwithstanding exercise limitations demonstrated as diminished exercise capacity when evaluated objectively (S2.2-12–S2.2-14). Thus, assessment of both subjective and objective exercise capacity is important (see NYHA classification system below). Exercise capacity is associated with prognosis (S2.2-15–S2.2-17). ■ Abnormal objective cardiac limitation to exercise is defined as an exercise maximum ventilatory equivalent of oxygen below the range expected for the specific CHD anatomic diagnosis (S2.2-18). ■ Expected norms for CPET values should take into account age, sex, and underlying congenital diagnosis. Published studies with institution-specific norms can be used as guides, bearing in mind variability among institutional norms and ranges.
Hypoxemia/hypoxia/cyanosis	See Section 3.15. for detailed definition of cyanosis. ■ Hypoxemia is defined as oxygen saturation measured by pulse oximetry at rest ≤90%. ■ Severe hypoxemia is defined as oxygen saturation at rest <85%. ■ In patients with normal or high hemoglobin concentrations, severe hypoxemia will be associated with visible cyanosis (which requires ≥5g/L desaturated hemoglobin to be appreciated). ■ The terms cyanosis and hypoxemia (or hypoxia) are sometimes used interchangeably. Such interchangeability would not apply; however, in the presence of anemia, severe hypoxemia can be present without visible cyanosis.
NYHA functional classification system (S2.3-31)	**Class Functional Capacity** I Patients with cardiac disease but resulting in no limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain. II Patients with cardiac disease resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain. III Patients with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes fatigue, palpitation, dyspnea, or anginal pain. IV Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. Symptoms of HF or the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort increases.
Pulmonary hypertension	Pulmonary hypertension is a broad term that encompasses pulmonary arterial hypertension, which is pulmonary hypertension with increased pulmonary vascular resistance. This document defines PH and PAH as they are used in the field of pulmonary hypertension. Pulmonary hypertension is defined as: ■ Mean PA pressure by right heart catheterization ≥25 mm Hg. PAH is defined as: ■ Mean PA pressure by right heart catheterization ≥25 mm Hg and a pulmonary capillary wedge pressure ≤15 mm Hg and pulmonary vascular resistance ≥3 Wood units (S2.2-20)
Shunt (hemodynamically significant shunt)	An intracardiac shunt is hemodynamically significant if: ■ There is evidence of chamber enlargement distal to the shunt ■ And/or evidence of sustained Qp:Qs ≥1.5:1 ■ An intracardiac shunt not meeting these criteria would be described as small or trivial
Venous and arterial stenosis	■ Aortic recoarctation after CoA repair ■ Supravalvular aortic obstruction ■ Venous baffle obstruction ■ Supravalvular pulmonary stenosis ■ Branch PA stenosis ■ Stenosis of cavopulmonary connection ■ Pulmonary vein stenosis

ACHD indicates adult congenital heart disease; AF, atrial fibrillation; AP, anatomic and physiologic; CHD, congenital heart disease; CoA, coarctation of the aorta; CPET, cardiopulmonary exercise test; HF, heart failure; ICD, implantable cardioverter-defibrillator; NYHA, New York Heart Association; PA, pulmonary artery; PAH, pulmonary arterial hypertension; Qp:Qs, pulmonary-systemic blood flow ratio; and VHD, valvular heart disease.

TABLE 4 ACHD AP Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)

CHD Anatomy*

I: Simple

Native disease

- Isolated small ASD
- Isolated small VSD
- Mild isolated pulmonic stenosis

Repaired conditions

- Previously ligated or occluded ductus arteriosus
- Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement
- Repaired VSD without significant residual shunt or chamber enlargement

II: Moderate Complexity

Repaired or unrepaired conditions

- Aorto-left ventricular fistula
- Anomalous pulmonary venous connection, partial or total
- Anomalous coronary artery arising from the pulmonary artery
- Anomalous aortic origin of a coronary artery from the opposite sinus
- AVSD (partial or complete, including primum ASD)
- Congenital aortic valve disease
- Congenital mitral valve disease
- Coarctation of the aorta
- Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
- Infundibular right ventricular outflow obstruction
- Ostium primum ASD
- Moderate and large unrepaired secundum ASD
- Moderate and large persistently patent ductus arteriosus
- Pulmonary valve regurgitation (moderate or greater)
- Pulmonary valve stenosis (moderate or greater)
- Peripheral pulmonary stenosis
- Sinus of Valsalva fistula/aneurysm
- Sinus venosus defect
- Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
- Supravalvar aortic stenosis
- Straddling atrioventricular valve
- Repaired tetralogy of Fallot
- VSD with associated abnormality and/or moderate or greater shunt

III: Great Complexity (or Complex)

- Cyanotic congenital heart defect (unrepaired or palliated, all forms)
- Double-outlet ventricle
- Fontan procedure
- Interrupted aortic arch
- Mitral atresia
- Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)
- Pulmonary atresia (all forms)
- TGA (classic or d-TGA; CCTGA or l-TGA)
- Truncus arteriosus
- Other abnormalities of atrioventricular and ventriculoarterial connection (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Physiological Stage

A

- NYHA FC I symptoms
- No hemodynamic or anatomic sequelae
- No arrhythmias
- Normal exercise capacity
- Normal renal/hepatic/pulmonary function

B

- NYHA FC II symptoms
- Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction)
- Mild valvular disease
- Trivial or small shunt (not hemodynamically significant)
- Arrhythmia not requiring treatment
- Abnormal objective cardiac limitation to exercise

Continued in the next column

TABLE 4 Continued

CHD Anatomy*

C

- NYHA FC III symptoms
- Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonic, or both)
- Moderate aortic enlargement
- Venous or arterial stenosis
- Mild or moderate hypoxemia/cyanosis
- Hemodynamically significant shunt
- Arrhythmias controlled with treatment
- Pulmonary hypertension (less than severe)
- End-organ dysfunction responsive to therapy

D

- NYHA FC IV symptoms
- Severe aortic enlargement
- Arrhythmias refractory to treatment
- Severe hypoxemia (almost always associated with cyanosis)
- Severe pulmonary hypertension
- Eisenmenger syndrome
- Refractory end-organ dysfunction

*This list is not meant to be comprehensive; other conditions may be important in individual patients.

ACHD indicates adult congenital heart disease; AP, anatomic and physiological; ASD, atrial septal defect; AVSD, atrioventricular septal defect; CCTGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; d-TGA, dextro-transposition of the great arteries; FC, functional class; HCM, hypertrophic cardiomyopathy; l-TGA, levo-transposition of the great arteries; NYHA, New York Heart Association; TGA, transposition of the great arteries; and VSD, ventricular septal defect.

Nevertheless, the added hemodynamic complexity brought by the ASD must be kept in mind.

Throughout this document, the ACHD AP classification is used to help guide resource utilization, including ACHD consultation and routine diagnostic studies.

3. GENERAL PRINCIPLES

See Online Data Supplements 1 and 2 for additional data supporting this section.

3.1. ACHD Program

Patients with complex CHD have generally better outcomes when cared for in an integrated, collaborative, and multidisciplinary program (S3.1-1). Many medical issues in patients with ACHD involve cardiac sequelae, and the diagnosis and management may require cardiac anesthesiologists, electrophysiologists, and interventional cardiologists; imaging services such as cardiovascular magnetic resonance (CMR)/cardiac computed tomography (CCT); and pulmonary hypertension services with expertise in ACHD (**Table 5**). Appropriate specialty care must be available to address pregnancy, acquired cardiovascular disease, and acute noncardiac illness complicating CHD, management of which is frequently more complicated in patients with ACHD.

Although individual providers may be community-based affiliates, ACHD programs are inpatient, outpatient, and hospital-based with staffing and expertise available on-site or accessible when needed (**Table 5**).

TABLE 5 Key Personnel and Services Recommended for ACHD Programs

Personnel
ACHD board-eligible/board-certified cardiologists
Congenital cardiac surgeons
Nurses/physician assistants/nurse practitioners
Cardiac anesthesiologists with CHD training/expertise
Multidisciplinary teams: ■ High-risk obstetrics ■ Pulmonary hypertension ■ HF/transplant ■ Genetics ■ Hepatology ■ Cardiac pathology ■ Rehabilitation services ■ Social services ■ Psychological services ■ Financial counselors
Services
Echocardiography, including TEE and intraoperative TEE*
CHD diagnostic and interventional catheterization*
CHD electrophysiology/pacing/ICD implantation*: ■ Exercise testing ■ Echocardiographic ■ Radionuclide ■ Cardiopulmonary
Cardiac imaging/radiology*: ■ CMR ■ CCT ■ Nuclear medicine
Information technology: ■ Data collection ■ Database support ■ Quality assessment review/protocols

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

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CHD, congenital heart disease; CMR, cardiovascular magnetic resonance; HF, heart failure; ICD, implantable cardioverter-defibrillator; and TEE, transesophageal echocardiography.

3.2. Access to Care

Recommendation for Access to Care
Referenced studies that support the recommendation are summarized in Online Data Supplement 3.

COR	LOE	RECOMMENDATION
I	B-NR C-EO	1. Physicians caring for patients with ACHD should support access to care by a) assuring smooth transitions for adolescents and young adults from pediatric to adult providers (S3.2-1, S3.2-2) (Level of Evidence: B-NR); and b) promoting awareness of the need for lifelong specialized care through outreach and educational programs (Level of Evidence: C-EO).

3.3. Delivery of Care

Recommendations for Delivery of Care
Referenced studies that support recommendations are summarized in Online Data Supplements 3, 4, and 5.

COR	LOE	RECOMMENDATIONS
I	B-NR	1. Patients with ACHD AP classification IB-D, IIA-D, and IIIA-D* should be managed in collaboration with an ACHD cardiologist (S3.3-1).
I	C-LD	2. Cardiac surgery, catheter-based interventional cardiac procedures, and electrophysiological procedures involving congenital heart lesions in patients with ACHD should be performed by operators with expertise in CHD procedures and in collaboration with an ACHD cardiologist (S3.3-1, S3.3-2).

*See Tables 3 and 4 for details on the ACHD Anatomic and Physiological classification system.

Table 6 addresses delivery of care where circumstances of ACHD expertise may improve patient outcomes.

TABLE 6 Delivery of Care: Circumstances Where ACHD Expertise May Improve Outcomes

Circumstance	Possible Solution	Rationale	Example
Care of patients in the lowest ACHD AP classification (IA)*	1. Face-to-face consultation with an ACHD cardiologist. 2. Collaborative care planning between an ACHD patient's general cardiologist or primary care provider and an ACHD cardiologist.	1. Patients in ACHD AP classification IA* are likely to be asymptomatic and not require frequent routine congenital cardiac care. 2. The very long-term outcomes of patients with ACHD AP classification IA* lesions have not been well described, although available data suggest that patients with simple CHD have higher cardiac mortality in long-term follow-up than age-matched controls (S3.3-3). 3. Consultation with an ACHD cardiologist should help to accurately assess the patient's ACHD AP class, provide information regarding potential long-term outcomes, and reinforce signs and symptoms that should prompt further evaluation.	Patients with small VSDs are thought to have excellent long-term survival, although complications (double-chamber RV, IE, aortic valve prolapse and aortic regurgitation) may manifest in adulthood; consequently, patients with small VSDs warrant lifelong follow-up (S3.3-4).
Cardiac imaging of patients with ACHD	Imaging studies should be performed and interpreted by individuals with expertise in CHD imaging.	1. The complexity and variability of lesions, repairs, and sequelae in CHD constrain the use of standard protocols and sequences and often require modification of plans during acquisition of images, as well as specialized skills in interpretation. Thus, CHD expertise is helpful for optimal quality and interpretation of cardiac imaging studies. 2. Use of a multimodality cardiac imaging approach can be used for patients with ACHD, accounting for patient-specific considerations, strengths and weaknesses of each modality, institutional resources, and expertise. 3. ACHD programs need a dedicated CMR service, and CMR expertise is integral to an ACHD program, as is expertise in ACHD CCT (S3.3-5, S3.3-6).	Although imaging of a patient with TOF may seem straightforward because many have familiar chamber and great vessel relationships, there are nuances to echocardiographic imaging of RV size and function, PR severity, and/or location of right ventricular outflow tract obstruction that affect clinical care and are thus best carried out by sonographers and echocardiographers with appropriate expertise. Similarly, expertise in congenital CMR is important in evaluating patients with TOF, as RV volumes and function are key components in evaluation for timing of pulmonary valve replacement (S3.3-7, S3.3-8).
Electrophysiological care of patients with ACHD	Perform procedures in electrophysiology laboratories equipped for 3D mapping and ablation and involve specialists experienced in the management of arrhythmias in patients with ACHD.	Examples of diagnostic questions best answered by electrophysiological study: a) evaluation of the conduction system in cases of suspected postsurgical conduction abnormalities b) evaluation of syncope c) diagnosis of the mechanism of supraventricular tachycardia or wide complex tachycardia d) programmed ventricular stimulation particularly in patients following repair of TOF and its variants (Section 4.4.1.) as well as preoperative assessment of arrhythmia substrates that may be amenable to operative intervention, such as an atrial maze procedure for atrial arrhythmias. The latter procedure is commonly used at the time of conversion of atriopulmonary connection Fontan, and may also be useful in other forms of repaired CHD with postoperative atrial arrhythmias such as TOF.	Bradyarrhythmia and tachyarrhythmias are common in TGA with atrial switch patients and may seem "straightforward," but the altered anatomy adds complexity to the procedures and emphasizes the need for specialized equipment and expertise to ensure the best chance for procedural success. For example: 1) pacemaker placement in a patient with TGA with atrial switch can be challenging because of the altered atrial anatomy and interatrial baffle that will necessitate placement of an atrial lead in the anatomic left atrium, often scarred such that tissue amenable to pacing is difficult to find; and 2) atrial flutter is a common arrhythmia in TGA with atrial switch, but the flutter circuit may be on the systemic side of the interatrial baffle and thus may require baffle puncture or retrograde approach to effectively ablate the circuit.
Diagnostic and interventional cardiac procedures, including electrophysiology procedures	1. Perform procedure in a hospital with cardiologists, anesthesiologists, surgeons, and other providers with expertise in the management of patients with ACHD. 2. Consultation with providers with ACHD expertise may be substituted if the procedure is urgent such that timely transfer is not feasible.	1. Patients with ACHD often have complex underlying cardiac anatomy and physiology. 2. The data obtained and the interventions performed during ACHD cardiac procedures are difficult to sort out without specialized knowledge of the CHD. 3. An ACHD program has additional resources such as cardiac anesthesia, congenital cardiac surgery, and specialty cardiac imaging, should the need for those services arise during or after the procedure.	In patients with CHD, the presence of anatomic and physiological complexity from the specific defect or surgical palliation, may change the overall care plan and procedural decision-making. Procedures that may seem straightforward, such as pacemaker implantation or ASD closure, may be more complex when accounting for the nuances imparted by CHD.

Continued in the next column

TABLE 6 Continued

Circumstance	Possible Solution	Rationale	Example
Administration of anesthesia for invasive procedures in patients with ACHD AP classification IB-D, IIA-D, and IIIA-D*	1. Performed by, or in collaboration with, an anesthesiologist with expertise in the management of patients with ACHD. 2. If clinical urgency precludes transfer, consultation with an anesthesiologist with ACHD expertise would be of benefit to on-site providers who do not have ACHD expertise.	1. ACHD-specific issues need to be addressed when considering anesthesia, including underlying cardiac physiology and hemodynamics, and the effects of anesthetic medications and ventilation strategies. 2. Many patients with ACHD have had surgeries in the past, which may have created or identified airway or vascular access concerns. Patients with ACHD can also have underlying restrictive and/or obstructive lung disease that should be considered (S3.3-9, S3.3-10).	The application of anesthesia for laparoscopic procedures can be especially challenging in Fontan patients. Significant cardiovascular and respiratory alterations may occur as a result of increased intra-abdominal pressure and decreased venous return. Abdominal insufflation may lead to lower preload and hypotension, while at the same time elevating systemic vascular resistance and compromising cardiac output. Elevations in pulmonary vascular resistance attributable to hypercarbia can be caused by either direct carbon dioxide absorption or hypoventilation (S3.3-11).
Patients with ACHD and pulmonary hypertension	1. Consultation with experts in pulmonary hypertension and ACHD to assist in the interpretation of diagnostic and invasive studies and determine the best course of management.	1. PAH imparts a poor prognosis compared with CHD without PAH. Because of the complexity of PAH in the setting of CHD, patients with ACHD benefit from the expertise of both ACHD providers and pulmonary hypertension providers (S3.3-12–S3.3-20).	Management of PAH in patients with shunts can be difficult. For example, in patients for whom PAH treatment is expected to allow subsequent closure of a shunt, cohort series demonstrate progression of pulmonary vascular resistance or late mortality if defects with associated pulmonary vascular resistance elevation beyond 2.5 Wood units (≥4 Wood units×m^2) or Qp:Qs ≥3 were closed (S3.3-21, S3.3-22). The utility of acute administration of pulmonary vasodilator therapy as a marker of reversibility of pulmonary vascular resistance remains uncertain. "Treat-to-repair" strategies involving use of PAH therapies to bring pulmonary vascular resistance into a range where repair can be considered have been applied, but the utility of such strategies also remains uncertain.

*See Tables 3 and 4 for details on the ACHD AP classification system.

3D indicates 3-dimensional; ACHD, adult congenital heart disease; AP, anatomic and physiological; ASD, atrial septal defect; CCT, cardiac computed tomography; CHD, congenital heart disease; CMR, cardiovascular magnetic resonance; IE, infective endocarditis; PAH, pulmonary arterial hypertension; PR, pulmonary regurgitation; Qp:Qs, pulmonary–systemic blood flow ratio; RV, right ventricle; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; and VSD, ventricular septal defect.

3.4. Evaluation of Suspected and Known CHD

3.4.1. Electrocardiogram

See also Table 7.

Recommendations for Electrocardiogram

COR	LOE	RECOMMENDATIONS
I	C-EO	1. A standard 12-lead electrocardiogram (ECG) is recommended in adults with CHD with serial assessment depending on the specific ACHD AP classification or when symptoms develop or worsen.
I	C-EO	2. Ambulatory electrocardiographic monitoring should be performed in patients with CHD who are at risk of tachyarrhythmia, bradyarrhythmia or heart block, or when symptoms possibly of arrhythmic origin develop.

TABLE 7 Use of ECGs in ACHD Evaluation

- Identification of sinus bradycardia or junctional rhythm in patients at risk of sinus node dysfunction (especially after the Mustard, Senning, Glenn, or Fontan procedure)
- Identification of clinically inapparent intra-atrial re-entry tachycardia in patients who have had atriotomy
- Identification of atrioventricular block in patients at risk for progression of atrioventricular conduction system disease (especially CCTGA)
- Evaluation of rhythm in patients with pacemakers
- Measurement of QRS duration in patients after repair of TOF and as part of CRT evaluation
- Preoperatively to compare with postoperative ECGs in patients undergoing heart surgery and noncardiac surgery
- Postoperatively to identify arrhythmias (e.g., atrial ectopic tachycardia, atrial flutter, AF, junctional ectopic tachycardia, atrioventricular block)
- Diagnosis of Wolff-Parkinson-White Syndrome in patients with Ebstein anomaly
- Initial evaluation of suspected acute coronary syndromes

ACHD indicates adult congenital heart disease; AF, atrial fibrillation; CCTGA, congenitally corrected transposition of the great arteries; CRT, cardiac resynchronization therapy; ECG, electrocardiogram; and TOF, tetralogy of Fallot.

3.4.2. Ionizing Radiation Principles

Recommendation for Ionizing Radiation Principles
Referenced studies that support the recommendation are summarized in Online Data Supplement 6.

COR	LOE	RECOMMENDATION
I	B-NR	1. Strategies to limit and monitor radiation exposure are recommended during imaging of patients with ACHD, with studies not involving ionizing radiation chosen whenever appropriate (S3.4.2-1–S3.4.2-4).

3.4.3. Echocardiography

Recommendations for Echocardiography
Referenced studies that support recommendations are summarized in Online Data Supplement 7.

COR	LOE	RECOMMENDATIONS
I	B-NR	1. Intraoperative TEE is recommended to guide surgical repair of CHD in adults (S3.4.3-1).
I	C-EO	2. Patients with ACHD should undergo transthoracic echocardiography (TTE) for initial assessment, with timing of serial assessment based on anatomic and physiological severity and clinical status.

3.4.4. CMR Imaging

See also **Tables 8 and 9**.

Recommendations for CMR Imaging
Referenced studies that support recommendations are summarized in Online Data Supplement 8.

COR	LOE	RECOMMENDATIONS
I	B-NR	1. In patients with ACHD who have or who are at risk of developing RV enlargement and dysfunction, serial CMR is recommended for quantitative assessment of RV size and function (S3.4.4-1–S3.4.4-3).
IIa	C-LD	2. CMR can be useful in the initial evaluation and serial assessment of selected patients with CHD based on anatomic complexity and clinical status (S3.4.4-1, S3.4.4-2, S3.4.4-4–S3.4.4-10).

TABLE 8 Circumstances Where CMR, CCT, TEE, and/or Cardiac Catheterization May be Superior to TTE

- Assessment of RV size and function in repaired TOF, systemic right ventricles, and other conditions associated with RV volume and pressure overload (S3.4.4-1, S3.4.4-3)
- Identification of anomalous pulmonary venous connections (S3.4.4-11)
  - Serial assessment of thoracic aortic aneurysms, especially when the dilation might extend beyond the echocardiographic windows (S3.4.4-7)
- Accurate assessment of PA pressure and pulmonary vascular resistance
- Assessment for recoarctation of the aorta
- Sinus venosus defects
- Vascular rings
- Evaluation of coronary anomalies
- Quantification of valvular regurgitation

CCT indicates cardiac computed tomography; CMR, cardiovascular magnetic resonance; PA, pulmonary artery; RV, right ventricular; TEE, transesophageal echocardiography; and TOF, tetralogy of Fallot.

TABLE 9 Comparison of Imaging Modalities Useful in ACHD Evaluation

	Radiation Exposure	Relative Cost	Ventricular Volumes/ Function	Valvular Structure/ Function	Coronary Anatomy and Course	Extracardiac Vascular Anatomy
Echocardiography	No	$	++	+++	+/-	+/-
CMR	No	$$	+++	++	++*	+++
CCT	Yes	$$	+*	+	+++	+++
Cardiac catheterization	Yes	$$	+	++	+++	++

$ indicates less expensive; $$, more expensive; +/-, possible value; +, good; ++, very good; and +++, excellent.
*In specific gated imaging protocols.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; and CMR, cardiovascular magnetic resonance.

3.4.5. Cardiac Computed Tomography

Recommendation for Cardiac Computed Tomography
Referenced studies that support the recommendation are summarized in Online Data Supplement 9.

COR	LOE	RECOMMENDATION
IIa	C-LD	1. CCT imaging can be useful in patients with ACHD when information that cannot be obtained by other diagnostic modalities is important enough to justify the exposure to ionizing radiation (S3.4.5-1, S3.4.5-2).

3.4.6. Cardiac Catheterization

Recommendations for Cardiac Catheterization
Referenced studies that support recommendations are summarized in Online Data Supplement 10.

COR	LOE	RECOMMENDATIONS
I	C-LD	1. Cardiac catheterization (hemodynamic and/or angiographic) in patients with ACHD AP classification II and III, or interventional cardiac catheterization in patients with ACHD AP classification I to III should be performed by, or in collaboration with, cardiologists with expertise in ACHD (S3.4.6-1–S3.4.6-4).
IIa	B-NR	2. In patients with a low or intermediate pretest probability of coronary artery disease (CAD), use of CT coronary angiography is reasonable to exclude significant obstructive CAD when cardiac catheterization has significant risk or because of patient preference (S3.4.6-5–S3.4.6-9).

3.4.7. Exercise Testing

Recommendations for Exercise Testing
Referenced studies that support recommendations are summarized in Online Data Supplement 11.

COR	LOE	RECOMMENDATIONS
IIa	B-NR	1. In patients with ACHD, cardiopulmonary exercise testing (CPET) can be useful for baseline functional assessment and serial testing (S3.4.7-1, S3.4.7-2).
IIa	C-LD	2. In symptomatic patients with ACHD, a 6-minute walk test can be useful to objectively assess symptom severity, functional capacity, and response to therapy (S3.4.7-3, S3.4.7-4).

3.5. Transition Education

Recommendation for Transition Education
Referenced studies that support the recommendation are summarized in Online Data Supplement 12.

COR	LOE	RECOMMENDATION
I	B-NR	1. Clinicians caring for patients with CHD should deliver developmentally appropriate transition education to adolescent and young patients with CHD, and to their families/support network (S3.5-1, S3.5-2).

3.6. Exercise and Sports

Recommendations for Exercise and Sports
Referenced studies that support recommendations are summarized in Online Data Supplement 13.

COR	LOE	RECOMMENDATIONS
I	C-LD	1. Clinicians should assess activity levels at regular intervals and counsel patients with ACHD about the types and intensity of exercise appropriate to their clinical status (S3.6-1–S3.6-9).
IIa	C-LD	2. CPET can be useful to guide activity recommendations for patients with ACHD (S3.6-10, S3.6-11).
IIa	B-NR	3. Cardiac rehabilitation can be useful to increase exercise capacity in patients with ACHD (S3.6-12, S3.6-13).

3.7. Mental Health and Neurodevelopmental Issues

Recommendations for Mental Health and Neurodevelopmental Issues
Referenced studies that support recommendations are summarized in Online Data Supplement 14.

COR	LOE	RECOMMENDATIONS
I	B-NR	1. Patients with ACHD should be evaluated for depression and anxiety (S3.7-1–S3.7-3)
IIa	B-NR	2. Referral for mental health evaluation and treatment is reasonable in patients with ACHD (S3.7-1–S3.7-4).
IIb	B-NR	3. Neurodevelopmental or neuropsychological testing may be considered in some patients with ACHD to guide therapies that enhance academic, behavioral, psychosocial, and adaptive functioning (S3.7-5–S3.7-9).

3.8. Endocarditis Prevention

Patients with ACHD have an increased risk of developing infective endocarditis (IE) (S3.8-1, S3.8-2). The most common pathogens responsible for IE include *Streptococcus viridans*, *Staphylococcus* species, and *Enterococcus* species. Despite advances in antimicrobial therapy and surgical techniques, IE remains a condition associated with significant morbidity and mortality. Numerous guidelines are available with recommendations on the prevention and diagnosis of IE (S3.8-3–S3.8-5). These guidelines include consistent descriptions of the patients at highest risk of adverse effects from endocarditis. Antibiotic prophylaxis continues to be recommended for patients with high-risk characteristics, which are often found in patients with ACHD (S3.8-2). These patients include:

- Those with previous IE;
- Patients with prosthetic valves (biological and mechanical, surgical and transcatheter);
- Patients within 6 months of placement of prosthetic material;
- Patients with residual intracardiac shunts at the site of or adjacent to previous repair with prosthetic material or devices; or
- Patients with uncorrected cyanotic heart disease.

See Online Data Supplement 15 for referenced studies.

3.9. Concomitant Syndromes

See also **Table 10**.

Recommendation for Concomitant Syndromes
Referenced studies that support the recommendation are summarized in Online Data Supplement 16.

COR	LOE	RECOMMENDATION
IIa	B-NR	1. Genetic testing for 22q11 deletions is reasonable for patients with conotruncal cardiac defects (S3.9-1, S3.9-2).

TABLE 10 Underlying Genetic Syndromes Commonly Associated With CHD (S3.9-3, S3.9-4)

Syndrome	Genetic Abnormality	Clinical Features	Common Cardiac Findings
DiGeorge syndrome (velocardiofacial syndrome)	22q11.2 deletion	Thymic and parathyroid hypoplasia, immunodeficiency, low-set ears, hypocalcemia, speech and learning disorders, renal anomalies, psychiatric disease 25%–75% have CHD, depending on age studied (S3.9-5, S3.9-6)	IAA type B, aortic arch anomalies, truncus arteriosus, TOF
Down syndrome	Trisomy 21	Developmental disability, characteristic facial features, hypotonia, palmar crease 40%–50% have CHD	ASD, VSD, AVSD, TOF
Holt-Oram syndrome (S3.9-7)	TBX5	Upper limb skeletal abnormalities 75% have CHD	ASD, VSD, MV disease
Klinefelter syndrome	47 XXY	Tall stature, hypoplastic testes, delayed puberty, developmental disability 50% have CHD	PDA, ASD, MV prolapse
Noonan syndrome (S3.9-8)	PTPN11, KRAS, SOS1 RAF1, NRAS, BRAF, MAP2K1	Facial anomalies, webbed neck, chest deformity, short stature, lymphatic abnormalities, bleeding abnormalities 80% have CHD	PS, ASD, HCM
Turner syndrome	45X	Short stature, webbed neck, lymphedema, primary amenorrhea 30% have CHD Risk of aortic dissection	Coarctation, BAV, aortic stenosis, hypoplastic left heart, ascending aortopathy
Williams syndrome	7q11.23 deletion	Elfin face, social personality, hearing loss, developmental delay, infantile hypercalcemia 50%–80% have CHD	Supravalvar aortic stenosis, peripheral PS

ASD indicates atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CHD, congenital heart disease; HCM, hypertrophic cardiomyopathy; IAA, interrupted aortic arch; MV, mitral valve; PDA, patent ductus arteriosus; PS, pulmonary stenosis; TOF, tetralogy of Fallot; and VSD, ventricular septal defect.

3.10. Noncardiac Medical Issues

Recommendation for Noncardiac Medical Issues
Referenced studies that support the recommendation are summarized in Online Data Supplement 18.

COR	LOE	RECOMMENDATION
I	C-LD	1. Patients with ACHD at risk for hepatitis C should be screened and vaccinated for viral hepatitis and treated as appropriate (S3.10-1).

3.11. Noncardiac Surgery

See also Table 11.

Recommendations for Noncardiac Surgery
Referenced studies that support recommendations are summarized in Online Data Supplement 18.

COR	LOE	RECOMMENDATIONS
I	C-LD	1. Optimization before and close surveillance after invasive procedures, regardless of the complexity of the anatomic defect or type of procedure is beneficial for patients with ACHD (S3.11-1–S3.11-4).
I	B-NR	2. In patients with ACHD AP classification IB-D, IIA-D, and IIIA-D* noncardiac surgical and interventional procedures should be performed in a hospital with or in consultation with experts in ACHD when possible (S3.11-1, S3.11-3, S3.11-5–S3.11-9).

*See Tables 3 and 4 for details on the ACHD AP classification system.

TABLE 11 ACHD Management Issues for Noncardiac Surgery

Clarify CHD diagnosis

- ≻ Clarify prior procedures, residua, sequelae, and current status, including ACHD AP classification
- ≻ Be aware that history obtained from only the patient and family may be faulty or incomplete
- ≻ Obtain and review old records to ensure accurate understanding of past procedures and clinical course
- ≻ Complete additional investigations required to define ACHD AP classification
- ≻ Develop management strategies to minimize risk and optimize outcome

Factors associated with increased risk of perioperative morbidity and mortality (S3.11-10):

- Cyanosis
- Congestive HF
- Poor general health
- Younger age
- Pulmonary hypertension
- Operations on the respiratory and nervous systems
- Complex CHD
- Urgent/emergency procedures

Issues to consider:

- Endocarditis prophylaxis
- Complications related to underlying hemodynamics
- Abnormal venous and/or arterial anatomy affecting venous and arterial access
- Persistent shunts
- Valvular disease
- Arrhythmias, including bradyarrhythmias
- Erythrocytosis
- Pulmonary vascular disease
- Meticulous line care (also consider air filters for intravenous lines) to reduce risk of paradoxic embolus in patients who are cyanotic because of right-to-left shunts
- Adjustment of anticoagulant volume in tubes for some blood work in cyanotic patients
- Prevention of venous thrombosis
- Monitoring of renal and liver function
- Periprocedure anticoagulation
- Possible need for nonconventional drug dosing
- Increased prevalence of hepatitis C infection because of prior procedures and remote blood transfusions
- Developmental disability

ACHD indicates adult congenital heart disease; AP, anatomic and physiological; CHD, congenital heart disease; and HF, heart failure.

3.12. Pregnancy, Reproduction, and Sexual Health

3.12.1. Pregnancy

Recommendations for Pregnancy
Referenced studies that support recommendations are summarized in Online Data Supplement 19.

COR	LOE	RECOMMENDATIONS
I	C-LD	1. Women with CHD should receive prepregnancy counseling with input from an ACHD cardiologist to determine maternal cardiac, obstetrical and fetal risks, and potential long-term risks to the mother (S3.12.1-1–S3.12.1-4).
I	C-LD	2. An individualized plan of care that addresses expectations and contingencies should be developed for and with women with CHD who are pregnant or who may become pregnant and shared with the patient and all caregivers (S3.12.1-2, S3.12.1-3).
I	B-NR	3. Women with CHD receiving chronic anticoagulation should be counseled, ideally before conception, on the risks and benefits of specific anticoagulants during pregnancy (S3.12.1-5, S3.12.1-6).
I	B-NR	4. Women with ACHD AP classification IB-D, IIA-D, and IIIA-D* should be managed collaboratively during pregnancy by ACHD cardiologists, obstetricians, and anesthesiologists experienced in ACHD (S3.12.1-2, S3.12.1-7, S3.12.1-8).
I	C-EO	5. In collaboration with an ACHD cardiologist to ensure accurate assessment of pregnancy risk, patients at high risk of maternal morbidity or mortality, including women with pulmonary arterial hypertension (PAH), Eisenmenger syndrome, severe systemic ventricular dysfunction, severe left-sided obstructive lesions, and/or ACHD AP classification ID, IID, and IIID* should be counseled against becoming pregnant or be given the option of terminating pregnancy.
I	B-NR	6. Men and women of childbearing age with CHD should be counseled on the risk of CHD recurrence in offspring (S3.12.1-9).
IIa	B-NR	7. Exercise testing can be useful for risk assessment in women with ACHD AP classification IC-D, IIA-D, and IIIA-D* who are considering pregnancy (S3.12.1-10, S3.12.1-11).
IIa	B-NR	8. When either parent has CHD, it is reasonable to perform fetal echocardiography (S3.12.1-12, S3.12.1-13).

*See Tables 3 and 4 for the ACHD AP classification system.

3.12.2. Contraception

Recommendations for Contraception
Referenced studies that support recommendations are summarized in Online Data Supplement 20.

COR	LOE	RECOMMENDATIONS
I	C-LD	1. Women of childbearing potential with CHD should be counseled about the risks associated with pregnancy and appropriate contraceptive options (S3.12.2-1–S3.12.2-3).
III: Harm	B-NR	2. Estrogen-containing contraceptives are potentially harmful for women with CHD at high risk of thromboembolic events (e.g., cyanosis, Fontan physiology, mechanical valves, prior thrombotic events, PAH) (S3.12.2-4, S3.12.2-5).

3.13. Heart Failure and Transplant

3.13.1. Heart Failure

Recommendation for Heart Failure
Referenced studies that support the recommendation are summarized in Online Data Supplement 22.

COR	LOE	RECOMMENDATION
I	C-LD	1. Consultation with ACHD and HF specialists is recommended for patients with ACHD and HF or severe ventricular dysfunction (S3.13.1-1–S3.13.1-4).

3.13.2. Heart Transplant

Because of the prevalence of HF among patients with CHD, heart transplantation is increasingly being considered as a therapeutic option. Data on proper timing of transplantation are limited, particularly for individual lesions. Larger studies based on transplant databases do not allow for analysis based on the type of CHD (S3.13.2-1–S3.13.2-4). Currently, patients with ACHD may have fewer mechanical circulatory devices (e.g., ventricular-assist devices), which may lower their listing status and hence potential for organ receipt (S3.13.2-1, S3.13.2-2, S3.13.2-4–S3.13.2-7).

Although specific criteria for timing of referral for transplantation are desirable, universal recommendations cannot be made based on current data. Generally, published data show that immediate and early post-transplantation risk is higher in ACHD than in acquired heart disease because of increased perioperative mortality (S3.13.2-2). However, once beyond the perioperative period, patients with ACHD do as well as or better than those with acquired heart disease, with expected 10-year survival equivalent to or better than that of patients without ACHD (S3.13.2-2-4, S3.13.2-6, S3.13.2-7). Risks for poor outcomes include single ventricle anatomy, anatomic complexity, protein-losing enteropathy, or high titers of panel reactive antibodies (S3.13.2-8, S3.13.2-9). The current allocation system puts patients with ACHD at a disadvantage. Rather than priority dictated by the usual accepted risk markers, patients with ACHD are often listed by "exception," a process that requires the clinician to argue that the patient warrants higher priority than would be evident by applying the used risk markers. There is also significant mortality for patients with ACHD while on the waitlist (S3.13.2-10, S3.13.2-11). Surgical alternatives to transplantation exist for some patients with CHD (e.g., valve replacement, shunt closure), but these patients are at high risk of perioperative mortality (S3.13.2-12). Ideally, providers will consider early referral to a transplant center with expertise in ACHD transplantation when transplantation becomes a relevant clinical consideration. Additionally, it is advisable to consider options for transplantation or ventricular assist device as a backup before other high-risk surgery is pursued.

See Online Data Supplement 23 for referenced studies.

3.14. Palliative Care

Recommendation for Palliative Care
Referenced studies that support the recommendation are summarized in Online Data Supplement 24.

COR	LOE	RECOMMENDATION
IIa	B-NR	1. Discussion of end-of-life issues and advance directives can be beneficial for patients with ACHD or their surrogates (S3.14-1–S3.14-3).

3.15. Cyanosis

Table 12.

TABLE 12 Specific Management Practices for Cyanotic CHD

- Recording clinical oxygen saturation at rest (>5 min) rather than immediately after effort (e.g., walking into a clinic examination room).
- Meticulous intravenous care to avoid air or particulate matter, which may include use of air/particulate filters on all intravenous access lines, when feasible, and careful de-airing of all lines.
- Cerebral imaging for any new headache or neurologic sign to assess for possible cerebral abscess, hemorrhage, or stroke.
- Measurement of serum uric acid and treatment with allopurinol in a patient with a history of gout.
- Supplemental oxygen as needed for symptom relief but not to a target oxygen saturation level and not if there is no demonstrable symptomatic benefit.
- Avoidance of or cautious use of therapies that may reduce the patient's hypoxia-mediated drive to ventilation, such as narcotics or, in rare circumstances, excess supplemental oxygen (S3.15-1).
- Anesthesia by providers with expertise in anesthesia for patients with ACHD for any noncardiac surgery.
- Non-estrogen–containing birth control for women of child-bearing potential (intrauterine device may be a preferred option). Avoidance of birth control entirely is not a safe acceptable option.
- Patients can travel safely on commercial airlines without undue risk (S3.15-2). Preflight simulation testing or mandated supplemental oxygen are not usually indicated, although adequate hydration and movement during the flight are appropriate.
- Measurement of coagulation parameters (e.g., activated partial thromboplastin time, international normalized ratio, thrombin time) in a patient with an elevated hematocrit >55% requires adjustment of anticoagulant volume in the blood collection vials to account for reduced plasma volume in the draw (S3.15-3).

See Online Data Supplement 25 for referenced studies.

ACHD indicates adult congenital heart disease and CHD, congenital heart disease.

3.16. Pharmacological Therapy for ACHD

Patients with ACHD are commonly excluded from clinical trials, and there are few data to guide pharmacological therapies. Although it may be tempting to extrapolate from management guidelines developed for patients without CHD (e.g., HF guidelines) (S3.16-1), treatments may not have the same benefit in the heterogeneous population of patients with ACHD and in some cases may cause harm. The evaluation of new symptoms in a patient with ACHD must be tailored to the patient's anatomy, surgical repair, and physiology. Before considering pharmacological therapies, evaluation for residual shunts, baffle stenosis, valvular or conduit dysfunction, and collateral vessels, any of which may be amenable to interventions, is an important consideration.

The literature documenting pharmacological therapies for patients with ACHD is limited to small studies with limited duration of drug administration and follow-up. Additionally, the endpoints used are often surrogate markers that have not been validated for clinical decision-making, and studies are also often underpowered. However, studies in patients with ACHD do exist and evaluate conventional pharmacological therapy, especially for HF and for arrhythmia, including beta blockers, angiotensin-converting enzyme inhibitors, angiotensin-receptor blockers, and aldosterone antagonists, although results vary (S3.16-2–S3.16-9).

Pharmacological therapies in patients with ACHD are often directed to specific conditions (i.e., beta blockers for arrhythmia treatment). However, there are limited data examining the benefits of beta blockers in specific ACHD populations. Results from a small study indicate that beta-blocker therapy may have potential to improve functional class in patients with a systemic right ventricle and a pacemaker (S3.16-2). Angiotensin-converting enzyme inhibitors and angiotensin-receptor blockers have also been assessed in small studies in specific ACHD populations in which no significant benefit on ventricular function or exercise capacity has been proven (S3.16-6–S3.16-8). Data from 1 small trial with a short follow-up interval in patients with a systemic right ventricle suggest that eplerenone may be associated with reduced myocardial fibrosis, as assessed by imaging (S3.16-3).

Some pharmacological therapies affecting the pulmonary vasculature (e.g., endothelin-receptor antagonists and phosphodiesterase type-5 [PDE-5] inhibitors) have a beneficial effect on long-term outcomes in patients with Eisenmenger syndrome (S3.16-10). Similarly, there are limited data on the use of pulmonary vasodilator therapy in Fontan patients, in whom the pulmonary vascular resistance may be abnormal (S3.16-11–S3.16-13). Because of the lack of data, clinical recommendations regarding pharmacological therapy for patients with ACHD are unsupported. Individualized care is needed, recognizing the potential benefits and risks of the therapy relative to patient-specific anatomic and physiological issues.

See Online Data Supplement 23 for referenced studies.

4. SPECIFIC LESIONS

4.1. Shunt Lesions

4.1.1. Atrial Septal Defect

See also **Table 13 and Figure 1**.

Recommendations for Atrial Septal Defect
Referenced studies that support recommendations are summarized in Online Data Supplement 26 and the ERC systematic review report (S4.1.1-1).

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-EO	1. Pulse oximetry at rest and during exercise is recommended for evaluation of adults with unrepaired or repaired ASD with residual shunt to determine the direction and magnitude of the shunt.
I	B-NR	2. CMR, CCT, and/or TEE are useful to evaluate pulmonary venous connections in adults with ASD (S4.1.1-2–S4.1.1-4).
I	B-NR	3. Echocardiographic imaging is recommended to guide percutaneous ASD closure (S4.1.1-5, S4.1.1-6).

(continued)

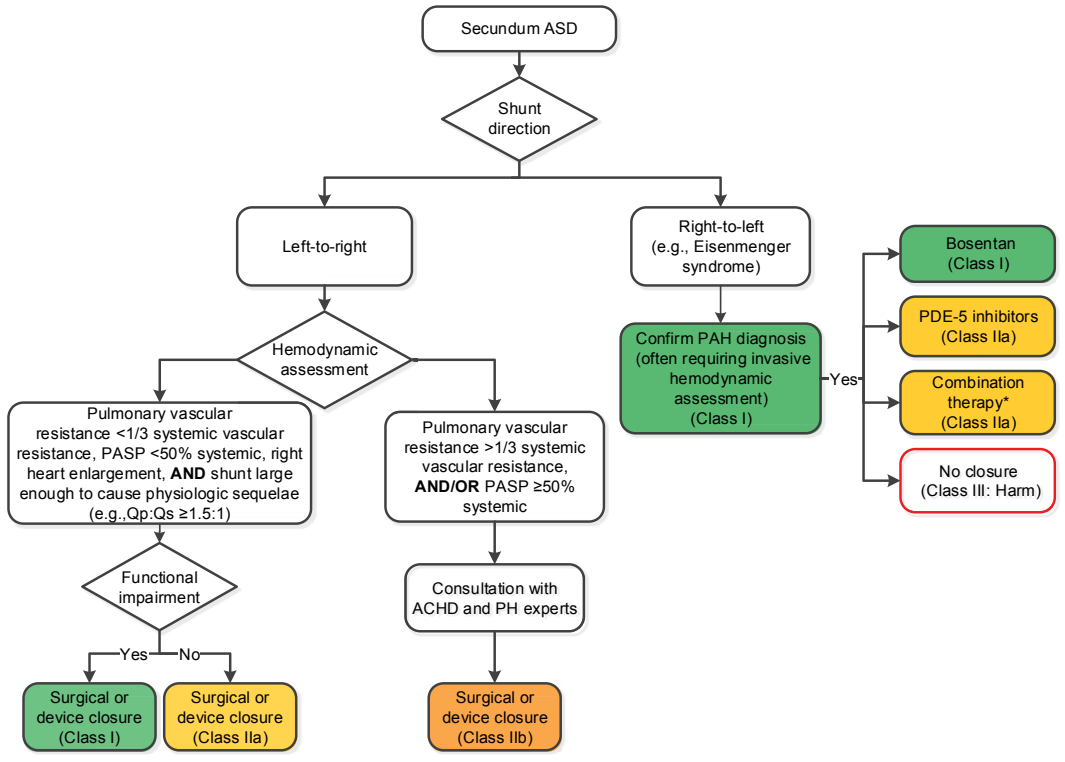
	Therapeutic
I / B-NR[SR]	4. In adults with isolated secundum ASD causing impaired functional capacity, right atrial and/or RV enlargement, and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., pulmonary-systemic blood flow ratio [Qp:Qs] ≥1.5:1) without cyanosis at rest or during exercise, transcatheter or surgical closure to reduce RV volume and improve exercise tolerance is recommended, provided that systolic PA pressure is less than 50% of systolic systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance (S4.1.1-7–S4.1.1-12).
I / B-NR	5. Adults with primum ASD, sinus venosus defect or coronary sinus defect causing impaired functional capacity, right atrial and/or RV enlargement and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs ≥1.5:1) without cyanosis at rest or during exercise, should be surgically repaired unless precluded by comorbidities, provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance (S4.1.1-13, S4.1.1-14).
IIa / C-LD[SR]	6. In asymptomatic adults with isolated secundum ASD, right atrial and RV enlargement, and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs 1.5:1 or greater), without cyanosis at rest or during exercise, transcatheter or surgical closure is reasonable to reduce RV volume and/or improve functional capacity, provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary vascular resistance is less than one third systemic resistance (S4.1.1-7–S4.1.1-10, S4.1.1-12).
IIa / C-LD	7. Surgical closure of a secundum ASD in adults is reasonable when a concomitant surgical procedure is being performed and there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs 1.5:1 or greater) and right atrial and RV enlargement without cyanosis at rest or during exercise (S4.1.1-15–S4.1.1-18).
IIb / B-NR	8. Percutaneous or surgical closure may be considered for adults with ASD when net left-to-right shunt (Qp:Qs) is 1.5:1 or greater, PA systolic pressure is 50% or more of systemic arterial systolic pressure, and/or pulmonary vascular resistance is greater than one third of the systemic resistance (S4.1.1-19, S4.1.1-20).
III: Harm / C-LD	9. ASD closure should not be performed in adults with PA systolic pressure greater than two thirds systemic, pulmonary vascular resistance greater than two thirds systemic, and/or a net right-to-left shunt (S4.1.1-21, S4.1.1-22).

TABLE 13 ASD: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36–60	24	6–12	3–6
ECG	36–60	24	12	12
TTE	36–60	24	12	12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Exercise test†	As needed	As needed	12–24	6–12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on the clinical indication.
ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

FIGURE 1 Secundum ASD



*Combination therapy with bosentan and PDE-5 inhibitor if symptomatic improvement does not occur with either alone. ACHD indicates adult congenital heart disease; ASD, atrial septal defect; PAH, pulmonary artery hypertension; PASP, pulmonary artery systolic pressure; PDE-5, phosphodiesterase type-5 inhibitors; PH, pulmonary hypertension; and Qp:Qs, pulmonary-systemic blood flow ratio.

4.1.2. Anomalous Pulmonary Venous Connections

Recommendations for Anomalous Pulmonary Venous Connections
Referenced studies that support recommendations are summarized in Online Data Supplement 27.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. CMR or CTA is recommended for evaluation of partial anomalous pulmonary venous connection (S4.1.2-1–S4.1.2-4).
IIa	B-NR	2. Cardiac catheterization can be useful in adults with partial anomalous pulmonary venous connection to further define hemodynamics (S4.1.2-5, S4.1.2-6).
		Therapeutic
I	B-NR	3. Surgical repair is recommended for patients with partial anomalous pulmonary venous connection when functional capacity is impaired and RV enlargement is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs ≥1.5:1), PA systolic pressure is less than 50% systemic pressure and pulmonary vascular resistance is less than one third of systemic resistance (S4.1.2-5).

(continued)

I	B-NR	4. Repair of partial anomalous pulmonary venous connection is recommended at the time of closure of a sinus venosus defect or ASD (S4.1.2-7).
I	B-NR	5. Repair of a scimitar vein is recommended in adults when functional capacity is impaired, evidence of RV volume overload is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs ≥1.5:1), PA systolic pressure is less than 50% systemic pressure and pulmonary vascular resistance is less than one third systemic (S4.1.2-5, S4.1.2-8, S4.1.2-9).
IIa	B-NR	6. Surgery can be useful for right- or left-sided partial anomalous pulmonary venous connection in asymptomatic adults with RV volume overload, net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs ≥1.5:1), pulmonary pressures less than 50% systemic and pulmonary vascular resistance less than one third systemic (S4.1.2-5).
IIa	B-NR	7. Surgery can be useful for repair of a scimitar vein in adults with evidence of RV volume overload, with Qp:Qs 1.5:1 or greater (S4.1.2-5, S4.1.2-9).

4.1.3. Ventricular Septal Defect

See also **Table 14 and Figure 2**.

Recommendations for Ventricular Septal Defect
Referenced studies that support recommendations are summarized in Online Data Supplement 28.

COR	LOE	RECOMMENDATIONS
		Therapeutic
I	B-NR	1. Adults with a VSD and evidence of left ventricular volume overload and hemodynamically significant shunts (Qp:Qs ≥1.5:1) should undergo VSD closure, if PA systolic pressure is less than 50% systemic and pulmonary vascular resistance is less than one third systemic (S4.1.3-1).
IIa	C-LD	2. Surgical closure of perimembranous or supracristal VSD is reasonable in adults when there is worsening aortic regurgitation (AR) caused by VSD (S4.1.3-1, S4.1.3-2).
IIb	C-LD	3. Surgical closure of a VSD may be reasonable in adults with a history of IE caused by VSD if not otherwise contraindicated (S4.1.3-3).
IIb	C-LD	4. Closure of a VSD may be considered in the presence of a net left-to-right shunt (Qp:Qs ≥1.5:1) when PA systolic pressure is 50% or more than systemic and/or pulmonary vascular resistance is greater than one third systemic (S4.1.3-4–S4.1.3-6).
III: Harm	C-LD	5. VSD closure should not be performed in adults with severe PAH with PA systolic pressure greater than two thirds systemic, pulmonary vascular resistance greater than two thirds systemic and/or a net right-to-left shunt (S4.1.3-7–S4.1.3-9).

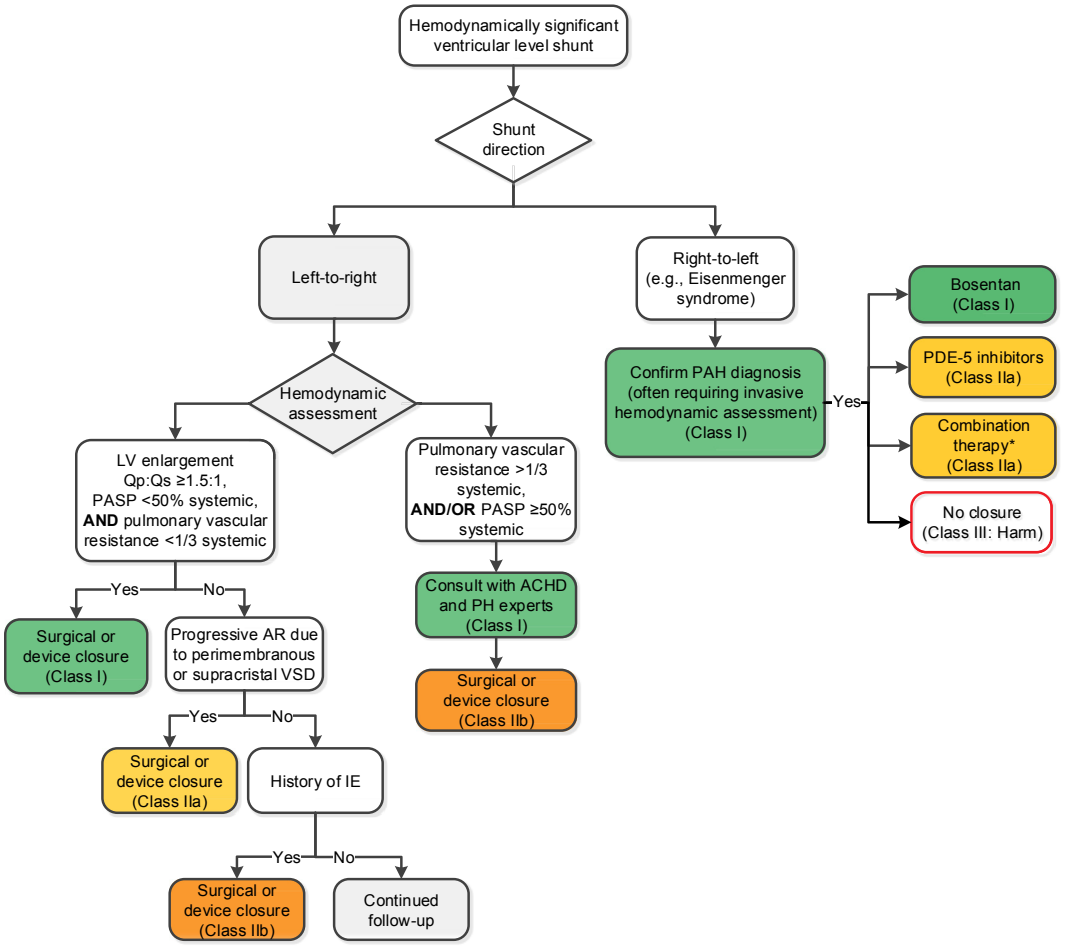
TABLE 14 VSD: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36	24	6–12	3–6
ECG	36	24	12	12
TTE	36	24	12	12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Exercise test†	As needed	As needed	12–24	6–12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on the clinical circumstance.

ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; TTE, transthoracic echocardiogram; and VSD, ventricular septal defect.

FIGURE 2 Hemodynamically Significant Ventricular Level Shunt



*Combination therapy with bosentan and PDE-5 inhibitor, if symptomatic improvement does not occur with either alone. ACHD indicates adult congenital heart disease; AR, aortic regurgitation; IE, infective endocarditis; LV, left ventricular; PAH, pulmonary artery hypertension; PASP, pulmonary artery systolic pressure; PDE-5, phosphodiesterase type-5 inhibitors; PH, pulmonary hypertension; Qp:Qs, pulmonary-systemic blood flow ratio; and VSD, ventricular septal defect.

4.1.4. Atrioventricular Septal Defect

See also **Table 15**.

Recommendations for Atrioventricular Septal Defect
Referenced studies that support recommendations are summarized in Online Data Supplement 29.

COR	LOE	RECOMMENDATIONS
		Diagnostic
IIa	C-EO	1. Cardiac catheterization can be useful in adults with atrioventricular septal defect when pulmonary hypertension is suspected.
		Therapeutic
I	C-LD	2. Surgery for severe left atrioventricular valve regurgitation is recommended per GDMT indications for mitral regurgitation (S4.1.4-1–S4.1.4-4).
I	C-EO	3. Surgery for primary repair of atrioventricular septal defect or closure of residual shunts in adults with repaired atrioventricular septal defect is recommended when there is a net left-to-right shunt (Qp:Qs ≥1.5:1), PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic.
IIa	C-EO	4. Operation for discrete LVOT obstruction in adults with atrioventricular septal defect is reasonable with a maximum gradient of 50 mm Hg or greater, a lesser gradient if HF symptoms are present, or if concomitant moderate-to-severe mitral or AR are present.
IIb	C-EO	5. Surgery for primary repair of atrioventricular septal defect or closure of residual shunts in adults with repaired atrioventricular septal defect may be considered in the presence of a net left-to-right shunt (Qp:Qs ≥1.5:1), if PA systolic pressure is 50% or more systemic, and/or pulmonary vascular resistance is greater than one third systemic.
III: Harm	C-LD	6. Surgery for primary repair of atrioventricular septal defect or closure of residual shunts in adults with repaired atrioventricular septal defect should not be performed with PA systolic pressure greater than two thirds systemic, pulmonary vascular resistance greater than two thirds systemic, or a net right-to-left shunt (S4.1.4-5, S4.1.4-6).

TABLE 15 AVSD: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24–36	24	6–12	3–6
ECG	24–36	24	12	12
TTE	24–36	24	12	12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Exercise test†	As needed	As needed	12–24	6–12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on the clinical indication.

ACHD indicates adult congenital heart disease; AVSD, atrioventricular septal defect; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.1.5. Patent Ductus Arteriosus

See also **Table 16**.

Recommendations for Patent Ductus Arteriosus
Referenced studies that support recommendations are summarized in Online Data Supplement 30.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-EO	1. Measurement of oxygen saturation should be performed in feet and both hands in adults with a PDA to assess for the presence of right-to-left shunting.

(continued)

IIa	C-EO	2. In addition to the standard diagnostic tools, cardiac catheterization can be useful in patients with PDA and suspected pulmonary hypertension (Section 3.5).
		Therapeutic
I	C-LD	3. PDA closure in adults is recommended if left atrial or LV enlargement is present and attributable to PDA with net left-to-right shunt, PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic (S4.1.5-1–S4.1.5-3).
IIb	B-NR	4. PDA closure in adults may be considered in the presence of a net left-to-right shunt if PA systolic pressure is 50% or greater systemic, and/or pulmonary vascular resistance is greater than one third systemic (S4.1.5-3, S4.1.5-4).
III: Harm	C-LD	5. PDA closure should not be performed in adults with a net right-to-left shunt and PA systolic pressure greater than two thirds systemic or pulmonary vascular resistance greater than two thirds systemic (S4.1.5-5).

TABLE 16 PDA: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36–60	24	6–12	3–6
ECG	36–60	24	12	12
TTE	36–60	24	12	12
Pulse oximetry†	As needed	As needed	Each visit	Each visit
Exercise test‡	As needed	As needed	12–24	6–12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†Upper and lower extremity.
‡6-minute walk test or CPET, depending on the clinical indication.

ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; PDA, patent ductus arteriosus; and TTE, transthoracic echocardiogram.

4.2. Left-Sided Obstructive Lesions

4.2.1. Cor Triatriatum

Recommendations for Cor Triatriatum
Referenced studies that support recommendations are summarized in Online Data Supplement 31.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. Adults presenting with cor triatriatum sinister should be evaluated for other congenital abnormalities, particularly ASD, VSD, and anomalous pulmonary venous connection (S4.2.1-1).
IIa	B-NR	2. In adults with prior repair of cor triatriatum sinister and recurrent symptoms, it is reasonable to evaluate for pulmonary vein stenosis (S4.2.1-2).
		Therapeutic
I	B-NR	3. Surgical repair is indicated for adults with cor triatriatum sinister for symptoms attributable to the obstruction or a substantial gradient across the membrane (S4.2.1-3)

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation.

4.2.2. Congenital Mitral Stenosis

See also **Table 17**.

Recommendation for Congenital Mitral Stenosis
Referenced studies that support the recommendation are summarized in Online Data Supplement 32.

COR	LOE	RECOMMENDATION
I	B-NR	1. Adults with congenital mitral stenosis or a parachute mitral valve should be evaluated for other left-sided obstructive lesions (S4.2.2-1, S4.2.2-2).

TABLE 17 Congenital Mitral Stenosis: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE	24	24	12	12
Exercise test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on the clinical indication.
ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.2.3. Subaortic Stenosis

See also **Table 18**.

Recommendations for Subaortic Stenosis
Referenced studies that support recommendations are summarized in Online Data Supplement 33.

COR	LOE	RECOMMENDATIONS
		Diagnostic
IIb	C-LD	1. Stress testing for adults with LVOT obstruction to determine exercise capacity, symptoms, electrocardiographic changes, or arrhythmias may be reasonable in the presence of otherwise equivocal indications for intervention (S4.2.3-1, S4.2.3-2).
		Therapeutic
I	C-EO	2. Surgical intervention is recommended for adults with subAS, a maximum gradient 50 mm Hg or more and symptoms attributable to the subAS.
I	C-LD	3. Surgical intervention is recommended for adults with subAS and less than 50 mm Hg maximum gradient and HF or ischemic symptoms, and/or LV systolic dysfunction attributable to subAS (S4.2.3-3).
IIb	C-LD	4. To prevent the progression of AR, surgical intervention may be considered for asymptomatic adults with subAS and at least mild AR and a maximum gradient of 50 mm Hg or more (S4.2.3-4–S4.2.3-6).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 18** for routine testing and follow-up intervals.

TABLE 18 Subaortic Stenosis: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE	24	24	12	12
Exercise test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on the clinical indication.
ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.2.4. Congenital Valvular Aortic Stenosis

See also Table 19.

Recommendations for Congenital Valvular Aortic Stenosis
Referenced studies that support recommendations are summarized in Online Data Supplement 34.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. Adults with bicuspid aortic valve should be evaluated for coarctation of the aorta by clinical examination and imaging studies (S4.2.4-1).
IIa	B-NR	2. It is reasonable to screen first-degree relatives of patients with bicuspid aortic valve or unicuspid aortic valve with echocardiography for valve disease and aortopathy (S4.2.4-2–S4.2.4-4).
		Therapeutic
IIb	B-NR	3. In adults with bicuspid aortic valve stenosis and a noncalcified valve with no more than mild AR meeting indications for intervention per GDMT (S4.2.4-5), it may be reasonable to treat with balloon valvuloplasty (S4.2.4-6).

See Section 3.4 for recommendations on diagnostic evaluation; and Table 19 for routine testing and follow-up intervals.

TABLE 19 Congenital Aortic Stenosis: Routine Follow-Up and Testing Intervals*

Stage	Frequency of Echocardiogram
Progressive (Stage B)	Every 3–5 y (mild severity, Vmax 2.0–2.9 m/s) Every 1–2 y (moderate severity, Vmax 3.0–3.9 m/s)
Severe (Stage C)	Every 6–12 mo (Vmax ≥4.0 m/s)
Aortic dilation >4.5 cm	Every 12 mo (echocardiogram, MRI or CT)

*Modified from existing GDMT for valvular heart disease (S4.2.4-5).
CT indicates computed tomography; GDMT, guideline-directed management and therapy; MRI, magnetic resonance imaging; and Vmax, maximum velocity.

4.2.4.1. Turner Syndrome

Recommendations for Turner Syndrome
Referenced studies that support recommendations are summarized in Online Data Supplement 35.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. Women with Turner syndrome should be evaluated for bicuspid aortic valve, coarctation of the aorta, and enlargement of the ascending aorta (S4.2.4.1-1).

(continued)

COR	LOE	RECOMMENDATIONS
		Therapeutic
IIa	B-NR	2. Prophylactic replacement of the aortic root or ascending aorta in adults with Turner syndrome is reasonable when the aortic diameter is 2.5 cm/m^2 or greater (S4.2.4.1-2).

4.2.5. Supravalvular Aortic Stenosis

See also **Table 20**.

Recommendations for Supravalvular Aortic Stenosis
Referenced studies that support recommendations are summarized in Online Data Supplement 37.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-LD	1. Aortic imaging using TTE, TEE, CMR, or CTA is recommended in adults with Williams syndrome or patients suspected of having supravalvular aortic stenosis (S4.2.5-1).
I	C-LD	2. Coronary imaging is recommended in patients with Williams syndrome and supravalvular aortic stenosis presenting with symptoms of coronary ischemia (S4.2.5-2–S4.2.5-4).
		Therapeutic
I	B-NR	3. Surgical repair is recommended for adults with supravalvular aortic stenosis (discrete or diffuse) and symptoms or decreased LV systolic function deemed secondary to aortic obstruction (S4.2.5-5–S4.2.5-8).
I	C-LD	4. Coronary artery revascularization is recommended in symptomatic adults with supravalvular aortic stenosis and coronary ostial stenosis (S4.2.5-4, S4.2.5-9).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 20** for routine testing and follow-up intervals.

TABLE 20 Supravalvular Aortic Stenosis: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE†	24	24	12	12
CMR‡/CCT§	36–60	36–60	36–60	36–60
Exercise test‖	As needed	24	24	12

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of aortic anatomy. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§If CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on the clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.2.6. Coarctation of the Aorta

See also **Table 21**.

Recommendations for Coarctation of the Aorta
Referenced studies that support recommendations are summarized in Online Data Supplement 38.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. Initial and follow-up aortic imaging using CMR or CTA is recommended in adults with coarctation of the aorta, including those who have had surgical or catheter intervention (S4.2.6-1-S4.2.6-3).
I	C-EO	2. Resting blood pressure should be measured in upper and lower extremities in all adults with coarctation of the aorta.
IIa	C-LD	3. Ambulatory blood pressure monitoring in adults with coarctation of the aorta can be useful for diagnosis and management of hypertension (S4.2.6-4).
IIb	B-NR	4. Screening for intracranial aneurysms by magnetic resonance angiography or CTA may be reasonable in adults with coarctation of the aorta (S4.2.6-5, S4.2.6-6).
IIb	C-LD	5. Exercise testing to evaluate for exercise-induced hypertension may be reasonable in adults with coarctation of the aorta who exercise (S4.2.6-4, S4.2.6-7).
		Therapeutic
I	B-NR	6. Surgical repair or catheter-based stenting is recommended for adults with hypertension and significant native or recurrent coarctation of the aorta (S4.2.6-1, S4.2.6-2, S4.2.6-8-S4.2.6-12).
I	C-EO	7. GDMT is recommended for treatment of hypertension in patients with coarctation of the aorta (S4.2.6-13).
IIb	B-NR	8. Balloon angioplasty for adults with native and recurrent coarctation of the aorta may be considered if stent placement is not feasible and surgical intervention is not an option (S4.2.6-14).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 21** for routine testing and follow-up intervals.

TABLE 21 CoA: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A*(mo)	Physiological Stage B*(mo)	Physiological Stage C*(mo)	Physiological Stage D*(mo)
Outpatient ACHD cardiologist	24	24	6-12	3-6
ECG	24	24	12	12
TTE†	24	24	12	12
CMR‡/CCT§	36-60	36-60	12-24	12-24
Exercise test‖	36	24	24	12

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of aortic size and aortic arch/coarctation repair site anatomy. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible and to evaluate cross-sectional imaging status-post-stent therapy for coarctation of the aorta; the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on the clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; CoA, coarctation of the aorta; CPET, cardiopulmonary exercise; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.3. Right-Sided Lesions

4.3.1. Valvular Pulmonary Stenosis

See also Tables 22 and 23.

Recommendations for Valvular Pulmonary Stenosis
Referenced studies that support recommendations are summarized in Online Data Supplement 39.

COR	LOE	RECOMMENDATIONS
I	B-NR	1. In adults with moderate or severe valvular pulmonary stenosis and otherwise unexplained symptoms of HF, cyanosis from interatrial right-to-left communication, and/or exercise intolerance, balloon valvuloplasty is recommended (S4.3.1-1–S4.3.1-4).
I	B-NR	2. In adults with moderate or severe valvular pulmonary stenosis and otherwise unexplained symptoms of HF, cyanosis, and/or exercise intolerance who are ineligible for or who failed balloon valvuloplasty, surgical repair is recommended (S4.3.1-1, S4.3.1-5–S4.3.1-8)
IIa	C-EO	3. In asymptomatic adults with severe valvular pulmonary stenosis, intervention is reasonable.

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; Table 23 for routine testing and follow-up intervals; and Figure 3 for a diagnostic and treatment algorithm for isolated PR after repair of PS.

TABLE 22 Severity of RVOT Obstruction

Mild	Peak gradient <36 mm Hg (peak velocity <3 m/s)
Moderate	Peak gradient 36–64 mm Hg (peak velocity 3–4 m/s)
Severe	Peak gradient 64 mm Hg (peak velocity >4 m/s); mean gradient >35 mm Hg

Estimations of RV systolic pressure by TR velocity is part of the echocardiographic assessment of RV obstruction, as Doppler measurements across the RV obstruction itself may be unreliable.

RV indicates right ventricular; RVOT, right ventricular outflow tract; and TR, tricuspid regurgitation.

TABLE 23 Valvular PS: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36–60	24	6–12	3–6
ECG	36–60	24	12	12
TTE	36–60	24	12	12
Exercise test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on clinical indication.

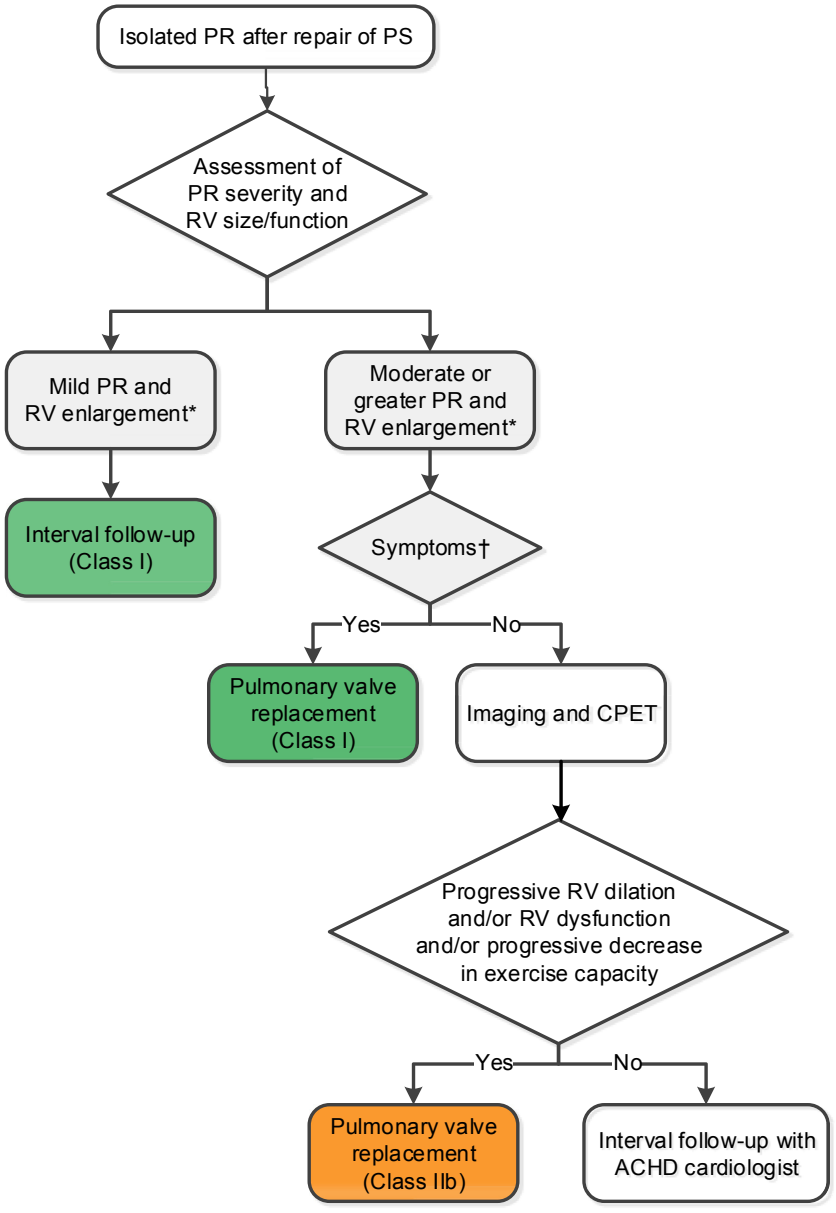
ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; PS, pulmonary stenosis; and TTE, transthoracic echocardiogram.

4.3.1.1. Isolated PR After Repair of PS

Recommendations for Isolated PR After Repair of Pulmonary Stenosis

COR	LOE	RECOMMENDATIONS
I	C-EO	1. In symptomatic patients with moderate or greater PR resulting from treated isolated pulmonary stenosis, with RV dilation or RV dysfunction, pulmonary valve replacement is recommended.
I	C-EO	2. For asymptomatic patients with residual PR resulting from treatment of isolated pulmonary stenosis with a dilated right ventricle, serial follow-up is recommended.
IIb	C-EO	3. In asymptomatic patients with moderate or greater PR resulting from treatment of isolated pulmonary stenosis with progressive RV dilation and/or RV dysfunction, pulmonary valve replacement may be reasonable.

FIGURE 3 **Isolated PR After Repair of PS**



*Significant PR causes RV dilation. If a patient has moderate or greater PR and normal RV size, most likely the estimation of PR severity is inaccurate or there may be restrictive RV physiology, which would warrant further investigation. †Symptoms may include dyspnea, chest pain, and/or exercise intolerance referable to PR or otherwise unexplained. ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; PR, pulmonary regurgitation; PS, pulmonary stenosis; and RV, right ventricular.

4.3.2. Branch and Peripheral Pulmonary Stenosis

See also **Table 24**.

Recommendations for Branch and Peripheral PS
Referenced studies that support recommendations are summarized in Online Data Supplement 40.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. For adults with peripheral or branch PS, ongoing surveillance is recommended (S4.3.2-1, S4.3.2-2).
		Therapeutic
IIa	B-NR	2. In adults with peripheral or branch PA stenosis, PA dilation and stenting can be useful (S4.3.2-2, S4.3.2-3).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 24** for routine testing and follow-up intervals.

TABLE 24 Branch and Peripheral PS: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24–36	24	6–12	3–6
ECG	24–36	24	12	12
TTE†	24–36	24	12	12
CMR‡/CCT§	36–60	36–60	24–36	24–36
Exercise test‖	36	24	24	12

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of branch PS. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible and to evaluate cross-sectional imaging status and post-stent therapy for peripheral PS; the frequency should be weighed against radiation exposure.
‖6-minute walk test or cardiopulmonary exercise test, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; ECG, electrocardiogram; PS, pulmonary stenosis; and TTE, transthoracic echocardiogram.

4.3.3. Double-Chambered Right Ventricle

See also **Table 25**.

Recommendations for Double-Chambered Right Ventricle
Referenced studies that support recommendations are summarized in Online Data Supplement 41.

COR	LOE	RECOMMENDATIONS
I	C-LD	1. Surgical repair for adults with double-chambered right ventricle and moderate or greater outflow obstruction is recommended in patients with otherwise unexplained symptoms of HF, cyanosis, or exercise limitation (S4.3.3-1–S4.3.3-3) (Table 22).
IIb	C-LD	2. Surgical repair for adults with double-chambered right ventricle with a severe gradient may be considered in asymptomatic patients (S4.3.3-3, S4.3.3-4) (Table 22).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 25** for routine testing and follow-up intervals.

TABLE 25 Double-Chambered Right Ventricle: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24–36	24	6–12	3–6
ECG	24–36	24	12	12
TTE	24–36	24	12	12
Exercise test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†6-minute walk test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.3.4. Ebstein Anomaly

See also Table 26.

Recommendations for Ebstein Anomaly
Referenced studies that support recommendations are summarized in Online Data Supplement 42.

COR	LOE	RECOMMENDATIONS
		Diagnostic
IIa	B-NR	1. In adults with Ebstein anomaly, CMR can be useful to determine anatomy, RV dimensions, and systolic function (S4.3.4-1, S4.3.4-2).
IIa	B-NR	2. In adults with Ebstein anomaly, TEE can be useful for surgical planning if TTE images are inadequate to evaluate tricuspid valve morphology and function (S4.3.4-1).
IIa	B-NR	3. Electrophysiological study with or without catheter ablation can be useful in the diagnostic evaluation of adults with Ebstein anomaly and ventricular preexcitation but without supraventricular tachycardia (S4.3.4-3, S4.3.4-4).
IIa	B-NR	4. In adults with Ebstein anomaly, electrophysiological study (and catheter ablation, if needed) is reasonable before surgical intervention on the tricuspid valve even in the absence of preexcitation or supraventricular tachycardia (S4.3.4-5).
		Therapeutic
I	B-NR	5. Surgical repair or reoperation for adults with Ebstein anomaly and significant TR is recommended when one or more of the following are present: HF symptoms, objective evidence of worsening exercise capacity, progressive RV systolic dysfunction by echocardiography or CMR (S4.3.4-6–S4.3.4-10).
I	C-LD	6. Catheter ablation is recommended for adults with Ebstein anomaly and high-risk pathway conduction or multiple accessory pathways (S4.3.4-3, S4.3.4-11, S4.3.4-12).
IIa	B-NR	7. Surgical repair or reoperation for adults with Ebstein anomaly and significant TR can be beneficial in the presence of progressive RV enlargement, systemic desaturation from right-to-left atrial shunt, paradoxical embolism, and/or atrial tachyarrhythmias (S4.3.4-11, S4.3.4-13, S4.3.4-14).
IIb	B-NR	8. Bidirectional superior cavopulmonary (Glenn) anastomosis at time of Ebstein anomaly repair may be considered for adults when severe RV dilation or severe RV systolic dysfunction is present, LV function is preserved, and left atrial pressure and LV end diastolic pressure are not elevated (S4.3.4-6, S4.3.4-15).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and Table 26 for routine testing and follow-up intervals.

TABLE 26 Ebstein Anomaly: Routine and Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12–24	12	6–12	3–6
ECG	12–24	12	12	12
CXR	As needed	As needed	12–24	12–24
TTE†	12–24	12–24	12	12
Pulse oximetry	24	12	Each visit	Each visit
Holter monitor	As needed	As needed	24	12–24
CMR‡/CCT§	60	36	24–36	12–24
Exercise test‖	36	24–36	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of right ventricular size and function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible; the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; CPET, cardiopulmonary exercise test; CXR, chest x ray; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.3.5. Tetralogy of Fallot

See also Table 27.

Recommendations for TOF
Referenced studies that support recommendations are summarized in Online Data Supplement 43. (See Section 4.3.6. for recommendations regarding evaluation and management of right ventricle-to-PA conduits.)

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. CMR is useful to quantify ventricular size and function, pulmonary valve function, pulmonary artery anatomy and left heart abnormalities in patients with repaired TOF (S4.3.5-1).
I	B-NR	2. Coronary artery compression testing is indicated before right ventricle–to-PA conduit stenting or transcatheter valve placement in repaired TOF (S4.3.5-2).
IIa	B-NR	3. Programmed ventricular stimulation can be useful to risk stratify adults with TOF and additional risk factors for SCD (S4.3.5-3–S4.3.5-8).
IIa	C-EO	4. In patients with repaired TOF, cardiac catheterization with angiography, if indicated, is reasonable to assess hemodynamics when adequate data cannot be obtained noninvasively in the setting of an arrhythmia, HF, unexplained ventricular dysfunction, suspected pulmonary hypertension or cyanosis.
		Therapeutic
I	B-NR	5. Pulmonary valve replacement (surgical or percutaneous) for relief of symptoms is recommended for patients with repaired TOF and moderate or greater PR with cardiovascular symptoms not otherwise explained (S4.3.5-9–S4.3.5-11).
IIa	B-NR	6. Pulmonary valve replacement (surgical or percutaneous) is reasonable for preservation of ventricular size and function in asymptomatic patients with repaired TOF and ventricular enlargement or dysfunction and moderate or greater PR (S4.3.5-1, S4.3.5-9, S4.3.5-12–S4.3.5-14).
IIa	B-NR	7. Primary prevention ICD therapy is reasonable in adults with TOF and multiple risk factors for SCD (S4.3.5-15–S4.3.5-17).

(continued)

IIb	C-EO

8. Surgical pulmonary valve replacement may be reasonable for adults with repaired TOF and moderate or greater PR with other lesions requiring surgical interventions.

IIb	C-EO

9. Pulmonary valve replacement, in addition to arrhythmia management, may be considered for adults with repaired TOF and moderate or greater PR and ventricular tachyarrhythmia.

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; **Figure 4** for a diagnostic and treatment algorithm for repaired TOF with residual PR; and **Table 27** for routine testing and follow-up intervals.

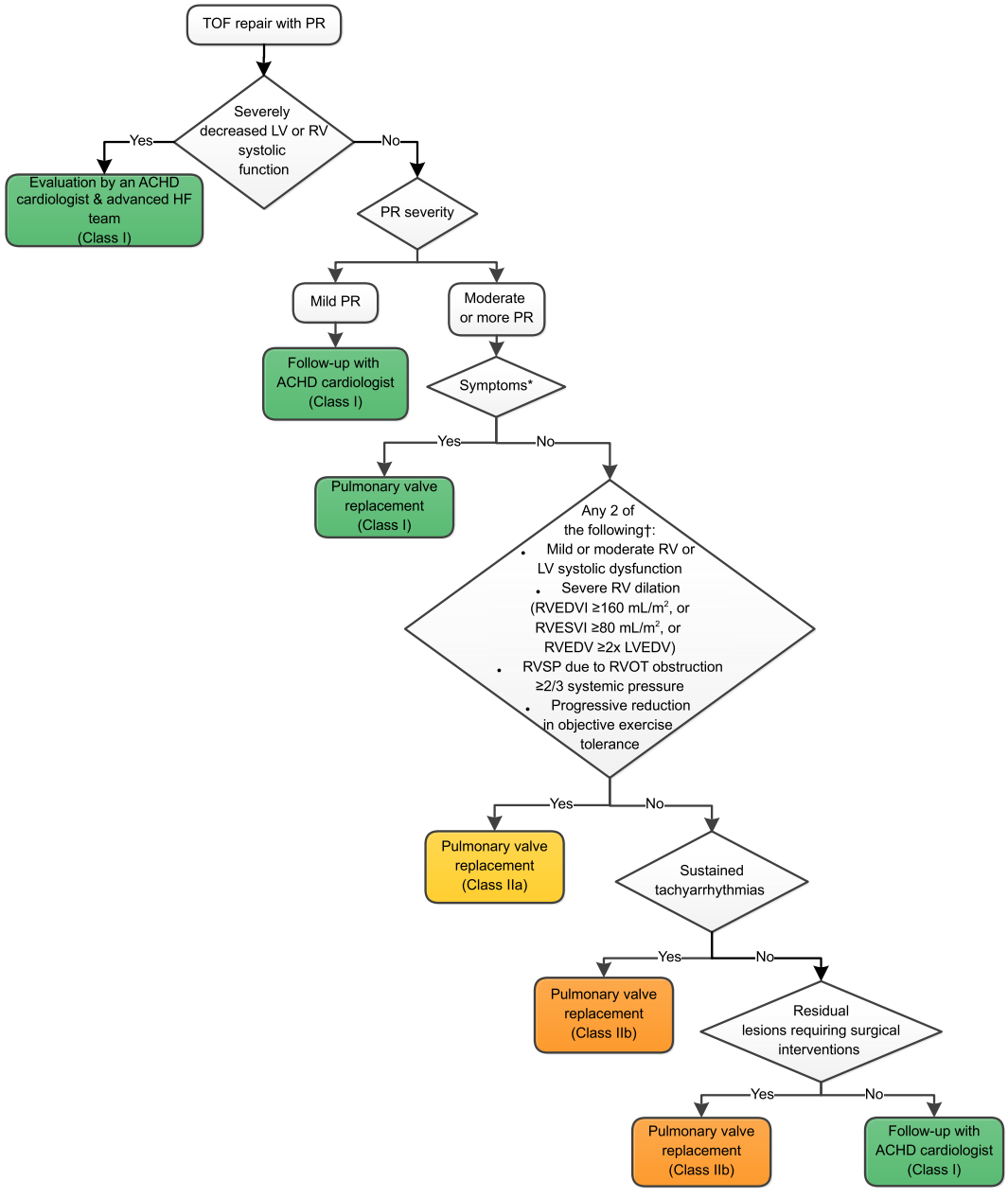
TABLE 27 TOF: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12–24	12	6–12	3–6
ECG	24	12	12	12
TTE†	24	12–24	12	6–12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Holter monitor	As needed	As needed	12–24	12–24
CMR‡/CCT§	36	24–36	12–24	12–24
Exercise test‖	36–60	24–60	12–24	12–24

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of right ventricular size and function, pulmonary valve function, pulmonary artery anatomy and left heart abnormalities. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible and to evaluate origin and course of the coronary arteries, and cross-sectional imaging status—post-stent therapy. If cardiac CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; TOF, tetralogy of Fallot; and TTE, transthoracic echocardiogram.

FIGURE 4 Pulmonary Valve Replacement in Patients With TOF Repair and PR



*Symptoms may included dyspnea, chest pain, and/or exercise intolerance referable to PR or otherwise unexplained. ACHD indicates adult congenital heart disease; HF, heart failure; LV, left ventricular; LVEDV, left ventricular end diastolic volume; PR, pulmonary regurgitation; RV, right ventricular; RVEDV, right ventricular end diastolic volume; RVEDVI, right ventricular end diastolic volume index; RVESVI, right ventricular end systolic volume index; RVOT, right ventricular outflow tract; RVSP, right ventricular systolic pressure; and TOF, tetralogy of Fallot. †See recommendation 6 supporting text in the full text of the "2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease" (P-9).

4.3.6. Right Ventricle-to-Pulmonary Artery Conduit

See also **Table 28**.

Recommendations for Right Ventricle-to-PA Conduit
Referenced studies that support recommendations are summarized in Online Data Supplement 44.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. Coronary artery compression testing with simultaneous coronary angiography and high-pressure balloon dilation in the conduit is indicated before right ventricle–to-PA conduit stenting or transcatheter valve placement (S4.3.6-1, S4.3.6-2).
I	B-NR	2. In patients with stented right ventricle–to-PA conduits and worsening PS or PR, evaluation for conduit complications should be performed, including fluoroscopy to evaluate for stent fracture and blood cultures to assess for IE (S4.3.6-3, S4.3.6-4).
IIa	C-LD	3. In adults with right ventricle–to-PA conduit and arrhythmia, congestive HF, unexplained ventricular dysfunction or cyanosis cardiac catheterization is reasonable to assess the hemodynamics (S4.3.6-5, S4.3.6-6).
		Therapeutic
IIa	B-NR	4. Right ventricle–to-PA conduit intervention is reasonable for adults with right ventricle–to-PA conduit and moderate or greater PR or moderate or greater stenosis (Table 22) with reduced functional capacity or arrhythmia (S4.3.6-7–S4.3.6-11).
IIb	B-NR	5. Right ventricle–to-PA conduit intervention may be reasonable for asymptomatic adults with right ventricle–to-PA conduit and severe stenosis or severe regurgitation with reduced RV ejection fraction or RV dilation (S4.3.6-12–S4.3.6-14).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 28** for routine testing and follow-up intervals.

TABLE 28 Right Ventricle-to-PA Conduit: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12–24	12	6–12	3–6
ECG	12–24	12	12	12
TTE†	12–24	12	12	12
CMR‡/CCT§	36–60	36–60	12–24	12–24
Exercise test‖	As needed	As needed	12–24	12–24

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of right ventricular size and function and valvular function, conduit anatomy and pulmonary artery anatomy. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible and to evaluate cross-sectional imaging status–post-stent therapy. If CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; PA, pulmonary artery; and TTE, transthoracic echocardiogram.

4.4. Complex Lesions

4.4.1. Transposition of the Great Arteries

4.4.1.1. Transposition of the Great Arteries With Atrial Switch

See also **Table 29**.

Recommendations for d-TGA With Atrial Switch
Referenced studies that support recommendations are summarized in Online Data Supplement 45.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-EO	1. Ambulatory monitoring for bradycardia or sinus node dysfunction is recommended for adults with d-TGA with atrial switch, especially if treated with beta blockers or other rate-slowing agents.
I	C-EO	2. Adults with d-TGA with atrial switch repair should undergo annual imaging with either echocardiography or CMR to evaluate for common long-term complications of the atrial switch.
IIa	C-LD	3. Assessment for a communication through the interatrial baffle or venous stenosis is reasonable for adults with d-TGA with atrial switch, particularly if transvenous pacemaker/ICD implantation is considered or leads are already present (S4.4.1.1-1).
		Therapeutic
I	B-NR	4. GDMT with appropriate attention to the need for anticoagulation is recommended to promptly restore sinus rhythm for adults with d-TGA with atrial switch repair presenting with atrial arrhythmia (S4.4.1.1-2).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; **Table 29** for routine testing and follow-up intervals; and Online Data Supplement 25 for referenced studies.

TABLE 29 d-TGA With Atrial Switch: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12	12	6–12	3–6
ECG	12	12	6–12	6–12
TTE†	12–24	12–24	12	12
Pulse oximetry	12	12	Each visit	Each visit
Holter monitor	24	24	12	12
CMR‡/CCT§	24–36	24	12–24	12–24
Exercise test‖	36	36	24	12

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of ventricular size and function, systemic and venous baffle obstruction and leaks, and valvular function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined anatomic and physiological findings.
§CCT may be used if CMR is not feasible and to evaluate cross-sectional imaging status-post-stent therapy. If CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute exercise test or cardiopulmonary exercise test, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; d-TGA, dextro-transposition of the great arteries; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.4.1.2. Transposition of the Great Arteries With Arterial Switch

See also **Table 30**.

Recommendations for d-TGA With Arterial Switch
Referenced studies that support recommendations are summarized in Online Data Supplement 46.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-LD	1. Baseline and serial imaging with either echocardiography or CMR should be performed in adults with d-TGA with arterial switch who have neoaortic dilation, valve dysfunction or PA or branch PA stenosis or ventricular dysfunction (S4.4.1.2-1–S4.4.1.2-3).
I	C-EO	2. Coronary revascularization for adults with d-TGA with arterial switch should be planned by surgeons or interventional cardiologists with expertise in revascularization in collaboration with ACHD providers to ensure coronary and pulmonary artery anatomy are understood.
IIa	B-NR	3. It is reasonable to perform anatomic evaluation of coronary artery patency (catheter angiography, or CT or MR angiography) in asymptomatic adults with d-TGA with arterial switch (S4.4.1.2-4, S4.4.1.2-5).
IIa	C-EO	4. Physiological tests of myocardial perfusion for adults with d-TGA after arterial switch can be beneficial for assessing symptoms suggestive of myocardial ischemia.
IIa	C-EO	5. GDMT is reasonable to determine the need for coronary revascularization for adults with d-TGA after arterial switch (S4.4.1.2-6–S4.4.1.2-8).
		Therapeutic
IIa	C-EO	6. GDMT is reasonable to determine indications for aortic valve replacement in adults with d-TGA after arterial switch with severe neoaortic valve regurgitation (S4.4.1.2-6).
IIa	C-EO	7. Catheter or surgical intervention for PS is reasonable in adults with d-TGA after arterial switch with symptoms of HF or decreased exercise capacity attributable to PS.

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 30** for routine testing and follow-up intervals.

TABLE 30 d-TGA With Arterial Switch: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12–24	12	6–12	3–6
ECG	12–24	12–24	12	6
TTE†	12–24	12–24	12	12
CMR‡/CCT§	36–60	24–36	12–24	12–24
Exercise test‖	36–60	36–60	24–36	12–24

*See ACHD AP classification **Table 4**.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of neoaortic size, the origin and proximal course of the coronary arteries, branch pulmonary arteries, ventricular function and valvular function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT or catheterization once to establish knowledge of coronary artery anatomy and then as warranted by clinical condition. CCT may be used if CMR is not feasible and to evaluate coronary artery anatomy and cross-sectional imaging status-post stent therapy. If CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute exercise test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; CPET, cardiopulmonary exercise test; d-TGA, dextro-transposition of the great arteries; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.4.1.3. Congenitally Corrected Transposition of the Great Arteries

See also **Table 31**.

Recommendations for Congenitally Corrected Transposition of the Great Arteries
Referenced studies that support recommendations are summarized in Online Data Supplement 47.

COR	LOE	RECOMMENDATIONS
		Diagnostic
IIa	C-LD	1. CMR is reasonable in adults with CCTGA to determine systemic RV dimensions and systolic function (S4.4.1.3-1, S4.4.1.3-2).
		Therapeutic
I	B-NR	2. Tricuspid valve replacement is recommended for symptomatic adults with CCTGA and severe TR, and preserved or mildly depressed systemic ventricular function (S4.4.1.3-3, S4.4.1.3-4).
IIa	C-LD	3. Tricuspid valve replacement is reasonable for asymptomatic adults with CCTGA and severe TR with dilation or mild dysfunction of the systemic ventricle (S4.4.1.3-3).
IIb	B-NR	4. Conduit intervention/replacement may be considered for adults with CCTGA and symptomatic subpulmonary left ventricle-to-PA conduit dysfunction, recognizing that unloading the subpulmonary ventricle may have a detrimental impact on systemic atrioventricular valve function (S4.4.1.3-5).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 31** for routine testing and follow-up intervals.

TABLE 31 CCTGA: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12	12	6–12	3–6
ECG	12	12	12	12
TTE†	12–24	12	12	12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Holter monitor	12–60	12–60	12–36	12
CMR‡/CCT§	36–60	36–60	12–24	12
Exercise test‖	36–60	36–60	12–24	12

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of ventricular size and function and valvular function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible. If CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CCTGA, congenitally corrected transposition of the great arteries; CMR, cardiovascular magnetic resonance imaging; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.4.2. Fontan Palliation of Single Ventricle Physiology (Including Tricuspid Atresia and Double Inlet Left Ventricle)

See also **Table 32**.

Recommendations for Fontan Palliation of Single Ventricle Physiology
Referenced studies that support recommendations are summarized in Online Data Supplement 48.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-LD	1. New presentation of an atrial tachyarrhythmia in adults with Fontan palliation should be managed promptly and include prevention of thromboembolic events and consultation with an electrophysiologist with CHD expertise (S4.4.2-1, S4.4.2-2).
I	C-EO	2. Adults after Fontan palliation should be evaluated annually with either echocardiography or CMR.
I	C-EO	3. Cardiac catheterization should be performed in adults before initial Fontan surgery or revision of a prior Fontan connection to assess suitability of preintervention hemodynamics for Fontan physiology or revision of a prior Fontan connection.
I	C-EO	4. New onset or worsening atrial tachyarrhythmias in adults with single ventricle after Fontan palliation should prompt a search for potential hemodynamic abnormalities, which may necessitate imaging and/or cardiac catheterization.
IIa	B-R	5. In adults with Fontan palliation, it is reasonable to encourage a regular exercise program appropriate to their abilities (S4.4.2-3–S4.4.2-5).
IIa	C-LD	6. Imaging of the liver (ultrasonography, CMR, CT) and laboratory evaluation of liver function for fibrosis, cirrhosis, and/or hepatocellular carcinoma are reasonable in adults after Fontan palliation (S4.4.2-6).
IIa	C-EO	7. In adults after Fontan palliation, it is reasonable to perform biochemical and hematological testing on an annual basis especially for liver and renal function.
IIa	C-LD	8. Cardiac catheterization can be useful to evaluate a symptomatic adult after Fontan palliation when noninvasive testing is insufficient to guide therapy (S4.4.2-7, S4.4.2-8).
IIa	C-LD	9. Evaluation for cardiac transplantation is reasonable in adults with Fontan palliation and signs and symptoms of protein-losing enteropathy (S4.4.2-9–S4.4.2-12).
IIb	C-EO	10. It may be reasonable to perform catheterization in asymptomatic adults after Fontan palliation to evaluate hemodynamics, oxygenation and cardiac function to guide optimal medical, interventional and/or surgical therapy.
		Therapeutic
I	C-EO	11. Anticoagulation with a vitamin K antagonist is recommended for adults with Fontan palliation with known or suspected thrombus, thromboembolic events, or prior atrial arrhythmia, and no contraindications to anticoagulation.
IIa	C-LD	12. Catheter ablation can be useful in adults after Fontan palliation with intra-atrial reentrant tachycardia or focal atrial tachycardia (S4.4.2-13–S4.4.2-15).
IIa	C-LD	13. Fontan revision surgery, including arrhythmia surgery as indicated, is reasonable for adults with atriopulmonary Fontan connections with recurrent atrial tachyarrhythmias refractory to pharmacological therapy and catheter ablation who have preserved systolic ventricular function and severe atrial dilation (S4.4.2-16–S4.4.2-18).
IIa	B-R	14. Pulmonary vasoactive medications can be beneficial to improve exercise capacity in adults with Fontan repair (S4.4.2-19–S4.4.2-25).

(continued)

IIb	B-NR	15. Antiplatelet therapy or anticoagulation with a vitamin K antagonist may be considered in adults after Fontan palliation without known or suspected thrombus, thromboembolic events, or prior arrhythmia (S4.4.2-26).
IIb	C-LD	16. Reoperation or intervention for structural/anatomic abnormalities in a Fontan palliated patient with symptoms or with failure of the Fontan circulation may be considered (S4.4.2-27).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and Table 32 for routine testing and follow-up intervals.

TABLE 32 Fontan Palliation: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12	12	6	3-6
ECG	12	12	6–12	6
TTE†	12	12	12	12
Pulse oximetry	12	12	Each visit	Each visit
Holter monitor	12	12	12	12
CMR‡/CCT§	36	24	24	24
Exercise test‖	36	24	12	12

*See Tables 3 and 4 for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of the long-term sequelae of Fontan palliation: thrombosis, right-to-left shunts (e.g., fenestration. intrapulmonary atrioventricular malformation), obstructive lesion, systemic atrioventricular valve dysfunction, ventricular size and function, collateral burden, and branch pulmonary artery obstruction. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§CCT may be used if CMR is not feasible and to evaluate cross-sectional imaging status-post stent therapy. CCT with contrast injection in Fontan patients can be misleading; therefore, it should be done only when clinically indicated and when it can be appropriately protocoled and interpreted. If CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.
‖6-minute walk test or CPET, depending on clinical indication.
ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.4.3. Double Outlet Right Ventricle

Double outlet right ventricle is an anatomic descriptor that includes abnormalities similar to TOF in some patients (when the aorta is closely related to the VSD) and similar to d-TGA with a VSD in others (when the PA is more closely related to the VSD than the aorta). Repairs are predicated on the underlying anatomy and may involve VSD closure with relief of PS, right ventricle-to-PA conduit, or Rastelli-type repair. In severe cases, single-ventricle physiology may be present. Consequently, recommendations for the management of a patient with double outlet right ventricle can generally be inferred in the recommendations for the lesion with the most similar anatomy and physiology (e.g., TOF can reasonably be based on the recommendations in Section 4.4.1, recognizing that a patient with double outlet right ventricle is more likely to have residual LVOT obstruction).

4.4.4. Severe PAH and Eisenmenger Syndrome

4.4.4.1. Severe PAH

See also Table 33.

Recommendations for Severe PAH
Referenced studies that support recommendations are summarized in Online Data Supplement 49.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	B-NR	1. Patients with ACHD with pulmonary vascular resistance 2.5 Wood units or greater (≥4 Wood units×m^2) should be assessed collaboratively by an ACHD cardiologist and an expert in pulmonary hypertension to develop a management plan (S4.4.4.1-1–S4.4.4.1-17).

(continued)

COR	LOE	RECOMMENDATIONS
I	B-NR	2. Adults with septal or great artery shunts should undergo periodic screening for pulmonary hypertension with TTE (S4.4.4.1-1–S4.4.4.1-18).
I	B-NR	3. Cardiac catheterization to assess pulmonary vascular hemodynamics is recommended for adults with septal or great artery shunts and clinical symptoms, signs, or echocardiographic findings suggestive of pulmonary hypertension (S4.4.4.1-1, S4.4.4.1-2, S4.4.4.1-4, S4.4.4.1-6, S4.4.4.1-7, S4.4.4.1-11, S4.4.4.1-12, S4.4.4.1-15–S4.4.4.1-18).
I	B-NR	4. In adults with septal or great artery shunts, cardiac catheterization with hemodynamics (performed before or at time of closure) is beneficial to assess suitability for closure (S4.4.4.1-1–S4.4.4.1-17).
I	C-EO	5. BNP, chest x-ray, 6-minute walk test, and cardiac catheterization are useful for initial and follow-up evaluation of patients with ACHD with PAH.

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 33** for routine testing and follow-up intervals.

TABLE 33 Pulmonary Hypertension and Eisenmenger Syndrome: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	6–12	3–6
ECG	12	12
TTE†	12	12
Pulse oximetry	Each visit	Each visit
CMR‡	As needed	As needed
Exercise test§	6–12	6–12
Cardiac catheterization‖	As needed	As needed

*See **Tables 3 and 4** for details on the ACHD AP classification system.
†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.
‡CMR may be indicated for assessment of right ventricular function and CHD anatomy not clarified with TTE. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.
§6-minute walk test or CPET, depending on clinical indication.
‖Cardiac catheterization should be performed at baseline and as needed.

ACHD indicates adult congenital heart disease; CMR, cardiovascular magnetic resonance; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; and TTE, transthoracic echocardiogram.

4.4.4.2. Eisenmenger Syndrome

Recommendations for Eisenmenger Syndrome
Referenced studies that support recommendations are summarized in Online Data Supplement 50.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-EO	1. When evaluating adults with presumed Eisenmenger syndrome, clinicians should confirm diagnostic imaging and cardiac catheterization data accuracy and exclude other potential contributors to right-to-left shunting or pulmonary hypertension.
		Therapeutic
I	A	2. Bosentan is beneficial in symptomatic adults with Eisenmenger syndrome with ASD or VSD (S4.4.4.2-1–S4.4.4.2-3).
IIa	B-R	3. In symptomatic adults with Eisenmenger syndrome, bosentan and PDE-5 inhibitors are reasonable in combination if symptomatic improvement does not occur with either medication alone (S4.4.4.2-1, S4.4.4.2-4–S4.4.4.2-6).

(continued)

IIa	C-EO B-NR	4. Bosentan is a reasonable therapy to treat symptomatic adults with Eisenmenger syndrome with 1 of the following: shunts other than ASD/VSD (e.g., PDA, aortopulmonary window) (Level of Evidence C-EO), or complex congenital heart lesions (S4.4.4.2-1, S4.4.4.2-7) or Down syndrome (S4.4.4.2-4, S4.4.4.2-5, S4.4.4.2-8–S4.4.4.2-10) (Level of Evidence B-NR).
IIa	B-NR	5. It is reasonable to use PDE-5 inhibitors (e.g., sildenafil, tadalafil) to treat symptomatic adults with Eisenmenger syndrome with ASD, VSD, or great artery shunt (S4.4.4.2-1, S4.4.4.2-11–S4.4.4.2-16).

See Section 3.3 for recommendations on who should perform surgeries, cardiac catheterization, and other procedures in these patients; Section 3.4 for recommendations on diagnostic evaluation; and **Table 33** for routine testing and follow-up intervals.

4.4.5. Coronary Anomalies

Table 34.

TABLE 34 Factors That May Relate to the Clinical Importance of AAOCA and Risk of SCD

Age	AAOCA is more commonly invoked as the cause of SCD in patients <35 y of age than in patients >35 y of age, in whom atherosclerotic coronary disease becomes a more prevalent cause. However, death has been attributed to AAOCA in patients of all ages; there does not seem to be an age beyond which the AAOCA may not be relevant, even in the setting of atherosclerotic coronary disease and other concomitant conditions (S4.4.5-1, S4.4.5-2).
Anatomy of coronary ostium and proximal coronary course	Slit-like/fish-mouth-shaped orifice, acute angle takeoff, intramural course, interarterial course and hypoplasia of the proximal coronary artery have all been proposed as reasons for symptoms, ischemia and SCD in patients with AAOCA. The slit-like orifice is more commonly seen in anomalous right coronary artery arising from the left sinus. Each of these anatomic findings offers a pathophysiological mechanism for intermittent ischemia, particularly at times of high cardiac output and/or increased aortic wall tension, such as during exercise (S4.4.5-3–S4.4.5-6).
Anomalous origin	Left coronary artery arising from the right cusp is less common than the right coronary artery arising from the left cusp but is more often found in autopsy series of SCD (S4.4.5-1, S4.4.5-7, S4.4.5-8). This suggests that anomalous origin of the left coronary artery from the right cusp is more likely to cause SCD than anomalous origin of the right coronary artery from the left cusp. This may be due either to anatomic features that make anomalous aortic origin of the left coronary artery prone to coronary compromise or because a larger proportion of myocardium is supplied by the left coronary artery, or both.
Exercise	Autopsy series suggest most patients die during, or in close temporal association with, exercise (S4.4.5-8–S4.4.5-10).
Ischemia	Autopsy series demonstrate myocardial fibrosis in a significant number of patients whose deaths were attributed to AAOCA, particularly in patients with anomalous left coronary artery arising from the right cusp (S4.4.5-10). Surgical series describe patients with ischemia or MI before surgical repair in the absence of other CAD, suggesting a relation of the coronary anomaly to the ischemia (S4.4.5-11). This suggests that had perfusion imaging been obtained before SCD, ischemia would have been found in such patients (S4.4.5-12, S4.4.5-13). However, other data indicate that a normal stress test does not preclude a SCD event, with the proviso that most of those studies used only stress ECG, rather than the more sensitive and specific modalities of nuclear perfusion imaging or stress echocardiography. In addition, postoperative studies have shown that ischemia may be found after surgical repair in the distribution not supplied by the abnormal coronary artery and may not persist on repeat testing (S4.4.5-14).
Symptoms	In autopsy and surgical series, a significant number of patients reported cardiovascular symptoms, including before SCD events (S4.4.5-9, S4.4.5-15–S4.4.5-18). Symptoms are more commonly reported in patients in whom the left coronary artery arises from the right sinus. Surgical series have described improvement in symptoms after surgical repair (S4.4.5-6, S4.4.5-8–S4.4.5-10, S4.4.5-15, S4.4.5-16).

AAOCA indicates anomalous aortic origin of the coronary artery; CAD, coronary artery disease; ECG, electrocardiogram; MI, myocardial infarction; and SCD, sudden cardiac death.

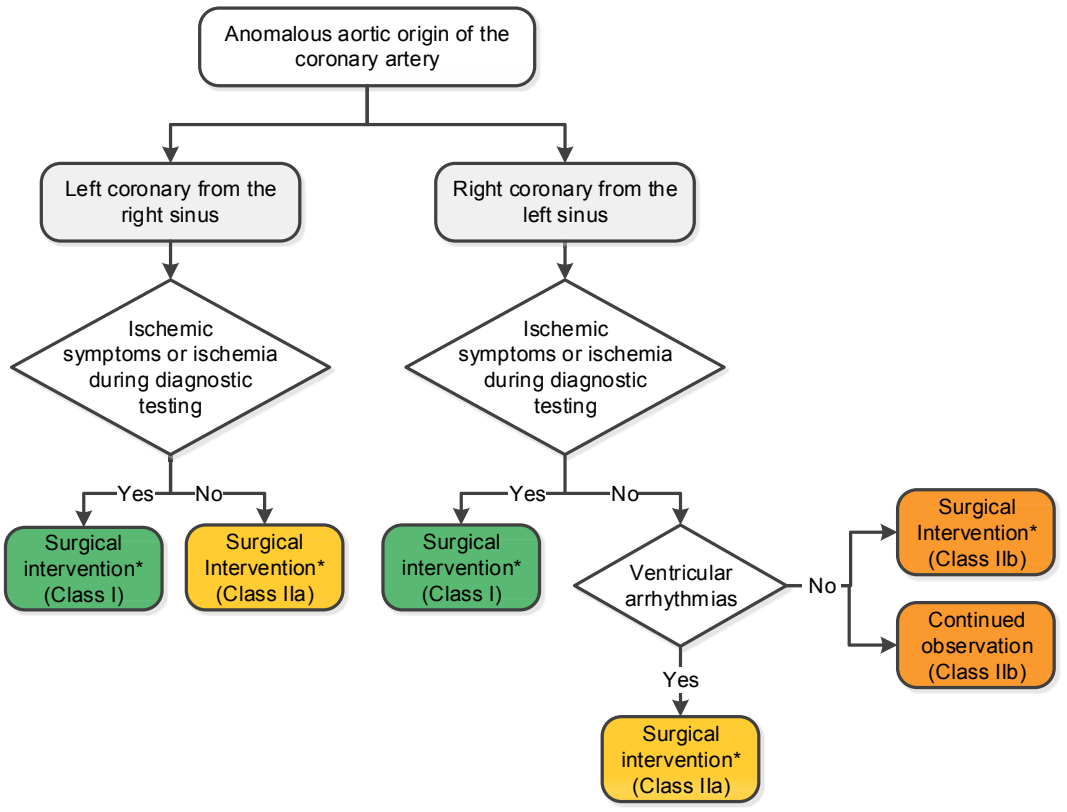
4.4.5.1. Anomalous Coronary Artery Evaluation

Table 34.

Recommendations for Anomalous Coronary Artery Evaluation
Referenced studies that support recommendations are summarized in Online Data Supplement 51.

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-LD	1. Coronary angiography, using catheterization, CT, or CMR, is recommended for evaluation of anomalous coronary artery (S4.4.5.1-1–S4.4.5.1-3).
I	C-LD	2. Anatomic and physiological evaluation should be performed in patients with anomalous aortic origin of the left coronary from the right sinus and/or right coronary from the left sinus (S4.4.5.1-4–S4.4.5.1-9).

FIGURE 5 Anomalous Aortic Origin of the Coronary Artery



*Surgical intervention to involve unroofing or coronary revascularization for patients with concomitant fixed obstruction.

4.4.5.2. Anomalous Aortic Origin of Coronary Artery

Figure 5.

Recommendations for Anomalous Aortic Origin of Coronary Artery
Referenced studies that support recommendations are summarized in Online Data Supplement 51.

COR	LOE	RECOMMENDATIONS
		Therapeutic
I	B-NR	1. Surgery is recommended for AAOCA from the left sinus or AAOCA from the right sinus for symptoms or diagnostic evidence consistent with coronary ischemia attributable to the anomalous coronary artery (S4.4.5.2-1-S4.4.5.2-3).
IIa	C-LD	2. Surgery is reasonable for anomalous aortic origin of the left coronary artery from the right sinus in the absence of symptoms or ischemia (S4.4.5.2-4-S4.4.5.2-6).
IIa	C-EO	3. Surgery for AAOCA is reasonable in the setting of ventricular arrhythmias.
IIb	B-NR	4. Surgery or continued observation may be reasonable for asymptomatic patients with an anomalous left coronary artery arising from the right sinus or right coronary artery arising from the left sinus without ischemia or anatomic or physiological evaluation suggesting potential for compromise of coronary perfusion (e.g., intramural course, fish-mouth-shaped orifice, acute angle) (S4.4.5.2-4-S4.4.5.2-6).

4.4.5.3. Anomalous Coronary Artery Arising From the PA

Recommendations for Anomalous Coronary Artery Arising From the PA
Referenced studies that support recommendations are summarized in Online Data Supplement 51.

COR	LOE	RECOMMENDATIONS
		Therapeutic
I	B-NR	1. Surgery is recommended for anomalous left coronary artery from the PA (S4.4.5.3-1–S4.4.5.3-7).
I	C-EO	2. In a symptomatic adult with anomalous right coronary artery from the PA with symptoms attributed to the anomalous coronary, surgery is recommended.
IIa	C-EO	3. Surgery for anomalous right coronary artery from the PA is reasonable in an asymptomatic adult with ventricular dysfunction or with myocardial ischemia attributed to anomalous right coronary artery from the PA.

5. EVIDENCE GAPS AND FUTURE DIRECTIONS

Table 35.

TABLE 35 High-Impact Research Questions in ACHD

General	
Pathophysiology	■ What are the mechanisms of heart failure that can be prevented, reversed, or treated? ■ Why does the systemic right ventricle fail? ■ Will all patients with Fontan physiology develop clinically important cirrhosis, and how can we prevent this? ■ Who is at risk of aortic rupture and dilation? ■ Are patients with manipulated coronary arteries (e.g., ASO, Ross repair) at risk of premature coronary artery disease? ■ What is the impact of radiation exposure on long-term health? ■ Can we predict who will develop pulmonary hypertension/pulmonary vascular disease?
Medical and surgical treatment	■ How can we modify current CHD surgical procedures to prevent or reduce later development of heart failure and/or arrhythmias? ■ Which patients with ACHD can use direct oral anticoagulants instead of warfarin? ■ What is the best algorithm for contraception choices? ■ Beyond those with severe PAH, which patients will benefit from PAH therapies? ■ Do patients with ACHD with systemic right ventricles and HF benefit from standard therapies (beta blockers, ACE inhibitors/ARBs, aldosterone antagonist)? Which one(s)? ■ What medical therapies benefit patients with failing Fontan physiology? ■ Do asymptomatic patients with ACHD with PAH benefit from PAH-specific therapy? ■ Who will benefit from ventricular assist devices? ■ What should be the threshold(s) for aortic aneurysm surgery? ■ What pacing and resynchronization strategies are of most benefit, and when should they be used?
Outcomes/risk assessment	■ What criteria should determine transplantation eligibility? ■ Which patients benefit from primary prevention ICDs? ■ How can we risk stratify for SCD in patients with systemic right ventricles? ■ What operative risk score predicts outcomes in ACHD reoperations? ■ What HF risk score predicts outcomes in patients with ACHD? ■ Is there a level of exercise where risk exceeds benefit? ■ What is the rate and/or risk of endocarditis?
Assessment	■ Who is at high risk of neurodevelopmental abnormalities and would benefit from neuropsychiatric evaluation and treatment? ■ Who should be screened for anxiety and depression, what treatment is most effective, and are there differences compared with non-patients with ACHD? ■ What is the standard protocol for assessing right ventricular size and function by CMR imaging? ■ Which biomarkers are predictive of mortality and morbidity?
Disease-specific	
Coarctation of the aorta	■ Which measure of hypertension—resting, exercise, or ambulatory—best predicts outcomes? ■ Is there an optimal antihypertensive regimen? ■ What should blood pressure goals be? ■ How often should patients be screened for thoracic aneurysm? ■ Should exercise-induced hypertension be treated? ■ What criteria warrant reintervention in recoarctation? ■ Is long-term outcome better with medical therapy or catheter intervention for less than severe recoarctation? ■ Should patients be screened for intracranial aneurysm, and if so, how often?

Continued in the next column

TABLE 35 Continued

Ebstein anomaly	■ What is the indication for surgery in the asymptomatic patient? ■ Who should have a Glenn shunt at the time of tricuspid valve surgery? ■ Should surgeons attempt tricuspid valve repair or routinely perform replacement in all patients?
TOF	■ What is the optimal timing for pulmonary valve replacement in asymptomatic patients with TOF? ■ Do pulmonary valve replacement and ventricular tachycardia ablation decrease the risk of SCD? ■ Who needs a primary prevention ICD, and does this strategy reduce mortality? ■ Is there a role for PAH therapies in TOF? ■ Why does left ventricular dysfunction develop?
TGA/systemic right ventricle	■ Who benefits from ACE inhibitors/ARBs/beta blockers/spironolactone? ■ Who needs a primary prevention ICD, and does this strategy prevent mortality? ■ What imaging findings predict mortality/morbidity? ■ In CCTGA with VSD/PS, does the double switch have better long-term outcomes than VSD closure and left ventricle-to-PA conduit? ■ When should tricuspid valve replacement be performed? ■ What is the role of cardiac resynchronization therapy in patients with systemic right ventricle?
ASO	■ What are the long-term outcomes after ASO? ■ How should the possibility of asymptomatic coronary disease (ostial, compression) and ischemia be assessed?
Single ventricle/Fontan	■ Is warfarin or aspirin beneficial in patients with a Fontan? ■ Are PAH therapies beneficial? ■ Is exercise capacity predictive of mortality? ■ What liver screening is appropriate and at what intervals? ■ How is protein-losing enteropathy best medically treated? ■ Why do some patients fail with preserved ejection fraction, whereas other have decreased ejection fraction? ■ What are the long-term outcomes of hypoplastic left heart syndrome? ■ What is ideal timing for heart transplantation in single ventricle Fontan patients, and should liver issues prompt earlier transplantation than might be felt necessary from a cardiac perspective? ■ Which has better long-term outcomes, the Fontan operation or bidirectional Glenn alone?
Coronary anomalies	■ Does surgical intervention in anomalous aortic origin of coronary arteries improve survival?

ACE indicates angiotensin-converting enzyme; ACHD, adult congenital heart disease; ARB, angiotensin-receptor blocker; ASO, arterial switch operation; CCTGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; CMR, cardiac magnetic resonance; ICD, implantable cardioverter-defibrillator; HF, heart failure; PA, pulmonary artery; PAH, pulmonary artery hypertension; PS, pulmonary stenosis; SCD, sudden cardiac death; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; and VSD, ventricular septal defect.

PRESIDENTS AND STAFF

American College of Cardiology
C. Michael Valentine, MD, FACC, President
Cathleen C. Gates, Interim Chief Executive Officer and Chief Operating Officer
William J. Oetgen, MD, MBA, FACC, Executive Vice President, Science, Education, Quality, and Publications
MaryAnne Elma, MPH, Senior Director, Science, Education, Quality, and Publishing
Amelia Scholtz, PhD, Publications Manager, Science, Education, Quality, and Publishing

American College of Cardiology/American Heart Association
Katherine A. Sheehan, PhD, Director, Guideline Strategy and Operations
Abdul R. Abdullah, MD, Senior Manager, Guideline Science

American Heart Association
Ivor Benjamin, MD, FAHA, President
Nancy Brown, Chief Executive Officer
Rose Marie Robertson, MD, FAHA, Chief Science and Medical Officer
Gayle R. Whitman, PhD, RN, FAHA, FAAN, Senior Vice President, Office of Science Operations
Prashant Nedungadi, PhD, Science and Medicine Advisor, Office of Science Operations
Jody Hundley, Production and Operations Manager, Scientific Publications, Office of Science Operations

REFERENCES

PREAMBLE

P-1. Committee on Standards for Developing Trustworthy Clinical Practice Guidelines, Institute of Medicine (U.S.). Clinical Practice Guidelines We Can Trust. Washington, DC: The National Academies Press, 2011.

P-2. Committee on Standards for Systematic Reviews of Comparative Effectiveness Research, Institute of Medicine (U.S.). Finding What Works in Health Care: Standards for Systematic Reviews. Washington, DC: The National Academies Press, 2011.

P-3. Anderson JL, Heidenreich PA, Barnett PG, et al. ACC/AHA statement on cost/value methodology in clinical practice guidelines and performance measures: a report of the American College of Cardiology/American Heart Association Task Force on Performance Measures and Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:2304–22.

P-4. ACCF/AHA Task Force on Practice Guidelines. Methodology Manual and Policies From the ACCF/AHA Task Force on Practice Guidelines. American College of Cardiology and American Heart Association, 2010. Available at: http://assets.cardiosource.com/Methodology_Manual_for_ACC_AHA_Writing_Committees.pdf and http://professional.heart.org/idc/groups/ahamah-public/@wcm/@sop/documents/downloadable/ucm_319826.pdf. Accessed September 15, 2017.

P-5. Halperin JL, Levine GN, Al-Khatib SM, et al. Further evolution of the ACC/AHA clinical practice guideline recommendation classification system: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2016;67:1572–4.

P-6. Jacobs AK, Kushner FG, Ettinger SM, et al. ACCF/AHA clinical practice guideline methodology summit report: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;61:213–65.

P-7. Jacobs AK, Anderson JL, Halperin JL, et al. The evolution and future of ACC/AHA clinical practice guidelines: a 30-year journey: a report of the American College of Cardiology/American Heart Association Task Force on practice guidelines. J Am Coll Cardiol. 2014;64:1373–84.

P-8. Arnett DK, Goodman RA, Halperin JL, et al. AHA/ACC/HHS strategies to enhance application of clinical practice guidelines in patients with cardiovascular disease and comorbid conditions: from the American Heart Association, American College of Cardiology, and US Department of Health and Human Services. J Am Coll Cardiol. 2014;64:1851–6.

P-9. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2018 Aug 10 [E-pub ahead of print].

1. INTRODUCTION

1.1. Methodology and Evidence Review

S1.1-1. Zaragoza-Macias E, Zaidi A, Dendukuri N, et al. Medical therapy for systemic right ventricles: a systematic review (part 1) for the 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2018 Aug 9 [E-pub ahead of print].

S1.1-2. Oster M, Bhatt A, Zaragoza-Macias E, et al. Interventional therapy versus medical therapy for secundum atrial septal defect: a systematic review (part 2) for the 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2018 Aug 9 [E-pub ahead of print].

1.4. Scope of the Guideline

S1.4-1. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol. 2008;52:e143–263.

S1.4-2. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:e57–185.

S1.4-3. Moons P, Bovijn L, Budts W, et al. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation. 2010;122:2264–72.

S1.4-4. Gilboa SM, Salemi JL, Nembhard WN, et al. Mortality resulting from congenital heart disease among children and adults in the United States, 1999 to 2006. Circulation. 2010;122:2254–63.

S1.4-5. van der Bom T, Bouma BJ, Meijboom FJ, et al. The prevalence of adult congenital heart disease, results from a systematic review and evidence based calculation. Am Heart J. 2012;164:568–75.

S1.4-6. Hoffman JIE, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. Am Heart J. 2004;147:425–39.

S1.4-7. Lien WP, Chen JJ, Chen JH, et al. Frequency of various congenital heart diseases in Chinese adults: analysis of 926 consecutive patients over 13 years of age. Am J Cardiol. 1986;57:840–4.

S1.4-8. Samánek M, Slavík Z, Zborilová B, et al. Prevalence, treatment, and outcome of heart disease in live-born children: a prospective analysis of 91,823 live-born children. Pediatr Cardiol. 1989;10:205–11.

S1.4-9. Mylotte D, Pilote L, Ionescu-Ittu R, et al. Specialized adult congenital heart disease care: the impact of policy on mortality. Circulation. 2014;129:1804–12.

S1.4-10. Shen W-K, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2017;70:e39–110.

S1.4-11. Page RL, Joglar JA, Caldwell MA, et al. 2015 ACC/AHA/HRS guideline for the management of adult patients with supraventricular tachycardia: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2016;67:e27–115.

S1.4-12. Callaway CW, Donnino MW, Fink EL, et al. Part 8: post-cardiac arrest care: 2015 American Heart Association guidelines update for cardiopulmonary resuscitation and emergency cardiovascular care. Circulation. 2015;132:S465–82.

S1.4-13. Amsterdam EA, Wenger NK, Brindis RG, et al. 2014 AHA/ACC guideline for the management of patients with non-ST-elevation acute coronary syndromes: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;64:e139–228.

S1.4-14. Fleisher LA, Fleischmann KE, Auerbach AD, et al. 2014 ACC/AHA guideline on perioperative cardiovascular evaluation and management of patients undergoing noncardiac surgery: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;64:e77–137.

S1.4-15. January CT, Wann LS, Alpert JS, et al. 2014 AHA/ACC/HRS guideline for the management of patients with atrial fibrillation: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2014;64:e1–76.

S1.4-16. Fihn SD, Blankenship JC, Alexander KP, et al. 2014 ACC/AHA/AATS/PCNA/SCAI/STS focused update of the guideline for the diagnosis and management of patients with stable ischemic heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, and the American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2014;64:1929–49.

S1.4-17. Fihn SD, Gardin JM, Abrams J, et al. 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS guideline for the diagnosis and management of patients with stable ischemic heart disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, and the American College of Physicians, American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2012;60:e44–164.

S1.4-18. Goff DC Jr., Lloyd-Jones DM, Bennett G, et al. 2013 ACC/AHA guideline on the assessment of

cardiovascular risk: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63(25 Pt B):2935–59.

S1.4-19. Stone NJ, Robinson JG, Lichtenstein AH, et al. 2013 ACC/AHA guideline on the treatment of blood cholesterol to reduce atherosclerotic cardiovascular risk in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63(25 Pt B):2889–934.

S1.4-20. Jensen MD, Ryan DH, Apovian CM, et al. 2013 AHA/ACC/TOS guideline for the management of overweight and obesity in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines and The Obesity Society. J Am Coll Cardiol. 2014;63(25 Pt B):2985–3023.

S1.4-21. Eckel RH, Jakicic JM, Ard JD, et al. 2013 AHA/ACC guideline on lifestyle management to reduce cardiovascular risk: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63(25 Pt B):2960–84.

S1.4-22. Nishimura RA, Otto CM, Bonow RO, et al. 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2017;70:252–89.

S1.4-23. Whelton PK, Carey RM, Aronow WS, et al. 2017 ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA guideline for the prevention, detection, evaluation, and management of high blood pressure in adults: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2018;71:e127–248.

S1.4-24. Svensson LG, Adams DH, Bonow RO, et al. Aortic valve and ascending aorta guidelines for management and quality measures. Ann Thorac Surg. 2013;95:S1–66.

S1.4-25. O'Gara PT, Kushner FG, Ascheim DD, et al. 2013 ACCF/AHA guideline for the management of ST-elevation myocardial infarction: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;61:e78–140.

S1.4-26. Yancy CW, Jessup M, Bozkurt B, et al. 2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. Circulation. 2017;136:e137–61.

S1.4-27. Tracy CM, Epstein AE, Darbar D, et al. 2012 ACCF/AHA/HRS focused update of the 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2012;60:1297–313.

S1.4-28. Hillis LD, Smith PK, Anderson JL, et al. 2011 ACCF/AHA guideline for coronary artery bypass graft surgery: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2011;58:e123–210.

S1.4-29. Levine GN, Bates ER, Blankenship JC, et al. 2011 ACCF/AHA/SCAI guideline for percutaneous coronary intervention: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Society for Cardiovascular Angiography and Interventions. J Am Coll Cardiol. 2011;58:e44–122.

S1.4-30. Smith SC Jr., Benjamin EJ, Bonow RO, et al. AHA/ACCF secondary prevention and risk reduction therapy for patients with coronary and other atherosclerotic vascular disease: 2011 update: a guideline from the American Heart Association and American College of Cardiology Foundation. J Am Coll Cardiol. 2011;58:2432–46.

S1.4-31. Mosca L, Benjamin EJ, Berra K, et al. Effectiveness-based guidelines for the prevention of cardiovascular disease in women—2011 update: a guideline from the American Heart Association. Circulation. 2011;123:1243–62.

S1.4-32. Baumgartner H, Bonhoeffer P, De Groot NMS, et al. ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010;31:2915–57.

S1.4-33. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. J Am Coll Cardiol. 2010;55:e27–129.

S1.4-34. Silversides CK, Marelli A, Beauchesne L, et al. Canadian Cardiovascular Society 2009 consensus conference on the management of adults with congenital heart disease: executive summary. Can J Cardiol. 2010;26:143–50.

S1.4-35. Habib G, Hoen B, Tornos P, et al. Guidelines on the prevention, diagnosis, and treatment of infective endocarditis (new version 2009): the Task Force on the Prevention, Diagnosis, and Treatment of Infective Endocarditis of the European Society of Cardiology (ESC). Eur Heart J. 2009;30:2369–413.

S1.4-36. Cohen MS, Eidem BW, Cetta F, et al. Multimodality imaging guidelines of patients with transposition of the great arteries: a report from the American Society of Echocardiography. Developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. J Am Soc Echocardiogr. 2016;29:571–621.

S1.4-37. Lang RM, Badano LP, Mor-Avi V, et al. Recommendations for cardiac chamber quantification by echocardiography in adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. J Am Soc Echocardiogr. 2015;28:1–39.e14.

S1.4-38. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease. Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Heart Rhythm. 2014;11:e102–65.

S1.4-39. Valente AM, Cook S, Festa P, et al. Multimodality imaging guidelines for patients with repaired tetralogy of Fallot: a report from the American Society of Echocardiography. Developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology. J Am Soc Echocardiogr. 2014;27:111–41.

S1.4-40. Boodhwani M, Andelfinger G, Leipsic J, et al. Canadian Cardiovascular Society position statement on the management of thoracic aortic disease. Can J Cardiol. 2014;30:577–89.

S1.4-41. Longmuir PE, Brothers JA, de Ferranti SD, et al. Promotion of physical activity for children and adults with congenital heart disease: a scientific statement from the American Heart Association. Circulation. 2013;127:2147–59.

S1.4-42. Marino BS, Lipkin PH, Newburger JW, et al. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. Circulation. 2012;126:1143–72.

S1.4-43. European Society of Gynecology (ESG), Association for European Paediatric Cardiology (AEPC), German Society for Gender Medicine (DGesGM), et al. ESC guidelines on the management of cardiovascular diseases during pregnancy: the Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). Eur Heart J. 2011;32:3147–97.

S1.4-44. Sable C, Foster E, Uzark K, et al. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues: a scientific statement from the American Heart Association. Circulation. 2011;123:1454–85.

S1.4-45. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association. Developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc., and the Pulmonary Hypertension Association. Circulation. 2009;119:2250–94.

S1.4-46. Wilson W, Taubert KA, Gewitz M, 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.

2. BACKGROUND AND PATHOPHYSIOLOGY

2.1. Anatomic and Physiological Terms

S2.1-1. Giroud JM, Jacobs JP, Spicer D, et al. Report from the International Society for Nomenclature of Paediatric and Congenital Heart Disease: creation of a visual encyclopedia illustrating the terms and definitions of the international pediatric and congenital cardiac code. World J Pediatr Congenit Heart Surg. 2010;1:300–13.

S2.1-2. Bergersen L, Giroud JM, Jacobs JP, et al. Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease:

cardiovascular catheterisation for congenital and paediatric cardiac disease (Part 2 - Nomenclature of complications associated with interventional cardiology). Cardiol Young. 2011;21:260–5.

S2.1-3. Bergersen L, Everett AD, Giroud JM, et al. Report from the International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (Part 1 - Procedural nomenclature). Cardiol Young. 2011;21:252–9.

S2.1-4. Franklin RCG, Jacobs JP, Krogmann ON, et al. Nomenclature for congenital and paediatric cardiac disease: historical perspectives and The International Pediatric and Congenital Cardiac Code. Cardiol Young. 2008;18 suppl 2:70–80.

S2.1-5. Franklin RCG, Béland MJ, Krogmann ON. Mapping and coding of nomenclatures for paediatric and congenital heart disease. Cardiol Young. 2006;16:105–6.

S2.1-6. International Society for Nomenclature of Paediatric and Congenital Heart Disease. International Paediatric and Congenital Cardiac Code. Available at: http://ipccc.net/. Accessed: May 29, 2018.

2.2. Severity of ACHD

S2.2-1. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol. 2008;52:e143–263.

S2.2-2. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:e57–185.

S2.2-3. Boodhwani M, Andelfinger G, Leipsic J, et al. Canadian Cardiovascular Society position statement on the management of thoracic aortic disease. Can J Cardiol. 2014;30:577–89.

S2.2-4. Svensson LG, Adams DH, Bonow RO, et al. Aortic valve and ascending aorta guidelines for management and quality measures. Ann Thorac Surg. 2013;95:S1–66.

S2.2-5. Verheugt CL, Uiterwaal CSPM, van der Velde ET, et al. Mortality in adult congenital heart disease. Eur Heart J. 2010;31:1220–9.

S2.2-6. Khairy P, Aboulhosn J, Gurvitz MZ, et al. Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. Circulation. 2010;122:868–75.

S2.2-7. Khairy P, Fernandes SM, Mayer JE Jr., et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation. 2008;117:85–92.

S2.2-8. Khairy P, Harris L, Landzberg MJ, et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. Circulation. 2008;117:363–70.

S2.2-9. Dimopoulos K, Diller G-P, Koltsida E, et al. Prevalence, predictors, and prognostic value of renal dysfunction in adults with congenital heart disease. Circulation. 2008;117:2320–8.

S2.2-10. Dimopoulos K, Diller G-P, Giannakoulas G, et al. Anemia in adults with congenital heart disease relates to adverse outcome. J Am Coll Cardiol. 2009;54:2093–100.

S2.2-11. Dimopoulos K, Diller G-P, Petraco R, et al. Hyponatraemia: a strong predictor of mortality in adults with congenital heart disease. Eur Heart J. 2010;31:595–601.

S2.2-12. Diller G-P, Dimopoulos K, Okonko D, et al. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. Circulation. 2005;112:828–35.

S2.2-13. Buys R, Cornelissen V, Van De Bruaene A, et al. Measures of exercise capacity in adults with congenital heart disease. Int J Cardiol. 2011;153:26–30.

S2.2-14. Gratz A, Hess J, Hager A. Self-estimated physical functioning poorly predicts actual exercise capacity in adolescents and adults with congenital heart disease. Eur Heart J. 2009;30:497–504.

S2.2-15. Dimopoulos K, Okonko DO, Diller G-P, et al. Abnormal ventilatory response to exercise in adults with congenital heart disease relates to cyanosis and predicts survival. Circulation. 2006;113:2796–802.

S2.2-16. Inuzuka R, Diller G-P, Borgia F, et al. Comprehensive use of cardiopulmonary exercise testing identifies adults with congenital heart disease at increased mortality risk in the medium term. Circulation. 2012;125:250–9.

S2.2-17. Müller J, Hager A, Diller G-P, et al. Peak oxygen uptake, ventilatory efficiency and QRS-duration predict event free survival in patients late after surgical repair of tetralogy of Fallot. Int J Cardiol. 2015;196:158–64.

S2.2-18. Kempny A, Dimopoulos K, Uebing A, et al. Reference values for exercise limitations among adults with congenital heart disease. Relation to activities of daily life—single centre experience and review of published data. Eur Heart J. 2012;33:1386–96.

S2.2-19. New York Heart Association. In: Dolgin M, editor. Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Great Vessels. 9th ed. Boston, MA: Little, Brown and Co, 1994.

S2.2-20. Hoeper MM, Bogaard HJ, Condliffe R, et al. Definitions and diagnosis of pulmonary hypertension. J Am Coll Cardiol. 2013;62:D42–50.

2.3. The ACHD AP Classification

S2.3-1. Verheugt CL, Uiterwaal CSPM, van der Velde ET, et al. Mortality in adult congenital heart disease. Eur Heart J. 2010;31:1220–9.

S2.3-2. Diller G-P, Dimopoulos K, Okonko D, et al. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. Circulation. 2005;112:828–35.

S2.3-3. Dimopoulos K, Diller G-P, Koltsida E, et al. Prevalence, predictors, and prognostic value of renal dysfunction in adults with congenital heart disease. Circulation. 2008;117:2320–8.

S2.3-4. Dimopoulos K, Okonko DO, Diller G-P, et al. Abnormal ventilatory response to exercise in adults with congenital heart disease relates to cyanosis and predicts survival. Circulation. 2006;113:2796–802.

S2.3-5. Hebson CL, McCabe NM, Elder RW, et al. Hemodynamic phenotype of the failing Fontan in an adult population. Am J Cardiol. 2013;112:1943–7.

S2.3-6. Khairy P, Aboulhosn J, Gurvitz MZ, et al. Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. Circulation. 2010;122:868–75.

S2.3-7. Khairy P, Harris L, Landzberg MJ, et al. Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study. Circ Arrhythm Electrophysiol. 2008;1:250–7.

S2.3-8. Inuzuka R, Diller G-P, Borgia F, et al. Comprehensive use of cardiopulmonary exercise testing identifies adults with congenital heart disease at increased mortality risk in the medium term. Circulation. 2012;125:250–9.

S2.3-9. Müller J, Hager A, Diller G-P, et al. Peak oxygen uptake, ventilatory efficiency and QRS-duration predict event free survival in patients late after surgical repair of tetralogy of Fallot. Int J Cardiol. 2015;196:158–64.

S2.3-10. Stefanescu A, Macklin EA, Lin E, et al. Usefulness of the Seattle Heart Failure Model to identify adults with congenital heart disease at high risk of poor outcome. Am J Cardiol. 2014;113:865–70.

S2.3-11. Giannakoulas G, Dimopoulos K, Engel R, et al. Burden of coronary artery disease in adults with congenital heart disease and its relation to congenital and traditional heart risk factors. Am J Cardiol. 2009;103:1445–50.

S2.3-12. Giannakoulas G, Dimopoulos K, Bolger AP, et al. Usefulness of natriuretic peptide levels to predict mortality in adults with congenital heart disease. Am J Cardiol. 2010;105:869–73.

S2.3-13. Khairy P, Fernandes SM, Mayer JE Jr., et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation. 2008;117:85–92.

S2.3-14. Khairy P, Harris L, Landzberg MJ, et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. Circulation. 2008;117:363–70.

S2.3-15. Dimopoulos K, Diller G-P, Giannakoulas G, et al. Anemia in adults with congenital heart disease relates to adverse outcome. J Am Coll Cardiol. 2009;54:2093–100.

S2.3-16. Dimopoulos K, Diller G-P, Petraco R, et al. Hyponatraemia: a strong predictor of mortality in adults with congenital heart disease. Eur Heart J. 2010;31:595–601.

S2.3-17. Yancy CW, Jessup M, Bozkurt B, et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. Circulation. 2013;128:e240–327.

3. GENERAL PRINCIPLES

3.1. ACHD Program

S3.1-1. Mylotte D, Pilote L, Ionescu-Ittu R, et al. Specialized adult congenital heart disease care: the impact of policy on mortality. Circulation. 2014;129:1804–12.

3.2. Access to Care

S3.2-1. Gurvitz M, Valente AM, Broberg C, et al. Prevalence and predictors of gaps in care among adult congenital heart disease patients: HEART-ACHD (The Health, Education, and Access Research Trial). J Am Coll Cardiol. 2013;61:2180–4.

S3.2-2. Mackie AS, Islam S, Magill-Evans J, et al. Healthcare transition for youth with heart disease: a clinical trial. Heart. 2014;100:1113–8.

3.3. Delivery of Care

S3.3-1. Mylotte D, Pilote L, Ionescu-Ittu R, et al. Specialized adult congenital heart disease care: the impact of policy on mortality. Circulation. 2014;129:1804–12.

S3.3-2. Karamlou T, Diggs BS, Person T, et al. National practice patterns for management of adult congenital heart disease: operation by pediatric heart surgeons decreases in-hospital death. Circulation. 2008;118:2345–52.

S3.3-3. Videbæk J, Laursen HB, Olsen M, et al. Long-term nationwide follow-up study of simple congenital heart disease diagnosed in otherwise healthy children. Circulation. 2016;133:474–83.

S3.3-4. Karonis T, Scognamiglio G, Babu-Narayan SV, et al. Clinical course and potential complications of small ventricular septal defects in adulthood: late development of left ventricular dysfunction justifies lifelong care. Int J Cardiol. 2016;208:102–6.

S3.3-5. Bonello B, Kilner PJ. Review of the role of cardiovascular magnetic resonance in congenital heart disease, with a focus on right ventricle assessment. Arch Cardiovasc Dis. 2012;105:605–13.

S3.3-6. Kilner PJ. Imaging congenital heart disease in adults. Br J Radiol. 2011;84 Spec No 3:S258–68.

S3.3-7. Cohen MS, Eidem BW, Cetta F, et al. Multimodality imaging guidelines of patients with transposition of the great arteries: a report from the American Society of Echocardiography. Developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. J Am Soc Echocardiogr. 2016;29:571–621.

S3.3-8. Valente AM, Cook S, Festa P, et al. Multimodality imaging guidelines for patients with repaired tetralogy of Fallot: a report from the American Society of Echocardiography. Developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology. J Am Soc Echocardiogr. 2014;27:111–41.

S3.3-9. Ginde S, Bartz PJ, Hill GD, et al. Restrictive lung disease is an independent predictor of exercise intolerance in the adult with congenital heart disease. Congenit Heart Dis. 2013;8:246–54.

S3.3-10. Alonso-Gonzalez R, Borgia F, Diller G-P, et al. Abnormal lung function in adults with congenital heart disease: prevalence, relation to cardiac anatomy, and association with survival. Circulation. 2013;127:882–90.

S3.3-11. Zach KJ, Ramakrishna H, Chandrasekaran K, et al. Laparoscopic colectomy in an adult with single ventricle physiology: anesthetic implications and management. Ann Card Anaesth. 2015;18:252–6.

S3.3-12. Duffels MGJ, Engelfriet PM, Berger RMF, et al. Pulmonary arterial hypertension in congenital heart disease: an epidemiologic perspective from a Dutch registry. Int J Cardiol. 2007;120:198–204.

S3.3-13. Lowe BS, Therrien J, Ionescu-Ittu R, et al. Diagnosis of pulmonary hypertension in the congenital heart disease adult population impact on outcomes. J Am Coll Cardiol. 2011;58:538–46.

S3.3-14. Van De Bruaene A, Delcroix M, Pasquet A, et al. The importance of pulmonary artery pressures on late atrial arrhythmia in transcatheter and surgically closed ASD type secundum. Int J Cardiol. 2011;152:192–5.

S3.3-15. Barst RJ, Ivy DD, Foreman AJ, et al. Four- and seven-year outcomes of patients with congenital heart disease-associated pulmonary arterial hypertension (from the REVEAL Registry). Am J Cardiol. 2014;113:147–55.

S3.3-16. D'Alto M, Romeo E, Argiento P, et al. Therapy for pulmonary arterial hypertension due to congenital heart disease and Down's syndrome. Int J Cardiol. 2013;164:323–6.

S3.3-17. Engelfriet PM, Duffels MGJ, Möller T, et al. Pulmonary arterial hypertension in adults born with a heart septal defect: the Euro Heart Survey on adult congenital heart disease. Heart. 2007;93:682–7.

S3.3-18. Manes A, Palazzini M, Leci E, et al. Current era survival of patients with pulmonary arterial hypertension associated with congenital heart disease: a comparison between clinical subgroups. Eur Heart J. 2014;35:716–24.

S3.3-19. O'Donnell C, Ruygrok PN, Whyte K, et al. Progressive pulmonary hypertension post atrial septal defect device closure—early symptomatic improvement may not predict outcome. Heart Lung Circ. 2010;19:713–6.

S3.3-20. Van De Bruaene A, Moons P, Belmans A, et al. Predictive model for late atrial arrhythmia after closure of an atrial septal defect. Int J Cardiol. 2013;164:318–22.

S3.3-21. D'Alto M, Romeo E, Argiento P, et al. Hemodynamics of patients developing pulmonary arterial hypertension after shunt closure. Int J Cardiol. 2013;168:3797–801.

S3.3-22. Steele PM, Fuster V, Cohen M, et al. Isolated atrial septal defect with pulmonary vascular obstructive disease—long-term follow-up and prediction of outcome after surgical correction. Circulation. 1987;76:1037–42.

3.4. Evaluation of Suspected and Known CHD

3.4.2. Ionizing Radiation Principles

S3.4.2-1. Glatz AC, Purrington KS, Klinger A, et al. Cumulative exposure to medical radiation for children requiring surgery for congenital heart disease. J Pediatr. 2014;164:789–94.e10.

S3.4.2-2. Johnson JN, Hornik CP, Li JS, et al. Cumulative radiation exposure and cancer risk estimation in children with heart disease. Circulation. 2014;130:161–7.

S3.4.2-3. Yakoumakis E, Kostopoulou H, Makri T, et al. Estimation of radiation dose and risk to children undergoing cardiac catheterization for the treatment of a congenital heart disease using Monte Carlo simulations. Pediatr Radiol. 2013;43:339–46.

S3.4.2-4. Andreassi MG, Ait-Ali L, Botto N, et al. Cardiac catheterization and long-term chromosomal damage in children with congenital heart disease. Eur Heart J. 2006;27:2703–8.

3.4.3. Echocardiography

S3.4.3-1. Randolph GR, Hagler DJ, Connolly HM, et al. Intraoperative transesophageal echocardiography during surgery for congenital heart defects. J Thorac Cardiovasc Surg. 2002;124:1176–82.

3.4.4. CMR Imaging

S3.4.4-1. Puchalski MD, Williams RV, Askovich B, et al. Assessment of right ventricular size and function: echo versus magnetic resonance imaging. Congenit Heart Dis. 2007;2:27–31.

S3.4.4-2. Blalock SE, Banka P, Geva T, et al. Interstudy variability in cardiac magnetic resonance imaging measurements of ventricular volume, mass, and ejection fraction in repaired tetralogy of Fallot: a prospective observational study. J Magn Reson Imaging. 2013;38:829–35.

S3.4.4-3. Crean AM, Maredia N, Ballard G, et al. 3D Echo systematically underestimates right ventricular volumes compared to cardiovascular magnetic resonance in adult congenital heart disease patients with moderate or severe RV dilatation. J Cardiovasc Magn Reson. 2011;13:78.

S3.4.4-4. Bonello B, Kilner PJ. Review of the role of cardiovascular magnetic resonance in congenital heart disease, with a focus on right ventricle assessment. Arch Cardiovasc Dis. 2012;105:605–13.

S3.4.4-5. Kilner PJ. Imaging congenital heart disease in adults. Br J Radiol. 2011;84 Spec No 3:S258–68.

S3.4.4-6. Festa P, Ait-Ali L, Cerillo AG, et al. Magnetic resonance imaging is the diagnostic tool of choice in the preoperative evaluation of patients with partial anomalous pulmonary venous return. Int J Cardiovasc Imaging. 2006;22:685–93.

S3.4.4-7. van der Linde D, Rossi A, Yap SC, et al. Ascending aortic diameters in congenital aortic stenosis: cardiac magnetic resonance versus transthoracic echocardiography. Echocardiography. 2013;30:497–504.

S3.4.4-8. Arheden H, Holmqvist C, Thilen U, et al. Left-to-right cardiac shunts: comparison of measurements obtained with MR velocity mapping and with radionuclide angiography. Radiology. 1999;211:453–8.

S3.4.4-9. Brenner DJ, Hall EJ. Computed tomography—an increasing source of radiation exposure. N Engl J Med. 2007;357:2277–84.

S3.4.4-10. Teo KSL, Disney PJ, Dundon BK, et al. Assessment of atrial septal defects in adults comparing cardiovascular magnetic resonance with transoesophageal echocardiography. J Cardiovasc Magn Reson. 2010;12:44.

S3.4.4-11. Prasad SK, Soukias N, Hornung T, et al. Role of magnetic resonance angiography in the diagnosis of major aortopulmonary collateral arteries and partial anomalous pulmonary venous drainage. Circulation. 2004;109:207–14.

3.4.5. Cardiac Computed Tomography

S3.4.5-1. Brenner DJ, Hall EJ. Computed tomography—an increasing source of radiation exposure. N Engl J Med. 2007;357:2277–84.

S3.4.5-2. Ghoshhajra BB, Sidhu MS, El-Sherief A, et al. Adult congenital heart disease imaging with second-generation dual-source computed tomography: initial experiences and findings. Congenit Heart Dis. 2012;7:516–25.

3.4.6. Cardiac Catheterization

S3.4.6-1. Moore JW, Vincent RN, Beekman RH 3rd, et al. Procedural results and safety of common interventional procedures in congenital heart disease: initial report from the National Cardiovascular Data Registry. J Am Coll Cardiol. 2014;64:2439–51.

S3.4.6-2. Learn CP, Holzer RJ, Daniels CJ, et al. Adverse events rates and risk factors in adults undergoing cardiac catheterization at pediatric hospitals—results from the C3PO. Catheter Cardiovasc Interv. 2013;81:997–1005.

S3.4.6-3. Sutton NJ, Greenberg MA, Menegus MA, et al. Caring for the adult with congenital heart disease in an adult catheterization laboratory by pediatric interventionalists—safety and efficacy. Congenit Heart Dis. 2013;8:111–6.

S3.4.6-4. Opotowsky AR, Landzberg MJ, Kimmel SE, et al. Percutaneous closure of patent foramen ovale and atrial septal defect in adults: the impact of clinical variables and hospital procedure volume on in-hospital adverse events. Am Heart J. 2009;157:867–74.

S3.4.6-5. Reant P, Brunot S, Lafitte S, et al. Predictive value of noninvasive coronary angiography with multidetector computed tomography to detect significant coronary stenosis before valve surgery. Am J Cardiol. 2006;97:1506–10.

S3.4.6-6. Gilard M, Cornily J-C, Pennec P-Y, et al. Accuracy of multislice computed tomography in the preoperative assessment of coronary disease in patients with aortic valve stenosis. J Am Coll Cardiol. 2006;47:2020–4.

S3.4.6-7. Manghat NE, Morgan-Hughes GJ, Broadley AJ, et al. 16-Detector row computed tomographic coronary angiography in patients undergoing evaluation for aortic valve replacement: comparison with catheter angiography. Clin Radiol. 2006;61:749–57.

S3.4.6-8. Meijboom WB, Mollet NR, Van Mieghem CAG, et al. Pre-operative computed tomography coronary angiography to detect significant coronary artery disease in patients referred for cardiac valve surgery. J Am Coll Cardiol. 2006;48:1658–65.

S3.4.6-9. Galas A, Hryniewiecki T, Kępka C, et al. May dual-source computed tomography angiography replace invasive coronary angiography in the evaluation of patients referred for valvular disease surgery? Kardiol Pol. 2012;70:877–82.

3.4.7. Exercise Testing

S3.4.7-1. Diller G-P, Dimopoulos K, Okonko D, et al. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. Circulation. 2005;112:828–35.

S3.4.7-2. Hager A, Hess J. Comparison of health related quality of life with cardiopulmonary exercise testing in adolescents and adults with congenital heart disease. Heart. 2005;91:517–20.

S3.4.7-3. Ross RM, Murthy JN, Wollak ID, et al. The six minute walk test accurately estimates mean peak oxygen uptake. BMC Pulm Med. 2010;10:31.

S3.4.7-4. Gungor H, Fatih Ayik M, Engin C, et al. Transthoracic echocardiographic and cardiopulmonary exercise testing parameters in Eisenmenger's syndrome. Association with six-minute walk test distance. Herz. 2014;39:633–7.

3.5. Transition Education

S3.5-1. Mackie AS, Islam S, Magill-Evans J, et al. Healthcare transition for youth with heart disease: a clinical trial. Heart. 2014;100:1113–8.

S3.5-2. Goossens E, Van Deyk K, Zupancic N, et al. Effectiveness of structured patient education on the knowledge level of adolescents and adults with congenital heart disease. Eur J Cardiovasc Nurs. 2014;13:63–70.

3.6. Exercise and Sports

S3.6-1. Pemberton VL, McCrindle BW, Barkin S, et al. Report of the National Heart, Lung, and Blood Institute's Working Group on obesity and other cardiovascular risk factors in congenital heart disease. Circulation. 2010;121:1153–9.

S3.6-2. Myers J, Kaykha A, George S, et al. Fitness versus physical activity patterns in predicting mortality in men. Am J Med. 2004;117:912–8.

S3.6-3. Warburton DER, Nicol CW, Bredin SSD. Health benefits of physical activity: the evidence. CMAJ. 2006;174:801–9.

S3.6-4. King AC, Taylor CB, Haskell WL, et al. Influence of regular aerobic exercise on psychological health: a randomized, controlled trial of healthy middle-aged adults. Health Psychol. 1989;8:305–24.

S3.6-5. Reybrouck T, Mertens L. Physical performance and physical activity in grown-up congenital heart disease. Eur J Cardiovasc Prev Rehabil. 2005;12:498–502.

S3.6-6. Duppen N, Takken T, Hopman MTE, et al. Systematic review of the effects of physical exercise training programmes in children and young adults with congenital heart disease. Int J Cardiol. 2013;168:1779–87.

S3.6-7. Dua JS, Cooper AR, Fox KR, et al. Exercise training in adults with congenital heart disease: feasibility and benefits. Int J Cardiol. 2010;138:196–205.

S3.6-8. Holloway TM, Chesssex C, Grace SL, et al. A call for adult congenital heart disease patient participation in cardiac rehabilitation. Int J Cardiol. 2011;150:345–6.

S3.6-9. Sandberg C, Pomeroy J, Thilén U, et al. Habitual physical activity in adults with congenital heart disease compared with age- and sex-matched controls. Can J Cardiol. 2016;32:547–53.

S3.6-10. Diller G-P, Dimopoulos K, Okonko D, et al. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. Circulation. 2005;112:828–35.

S3.6-11. Kempny A, Dimopoulos K, Uebing A, et al. Reference values for exercise limitations among adults with congenital heart disease. Relation to activities of daily life—single centre experience and review of published data. Eur Heart J. 2012;33:1386–96.

S3.6-12. Westhoff-Bleck M, Schieffer B, Tegtbur U, et al. Aerobic training in adults after atrial switch procedure for transposition of the great arteries improves exercise capacity without impairing systemic right ventricular function. Int J Cardiol. 2013;170:24–9.

S3.6-13. Tikkanen AU, Opotowsky AR, Bhatt AB, et al. Physical activity is associated with improved aerobic exercise capacity over time in adults with congenital heart disease. Int J Cardiol. 2013;168:4685–91.

3.7. Mental Health and Neurodevelopmental Issues

S3.7-1. Kovacs AH, Saidi AS, Kuhl EA, et al. Depression and anxiety in adult congenital heart disease: predictors and prevalence. Int J Cardiol. 2009;137:158–64.

S3.7-2. Kovacs AH, Sears SF, Saidi AS. Biopsychosocial experiences of adults with congenital heart disease: review of the literature. Am Heart J. 2005;150:193–201.

S3.7-3. van Rijen EHM, Utens EMWJ, Roos-Hesselink JW, et al. Psychosocial functioning of the adult with congenital heart disease: a 20-33 years follow-up. Eur Heart J. 2003;24:673–83.

S3.7-4. Callus E, Quadri E, Ricci C, et al. Update on psychological functioning in adults with congenital heart disease: a systematic review. Expert Rev Cardiovasc Ther. 2013;11:785–91.

S3.7-5. Amianto F, Bergui G, Abbate-Daga G, et al. Growing up with a congenital heart disease: neurocognitive, psychopathological and quality of life outcomes. Panminerva Med. 2011;53:109–27.

S3.7-6. Granberg M, Rydberg A, Fisher AG. Activities in daily living and schoolwork task performance in children with complex congenital heart disease. Acta Paediatr. 2008;97:1270–4.

S3.7-7. Karsdorp PA, Everaerd W, Kindt M, et al. Psychological and cognitive functioning in children and adolescents with congenital heart disease: a meta-analysis. J Pediatr Psychol. 2007;32:527–41.

S3.7-8. van der Rijken REA, Maassen BAM, Walk TLM, et al. Outcome after surgical repair of congenital cardiac malformations at school age. Cardiol Young. 2007;17:64–71.

S3.7-9. Kirshbom PM, Flynn TB, Clancy RR, et al. Late neurodevelopmental outcome after repair of total anomalous pulmonary venous connection. J Thorac Cardiovasc Surg. 2005;129:1091–7.

3.8. Endocarditis Prevention

S3.8-1. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014;129:e521–643.

S3.8-2. Verheugt CL, Uiterwaal CSPM, van der Velde ET, et al. Turning 18 with congenital heart disease: prediction of infective endocarditis based on a large population. Eur Heart J. 2011;32:1926–34.

S3.8-3. Habib G, Hoen B, Tornos P, et al. Guidelines on the prevention, diagnosis, and treatment of infective endocarditis (new version 2009): the Task Force on the Prevention, Diagnosis, and Treatment of Infective Endocarditis of the European Society of Cardiology (ESC). Eur Heart J. 2009;30:2369–413.

S3.8-4. Wilson W, Taubert KA, Gewitz M, 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.

S3.8-5. Allen U. Infective endocarditis: updated guidelines. Paediatr Child Health. 2010;15:205–12.

3.9. Concomitant Syndromes

S3.9-1. van Engelen K, Topf A, Keavney BD, et al. 22q11.2 Deletion syndrome is under-recognised in adult patients with tetralogy of Fallot and pulmonary atresia. Heart. 2010;96:621–4.

S3.9-2. Fung WLA, Chow EWC, Webb GD, et al. Extracardiac features predicting 22q11.2 deletion syndrome in adult congenital heart disease. Int J Cardiol. 2008;131:51–8.

S3.9-3. Lin AE, Basson CT, Goldmuntz E, et al. Adults with genetic syndromes and cardiovascular

abnormalities: clinical history and management. Genet Med. 2008;10:469–94.

S3.9-4. Richards AA, Garg V. Genetics of congenital heart disease. Curr Cardiol Rev. 2010;6:91–7.

S3.9-5. Kitsiou-Tzeli S, Kolialexi A, Fryssira H, et al. Detection of 22q11.2 deletion among 139 patients with Di George/velocardiofacial syndrome features. In Vivo. 2004;18:603–8.

S3.9-6. Bassett AS, Chow EWC, Husted J, et al. Clinical features of 78 adults with 22q11 deletion syndrome. Am J Med Genet A. 2005;138:307–13.

S3.9-7. Barisic I, Boban L, Greenlees R, et al. Holt Oram syndrome: a registry-based study in Europe. Orphanet J Rare Dis. 2014;9:156.

S3.9-8. Pierpont MEM, Magoulas PL, Adi S, et al. Cardio-facio-cutaneous syndrome: clinical features, diagnosis, and management guidelines. Pediatrics. 2014;134:e1149–62.

3.10. Noncardiac Medical Issues

S3.10-1. Wang A, Book WM, McConnell M, et al. Prevalence of hepatitis C infection in adult patients who underwent congenital heart surgery prior to screening in 1992. Am J Cardiol. 2007;100:1307–9.

3.11. Noncardiac Surgery

S3.11-1. Ammash NM, Connolly HM, Abel MD, et al. Noncardiac surgery in Eisenmenger syndrome. J Am Coll Cardiol. 1999;33:222–7.

S3.11-2. Maxwell BG, Posner KL, Wong JK, et al. Factors contributing to adverse perioperative events in adults with congenital heart disease: a structured analysis of cases from the closed claims project. Congenit Heart Dis. 2015;10:21–9.

S3.11-3. Maxwell BG, Wong JK, Lobato RL. Perioperative morbidity and mortality after noncardiac surgery in young adults with congenital or early acquired heart disease: a retrospective cohort analysis of the National Surgical Quality Improvement Program database. Am Surg. 2014;80:321–6.

S3.11-4. Maxwell BG, Williams GD, Ramamoorthy C. Knowledge and attitudes of anesthesia providers about noncardiac surgery in adults with congenital heart disease. Congenit Heart Dis. 2014;9:45–53.

S3.11-5. Eagle SS, Daves SM. The adult with Fontan physiology: systematic approach to perioperative management for noncardiac surgery. J Cardiothorac Vasc Anesth. 2011;25:320–34.

S3.11-6. Maxwell BG, Wong JK, Kin C, et al. Perioperative outcomes of major noncardiac surgery in adults with congenital heart disease. Anesthesiology. 2013;119:762–9.

S3.11-7. Maxwell BG, Wong JK, Sheikh AY, et al. Heart transplantation with or without prior mechanical circulatory support in adults with congenital heart disease. Eur J Cardiothorac Surg. 2014;45:842–6.

S3.11-8. Rabbitts JA, Groenewald CB, Mauermann WJ, et al. Outcomes of general anesthesia for noncardiac surgery in a series of patients with Fontan palliation. Paediatr Anaesth. 2013;23:180–7.

S3.11-9. Mylotte D, Quenneville SP, Kotowycz MA, et al. Long-term cost-effectiveness of transcatheter versus surgical closure of secundum atrial septal defect in adults. Int J Cardiol. 2014;172:109–14.

S3.11-10. Warner MA, Lunn RJ, O'Leary PW, et al. Outcomes of noncardiac surgical procedures in children and adults with congenital heart disease. Mayo Perioperative Outcomes Group. Mayo Clin Proc. 1998;73:728–34.

3.12. Pregnancy, Reproduction, and Sexual Health

3.12.1. Pregnancy

S3.12.1-1. Balint OH, Siu SC, Mason J, et al. Cardiac outcomes after pregnancy in women with congenital heart disease. Heart. 2010;96:1656–61.

S3.12.1-2. Roos-Hesselink JW, Ruys TPE, Stein JI, et al. Outcome of pregnancy in patients with structural or ischaemic heart disease: results of a registry of the European Society of Cardiology. Eur Heart J. 2013;34:657–65.

S3.12.1-3. Ouyang DW, Khairy P, Fernandes SM, et al. Obstetric outcomes in pregnant women with congenital heart disease. Int J Cardiol. 2010;144:195–9.

S3.12.1-4. Egidy Assenza G, Cassater D, Landzberg M, et al. The effects of pregnancy on right ventricular remodeling in women with repaired tetralogy of Fallot. Int J Cardiol. 2013;168:1847–52.

S3.12.1-5. Chan WS, Anand S, Ginsberg JS. Anticoagulation of pregnant women with mechanical heart valves: a systematic review of the literature. Arch Intern Med. 2000;160:191–6.

S3.12.1-6. Cutts BA, Dasgupta D, Hunt BJ. New directions in the diagnosis and treatment of pulmonary embolism in pregnancy. Am J Obstet Gynecol. 2013;208:102–8.

S3.12.1-7. Siu SC, Sermer M, Colman JM, et al. Prospective multicenter study of pregnancy outcomes in women with heart disease. Circulation. 2001;104:515–21.

S3.12.1-8. Greutmann M, gnskipK. VK, Brooks R, et al. Pregnancy outcome in women with congenital heart disease and residual haemodynamic lesions of the right ventricular outflow tract. Eur Heart J. 2010;31:1764–70.

S3.12.1-9. Gill HK, Splitt M, Sharland GK, et al. Patterns of recurrence of congenital heart disease: an analysis of 6,640 consecutive pregnancies evaluated by detailed fetal echocardiography. J Am Coll Cardiol. 2003;42:923–9.

S3.12.1-10. Ohuchi H, Tanabe Y, Kamiya C, et al. Cardiopulmonary variables during exercise predict pregnancy outcome in women with congenital heart disease. Circ J. 2013;77:470–6.

S3.12.1-11. Lui GK, Silversides CK, Khairy P, et al. Heart rate response during exercise and pregnancy outcome in women with congenital heart disease. Circulation. 2011;123:242–8.

S3.12.1-12. Siu SC, Colman JM, Sorensen S, et al. Adverse neonatal and cardiac outcomes are more common in pregnant women with cardiac disease. Circulation. 2002;105:2179–84.

S3.12.1-13. Holland BJ, Myers JA, Woods CR Jr. Prenatal diagnosis of critical congenital heart disease reduces risk of death from cardiovascular compromise prior to planned neonatal cardiac surgery: a meta-analysis. Ultrasound Obstet Gynecol. 2015;45:631–8.

3.12.2. Contraception

S3.12.2-1. Kovacs AH, Harrison JL, Colman JM, et al. Pregnancy and contraception in congenital heart disease: what women are not told. J Am Coll Cardiol. 2008;52:577–8.

S3.12.2-2. Vigl M, Kaemmerer M, Seifert-Klauss V, et al. Contraception in women with congenital heart disease. Am J Cardiol. 2010;106:1317–21.

S3.12.2-3. Kaemmerer M, Vigl M, Seifert-Klauss V, et al. Counseling reproductive health issues in women with congenital heart disease. Clin Res Cardiol. 2012;101:901–7.

S3.12.2-4. Lidegaard Ø, Løkkegaard E, Svendsen AL, et al. Hormonal contraception and risk of venous thromboembolism: national follow-up study. BMJ. 2009;339:b2890.

S3.12.2-5. Pijuan-Domènech A, Baró-Mariné F, Rojas-Torrijos M, et al. Usefulness of progesterone-only components for contraception in patients with congenital heart disease. Am J Cardiol. 2013;112:590–3.

3.13. Heart Failure and Transplant

3.13.1. Heart Failure

S3.13.1-1. Verheugt CL, Uiterwaal CSPM, van der Velde ET, et al. Mortality in adult congenital heart disease. Eur Heart J. 2010;31:1220–9.

S3.13.1-2. Zomer AC, Vaartjes I, van der Velde ET, et al. Heart failure admissions in adults with congenital heart disease; risk factors and prognosis. Int J Cardiol. 2013;168:2487–93.

S3.13.1-3. Oechslin EN, Harrison DA, Connelly MS, et al. Mode of death in adults with congenital heart disease. Am J Cardiol. 2000;86:1111–6.

S3.13.1-4. Zomer AC, Vaartjes I, Uiterwaal CSPM, et al. Circumstances of death in adult congenital heart disease. Int J Cardiol. 2012;154:168–72.

3.13.2. Heart Transplant

S3.13.2-1. Everitt MD, Donaldson AE, Stehlik J, et al. Would access to device therapies improve transplant outcomes for adults with congenital heart disease? Analysis of the United Network for Organ Sharing (UNOS). J Heart Lung Transplant. 2011;30:395–401.

S3.13.2-2. Davies RR, Russo MJ, Yang J, et al. Listing and transplanting adults with congenital heart disease. Circulation. 2011;123:759–67.

S3.13.2-3. Karamlou T, Hirsch J, Welke K, et al. A United Network for Organ Sharing analysis of heart transplantation in adults with congenital heart disease: outcomes and factors associated with mortality and retransplantation. J Thorac Cardiovasc Surg. 2010;140:161–8.

S3.13.2-4. Stout KK, Broberg CS, Book WM, et al. Chronic heart failure in congenital heart disease: a scientific statement from the American Heart Association. Circulation. 2016;133:770–801.

S3.13.2-5. Gelow JM, Song HK, Weiss JB, et al. Organ allocation in adults with congenital heart disease listed for heart transplant: impact of ventricular assist devices. J Heart Lung Transplant. 2013;32:1059–64.

S3.13.2-6. Goerler H, Simon A, Gohrbandt B, et al. Heart-lung and lung transplantation in grown-up congenital heart disease: long-term single centre experience. Eur J Cardiothorac Surg. 2007;32:926–31.

S3.13.2-7. Burchill LJ, Edwards LB, Dipchand AI, et al. Impact of adult congenital heart disease on survival and mortality after heart transplantation. J Heart Lung Transplant. 2014;33:1157–63.

S3.13.2-8. Greutmann M, Prêtre R, Furrer L, et al. Heart transplantation in adolescent and adult patients

with congenital heart disease: a case-control study. Transplant Proc. 2009;41:3821–6.

S3.13.2-9. Seddio F, Gorislavets N, Iacovoni A, et al. Is heart transplantation for complex congenital heart disease a good option? A 25-year single centre experience. Eur J Cardiothorac Surg. 2013;43:605–11.

S3.13.2-10. Alshawabkeh LI, Hu N, Carter KD, et al. Wait-list outcomes for adults with congenital heart disease listed for heart transplantation in the U.S. J Am Coll Cardiol. 2016;68:908–17.

S3.13.2-11. Krishnamurthy Y, Cooper LB, Lu D, et al. Trends and outcomes of patients with adult congenital heart disease and pulmonary hypertension listed for orthotopic heart transplantation in the United States. J Heart Lung Transplant. 2016;35:619–24.

S3.13.2-12. Harper AR, Crossland DS, Perri G, et al. Is alternative cardiac surgery an option in adults with congenital heart disease referred for thoracic organ transplantation? Eur J Cardiothorac Surg. 2013;43:344–51.

3.14. Palliative Care

S3.14-1. Greutmann M, Tobler D, Colman JM, et al. Facilitators of and barriers to advance care planning in adult congenital heart disease. Congenit Heart Dis. 2013;8:281–8.

S3.14-2. Tobler D, Greutmann M, Colman JM, et al. Knowledge of and preference for advance care planning by adults with congenital heart disease. Am J Cardiol. 2012;109:1797–800.

S3.14-3. Tobler D, Greutmann M, Colman JM, et al. End-of-life in adults with congenital heart disease: a call for early communication. Int J Cardiol. 2012;155:383–7.

3.15. Cyanosis

S3.15-1. Broberg CS, Van Woerkom RC, Swallow E, et al. Lung function and gas exchange in Eisenmenger syndrome and their impact on exercise capacity and survival. Int J Cardiol. 2014;171:73–7.

S3.15-2. Broberg CS, Uebing A, Cuomo L, et al. Adult patients with Eisenmenger syndrome report flying safely on commercial airlines. Heart. 2007;93:1599–603.

S3.15-3. Perloff JK, Child JS, Aboulhosn J, editors. Congenital Heart Disease in Adults. 3rd ed. Philadelphia, PA: Saunders/Elsevier, 2009.

3.16. Pharmacological Therapy for ACHD

S3.16-1. Yancy CW, Jessup M, Bozkurt B, et al. 2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. J Am Coll Cardiol. 2017;70:776–803.

S3.16-2. Doughan ARK, McConnell ME, Book WM. Effect of beta blockers (carvedilol or metoprolol XL) in patients with transposition of great arteries and dysfunction of the systemic right ventricle. Am J Cardiol. 2007;99:704–6.

S3.16-3. Dos L, Pujadas S, Estruch M, et al. Eplerenone in systemic right ventricle: double blind randomized clinical trial. The EVEDES Study. Int J Cardiol. 2013;168:5167–73.

S3.16-4. van der Bom T, Winter MM, Bouma BJ, et al. Effect of valsartan on systemic right ventricular function: a double-blind, randomized, placebo-controlled pilot trial. Circulation. 2013;127:322–30.

S3.16-5. Dore A, Houde C, Chan K-L, et al. Angiotensin receptor blockade and exercise capacity in adults with systemic right ventricles: a multicenter, randomized, placebo-controlled clinical trial. Circulation. 2005;112:2411–6.

S3.16-6. Babu-Narayan SV, Uebing A, Davlouros PA, et al. Randomised trial of ramipril in repaired tetralogy of Fallot and pulmonary regurgitation: the APPROPRIATE study (Ace inhibitors for Potential PRevention Of the deleterious effects of Pulmonary Regurgitation In Adults with repaired TEtralogy of Fallot). Int J Cardiol. 2012;154:299–305.

S3.16-7. Lester SJ, McElhinney DB, Viloria E, et al. Effects of losartan in patients with a systemically functioning morphologic right ventricle after atrial repair of transposition of the great arteries. Am J Cardiol. 2001;88:1314–6.

S3.16-8. Hechter SJ, Fredriksen PM, Liu P, et al. Angiotensin-converting enzyme inhibitors in adults after the Mustard procedure. Am J Cardiol. 2001;87:660–3. A11.

S3.16-9. Therrien J, Provost Y, Harrison J, et al. Effect of angiotensin receptor blockade on systemic right ventricular function and size: a small, randomized, placebo-controlled study. Int J Cardiol. 2008;129:187–92.

S3.16-10. Galiè N, Beghetti M, Gatzoulis MA, et al. Bosentan therapy in patients with Eisenmenger syndrome: a multicenter, double-blind, randomized, placebo-controlled study. Circulation. 2006;114:48–54.

S3.16-11. Giardini A, Balducci A, Specchia S, et al. Effect of sildenafil on haemodynamic response to exercise and exercise capacity in Fontan patients. Eur Heart J. 2008;29:1681–7.

S3.16-12. Hebert A, Mikkelsen UR, Thilen U, et al. Bosentan improves exercise capacity in adolescents and adults after Fontan operation: the TEMPO (Treatment With Endothelin Receptor Antagonist in Fontan Patients, a Randomized, Placebo-Controlled, Double-Blind Study Measuring Peak Oxygen Consumption) study. Circulation. 2014;130:2021–30.

S3.16-13. Goldberg DJ, French B, McBride MG, et al. Impact of oral sildenafil on exercise performance in children and young adults after the Fontan operation: a randomized, double-blind, placebo-controlled, crossover trial. Circulation. 2011;123:1185–93.

4. SPECIFIC LESIONS

4.1. Shunt Lesions

4.1.1. Atrial Septal Defect

S4.1.1-1. Oster M, Bhatt A, Zaragoza-Macias E, et al. Interventional therapy versus medical therapy for secundum atrial septal defect: a systematic review (part 2) for the 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2018 Aug 9 [E-pub ahead of print].

S4.1.1-2. Ammash NM, Seward JB, Warnes CA, et al. Partial anomalous pulmonary venous connection: diagnosis by transesophageal echocardiography. J Am Coll Cardiol. 1997;29:1351–8.

S4.1.1-3. Haramati LB, Moche IE, Rivera VT, et al. Computed tomography of partial anomalous pulmonary venous connection in adults. J Comput Assist Tomogr. 2003;27:743–9.

S4.1.1-4. Nordmeyer S, Berger F, Kuehne T, et al. Flow-sensitive four-dimensional magnetic resonance imaging facilitates and improves the accurate diagnosis of partial anomalous pulmonary venous drainage. Cardiol Young. 2011;21:528–35.

S4.1.1-5. Kardon RE, Sokoloski MC, Levi DS, et al. Transthoracic echocardiographic guidance of transcatheter atrial septal defect closure. Am J Cardiol. 2004;94:256–60.

S4.1.1-6. Zanchetta M, Rigatelli G, Pedon L, et al. Transcatheter atrial septal defect closure assisted by intracardiac echocardiography: 3-year follow-up. J Interv Cardiol. 2004;17:95–8.

S4.1.1-7. Attie F, Rosas M, Granados N, et al. Surgical treatment for secundum atrial septal defects in patients >40 years old. A randomized clinical trial. J Am Coll Cardiol. 2001;38:2035–42.

S4.1.1-8. Brochu M-C, Baril J-F, Dore A, et al. Improvement in exercise capacity in asymptomatic and mildly symptomatic adults after atrial septal defect percutaneous closure. Circulation. 2002;106:1821–6.

S4.1.1-9. Shah D, Azhar M, Oakley CM, et al. Natural history of secundum atrial septal defect in adults after medical or surgical treatment: a historical prospective study. Br Heart J. 1994;71:224–7.

S4.1.1-10. Roos-Hesselink JW, Meijboom FJ, Spitaels SEC, et al. Excellent survival and low incidence of arrhythmias, stroke and heart failure long-term after surgical ASD closure at young age. A prospective follow-up study of 21-33 years. Eur Heart J. 2003;24:190–7.

S4.1.1-11. Altındag T, Roos-Hesselink JW, Cuypers JA, et al. Transcatheter device closure of atrial septal defects in patients aged 40 years and older. Neth Heart J. 2010;18:537–42.

S4.1.1-12. Varma C, Benson LN, Silversides C, et al. Outcomes and alternative techniques for device closure of the large secundum atrial septal defect. Catheter Cardiovasc Interv. 2004;61:131–9.

S4.1.1-13. Du Z-D, Cao Q-L, Rhodes J, et al. Choice of device size and results of transcatheter closure of atrial septal defect using the amplatzer septal occluder. J Interv Cardiol. 2002;15:287–92.

S4.1.1-14. Gajjar TP, Hiremath CS, Desai NB. Surgical closure of sinus venosus atrial septal defect using a single patch—transcaval repair technique. J Card Surg. 2011;26:429–34.

S4.1.1-15. Du Z-D, Hijazi ZM, Kleinman CS, et al. Comparison between transcatheter and surgical closure of secundum atrial septal defect in children and adults: results of a multicenter nonrandomized trial. J Am Coll Cardiol. 2002;39:1836–44.

S4.1.1-16. Du Z-D, Koenig P, Cao Q-L, et al. Comparison of transcatheter closure of secundum atrial septal defect using the Amplatzer septal occluder associated with deficient versus sufficient rims. Am J Cardiol. 2002;90:865–9.

S4.1.1-17. Lopez K, Dalvi BV, Balzer D, et al. Transcatheter closure of large secundum atrial septal defects using the 40 mm Amplatzer septal occluder: results of an international registry. Catheter Cardiovasc Interv. 2005;66:580–4.

S4.1.1-18. Amin Z, Hijazi ZM, Bass JL, et al. Erosion of Amplatzer septal occluder device after closure of

secundum atrial septal defects: review of registry of complications and recommendations to minimize future risk. Catheter Cardiovasc Interv. 2004;63:496–502.

S4.1.1-19. Bradley EA, Chakinala M, Billadello JJ. Usefulness of medical therapy for pulmonary hypertension and delayed atrial septal defect closure. Am J Cardiol. 2013;112:1471–6.

S4.1.1-20. Cho YH, Jun T-G, Yang J-H, et al. Surgical strategy in patients with atrial septal defect and severe pulmonary hypertension. Heart Surg Forum. 2012;15:E111–5.

S4.1.1-21. Kouchoukos NT, Blackstone EH, Kirklin JW. Surgical implications of pulmonary hypertension in congenital heart disease. Adv Cardiol. 1978:225–31.

S4.1.1-22. Shohtsu A, Takeuchi S, Inoue T. Surgical indications and results for congenital cardiac anomalies associated with left to right shunt and severe pulmonary hypertension. Jpn Circ J. 1976;40:629–32.

4.1.2. Anomalous Pulmonary Venous Connections

S4.1.2-1. Festa P, Ait-Ali L, Cerillo AG, et al. Magnetic resonance imaging is the diagnostic tool of choice in the preoperative evaluation of patients with partial anomalous pulmonary venous return. Int J Cardiovasc Imaging. 2006;22:685–93.

S4.1.2-2. Nordmeyer S, Berger F, Kuehne T, et al. Flow-sensitive four-dimensional magnetic resonance imaging facilitates and improves the accurate diagnosis of partial anomalous pulmonary venous drainage. Cardiol Young. 2011;21:528–35.

S4.1.2-3. Dyme JL, Prakash A, Printz BF, et al. Physiology of isolated anomalous pulmonary venous connection of a single pulmonary vein as determined by cardiac magnetic resonance imaging. Am J Cardiol. 2006;98:107–10.

S4.1.2-4. Haramati LB, Moche IE, Rivera VT, et al. Computed tomography of partial anomalous pulmonary venous connection in adults. J Comput Assist Tomogr. 2003;27:743–9.

S4.1.2-5. Majdalany DS, Phillips SD, Dearani JA, et al. Isolated partial anomalous pulmonary venous connections in adults: twenty-year experience. Congenit Heart Dis. 2010;5:537–45.

S4.1.2-6. Sachweh JS, Daebritz SH, Hermanns B, et al. Hypertensive pulmonary vascular disease in adults with secundum or sinus venosus atrial septal defect. Ann Thorac Surg. 2006;81:207–13.

S4.1.2-7. Jemielity M, Perek B, Paluszkiewicz L, et al. Results of repair of partial anomalous pulmonary venous connection and sinus venosus atrial septal defect in adults. J Heart Valve Dis. 1998;7:410–4.

S4.1.2-8. Dusenbery SM, Geva T, Seale A, et al. Outcome predictors and implications for management of scimitar syndrome. Am Heart J. 2013;165:770–7.

S4.1.2-9. Brink J, Yong MS, d'Udekem Y, et al. Surgery for scimitar syndrome: the Melbourne experience. Interact Cardiovasc Thorac Surg. 2015;20:31–4.

4.1.3. Ventricular Septal Defect

S4.1.3-1. Corone P, Doyon F, Gaudeau S, et al. Natural history of ventricular septal defect. A study involving 790 cases. Circulation. 1977;55:908–15.

S4.1.3-2. Yoshimura N, Hori Y, Horii Y, et al. Comparison of magnetic resonance imaging with transthoracic echocardiography in the diagnosis of ventricular septal defect-associated coronary cusp prolapse. J Magn Reson Imaging. 2010;32:1099–103.

S4.1.3-3. Wu M-H, Wang J-K, Lin M-T, et al. Ventricular septal defect with secondary left ventricular-to-right atrial shunt is associated with a higher risk for infective endocarditis and a lower late chance of closure. Pediatrics. 2006;117:e262–7.

S4.1.3-4. D'Alto M, Romeo E, Argiento P, et al. Hemodynamics of patients developing pulmonary arterial hypertension after shunt closure. Int J Cardiol. 2013;168:3797–801.

S4.1.3-5. Janjua AM, Saleem K, Khan I, et al. Double flap patch closure of VSD with elevated pulmonary vascular resistance: an experience at AFIC/NIHD. J Coll Physicians Surg Pak. 2011;21:197–201.

S4.1.3-6. Talwar S, Keshri VK, Choudhary SK, et al. Unidirectional valved patch closure of ventricular septal defects with severe pulmonary arterial hypertension: hemodynamic outcomes. J Thorac Cardiovasc Surg. 2014;148:2570–5.

S4.1.3-7. Kouchoukos NT, Blackstone EH, Kirklin JW. Surgical implications of pulmonary hypertension in congenital heart disease. Adv Cardiol. 1978:225–31.

S4.1.3-8. Shohtsu A, Takeuchi S, Inoue T. Surgical indications and results for congenital cardiac anomalies associated with left to right shunt and severe pulmonary hypertension. Jpn Circ J. 1976;40:629–32.

S4.1.3-9. Cartmill TB, DuShane JW, McGoon DC, et al. Results of repair of ventricular septal defect. J Thorac Cardiovasc Surg. 1966;52:486–501.

4.1.4. Atrioventricular Septal Defect

S4.1.4-1. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014;129:e521–643.

S4.1.4-2. Bianchi G, Bevilacqua S, Solinas M, et al. In adult patients undergoing redo surgery for left atrioventricular valve regurgitation after atrioventricular septal defect correction, is replacement superior to repair? Interact Cardiovasc Thorac Surg. 2011;12:1033–9.

S4.1.4-3. Hoohenkerk GJF, Bruggemans EF, Koolbergen DR, et al. Long-term results of reoperation for left atrioventricular valve regurgitation after correction of atrioventricular septal defects. Ann Thorac Surg. 2012;93:849–55.

S4.1.4-4. Stulak JM, Burkhart HM, Dearani JA. Reoperations after repair of partial and complete atrioventricular septal defect. World J Pediatr Congenit Heart Surg. 2010;1:97–104.

S4.1.4-5. Kouchoukos NT, Blackstone EH, Kirklin JW. Surgical implications of pulmonary hypertension in congenital heart disease. Adv Cardiol. 1978:225–31.

S4.1.4-6. Shohtsu A, Takeuchi S, Inoue T. Surgical indications and results for congenital cardiac anomalies associated with left to right shunt and severe pulmonary hypertension. Jpn Circ J. 1976;40:629–32.

4.1.5. Patent Ductus Arteriosus

S4.1.5-1. Yan C, Zhao S, Jiang S, et al. Transcatheter closure of patent ductus arteriosus with severe pulmonary arterial hypertension in adults. Heart. 2007;93:514–8.

S4.1.5-2. Gamboa R, Rios-Méndez RE, Mollón FP, et al. Percutaneous closure of patent ductus arteriosus in adults using different devices. Rev Esp Cardiol. 2010;63:726–9.

S4.1.5-3. Jeong Y-H, Yun T-J, Song J-M, et al. Left ventricular remodeling and change of systolic function after closure of patent ductus arteriosus in adults: device and surgical closure. Am Heart J. 2007;154:436–40.

S4.1.5-4. Zabal C, García-Montes JA, Buendía-Hernández A, et al. Percutaneous closure of hypertensive ductus arteriosus. Heart. 2010;96:625–9.

S4.1.5-5. Kouchoukos NT, Blackstone EH, Kirklin JW. Surgical implications of pulmonary hypertension in congenital heart disease. Adv Cardiol. 1978:225–31.

4.2. Left-Sided Obstructive Lesions

4.2.1. Cor Triatriatum

S4.2.1-1. Ozyuksel A, Yildirim O, Avsar M, et al. Surgical correction of cor triatriatum sinister in the paediatric population: mid-term results in 15 cases. Eur J Cardiothorac Surg. 2015;47:e25–8.

S4.2.1-2. Yaroglu Kazanci S, Emani S, McElhinney DB. Outcome after repair of cor triatriatum. Am J Cardiol. 2012;109:412–6.

S4.2.1-3. Saxena P, Burkhart HM, Schaff HV, et al. Surgical repair of cor triatriatum sinister: the Mayo Clinic 50-year experience. Ann Thorac Surg. 2014;97:1659–63.

4.2.2. Congenital Mitral Stenosis

S4.2.2-1. Schaverien MV, Freedom RM, McCrindle BW. Independent factors associated with outcomes of parachute mitral valve in 84 patients. Circulation. 2004;109:2309–13.

S4.2.2-2. Marino BS, Kruge LE, Cho CJ, et al. Parachute mitral valve: morphologic descriptors, associated lesions, and outcomes after biventricular repair. J Thorac Cardiovasc Surg. 2009;137:385–93.e4.

4.2.3. Subaortic Stenosis

S4.2.3-1. Diller G-P, Dimopoulos K, Okonko D, et al. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. Circulation. 2005;112:828–35.

S4.2.3-2. Diller G-P, Dimopoulos K, Okonko D, et al. Heart rate response during exercise predicts survival in adults with congenital heart disease. J Am Coll Cardiol. 2006;48:1250–6.

S4.2.3-3. Trinchero R, Demarie D, Orzan F, et al. Fixed subaortic stenosis. Natural history of patients with mild obstruction and follow-up of operated patients. G Ital Cardiol. 1988;18:738–44.

S4.2.3-4. Oliver JM, González A, Gallego P, et al. Discrete subaortic stenosis in adults: increased prevalence and slow rate of progression of the obstruction and aortic regurgitation. J Am Coll Cardiol. 2001;38:835–42.

S4.2.3-5. Brauner R, Laks H, Drinkwater DCJ, et al. Benefits of early surgical repair in fixed subaortic stenosis. J Am Coll Cardiol. 1997;30:1835–42.

S4.2.3-6. Parry AJ, Kovalchin JP, Suda K, et al. Resection of subaortic stenosis; can a more aggressive approach be justified? Eur J Cardiothorac Surg. 1999;15:631–8.

4.2.4. Congenital Valvular Aortic Stenosis

S4.2.4-1. Fernandes SM, Sanders SP, Khairy P, et al. Morphology of bicuspid aortic valve in

children and adolescents. J Am Coll Cardiol. 2004; 44:1648–51.

S4.2.4-2. Biner S, Rafique AM, Ray I, et al. Aortopathy is prevalent in relatives of bicuspid aortic valve patients. J Am Coll Cardiol. 2009;53:2288–95.

S4.2.4-3. Tutar E, Ekici F, Atalay S, et al. The prevalence of bicuspid aortic valve in newborns by echocardiographic screening. Am Heart J. 2005;150:513–5.

S4.2.4-4. Huntington K, Hunter AG, Chan KL. A prospective study to assess the frequency of familial clustering of congenital bicuspid aortic valve. J Am Coll Cardiol. 1997;30:1809–12.

S4.2.4-5. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014;129:e521–643.

S4.2.4-6. Arora R, Jolly N, Bhat A, et al. Follow-up of balloon aortic valvuloplasty in young adults—a combined hemodynamic and Doppler echocardiographic study. Indian Heart J. 1989;41:314–7.

4.2.4.1. Turner Syndrome

S4.2.4.1-1. Sybert VP. Cardiovascular malformations and complications in Turner syndrome. Pediatrics. 1998;101:E11.

S4.2.4.1-2. Carlson M, Airhart N, Lopez L, et al. Moderate aortic enlargement and bicuspid aortic valve are associated with aortic dissection in Turner syndrome: report of the International Turner Syndrome Aortic Dissection Registry. Circulation. 2012;126:2220–6.

4.2.5. Supravalvular Aortic Stenosis

S4.2.5-1. Tani LY, Minich LL, Pagotto LT, et al. Usefulness of doppler echocardiography to determine the timing of surgery for supravalvar aortic stenosis. Am J Cardiol. 2000;86:114–6.

S4.2.5-2. Thiene G, Ho SY. Aortic root pathology and sudden death in youth: review of anatomical varieties. Appl Pathol. 1986;4:237–45.

S4.2.5-3. Stamm C, Li J, Ho SY, et al. The aortic root in supravalvular aortic stenosis: the potential surgical relevance of morphologic findings. J Thorac Cardiovasc Surg. 1997;114:16–24.

S4.2.5-4. Martin MM, Lemmer JH Jr., Shaffer E, et al. Obstruction to left coronary artery blood flow secondary to obliteration of the coronary ostium in supravalvular aortic stenosis. Ann Thorac Surg. 1988; 45:16–20.

S4.2.5-5. Bruno E, Rossi N, Thüer O, et al. Cardiovascular findings, and clinical course, in patients with Williams syndrome. Cardiol Young. 2003;13:532–6.

S4.2.5-6. Wren C, Oslizlok P, Bull C. Natural history of supravalvular aortic stenosis and pulmonary artery stenosis. J Am Coll Cardiol. 1990;15:1625–30.

S4.2.5-7. Eronen M, Peippo M, Hiippala A, et al. Cardiovascular manifestations in 75 patients with Williams syndrome. J Med Genet. 2002;39:554–8.

S4.2.5-8. Greutmann M, Tobler D, Sharma NC, et al. Cardiac outcomes in adults with supravalvar aortic stenosis. Eur Heart J. 2012;33:2442–50.

S4.2.5-9. Doty DB, Eastham CL, Hiratzka LF, et al. Determination of coronary reserve in patients with supravalvular aortic stenosis. Circulation. 1982;66: I186–92.

4.2.6. Coarctation of the Aorta

S4.2.6-1. Qureshi AM, McElhinney DB, Lock JE, et al. Acute and intermediate outcomes, and evaluation of injury to the aortic wall, as based on 15 years experience of implanting stents to treat aortic coarctation. Cardiol Young. 2007;17:307–18.

S4.2.6-2. Hager A, Kanz S, Kaemmerer H, et al. Coarctation Long-term Assessment (COALA): significance of arterial hypertension in a cohort of 404 patients up to 27 years after surgical repair of isolated coarctation of the aorta, even in the absence of restenosis and prosthetic material. J Thorac Cardiovasc Surg. 2007;134:738–45.

S4.2.6-3. Forbes TJ, Garekar S, Amin Z, et al. Procedural results and acute complications in stenting native and recurrent coarctation of the aorta in patients over 4 years of age: a multi-institutional study. Catheter Cardiovasc Interv. 2007;70:276–85.

S4.2.6-4. Morgan GJ, Lee KJ, Chaturvedi R, et al. Systemic blood pressure after stent management for arch coarctation implications for clinical care. J Am Coll Cardiol Intv. 2013;6:192–201.

S4.2.6-5. Cook SC, Hickey J, Maul TM, et al. Assessment of the cerebral circulation in adults with coarctation of the aorta. Congenit Heart Dis. 2013;8:289–95.

S4.2.6-6. Connolly HM, Huston J 3rd, Brown RD Jr., et al. Intracranial aneurysms in patients with coarctation of the aorta: a prospective magnetic resonance angiographic study of 100 patients. Mayo Clin Proc. 2003;78:1491–9.

S4.2.6-7. Correia AS, Gonçalves A, Paiva M, et al. Long-term follow-up after aortic coarctation repair: the unsolved issue of exercise-induced hypertension. Rev Port Cardiol. 2013;32:879–83.

S4.2.6-8. Holzer R, Qureshi S, Ghasemi A, et al. Stenting of aortic coarctation: acute, intermediate, and long-term results of a prospective multi-institutional registry—Congenital Cardiovascular Interventional Study Consortium (CCISC). Catheter Cardiovasc Interv. 2010;76:553–63.

S4.2.6-9. Chessa M, Carrozza M, Butera G, et al. Results and mid-long-term follow-up of stent implantation for native and recurrent coarctation of the aorta. Eur Heart J. 2005;26:2728–32.

S4.2.6-10. Toro-Salazar OH, Steinberger J, Thomas W, et al. Long-term follow-up of patients after coarctation of the aorta repair. Am J Cardiol. 2002;89:541–7.

S4.2.6-11. Carr JA. The results of catheter-based therapy compared with surgical repair of adult aortic coarctation. J Am Coll Cardiol. 2006;47:1101–7.

S4.2.6-12. Brown JW, Ruzmetov M, Hoyer MH, et al. Recurrent coarctation: is surgical repair of recurrent coarctation of the aorta safe and effective? Ann Thorac Surg. 2009;88:1923–30.

S4.2.6-13. Chobanian AV, Bakris GL, Black HR, et al. Seventh report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure. Hypertension. 2003;42:1206–52.

S4.2.6-14. Suárez de Lezo J, Pan M, Romero M, et al. Percutaneous interventions on severe coarctation of the aorta: a 21-year experience. Pediatr Cardiol. 2005; 26:176–89.

4.3. Right-Sided Lesions

4.3.1. Valvular Pulmonary Stenosis

S4.3.1-1. Voet A, Rega F, de Bruaene AV, et al. Long-term outcome after treatment of isolated pulmonary valve stenosis. Int J Cardiol. 2012;156:11–5.

S4.3.1-2. Kaul UA, Singh B, Tyagi S, et al. Long-term results after balloon pulmonary valvuloplasty in adults. Am Heart J. 1993;126:1152–5.

S4.3.1-3. Chen CR, Cheng TO, Huang T, et al. Percutaneous balloon valvuloplasty for pulmonic stenosis in adolescents and adults. N Engl J Med. 1996;335:21–5.

S4.3.1-4. Taggart NW, Cetta F, Cabalka AK, et al. Outcomes for balloon pulmonary valvuloplasty in adults: comparison with a concurrent pediatric cohort. Catheter Cardiovasc Interv. 2013;82:811–5.

S4.3.1-5. Kopecky SL, Gersh BJ, McGoon MD, et al. Long-term outcome of patients undergoing surgical repair of isolated pulmonary valve stenosis. Follow-up at 20-30 years. Circulation. 1988;78:1150–6.

S4.3.1-6. Hayes CJ, Gersony WM, Driscoll DJ, et al. Second natural history study of congenital heart defects. Results of treatment of patients with pulmonary valvar stenosis. Circulation. 1993;87:I28–37.

S4.3.1-7. Roos-Hesselink JW, Meijboom FJ, Spitaels SEC, et al. Long-term outcome after surgery for pulmonary stenosis (a longitudinal study of 22-33 years). Eur Heart J. 2006;27:482–8.

S4.3.1-8. Earing MG, Connolly HM, Dearani JA, et al. Long-term follow-up of patients after surgical treatment for isolated pulmonary valve stenosis. Mayo Clin Proc. 2005;80:871–6.

4.3.2. Branch and Peripheral Pulmonary Stenosis

S4.3.2-1. Hallbergson A, Lock JE, Marshall AC. Frequency and risk of in-stent stenosis following pulmonary artery stenting. Am J Cardiol. 2014;113:541–5.

S4.3.2-2. Gonzalez I, Kenny D, Slyder S, et al. Medium and long-term outcomes after bilateral pulmonary artery stenting in children and adults with congenital heart disease. Pediatr Cardiol. 2013;34:179–84.

S4.3.2-3. Kenny D, Amin Z, Slyder S, et al. Medium-term outcomes for peripheral pulmonary artery stenting in adults with congenital heart disease. J Interv Cardiol. 2011;24:373–7.

4.3.3. Double-Chambered Right Ventricle

S4.3.3-1. Hachiro Y, Takagi N, Koyanagi T, et al. Repair of double-chambered right ventricle: surgical results and long-term follow-up. Ann Thorac Surg. 2001;72: 1520–2.

S4.3.3-2. Kveselis D, Rosenthal A, Ferguson P, et al. Long-term prognosis after repair of double-chamber right ventricle with ventricular septal defect. Am J Cardiol. 1984;54:1292–5.

S4.3.3-3. Said SM, Burkhart HM, Dearani JA, et al. Outcomes of surgical repair of double-chambered right ventricle. Ann Thorac Surg. 2012;93:197–200.

S4.3.3-4. Kahr PC, Alonso-Gonzalez R, Kempny A, et al. Long-term natural history and postoperative outcome of double-chambered right ventricle—

experience from two tertiary adult congenital heart centres and review of the literature. Int J Cardiol. 2014;174:662–8.

4.3.4. Ebstein Anomaly

S4.3.4-1. Bharucha T, Anderson RH, Lim ZS, et al. Multiplanar review of three-dimensional echocardiography gives new insights into the morphology of Ebstein's malformation. Cardiol Young. 2010;20:49–53.

S4.3.4-2. Attenhofer Jost CH, Edmister WD, Julsrud PR, et al. Prospective comparison of echocardiography versus cardiac magnetic resonance imaging in patients with Ebstein's anomaly. Int J Cardiovasc Imaging. 2012;28:1147–59.

S4.3.4-3. Wei W, Zhan X, Xue Y, et al. Features of accessory pathways in adult Ebstein's anomaly. Europace. 2014;16:1619–25.

S4.3.4-4. Iturralde P, Nava S, Sálica G, et al. Electrocardiographic characteristics of patients with Ebstein's anomaly before and after ablation of an accessory atrioventricular pathway. J Cardiovasc Electrophysiol. 2006;17:1332–6.

S4.3.4-5. Shivapour JKL, Sherwin ED, Alexander ME, et al. Utility of preoperative electrophysiologic studies in patients with Ebstein's anomaly undergoing the cone procedure. Heart Rhythm. 2014;11:182–6.

S4.3.4-6. Raju V, Dearani JA, Burkhart HM, et al. Right ventricular unloading for heart failure related to Ebstein malformation. Ann Thorac Surg. 2014;98:167–73.

S4.3.4-7. Brown ML, Dearani JA, Danielson GK, et al. The outcomes of operations for 539 patients with Ebstein anomaly. J Thorac Cardiovasc Surg. 2008;135:1120–36. e1-7.

S4.3.4-8. Dearani JA, Said SM, Burkhart HM, et al. Strategies for tricuspid re-repair in Ebstein malformation using the cone technique. Ann Thorac Surg. 2013;96:202–8.

S4.3.4-9. Dearani JA, Said SM, O'Leary PW, et al. Anatomic repair of Ebstein's malformation: lessons learned with cone reconstruction. Ann Thorac Surg. 2013;95:220–6.

S4.3.4-10. Badiu CC, Schreiber C, Hörer J, et al. Early timing of surgical intervention in patients with Ebstein's anomaly predicts superior long-term outcome. Eur J Cardiothorac Surg. 2010;37:186–92.

S4.3.4-11. Khositseth A, Danielson GK, Dearani JA, et al. Supraventricular tachyarrhythmias in Ebstein anomaly: management and outcome. J Thorac Cardiovasc Surg. 2004;128:826–33.

S4.3.4-12. Zachariah JP, Walsh EP, Triedman JK, et al. Multiple accessory pathways in the young: the impact of structural heart disease. Am Heart J. 2013;165:87–92.

S4.3.4-13. Chauvaud S. Ebstein's malformation. Surgical treatment and results. Thorac Cardiovasc Surg. 2000;48:220–3.

S4.3.4-14. Chen JM, Mosca RS, Altmann K, et al. Early and medium-term results for repair of Ebstein anomaly. J Thorac Cardiovasc Surg. 2004;127:990–8.

S4.3.4-15. Malhotra SP, Petrossian E, Reddy VM, et al. Selective right ventricular unloading and novel technical concepts in Ebstein's anomaly. Ann Thorac Surg. 2009;88:1975–81.

4.3.5. Tetralogy of Fallot

S4.3.5-1. Knauth AL, Gauvreau K, Powell AJ, et al. Ventricular size and function assessed by cardiac MRI predict major adverse clinical outcomes late after tetralogy of Fallot repair. Heart. 2008;94:211–6.

S4.3.5-2. Morray BH, McElhinney DB, Cheatham JP, et al. Risk of coronary artery compression among patients referred for transcatheter pulmonary valve implantation: a multicenter experience. Circ Cardiovasc Interv. 2013;6:535–42.

S4.3.5-3. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease. Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Heart Rhythm. 2014;11:e102–65.

S4.3.5-4. Dietl CA, Cazzaniga ME, Dubner SJ, et al. Life-threatening arrhythmias and RV dysfunction after surgical repair of tetralogy of Fallot. Comparison between transventricular and transatrial approaches. Circulation. 1994;90:II7–12.

S4.3.5-5. Khairy P, Landzberg MJ, Gatzoulis MA, et al. Value of programmed ventricular stimulation after tetralogy of Fallot repair: a multicenter study. Circulation. 2004;109:1994–2000.

S4.3.5-6. Tsai SF, Chan DP, Ro PS, et al. Rate of inducible ventricular arrhythmia in adults with congenital heart disease. Am J Cardiol. 2010;106:730–6.

S4.3.5-7. Khairy P. Programmed ventricular stimulation for risk stratification in patients with tetralogy of Fallot: a Bayesian perspective. Nat Clin Pract Cardiovasc Med. 2007;4:292–3.

S4.3.5-8. Lucron H, Marçon F, Bosser G, et al. Induction of sustained ventricular tachycardia after surgical repair of tetralogy of Fallot. Am J Cardiol. 1999;83:1369–73.

S4.3.5-9. Ferraz Cavalcanti PE, Sá MPBO, Santos CA, et al. Pulmonary valve replacement after operative repair of tetralogy of Fallot: meta-analysis and meta-regression of 3,118 patients from 48 studies. J Am Coll Cardiol. 2013;62:2227–43.

S4.3.5-10. Gengsakul A, Harris L, Bradley TJ, et al. The impact of pulmonary valve replacement after tetralogy of Fallot repair: a matched comparison. Eur J Cardiothorac Surg. 2007;32:462–8.

S4.3.5-11. Discigil B, Dearani JA, Puga FJ, et al. Late pulmonary valve replacement after repair of tetralogy of Fallot. J Thorac Cardiovasc Surg. 2001;121:344–51.

S4.3.5-12. Oosterhof T, van Straten A, Vliegen HW, et al. Preoperative thresholds for pulmonary valve replacement in patients with corrected tetralogy of Fallot using cardiovascular magnetic resonance. Circulation. 2007;116:545–51.

S4.3.5-13. Lee C, Kim YM, Lee C-H, et al. Outcomes of pulmonary valve replacement in 170 patients with chronic pulmonary regurgitation after relief of right ventricular outflow tract obstruction: implications for optimal timing of pulmonary valve replacement. J Am Coll Cardiol. 2012;60:1005–14.

S4.3.5-14. Geva T. Indications for pulmonary valve replacement in repaired tetralogy of Fallot: the quest continues. Circulation. 2013;128:1855–7.

S4.3.5-15. Koyak Z, de Groot JR, Bouma BJ, et al. Symptomatic but not asymptomatic non-sustained ventricular tachycardia is associated with appropriate implantable cardioverter therapy in tetralogy of Fallot. Int J Cardiol. 2013;167:1532–5.

S4.3.5-16. Khairy P, Harris L, Landzberg MJ, et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. Circulation. 2008;117:363–70.

S4.3.5-17. Witte KK, Pepper CB, Cowan JC, et al. Implantable cardioverter-defibrillator therapy in adult patients with tetralogy of Fallot. Europace. 2008;10:926–30.

4.3.6. Right Ventricle-to-Pulmonary Artery Conduit

S4.3.6-1. Morray BH, McElhinney DB, Cheatham JP, et al. Risk of coronary artery compression among patients referred for transcatheter pulmonary valve implantation: a multicenter experience. Circ Cardiovasc Interv. 2013;6:535–42.

S4.3.6-2. Fraisse A, Assaidi A, Mauri L, et al. Coronary artery compression during intention to treat right ventricle outflow with percutaneous pulmonary valve implantation: incidence, diagnosis, and outcome. Catheter Cardiovasc Interv. 2014;83:E260–8.

S4.3.6-3. McElhinney DB, Benson LN, Eicken A, et al. Infective endocarditis after transcatheter pulmonary valve replacement using the Melody valve: combined results of 3 prospective North American and European studies. Circ Cardiovasc Interv. 2013;6:292–300.

S4.3.6-4. McElhinney DB, Cheatham JP, Jones TK, et al. Stent fracture, valve dysfunction, and right ventricular outflow tract reintervention after transcatheter pulmonary valve implantation: patient-related and procedural risk factors in the US Melody Valve Trial. Circ Cardiovasc Interv. 2011;4:602–14.

S4.3.6-5. Mainwaring RD, Pirolli T, Punn R, et al. Late repair of the native pulmonary valve in patients with pulmonary insufficiency after surgery for tetralogy of Fallot. Ann Thorac Surg. 2012;93:677–9.

S4.3.6-6. Bauer BS, Zachariah S, Levi D, et al. Evaluation of peak pressure gradients in patients after Melody valve implantation: a comparison of cardiac catheterization and doppler echocardiography. Echocardiography. 2015;32.

S4.3.6-7. McElhinney DB, Hellenbrand WE, Zahn EM, et al. Short- and medium-term outcomes after transcatheter pulmonary valve placement in the expanded multicenter US Melody valve trial. Circulation. 2010;122:507–16.

S4.3.6-8. Niemantsverdriet MBA, Ottenkamp J, Gauvreau K, et al. Determinants of right ventricular outflow tract conduit longevity: a multinational analysis. Congenit Heart Dis. 2008;3:176–84.

S4.3.6-9. Batra AS, McElhinney DB, Wang W, et al. Cardiopulmonary exercise function among patients undergoing transcatheter pulmonary valve implantation in the US Melody valve investigational trial. Am Heart J. 2012;163:280–7.

S4.3.6-10. Zahn EM, Hellenbrand WE, Lock JE, et al. Implantation of the Melody transcatheter pulmonary valve in patients with a dysfunctional right ventricular outflow tract conduit early results from the U.S. clinical trial. J Am Coll Cardiol. 2009;54:1722–9.

S4.3.6-11. Brown JW, Ruzmetov M, Rodefeld MD, et al. Right ventricular outflow tract reconstruction in Ross

patients: does the homograft fare better? Ann Thorac Surg. 2008;86:1607–12.

S4.3.6-12. Frigiola A, Tsang V, Nordmeyer J, et al. Current approaches to pulmonary regurgitation. Eur J Cardiothorac Surg. 2008;34:576–80.

S4.3.6-13. Lurz P, Nordmeyer J, Giardini A, et al. Early versus late functional outcome after successful percutaneous pulmonary valve implantation: are the acute effects of altered right ventricular loading all we can expect? J Am Coll Cardiol. 2011;57:724–31.

S4.3.6-14. Coats L, Khambadkone S, Derrick G, et al. Physiological and clinical consequences of relief of right ventricular outflow tract obstruction late after repair of congenital heart defects. Circulation. 2006; 113:2037–44.

4.4. Complex Lesions

4.4.1. Transposition of the Great Arteries

4.4.1.1. Transposition of the Great Arteries With Atrial Switch

S4.4.1.1-1. Khairy P, Landzberg MJ, Gatzoulis MA, et al. Transvenous pacing leads and systemic thromboemboli in patients with intracardiac shunts: a multicenter study. Circulation. 2006;113:2391–7.

S4.4.1.1-2. Khairy P, Harris L, Landzberg MJ, et al. Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study. Circ Arrhythm Electrophysiol. 2008;1:250–7.

4.4.1.2. Transposition of the Great Arteries With Arterial Switch

S4.4.1.2-1. Khairy P, Clair M, Fernandes SM, et al. Cardiovascular outcomes after the arterial switch operation for D-transposition of the great arteries. Circulation. 2013;127:331–9.

S4.4.1.2-2. Kempny A, Wustmann K, Borgia F, et al. Outcome in adult patients after arterial switch operation for transposition of the great arteries. Int J Cardiol. 2013;167:2588–93.

S4.4.1.2-3. Lim H-G, Kim W-H, Lee JR, et al. Long-term results of the arterial switch operation for ventriculo-arterial discordance. Eur J Cardiothorac Surg. 2013;43:325–34.

S4.4.1.2-4. Ou P, Khraiche D, Celermajer DS, et al. Mechanisms of coronary complications after the arterial switch for transposition of the great arteries. J Thorac Cardiovasc Surg. 2013;145:1263–9.

S4.4.1.2-5. Legendre A, Losay J, Touchot-Koné A, et al. Coronary events after arterial switch operation for transposition of the great arteries. Circulation. 2003;108 suppl 1:II186–90.

S4.4.1.2-6. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014;129:e521–643.

S4.4.1.2-7. Fihn SD, Gardin JM, Abrams J, et al. 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS guideline for the diagnosis and management of patients with stable ischemic heart disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, and the American College of Physicians, American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. Circulation. 2012;126:e354–471.

S4.4.1.2-8. Fihn SD, Blankenship JC, Alexander KP, et al. 2014 ACC/AHA/AATS/PCNA/SCAI/STS focused update of the guideline for the diagnosis and management of patients with stable ischemic heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, and the American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2014;64:1929–49.

4.4.1.3. Congenitally Corrected Transposition of the Great Arteries

S4.4.1.3-1. Lewis M, Ginns J, Rosenbaum M. Is systemic right ventricular function by cardiac MRI related to the degree of tricuspid regurgitation in congenitally corrected transposition of the great arteries? Int J Cardiol. 2014;174:586–9.

S4.4.1.3-2. Grewal J, Crean A, Garceau P, et al. Subaortic right ventricular characteristics and relationship to exercise capacity in congenitally corrected transposition of the great arteries. J Am Soc Echocardiogr. 2012;25:1215–21.

S4.4.1.3-3. Mongeon F-P, Connolly HM, Dearani JA, et al. Congenitally corrected transposition of the great arteries ventricular function at the time of systemic atrioventricular valve replacement predicts long-term ventricular function. J Am Coll Cardiol. 2011;57: 2008–17.

S4.4.1.3-4. Prieto LR, Hordof AJ, Secic M, et al. Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries. Circulation. 1998;98:997–1005.

S4.4.1.3-5. Buber J, McElhinney DB, Valente AM, et al. Tricuspid valve regurgitation in congenitally corrected transposition of the great arteries and a left ventricle to pulmonary artery conduit. Ann Thorac Surg. 2015; 99:1348–56.

4.4.2. Fontan Palliation of Single Ventricle Physiology (Including Tricuspid Atresia and Double Inlet Left Ventricle)

S4.4.2-1. Khairy P, Fernandes SM, Mayer JE Jr., et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation. 2008;117:85–92.

S4.4.2-2. Gatzoulis MA, Munk MD, Williams WG, et al. Definitive palliation with cavopulmonary or aortopulmonary shunts for adults with single ventricle physiology. Heart. 2000;83:51–7.

S4.4.2-3. Cordina R, O'Meagher S, Gould H, et al. Skeletal muscle abnormalities and exercise capacity in adults with a Fontan circulation. Heart. 2013;99:1530–4.

S4.4.2-4. Dulfer K, Duppen N, Blom NA, et al. Effects of exercise training on behavioral and emotional problems in adolescents with tetralogy of Fallot or a Fontan circulation: a randomized controlled trial. Int J Cardiol. 2014;172:e425–7.

S4.4.2-5. Inai K, Saita Y, Takeda S, et al. Skeletal muscle hemodynamics and endothelial function in patients after Fontan operation. Am J Cardiol. 2004; 93:792–7.

S4.4.2-6. Kiesewetter CH, Sheron N, Vettukattill JJ, et al. Hepatic changes in the failing Fontan circulation. Heart. 2007;93:579–84.

S4.4.2-7. Seddio F, Gorislavets N, Iacovoni A, et al. Is heart transplantation for complex congenital heart disease a good option? A 25-year single centre experience. Eur J Cardiothorac Surg. 2013;43:605–11.

S4.4.2-8. Hebson CL, McCabe NM, Elder RW, et al. Hemodynamic phenotype of the failing Fontan in an adult population. Am J Cardiol. 2013;112:1943–7.

S4.4.2-9. Davies RR, Sorabella RA, Yang J, et al. Outcomes after transplantation for "failed" Fontan: a single-institution experience. J Thorac Cardiovasc Surg. 2012;143:1183–1192, e4.

S4.4.2-10. Gamba A, Merlo M, Fiocchi R, et al. Heart transplantation in patients with previous Fontan operations. J Thorac Cardiovasc Surg. 2004;127:555–62.

S4.4.2-11. Jayakumar KA, Addonizio LJ, Kichuk-Chrisant MR, et al. Cardiac transplantation after the Fontan or Glenn procedure. J Am Coll Cardiol. 2004; 44:2065–72.

S4.4.2-12. Backer CL, Russell HM, Pahl E, et al. Heart transplantation for the failing Fontan. Ann Thorac Surg. 2013;96:1413–9.

S4.4.2-13. Triedman JK. Arrhythmias in adults with congenital heart disease. Heart. 2002;87:383–9.

S4.4.2-14. Yap S-C, Harris L, Silversides CK, et al. Outcome of intra-atrial re-entrant tachycardia catheter ablation in adults with congenital heart disease: negative impact of age and complex atrial surgery. J Am Coll Cardiol. 2010;56:1589–96.

S4.4.2-15. de Groot NMS, Atary JZ, Blom NA, et al. Long-term outcome after ablative therapy of postoperative atrial tachyarrhythmia in patients with congenital heart disease and characteristics of atrial tachyarrhythmia recurrences. Circ Arrhythm Electrophysiol. 2010;3:148–54.

S4.4.2-16. Aboulhosn J, Williams R, Shivkumar K, et al. Arrhythmia recurrence in adult patients with single ventricle physiology following surgical Fontan conversion. Congenit Heart Dis. 2010;5:430–4.

S4.4.2-17. Mavroudis C, Deal BJ, Backer CL, et al. J. Maxwell Chamberlain Memorial Paper for congenital heart surgery. 111 Fontan conversions with arrhythmia surgery: surgical lessons and outcomes. Ann Thorac Surg. 2007;84:1457–65.

S4.4.2-18. Takahashi K, Fynn-Thompson F, Cecchin F, et al. Clinical outcomes of Fontan conversion surgery with and without associated arrhythmia intervention. Int J Cardiol. 2009;137:260–6.

S4.4.2-19. Hebert A, Mikkelsen UR, Thilen U, et al. Bosentan improves exercise capacity in adolescents and adults after Fontan operation: the TEMPO (Treatment With Endothelin Receptor Antagonist in Fontan Patients, a Randomized, Placebo-Controlled, Double-Blind Study Measuring Peak Oxygen Consumption) study. Circulation. 2014;130:2021–30.

S4.4.2-20. Giardini A, Balducci A, Specchia S, et al. Effect of sildenafil on haemodynamic response to exercise and exercise capacity in Fontan patients. Eur Heart J. 2008;29:1681–7.

S4.4.2-21. Goldberg DJ, French B, McBride MG, et al. Impact of oral sildenafil on exercise performance in children and young adults after the Fontan operation: a randomized, double-blind, placebo-controlled, crossover trial. Circulation. 2011;123:1185–93.

S4.4.2-22. Ovaert C, Thijs D, Dewolf D, et al. The effect of bosentan in patients with a failing Fontan circulation. Cardiol Young. 2009;19:331–9.

S4.4.2-23. Bowater SE, Weaver RA, Thorne SA, et al. The safety and effects of bosentan in patients with a Fontan circulation. Congenit Heart Dis. 2012;7:243–9.

S4.4.2-24. Derk G, Houser L, Miner P, et al. Efficacy of endothelin blockade in adults with Fontan physiology. Congenit Heart Dis. 2015;10:E11–6.

S4.4.2-25. Schuuring MJ, Vis JC, van Dijk APJ, et al. Impact of bosentan on exercise capacity in adults after the Fontan procedure: a randomized controlled trial. Eur J Heart Fail. 2013;15:690–8.

S4.4.2-26. Potter BJ, Leong-Sit P, Fernandes SM, et al. Effect of aspirin and warfarin therapy on thromboembolic events in patients with univentricular hearts and Fontan palliation. Int J Cardiol. 2013;168:3940–3.

S4.4.2-27. Harper AR, Crossland DS, Perri G, et al. Is alternative cardiac surgery an option in adults with congenital heart disease referred for thoracic organ transplantation? Eur J Cardiothorac Surg. 2013;43:344–51.

4.4.4. Severe PAH and Eisenmenger Syndrome

4.4.4.1. Severe PAH

S4.4.4.1-1. D'Alto M, Romeo E, Argiento P, et al. Hemodynamics of patients developing pulmonary arterial hypertension after shunt closure. Int J Cardiol. 2013;168:3797–801.

S4.4.4.1-2. Barst RJ, Ivy DD, Foreman AJ, et al. Four- and seven-year outcomes of patients with congenital heart disease-associated pulmonary arterial hypertension (from the REVEAL Registry). Am J Cardiol. 2014;113:147–55.

S4.4.4.1-3. Duffels MGJ, Engelfriet PM, Berger RMF, et al. Pulmonary arterial hypertension in congenital heart disease: an epidemiologic perspective from a Dutch registry. Int J Cardiol. 2007;120:198–204.

S4.4.4.1-4. Engelfriet PM, Duffels MGJ, Möller T, et al. Pulmonary arterial hypertension in adults born with a heart septal defect: the Euro Heart Survey on adult congenital heart disease. Heart. 2007;93:682–7.

S4.4.4.1-5. Lowe BS, Therrien J, Ionescu-Ittu R, et al. Diagnosis of pulmonary hypertension in the congenital heart disease adult population impact on outcomes. J Am Coll Cardiol. 2011;58:538–46.

S4.4.4.1-6. Manes A, Palazzini M, Leci E, et al. Current era survival of patients with pulmonary arterial hypertension associated with congenital heart disease: a comparison between clinical subgroups. Eur Heart J. 2014;35:716–24.

S4.4.4.1-7. Gabriels C, De Meester P, Pasquet A, et al. A different view on predictors of pulmonary hypertension in secundum atrial septal defect. Int J Cardiol. 2014;176:833–40.

S4.4.4.1-8. O'Donnell C, Ruygrok PN, Whyte K, et al. Progressive pulmonary hypertension post atrial septal defect device closure—early symptomatic improvement may not predict outcome. Heart Lung Circ. 2010;19:713–6.

S4.4.4.1-9. Van De Bruaene A, Delcroix M, Pasquet A, et al. The importance of pulmonary artery pressures on late atrial arrhythmia in transcatheter and surgically closed ASD type secundum. Int J Cardiol. 2011;152:192–5.

S4.4.4.1-10. Van De Bruaene A, Moons P, Belmans A, et al. Predictive model for late atrial arrhythmia after closure of an atrial septal defect. Int J Cardiol. 2013;164:318–22.

S4.4.4.1-11. Berner M, Beghetti M, Spahr-Schopfer I, et al. Inhaled nitric oxide to test the vasodilator capacity of the pulmonary vascular bed in children with long-standing pulmonary hypertension and congenital heart disease. Am J Cardiol. 1996;77:532–5.

S4.4.4.1-12. Balint OH, Samman A, Haberer K, et al. Outcomes in patients with pulmonary hypertension undergoing percutaneous atrial septal defect closure. Heart. 2008;94:1189–93.

S4.4.4.1-13. Gorenflo M, Gu H, Xu Z. Peri-operative pulmonary hypertension in paediatric patients: current strategies in children with congenital heart disease. Cardiology. 2010;116:10–7.

S4.4.4.1-14. Huang J-B, Liu Y-L, Yu C-T, et al. Lung biopsy findings in previously inoperable patients with severe pulmonary hypertension associated with congenital heart disease. Int J Cardiol. 2011;151:76–83.

S4.4.4.1-15. Neutze JM, Ishikawa T, Clarkson PM, et al. Assessment and follow-up of patients with ventricular septal defect and elevated pulmonary vascular resistance. Am J Cardiol. 1989;63:327–31.

S4.4.4.1-16. Steele PM, Fuster V, Cohen M, et al. Isolated atrial septal defect with pulmonary vascular obstructive disease—long-term follow-up and prediction of outcome after surgical correction. Circulation. 1987;76:1037–42.

S4.4.4.1-17. Yong G, Khairy P, De Guise P, et al. Pulmonary arterial hypertension in patients with transcatheter closure of secundum atrial septal defects: a longitudinal study. Circ Cardiovasc Interv. 2009;2:455–62.

S4.4.4.1-18. Balzer DT, Kort HW, Day RW, et al. Inhaled nitric oxide as a preoperative test (INOP Test I): the INOP Test Study Group. Circulation. 2002;106:I76–81.

4.4.4.2. Eisenmenger Syndrome

S4.4.4.2-1. Dimopoulos K, Inuzuka R, Goletto S, et al. Improved survival among patients with Eisenmenger syndrome receiving advanced therapy for pulmonary arterial hypertension. Circulation. 2010;121:20–5.

S4.4.4.2-2. Gatzoulis MA, Beghetti M, Galiè N, et al. Longer-term bosentan therapy improves functional capacity in Eisenmenger syndrome: results of the BREATHE-5 open-label extension study. Int J Cardiol. 2008;127:27–32.

S4.4.4.2-3. Galiè N, Beghetti M, Gatzoulis MA, et al. Bosentan therapy in patients with Eisenmenger syndrome: a multicenter, double-blind, randomized, placebo-controlled study. Circulation. 2006;114:48–54.

S4.4.4.2-4. Diller G-P, Alonso-Gonzalez R, Dimopoulos K, et al. Disease targeting therapies in patients with Eisenmenger syndrome: response to treatment and long-term efficiency. Int J Cardiol. 2013;167:840–7.

S4.4.4.2-5. D'Alto M, Romeo E, Argiento P, et al. Therapy for pulmonary arterial hypertension due to congenital heart disease and Down's syndrome. Int J Cardiol. 2013;164:323–6.

S4.4.4.2-6. Iversen K, Jensen AS, Jensen TV, et al. Combination therapy with bosentan and sildenafil in Eisenmenger syndrome: a randomized, placebo-controlled, double-blinded trial. Eur Heart J. 2010;31:1124–31.

S4.4.4.2-7. Baptista R, Castro G, da Silva AM, et al. Long-term effect of bosentan in pulmonary hypertension associated with complex congenital heart disease. Rev Port Cardiol. 2013;32:123–9.

S4.4.4.2-8. Crepaz R, Romeo C, Montanaro D, et al. Long-term results of treatment with bosentan in adult Eisenmenger's syndrome patients with Down's syndrome related to congenital heart disease. BMC Cardiovasc Disord. 2013;13:74.

S4.4.4.2-9. Duffels MGJ, Vis JC, van Loon RLE, et al. Down patients with Eisenmenger syndrome: is bosentan treatment an option? Int J Cardiol. 2009;134:378–83.

S4.4.4.2-10. Serino G, Guazzi M, Micheletti A, et al. Effect of bosentan on exercise capacity and clinical worsening in patients with dual Down and Eisenmenger syndrome. Clin Med Insights Cardiol. 2013;7:29–34.

S4.4.4.2-11. Zhang Z-N, Jiang X, Zhang R, et al. Oral sildenafil treatment for Eisenmenger syndrome: a prospective, open-label, multicentre study. Heart. 2011;97:1876–81.

S4.4.4.2-12. Garg N, Tripathy N, Sinha N. Comparative efficacy of sildenafil in Eisenmenger's syndrome secondary to atrial septal defect versus ventricular septal defect: a cardiac catheterisation follow-up study. Cardiol Young. 2011;21:631–8.

S4.4.4.2-13. Tay ELW, Papaphylactou M, Diller G-P, et al. Quality of life and functional capacity can be improved in patients with Eisenmenger syndrome with oral sildenafil therapy. Int J Cardiol. 2011;149:372–6.

S4.4.4.2-14. Bharani A, Patel A, Saraf J, et al. Efficacy and safety of PDE-5 inhibitor tadalafil in pulmonary arterial hypertension. Indian Heart J. 2007;59:323–8.

S4.4.4.2-15. Mukhopadhyay S, Sharma M, Ramakrishnan S, et al. Phosphodiesterase-5 inhibitor in Eisenmenger syndrome: a preliminary observational study. Circulation. 2006;114:1807–10.

S4.4.4.2-16. Singh TP, Rohit M, Grover A, et al. A randomized, placebo-controlled, double-blind, crossover study to evaluate the efficacy of oral sildenafil therapy in severe pulmonary artery hypertension. Am Heart J. 2006;151. 851.e1–851.e5.

4.4.5. Coronary Anomalies

S4.4.5-1. Eckart RE, Shry EA, Burke AP, et al. Sudden death in young adults: an autopsy-based series of a population undergoing active surveillance. J Am Coll Cardiol. 2011;58:1254–61.

S4.4.5-2. Krasuski RA, Magyar D, Hart S, et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. Circulation. 2011;123:154–62.

S4.4.5-3. Angelini P, Uribe C, Monge J, et al. Origin of the right coronary artery from the opposite sinus of Valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. Catheter Cardiovasc Interv. 2015;86:199–208.

S4.4.5-4. Opolski MP, Pregowski J, Kruk M, et al. Prevalence and characteristics of coronary anomalies originating from the opposite sinus of Valsalva in 8,522 patients referred for coronary computed tomography angiography. Am J Cardiol. 2013;111:1361–7.

S4.4.5-5. Lee H-J, Hong YJ, Kim HY, et al. Anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course: subtypes and clinical importance. Radiology. 2012;262:101–8.

S4.4.5-6. Kaushal S, Backer CL, Popescu AR, et al. Intramural coronary length correlates with symptoms in patients with anomalous aortic origin of the coronary artery. Ann Thorac Surg. 2011;92:986–91.

S4.4.5-7. Maron BJ, Doerer JJ, Haas TS, et al. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. Circulation. 2009;119:1085–92.

S4.4.5-8. Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med. 2004; 141:829–34.

S4.4.5-9. Frescura C, Basso C, Thiene G, et al. Anomalous origin of coronary arteries and risk of sudden death: a study based on an autopsy population of congenital heart disease. Hum Pathol. 1998;29:689–95.

S4.4.5-10. Basso C, Maron BJ, Corrado D, et al. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol. 2000;35:1493–501.

S4.4.5-11. Davies JE, Burkhart HM, Dearani JA, et al. Surgical management of anomalous aortic origin of a coronary artery. Ann Thorac Surg. 2009;88:844–7.

S4.4.5-12. Kragel AH, Roberts WC. Anomalous origin of either the right or left main coronary artery from the aorta with subsequent coursing between aorta and pulmonary trunk: analysis of 32 necropsy cases. Am J Cardiol. 1988;62:771–7.

S4.4.5-13. Donaldson RM, Raphael M, Radley-Smith R, et al. Angiographic identification of primary coronary anomalies causing impaired myocardial perfusion. Cathet Cardiovasc Diagn. 1983;9:237–49.

S4.4.5-14. Brothers JA, McBride MG, Seliem MA, et al. Evaluation of myocardial ischemia after surgical repair of anomalous aortic origin of a coronary artery in a series of pediatric patients. J Am Coll Cardiol. 2007; 50:2078–82.

S4.4.5-15. Sharma V, Burkhart HM, Dearani JA, et al. Surgical unroofing of anomalous aortic origin of a coronary artery: a single-center experience. Ann Thorac Surg. 2014;98:941–5.

S4.4.5-16. Frommelt PC, Sheridan DC, Berger S, et al. Ten-year experience with surgical unroofing of anomalous aortic origin of a coronary artery from the opposite sinus with an interarterial course. J Thorac Cardiovasc Surg. 2011;142:1046–51.

S4.4.5-17. Mainwaring RD, Reddy VM, Reinhartz O, et al. Anomalous aortic origin of a coronary artery: medium-term results after surgical repair in 50 patients. Ann Thorac Surg. 2011;92:691–7.

S4.4.5-18. Mumtaz MA, Lorber RE, Arruda J, et al. Surgery for anomalous aortic origin of the coronary artery. Ann Thorac Surg. 2011;91:811–4.

4.4.5.1. Anomalous Coronary Artery Evaluation

S4.4.5.1-1. Basso C, Maron BJ, Corrado D, et al. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol. 2000;35:1493–501.

S4.4.5.1-2. Davies JE, Burkhart HM, Dearani JA, et al. Surgical management of anomalous aortic origin of a coronary artery. Ann Thorac Surg. 2009;88:844–7.

S4.4.5.1-3. Sharma V, Burkhart HM, Dearani JA, et al. Surgical unroofing of anomalous aortic origin of a coronary artery: a single-center experience. Ann Thorac Surg. 2014;98:941–5.

S4.4.5.1-4. Eckart RE, Shry EA, Burke AP, et al. Sudden death in young adults: an autopsy-based series of a population undergoing active surveillance. J Am Coll Cardiol. 2011;58:1254–61.

S4.4.5.1-5. Angelini P, Uribe C, Monge J, et al. Origin of the right coronary artery from the opposite sinus of Valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. Catheter Cardiovasc Interv. 2015;86:199–208.

S4.4.5.1-6. Opolski MP, Pregowski J, Kruk M, et al. Prevalence and characteristics of coronary anomalies originating from the opposite sinus of Valsalva in 8,522 patients referred for coronary computed tomography angiography. Am J Cardiol. 2013;111:1361–7.

S4.4.5.1-7. Lee H-J, Hong YJ, Kim HY, et al. Anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course: subtypes and clinical importance. Radiology. 2012;262:101–8.

S4.4.5.1-8. Kaushal S, Backer CL, Popescu AR, et al. Intramural coronary length correlates with symptoms in patients with anomalous aortic origin of the coronary artery. Ann Thorac Surg. 2011;92:986–91.

S4.4.5.1-9. Frescura C, Basso C, Thiene G, et al. Anomalous origin of coronary arteries and risk of sudden death: a study based on an autopsy population of congenital heart disease. Hum Pathol. 1998;29: 689–95.

4.4.5.2. Anomalous Aortic Origin of Coronary Artery

S4.4.5.2-1. Basso C, Maron BJ, Corrado D, et al. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol. 2000;35:1493–501.

S4.4.5.2-2. Davies JE, Burkhart HM, Dearani JA, et al. Surgical management of anomalous aortic origin of a coronary artery. Ann Thorac Surg. 2009;88:844–7.

S4.4.5.2-3. Sharma V, Burkhart HM, Dearani JA, et al. Surgical unroofing of anomalous aortic origin of a coronary artery: a single-center experience. Ann Thorac Surg. 2014;98:941–5.

S4.4.5.2-4. Frommelt PC, Sheridan DC, Berger S, et al. Ten-year experience with surgical unroofing of anomalous aortic origin of a coronary artery from the opposite sinus with an interarterial course. J Thorac Cardiovasc Surg. 2011;142:1046–51.

S4.4.5.2-5. Gulati R, Reddy VM, Culbertson C, et al. Surgical management of coronary artery arising from the wrong coronary sinus, using standard and novel approaches. J Thorac Cardiovasc Surg. 2007; 134:1171–8.

S4.4.5.2-6. Mainwaring RD, Reddy VM, Reinhartz O, et al. Anomalous aortic origin of a coronary artery: medium-term results after surgical repair in 50 patients. Ann Thorac Surg. 2011;92:691–7.

4.4.5.3. Anomalous Coronary Artery Arising From the PA

S4.4.5.3-1. Cochrane AD, Coleman DM, Davis AM, et al. Excellent long-term functional outcome after an operation for anomalous left coronary artery from the pulmonary artery. J Thorac Cardiovasc Surg. 1999;117: 332–42.

S4.4.5.3-2. Paridon SM, Farooki ZQ, Kuhns LR, et al. Exercise performance after repair of anomalous origin of the left coronary artery from the pulmonary artery. Circulation. 1990;81:1287–92.

S4.4.5.3-3. Lange R, Vogt M, Hörer J, et al. Long-term results of repair of anomalous origin of the left coronary artery from the pulmonary artery. Ann Thorac Surg. 2007;83:1463–71.

S4.4.5.3-4. el-Said GM, Ruzyllo W, Williams RL, et al. Early and late result of saphenous vein graft for anomalous origin of left coronary artery from pulmonary artery. Circulation. 1973;48:III2–6.

S4.4.5.3-5. Wilson CL, Dlabal PW, McGuire SA. Surgical treatment of anomalous left coronary artery from pulmonary artery: follow-up in teenagers and adults. Am Heart J. 1979;98:440–6.

S4.4.5.3-6. Yau JM, Singh R, Halpern EJ, et al. Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adult cases and a new diagnosis in a 53-year-old woman. Clin Cardiol. 2011;34: 204–10.

S4.4.5.3-7. Rajbanshi BG, Burkhart HM, Schaff HV, et al. Surgical strategies for anomalous origin of coronary artery from pulmonary artery in adults. J Thorac Cardiovasc Surg. 2014;148:220–4.

KEY WORDS ACC/AHA Clinical Practice Guidelines, arrhythmias, cardiac catheterization, cardiac defects, congenital heart disease, congenital heart surgery, unoperated/repaired heart defect

APPENDIX 1. AUTHOR RELATIONSHIPS WITH INDUSTRY AND OTHER ENTITIES (RELEVANT)—2018 AHA/ACC GUIDELINE FOR THE MANAGEMENT OF ADULTS WITH CONGENITAL HEART DISEASE* (FEBRUARY 2018)

Committee Member	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness	Voting Recusals by Section†
Karen K. Stout *(Chair)*	University of Washington—Director, Adult Congenital Heart Disease Program, Professor, Internal Medicine and Pediatrics	None	None	None	None	None	None	None
Curt J. Daniels *(Vice Chair)*	The Ohio State University Heart Center and Nationwide Children's Hospital—Director, Adult Congenital Heart Disease and Pulmonary Hypertension Program, Professor, Internal Medicine and Pediatrics	None	None	None	None	■ Actelion‡	None	4.1.1, 4.1.2, 4.1.3, 4.1.4, 4.1.5, 4.4.6.1, 4.4.6.2
Jamil A. Aboulhosn	UCLA Adult Congenital Heart Disease Center—Director	■ Actelion ■ GE Medical ■ Edward Lifesciences§ ■ Medtronic	None	None	■ Gore	■ United Therapeutics ■ Actelion ■ Medtronic ■ St. Jude ■ Edward Lifesciences	None	4.1.1, 4.1.2, 4.1.3, 4.1.4, 4.1.5, 4.2.6, 4.3.1.1, 4.3.2, 4.3.5, 4.4.6.1, 4.4.6.2
Biykem Bozkurt	Baylor College of Medicine—Professor of Medicine	None	None	None	None	■ Novartis	None	None
Craig S. Broberg	Oregon Health and Science University—Associate Professor of Medicine	None	None	None	None	■ Actelion	None	4.1.1, 4.1.2, 4.1.3, 4.1.4, 4.1.5, 4.4.6.1, 4.4.6.2
Jack M. Colman	University of Toronto—Professor of Medicine and Obstetrics & Gynecology, Toronto Congenital Cardiac Centre for Adults and Pregnancy and Heart Disease Program; University Health Network and Mount Sinai Hospital—Senior Attending Cardiologist	None	None	None	None	None	None	None
Stephen R. Crumb	Nationwide Children's Hospital—Nurse Practitioner and Coordinator, COACH and Pulmonary Hypertension Programs	None	None	None	None	None	None	None
Joseph A. Dearani	Mayo Clinic—Professor of Surgery and Chair, Division of Cardiovascular Surgery	None	None	None	None	■ Sorin (LivaNova)§‖ ■ Cormatrix‖	None	None
Stephanie Fuller	University of Pennsylvania Perelman School of Medicine—Associate Professor of Clinical Surgery	None	None	None	None	None	None	None

Continued on the next page

APPENDIX 1. CONTINUED

Committee Member	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness	Voting Recusals by Section†
Michelle Gurvitz	Harvard Medical School; Brigham and Women's Hospital—Instructor of Pediatrics, Assistant Professor	None	None	None	None	None	None	None
Paul Khairy	Montreal Heart Institute Adult Congenital Center—Director; Professor of Medicine	None	None	None	None	■ Boehringer Ingelheim§ ■ St. Jude Medical§ ■ Medtronic§ ■ Actelion§	None	4.1.1, 4.1.2, 4.1.3, 4.1.4, 4.1.5, 4.2.6, 4.3.1.1, 4.3.2, 4.3.5, 4.4.6.1, 4.4.6.2
Michael J. Landzberg	Boston Children's Hospital— Director, Adult Congenital Heart Service; Harvard Medical School—Associate Professor of Medicine	None	None	None	None	■ Gilead ■ Actelion	None	4.1.1, 4.1.2, 4.1.3, 4.1.4, 4.1.5, 4.4.6.1, 4.4.6.2
Arwa Saidi	University of Florida College of Medicine—Professor, Congenital Heart Center	None	None	None	None	■ Actelion§	None	4.1.1, 4.1.2, 4.1.3, 4.1.4, 4.1.5, 4.4.6.1, 4.4.6.2
Anne Marie Valente	Boston Children's Hospital, Brigham and Women's Hospital—Outpatient Director, Boston Adult Congenital Heart Disease and Pulmonary Hypertension Service; Harvard Medical School—Associate Professor of Medicine and Pediatrics	None	None	None	None	None	None	None
George F. Van Hare	Washington University School of Medicine—Director, Pediatric Cardiology; St. Louis Children's, Washington University Heart Center—Co-Director	None	None	None	None	None	None	None

This table represents the relationships of committee members with industry and other entities that were determined to be relevant to this document. These relationships were reviewed and updated in conjunction with all meetings and/or conference calls of the writing committee during the document development process. The table 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% of the voting stock or share of the business entity, or ownership of ≥$5,000 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. Relationships that exist with no financial benefit are also included for the purpose of transparency. Relationships in this table are modest unless otherwise noted. According to the ACC/AHA, a person has a *relevant* relationship IF: a) the *relationship or interest* relates to the same or similar subject matter, intellectual property or asset, topic, or issue addressed in the *document;* or b) the *company/entity* (with whom the relationship exists) makes a drug, drug class, or device addressed in the *document* or makes a competing drug or device addressed in the *document*; or c) the *person or a member of the person's household*, has a reasonable potential for financial, professional or other personal gain or loss as a result of the issues/content addressed in the *document*.

*The ACHD Guideline began in March 2014. Over the initial years of the CMS Open Payment System, understandably, there have been issues related to accurate reporting of food and beverage payments. For this reason, the ACC and AHA have not considered these minor charges relevant relationships with industry.

†Writing committee members are required to recuse themselves from voting on sections to which their specific relationships with industry and other entities may apply. Section numbers pertain to those in the full-text guideline.

‡No financial benefit.

§Significant relationship.

‖CMS reported payments to Dr. Dearani in 2016 related to research for the Sorin Group and Cormatrix; however, he disagrees with this report. The sections authored by Dr. Dearani have been reviewed, and it was affirmed that there was no implication of any influence of industry.

ACC indicates American College of Cardiology; AHA, American Heart Association; CMS, Centers for Medicare & Medicaid Services; COACH, Columbus Ohio Adult Congenital Heart; and UCLA, University of California, Los Angeles.

APPENDIX 2. REVIEWER RELATIONSHIPS WITH INDUSTRY AND OTHER ENTITIES (COMPREHENSIVE)—2018 AHA/ACC GUIDELINE FOR THE MANAGEMENT OF ADULTS WITH CONGENITAL HEART DISEASE (FEBRUARY 2018)

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Samuel J. Asirvatham	Official Reviewer—AHA	Mayo Clinic—Professor of Medicine and Pediatrics	■ Abiomed ■ AtriCure ■ Biosense Webster ■ Biotronik ■ Boston Scientific* ■ Medtronic ■ Sanofi-aventis ■ St. Jude Medical	None	None	None	None	None
Wendy M. Book	Official Reviewer—AHA	Emory University—Professor of Medicine and Director of Emory Adult Congenital Heart Center, Department of Medicine	None	None	None	None	■ Actelion	■ Defendant, congenital heart disease, 2015
Samuel S. Gidding	Official Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Nemours Cardiac Center DuPont Hospital for Children—Chief, Division of Pediatric Cardiology	None	None	None	None	None	None
Yuli Y. Kim	Official Reviewer—AHA	University of Pennsylvania—Assistant Professor of Medicine; Children's Hospital of Philadelphia—Medical Director, Philadelphia Adult Congenital Heart Center	None	None	None	None	None	None
Geetha Raghuveer	Official Reviewer—ACC Board of Governors	Children's Mercy Hospital—Pediatric Cardiologist; University of Missouri, Kansas City School of Medicine—Professor of Pediatrics	None	None	None	None	None	None
Carole A. Warnes	Official Reviewer—ACC Board of Trustees	Mayo Clinic—Professor of Medicine	None	None	None	None	None	None
Peter J. Bartz	Organizational Reviewer—ASE	Children's Hospital of Wisconsin—Associate Professor, Medical College of Wisconsin	None	None	None	None	None	None
Mitchell I. Cohen	Organizational Reviewer—HRS	Inova Fairfax Children's Hospital—Co-Director of the Heart Center and Chief, Pediatric Cardiology	None	None	None	None	None	None
Marshall L. Jacobs	Organizational Reviewer—AATS	Johns Hopkins School of Medicine—Professor of Surgery and Director, Pediatric Heart Surgery Outcomes Research	None	None	None	None	None	None
Larry A. Latson	Organizational Reviewer—SCAI	Joe DiMaggio Children's Hospital Heart Institute—Medical Director, Pediatric and Congenital Interventional Cardiology	■ Gore Medical	None	None	None	■ St. Jude Medical	None

Continued on the next page

APPENDIX 2. CONTINUED

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Constantine Mavroudis	Organizational Reviewer—STS	Florida Hospital for Children—Medical Director, Pediatric and Congenital Heart Center	None	None	None	None	None	None
Doff B. McElhinney	Organizational Reviewer—SCAI	Stanford University—Professor, Cardiothoracic Surgery and of Pediatrics	■ Medtronic*	None	None	None	None	None
Erwin N. Oechslin	Organizational Reviewer—ISACHD	University of Toronto—Professor of Medicine; Peter Munk Cardiac Centre—Director, Adult Congenital Heart Disease Program	■ Actelion	None	None	None	None	None
John K. Triedman	Organizational Reviewer—HRS	Boston Children's Hospital—Senior Associate in Cardiology; Harvard Medical School—Professor of Pediatrics	■ Biosense Webster	None	None	None	None	None
Sana M. Al-Khatib	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Duke University Medical Center—Professor of Medicine	None	None	None	None	None	None
Naser Ammash	Content Reviewer	Mayo Clinic—Professor of Medicine	None	None	None	None	None	None
Helmut Baumgartner	Content Reviewer	University of Muenster—Professor of Cardiology and Adult Congenital Heart Disease; University Hospital Muenster—Director, Division of Adult Congenital and Valvular Heart Disease, Department of Cardiovascular Medicine	None	None	None	None	None	None
James C. Blankenship	Content Reviewer—ACC Interventional Section Leadership Council	Geisinger Medical Center—Staff Physician and Director, Cardiac Catheterization Laboratory	None	None	None	None	■ Abbott Vascular† ■ Boston Scientific† ■ GlaxoSmithKline† ■ Takeda Pharmaceutical†	None
Ralph G. Brindis	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Northern California Kaiser Permanente—Senior Advisor, Cardiovascular Disease; University of California, San Francisco—Clinical Professor of Medicine	None	None	None	None	None	None
Robert M. Campbell	Content Reviewer	Emory University School of Medicine, Sibley Heart Center Cardiology—Professor of Pediatrics	None	None	None	None	None	None
Lesley H. Curtis	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Duke University School of Medicine—Professor of Medicine	None	None	None	None	■ Boston Scientific* ■ GE Healthcare* ■ GlaxoSmithKline* ■ Medtronic* ■ Novartis*	None

Continued on the next page

APPENDIX 2. CONTINUED

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Kristi K. Fitzgerald	Content Reviewer	Nemours Cardiac Center DuPont Hospital for Children—Genetic Counselor, Division of Pediatric Cardiology	None	None	None	None	None	None
Lee A. Fleisher	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	University of Pennsylvania Health System Department of Anesthesiology and Critical Care—Robert Dunning Dripps Professor of Anesthesiology	None	None	None	None	None	None
Federico Gentile	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Centro Cardiologico Gentile—Director, Cardiovascular Disease	None	None	None	None	None	None
Louise Harris	Content Reviewer	Toronto General Hospital—Professor of Medicine	■ St. Jude Medical	None	None	None	None	None
Mark A. Hlatky	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Stanford University School of Medicine—Professor of Health Research and Policy, and of Cardiovascular Medicine	None	None	None	None	None	None
Craig T. January	Content Reviewer	University of Wisconsin School of Medicine and Public Health—Professor, Division of Cardiovascular Medicine	None	None	None	None	None	None
José A. Joglar	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	UT Southwestern Medical Center—Professor of Internal Medicine	None	None	None	None	None	None
Thomas K. Jones	Content Reviewer	Seattle Children's Hospital—Professor of Pediatrics and Director, Cardiac Catheterization Laboratories	■ Gore Medical* ■ Medtronic*	None	None	None	■ Gore Medical* ■ Medtronic* ■ St. Jude Medical*	None
Brian E. Kogon	Content Reviewer	Emory University School of Medicine—Associate Professor, Surgery and Surgical Director of Emory Adult Congenital Heart Center and Chief of Pediatric Cardiac Surgery	None	None	None	None	None	None
Gautam Kumar	Content Reviewer—ACC Interventional Section Leadership Council	Emory University School of Medicine, Division of Cardiology—Associate Professor of Medicine	■ Abiomed	None	None	None	■ OrbusNeich Medical	None
Eric V. Krieger	Content Reviewer	University of Washington—Associate Professor of Medicine and Associate Director, Adult Congenital Heart Service	■ Actelion	None	None	None	None	None
Glenn N. Levine	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Baylor College of Medicine—Professor of Medicine; Michael E. DeBakey VA Medical Center—Director, Cardiac Care Unit	None	None	None	None	None	None

Continued on the next page

APPENDIX 2. CONTINUED

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
C. Huie Lin	Content Reviewer—SCAI	Houston Methodist DeBakey Heart & Vascular Center—Cardiologist	■ Gore Medical ■ ACI Clinical (DSMB)	■ Abiomed	None	■ St. Jude Medical	None	None
Massimo Mancone	Content Reviewer—ACC AIG	Sapienza University of Rome—Cardiology Consultant	None	None	None	None	None	None
Ariane Marelli	Content Reviewer	McGill University Health Center—Professor of Medicine and Director, MAUDE Unit	None	None	None	None	None	None
Koichiro Niwa	Content Reviewer	St. Luke's International Hospital—Director, Department of Cardiology	None	None	None	None	None	None
Matthew Oster	Content Reviewer	Emory University School of Medicine—Associate Professor; Children's Healthcare of Atlanta—Director, Children's Cardiac Outcomes Research Program at Sibley Heart Center	None	None	None	None	None	None
Catherine M. Otto	Content Reviewer	University of Washington School of Medicine—Professor of Medicine, Division of Cardiology and Director, Heart Valve Clinic	None	None	None	None	None	None
Richard L. Page	Content Reviewer	University of Wisconsin School of Medicine and Public Health—Chair, Department of Medicine	None	None	None	None	None	None
James Perry	Content Reviewer	Rady Children's Hospital; University of California, San Diego—Professor of Pediatrics, Affiliate Professor of Bioengineering, and Director, Electrophysiology and Adult Congenital Heart Programs	None	None	None	None	None	None
Susan J. Pressler	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	Indiana University School of Nursing—Professor and Director, Center for Enhancing Quality of Life in Chronic Illness	None	None	None	None	■ Pfizer†	None
Candice K. Silversides	Content Reviewer	University of Toronto—Associate Professor	None	None	None	None	None	None
Duminda N. Wijeysundera	Content Reviewer—ACC/AHA Task Force on Clinical Practice Guidelines	University of Toronto—Assistant Professor, Department of Anesthesia and Institute of Health Policy Management and Evaluation	None	None	None	None	None	None

Continued on the next page

APPENDIX 2. CONTINUED

Reviewer	Representation	Employment	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Ali N. Zaidi	Content Reviewer	Montefiore Einstein Center for Heart and Vascular Care—Director, Montefiore Adult Congenital Heart Disease Program; Albert Einstein College of Medicine—Associate Professor, Internal Medicine and Pediatrics	None	None	None	None	None	None
Elisa Zaragoza-Macias	Content Reviewer	PeaceHealth North Cascade Cardiology—Cardiologist	None	None	None	None	None	None

This table represents the relationships of reviewers with industry and other entities that were disclosed at the time of peer review and determined to be relevant to this document. 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% of the voting stock or share of the business entity, or ownership of ≥$5,000 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 that exist with no financial benefit are also included for the purpose of transparency. Relationships in this table are modest unless otherwise noted. Names are listed in alphabetical order within each category of review. Please refer to http://www.acc.org/guidelines/about-guidelines-and-clinical-documents/relationships-with-industry-policy for definitions of disclosure categories or additional information about the ACC/AHA Disclosure Policy for Writing Committees.

**Significant relationship.

††No financial benefit.

AATS indicates American Association for Thoracic Surgery; ACC, American College of Cardiology; AHA, American Heart Association; AIG, Assembly of International Governors; ASE, American Society of Echocardiography; DSMB, Data Safety Monitoring Board; HRS, Heart Rhythm Society; ISACHD, International Society for Adult Congenital Heart Disease; MAUDE, McGill Adult Unit for Congenital Heart Disease Excellence; SCAI, Society for Cardiovascular Angiography and Interventions; and STS, Society of Thoracic Surgeons; and UT, University of Texas.