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). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD)

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Preamble

Nearly one third of all major congenital anomalies are heart defects, with an estimated 9 per 1000 live births afflicted by congenital heart disease (CHD) worldwide.[1] Remarkable advances in care have resulted in impressive gains in survival such that over 90% of children with CHD in developed countries today are expected to survive into adulthood.[2] Consequently, the past decades have witnessed historical shifts in population demographics, as adults now outnumber children with CHD. Population-based estimates indicate that there are currently over 1 million adults with CHD in the United States alone, over 100,000 in Canada, and 1.8 million in Europe.[3–5] Rhythm disorders, which span the entire spectrum of brady- and tachyarrhythmias, are among the most prominent complications encountered by adults with CHD.[6] Arrhythmias range in symptomatology and significance, from inconsequential and benign to poorly tolerated and potentially fatal. Taken together, arrhythmias are a leading cause of morbidity, impaired quality of life, and mortality in adults with CHD.

In light of the unique issues, challenges, and considerations involved in managing arrhythmias in this growing, aging, and heterogeneous patient population,[7] it appears both timely and essential to critically appraise and synthesize optimal treatment strategies. The purpose of this consensus statement is, therefore, to define optimal conditions for the delivery of care regarding arrhythmias in adults with CHD and provide expert and, where possible, evidence-based recommendations on best practice procedures for the evaluation, diagnosis, and management of arrhythmias, including medical treatment, catheter-based interventions, device therapy, and surgical options.

1. Methodology and evidence

The Pediatric and Congenital Electrophysiology Society (PACES), in conjunction with the Heart Rhythm Society (HRS), appointed a 22-member writing committee from the United States, Canada, and Europe with complementary multidisciplinary expertise in pediatric and adult electrophysiology, adult CHD, and CHD surgery. The writing committee included representation from the American College of Cardiology (ACC), American Heart Association (AHA), European Heart Rhythm Association (EHRA), Canadian Heart Rhythm Society (CHRS), and International Society for Adult Congenital Heart Disease (ISACHD). The committee was divided into subgroups to review key aspects in the recognition and management of arrhythmias in adults with CHD. Experts in the topics under consideration were tasked with performing formal literature reviews, weighing the strength of evidence for or against diagnostic and therapeutic interventions, estimating expected health outcomes where relevant, and proposing practical clinical recommendations. Wherever possible, recommendations are evidence-based. However, unlike some practice guidelines, there is not a sizeable body of literature with definitive evidence to support most recommendations in this emerging field of cardiology. In order to maximize the value and credibility of consensus-based recommendations, a high-threshold (i.e., 80% or greater agreement among writing members) was required to constitute a consensus. Supportive evidence is indicated where appropriate, and variations in opinion are nuanced in the text. As a general recommendation, the committee strongly supports expanding the evidence base related to arrhythmias in adults with CHD through participation in research and clinical registries.

The consensus statement was organized by arrhythmia-related topics rather than by heart defect. Depending, in part, on the particular issue and available evidence, recommendations range from being broadly applicable to adults with CHD at large to a more focused lesion-specific scope.

Table 1.1 Classification of CHD complexity in adults

Complexity	Type of congenital heart disease in the adult patients
Simple	*Native disease*
	Isolated congenital aortic valve disease
	Isolated congenital mitral valve disease (except parachute valve, cleft leaflet)
	Small atrial septal defect
	Isolated small ventricular septal defect (no associated lesions)
	Mild pulmonary stenosis
	Small patent ductus arteriosus
	Repaired conditions
	Previously ligated or occluded ductus arteriosus
	Repaired secundum or sinus venosus atrial septal defect without residua
	Repaired ventricular septal defect without residua
Moderate	Aorto-left ventricular fistulas
	Anomalous pulmonary venous drainage, partial or total
	Atrioventricular septal defects, partial or complete
	Coarctation of the aorta
	Ebstein anomaly
	Infundibular right ventricular outflow obstruction of significance
	Ostium primum atrial septal defect
	Patent ductus arteriosus, not closed
	Pulmonary valve regurgitation, moderate to severe
	Pulmonary valve stenosis, moderate to severe
	Sinus of Valsalva fistula/aneurysm
	Sinus venosus atrial septal defect
	Subvalvular or supravalvular aortic stenosis
	Tetralogy of Fallot
	Ventricular septal defect with:
	Absent valve or valves
	Aortic regurgitation
	Coarctation of the aorta
	Mitral disease
	Right ventricular outflow tract obstruction
	Straddling tricuspid or mitral valve
	Subaortic stenosis
Severe/complex	Conduits, valved or nonvalved
	Cyanotic congenital heart disease, all forms
	Double-outlet ventricle
	Eisenmenger syndrome
	Fontan procedure
	Mitral atresia
	Single ventricle (also called double inlet or outlet, common, or primitive)
	Pulmonary atresia, all forms
	Pulmonary vascular obstructive disease
	Transposition of the great arteries
	Tricuspid atresia
	Truncus arteriosus/hemitruncus
	Other abnormalities of atrioventricular or ventriculoarterial connection not included above (e.g., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Adapted from Warnes CA, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol. 2008;52:1890–1947.[8]

The detailed index should assist the reader in rapidly locating sections of interest for specific heart defects. In addition, the writing committee retained the nomenclature for complexity of CHD (i.e., simple, moderate, complex/severe) proposed by the ACC/AHA task force on practice guidelines for adults with CHD,[8] summarized in Table 1.1.

Recommendations were subject to a previously described standardized classification process (Methodology Manual and Policies from the ACCHF and AHA Task Force on Practice Guidelines June 2010)[9] that ranked each item (Classes I, IIa, IIb, III) and its accompanying level of evidence (Levels A, B, C), as summarized in Table 1.2.

Table 1.2 Classification of recommendations and levels of evidence[9]

Classification of Recommendations	
Class I	Conditions for which there is evidence and/or general agreement that a given procedure or treatment plan is beneficial, useful, and effective
Class II	Conditions for which there is conflicting evidence and/or divergence of opinion about the usefulness/efficacy of a procedure or treatment
Class IIa	Weight of evidence/opinion is in favor of usefulness/efficacy
Class IIb	Usefulness/efficacy is less well established by evidence/opinion
Class III	Conditions for which there is conflicting evidence and/or general agreement that a procedure or treatment is not useful/effective and in some cases may be harmful
Levels of Evidence	
Level of evidence A	Data derived from multiple randomized clinical trials or meta-analyses
Level of evidence B	Data derived from a single randomized trial or nonrandomized studies
Level of evidence C	Only consensus opinion of experts, case studies, or standard of care

2. Document review and approval

The PACES/HRS Task Force made every effort to avoid all potential conflicts of interest relevant to this consensus statement, whether actual or perceived, among members of the writing committee. Members of the writing committee (Appendix 1) and peer reviewers (Appendix 2) were required to disclose all actual or potential direct or indirect conflicts. Committee members were obliged to refrain from voting on issues related to the potential conflict. The document was reviewed by the PACES executive committee, additional members of HRS, and official reviewers nominated by ACC, AHA, EHRA, CHRS, and ISACHD. All writing members approved this final version.

3. Epidemiology and scope of arrhythmias in adults with CHD

3.1. Changing mortality

The advent of cardiopulmonary bypass and early surgical innovations for CHD of the 1960s and 1970s, coupled with advances in clinical care, have culminated in an increasing and aging cohort with CHD.[10] Survival beyond the first year of life has risen from an estimated 25% 50 years ago to >90% expected survival into adulthood.[11,12] In a population-based cohort study of patients with CHD, an overall mortality reduction of 31% was observed from 1987 to 2005, largely driven by improved survival in infants.[2] Most notably, the median age of death in patients with severe forms of CHD increased from 2 to 23 years of age. The older adult with CHD can also anticipate a considerably longer life expectancy, with one study reporting a median age at death of 57 years in 2007 compared to 37 years in 2002.[13] Although causes of death appear to have remained more or less consistent over the past two decades, recent years have seen a shift in the profile of the patient at risk. While lesion severity and surgical results are major determinants of outcome in infants and children, heart failure, arrhythmias, and pulmonary hypertension become increasingly important in adulthood. Additional prognostic factors in older patients include systemic ventricular dysfunction, chronic renal disease, coronary artery disease, malignancies, and conventional risk factors such as diabetes, hypertension, and obesity.[10,14,15]

3.2. Spectrum of arrhythmias

Arrhythmias increase in prevalence as adults with CHD age and are the most frequent reason for hospital admission.[16,17] Along with heart failure, arrhythmias are the leading cause of death.[18–21] Factors associated with pre- and postoperative arrhythmias in CHD are schematically depicted in Figure 3.1.[22] Arrhythmias may reflect congenitally displaced or malformed sinus nodes or atrioventricular (AV) conduction systems, abnormal hemodynamics, primary myocardial disease, hypoxic tissue injury, residual or postoperative sequelae, and genetic influences.[23–25]

The entire spectrum of arrhythmias may be encountered in adults with CHD, with several subtypes often coexisting. Bradyarrhythmias may involve disorders of the sinus node, AV node, His–Purkinje system, or intra-atrial propagation. It has been estimated that approximately 50% of 20-year-olds with CHD will develop an atrial tachyarrhythmia during their lifetime.[26] Table 3.1 summarizes atrial tachyarrhythmias typically encountered in common forms of CHD.[27] Atrial tachyarrhythmias may be mediated by accessory pathways, AV node reentry, twin AV nodes,[28,29] macroreentrant circuits, automatic foci, or nonautomatic foci.[30] Intra-atrial reentry is the most common tachyarrhythmia in adults with CHD,[31–33] although the prevalence of atrial fibrillation is on the rise as the population ages.[31,34] Ventricular arrhythmias are thought to be the leading cause of sudden death in several subtypes of CHD, with an overall risk that is up to 100-fold higher than in age-matched controls.[18,19] Fortunately, the absolute incidence of these devastating events remains relatively low, at approximately 0.1% per year.

A tabular representation of approximate expected risks for atrial arrhythmia, ventricular arrhythmia, AV block, and ventricular dyssynchrony are summarized in Figure 3.2. The prevalence and mechanism of arrhythmias vary according to factors such as age, underlying anatomic defect, and method of surgical repair.[31] For example, while 3%–5% of patients with congenitally corrected transposition will be born with complete AV block, it is estimated that an additional 20% will develop complete heart block by adulthood.[35,36] For others, prior surgery in the region of the sinus node or its arterial supply (e.g., Mustard, Senning, Glenn, or Fontan) will leave them predisposed to later sinus node dysfunction.[32,37,38]

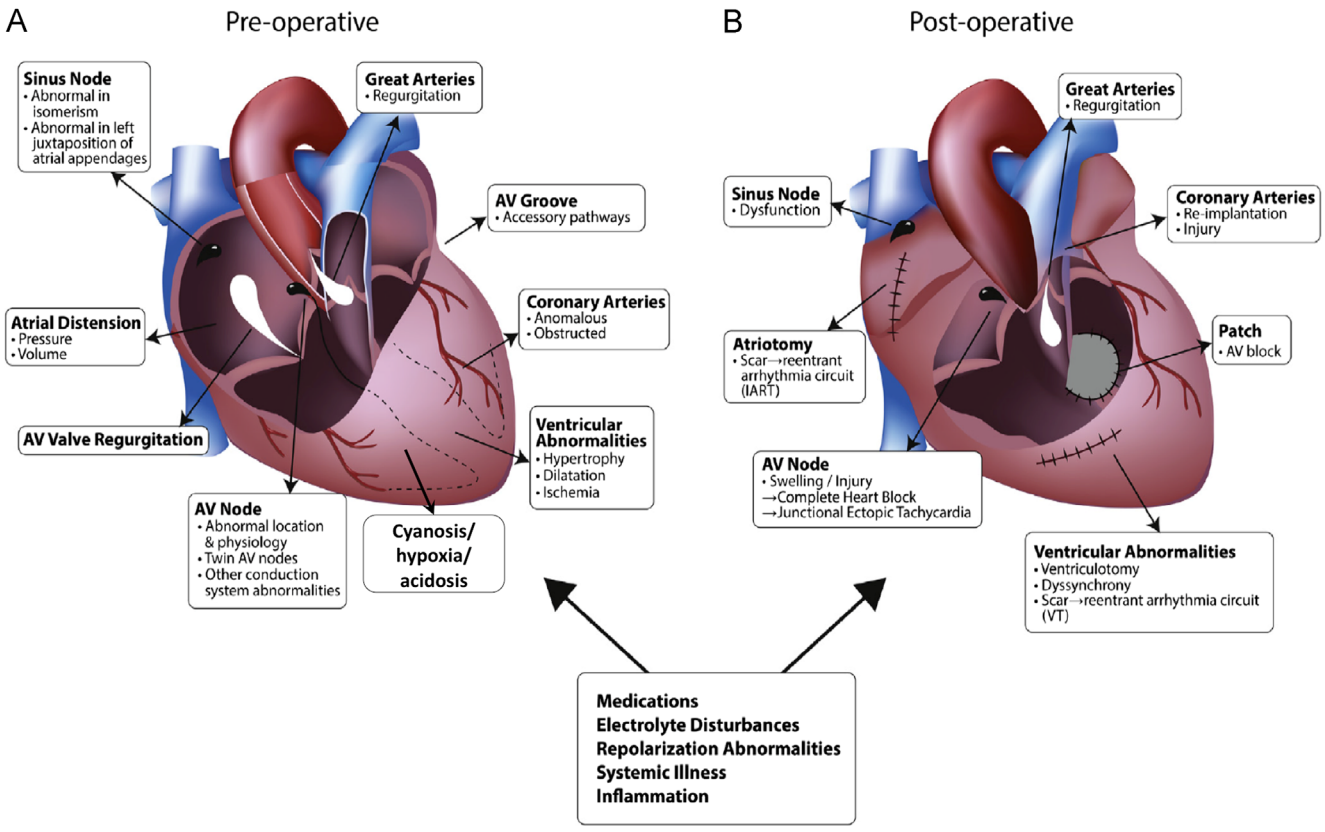

Figure 3.1 Schematic of factors leading to arrhythmias in (**A**) pre- and (**B**) postoperative congenital heart disease. AV = atrioventricular. (Reproduced with permission from Escudero C, et al. Electrophysiologic considerations in congenital heart disease and their relationship to heart failure. Can J Cardiol 2013;29(7):821–829.[22])

3.3. Heart failure and arrhythmogenesis

The relationship of heart failure to arrhythmogenesis and sudden cardiac death risk is increasingly appreciated.[22] Hemodynamic and electrophysiologic conditions that lead to heart failure, clinical arrhythmias, and adverse outcomes in adults with CHD often extend over several decades. These include long-standing effects of prior atrial or ventricular volume loading, scarring, patches, baffles and surgical barriers, electromechanical dyssynchrony, ongoing deleterious effects on cell–cell electrical coupling, and underlying genetic aspects. Inevitably, the incidence of arrhythmias in the adult CHD population far exceeds that seen in younger patients.

Unique forms of heart failure can also be encountered, including dysfunction of a systemic right ventricle or univentricular heart. Systemic left ventricular failure is often associated with congenital left-sided cardiac lesions. Left ventricular dysfunction in tetralogy of Fallot and Ebstein malformation of the tricuspid valve is more widely appreciated as a sequela associated with heightened risk for sudden cardiac death.[31,39–41] Right–left ventricular interactions are

Table 3.1 Summary of atrial tachyarrhythmias encountered in common forms of CHD

Congenital heart disease type	Tachyarrhythmia
Atrial septal defect	IART/AF with increasing age, particularly if late closure
Atrioventricular septal defect	IART/AF following surgical repair
Ebstein anomaly	IART; AV or atriofascicular (Mahaim) AP; sudden death if high risk or multiple APs; ectopic atrial tachycardia; AF
Left-sided obstructive lesions	IART/AF
TGA with intraatrial baffle	IART, NAFAT, AVNRT; VT/VF may be secondary to atrial arrhythmias
Congenitally corrected TGA	Accessory pathway if Ebstein-like systemic AV valve
Tetralogy of Fallot	IART; NAFAT along the anterolateral right atrium
Heterotaxy syndrome	Twin AV node-mediated reentrant tachycardia
Single ventricle with Fontan	IART; NAFAT; AF; may be poorly tolerated
Eisenmenger physiology	MAT; IART; AF

AF = atrial fibrillation; AP = accessory pathway; AV = atrioventricular; AVNRT = AV nodal reentrant tachycardia; IART = intraatrial reentrant tachycardia; MAT = multifocal atrial tachycardia; NAFAT = nonautomatic focal atrial tachycardia; TGA = transposition of the great arteries; VF = ventricular fibrillation; VT = ventricular tachycardia. Adapted with permission from Khairy P. In: Shenasa M et al (eds). *Cardiac Mapping*. Fourth Edition. Oxford, UK: Wiley-Blackwell; 2013:771–777.[27]

Complexity of CHD	Type of CHD	Prevalence (in CHD population)	Atrial Arrhythmia			Ventricular Arrhythmia	Other Pacing Needs		
			AT	AF	Other		SND	AV block	Dyssynchrony, heart failure
Simple	Patent ductus arteriosus	6-8%							
	Pulmonary stenosis	6-8%							
	Ventricular septal defect	30-32%							
	Secundum atrial septal defect	8-10%							
Moderate	Aortic coarctation	5-7%							
	Anomalous pulmonary venous return	0.5-2.5%							
	Atrioventricular septal defect	3-5%							
	Aortic stenosis	3-5%							
	Ebstein anomaly	0.5-1.5%							
	Tetralogy of Fallot	8-10%							
	Primum atrial septal defect	2-3%							
Severe	Truncus arteriosus	1.5-2%							
	Pulmonary atresia	2-2.5%							
	Double outlet right ventricle	1.5-2%							
	D-transposition of the great arteries	6-7%							
	L-transposition of the great arteries	1-2%							
	Hypoplastic left heart syndrome	3-4%							
	Other (heterotaxy, other single ventricles)	7-10%							

Figure 3.2 Approximate risk estimates for atrial tachycardia (AT), atrial fibrillation (AF), other supraventricular arrhythmias, ventricular arrhythmia, sinus node dysfunction (SND), atrioventricular (AV) block, and ventricular dyssynchrony are shown across various congenital heart defects (CHD) of simple, moderate, and severe complexity. The color-coded pattern ranges from minimal (*no shading*) to mild (*light blue*), moderate (*medium blue*), and high (*dark blue*) risk.

increasingly acknowledged, and subpulmonary right ventricular failure itself contributes to the complex interplay of factors associated with sudden death.[42,43] Ventricular dyssynchrony due to intrinsic or pacing-induced ventricular conduction delay can likewise have deleterious effects on systemic ventricular function. In adults with CHD, right bundle branch block (RBBB) is more common than left bundle branch block (LBBB), particularly in the setting of tetralogy of Fallot, ventricular septal defects, double-outlet right ventricle variants, Rastelli surgery, AV septal defects, and Ebstein malformation of the tricuspid valve. In most cases, RBBB is a complication of surgical repair.

3.4. Systemic right ventricle and univentricular heart

Adults with systemic right ventricles and atrial switch surgery (e.g., Mustard or Senning) have extensive atrial scarring, with a high incidence of atrial tachyarrhythmias.[44] Rapid AV conduction in the setting of an already compromised systemic right ventricle can result in induction of a secondary ventricular tachycardia.[45,46] Primary ventricular arrhythmias may also occur, most often in association with systemic right ventricular failure.[36,46–48] Myocardial oxygen supply–demand mismatch can increase over time, leading to ongoing fibrosis, worsening systemic ventricular function, and accrued risk of sudden death.[49] Adults with single ventricle physiology and Fontan palliation are also at risk for developing sinus node dysfunction and atrial tachyarrhythmias.[50,51] Atrial arrhythmias occur in up to 60% of Fontan recipients and are associated with substantial morbidity and mortality.[52] Approximately 90% of Fontan patients with heart failure–related deaths have concomitant atrial tachyarrhythmias.[51]

4. Delivery of care and ensuring access to care

4.1. Recommendations for the coordination and delivery of care for adults with CHD and arrhythmias

Recommendations	
Class I	1. Health care for adults with CHD and arrhythmias should be coordinated by regional adult CHD (ACHD) centers of excellence that serve the surrounding medical community as a resource for consultation and referral *(Level of evidence: C)*.[53] 2. A regional ACHD center that cares for adults with CHD and arrhythmias should be staffed by at least one cardiac electrophysiologist with expertise in CHD, in addition to associated CHD subspecialists in imaging, interventional cardiology, and cardiac surgery *(Level of evidence: C)*.[8,54,55] 3. Diagnostic and interventional catheter- and device-based electrophysiologic procedures in adults with moderate or complex CHD or complex arrhythmias should be performed in a regional ACHD center by a cardiac electrophysiologist with expertise in CHD, and in a laboratory with appropriate personnel and equipment *(Level of evidence: C)*.[7,56,57]

The 32nd Bethesda Conference report called attention to the need for health care professionals, patients and their families, and regulatory agencies to develop a strategic plan to improve care access and delivery to the adult with CHD.[53] Recognition and management of arrhythmias is an integral component of such specialized care.[18,19,24,58] Coordinating care across subspecialties and the development of training programs specific to arrhythmias in adults with CHD are considered key factors in ensuring access and delivery of high-quality care. Health care needs, particularly for adults with moderate and complex forms of CHD, should be coordinated by regional ACHD centers of excellence.[8,54,59] Personnel and services previously recommended for regional ACHD centers are summarized in Table 4.1.[8]

Table 4.1 Personnel and services recommended for regional ACHD centers

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

Reproduced with permission from Warnes CA, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol 2008;52:1890–1947.[8]

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

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

Table 4.2 Basic requirements for electrophysiologists with expertise in adult CHD

Completion of specialized fellowship training in adult or pediatric electrophysiology with demonstrated acquisition of required clinical competencies[62–65]
Formal affiliation with an established ACHD center[8,64]
Fundamental knowledge of congenital heart disease, including:
• Anatomy and physiology of simple, moderate, and complex forms of congenital heart disease
• Surgical procedures for congenital heart disease
• Natural and unnatural (postsurgical) short- and long-term arrhythmia sequelae
• Particularities essential to safely and effectively execute arrhythmia interventions, including an appreciation for complex access issues and displaced or malformed atrioventricular conduction systems[6,7,68]
Experience and skills in managing adults with congenital heart disease and arrhythmias,[64] including:
• Noninvasive testing
• Electrophysiologic studies
• Catheter ablation, including with 3-dimensional electroanatomic mapping systems and large-tip/irrigated catheters
• Intraoperative procedures
• Cardiac rhythm management devices

ACHD = adult congenital heart disease.

Because arrhythmias account for the majority of emergency room visits in adults with CHD, emergency care facilities should ideally have access to, and an affiliation with, a regional ACHD center.[60] The provision of support for local emergency centers is critically important considering that these centers may have little or no familiarity with CHD anatomy, hemodynamics, and complex management issues.[61] In other less urgent situations, coordination by a regional ACHD center should include the availability of consultation services for arrhythmia-related issues, with consideration given to transferring care whenever subspecialty expertise are required, including for electrophysiologic studies, catheter ablation, or device implantation.[8]

Although detailed recommendations regarding training and skills required to qualify as an electrophysiologist with expertise in adult CHD are beyond the scope of this consensus document, basic competencies are summarized in Table 4.2.[62–65] Currently, there is a paucity of formally trained adult CHD electrophysiologists, and, therefore, close collaborations between adult and pediatric electrophysiologists and ACHD specialists may be required to deliver high-quality care to adults with CHD and arrhythmias. These arrangements are viewed by the committee as acceptable methods of optimizing quality of care. In certain circumstances, a broader team approach to managing adults with CHD and arrhythmias may be beneficial, including interventional cardiologists, heart failure specialists, and/or adult CHD surgeons. Examples include hybrid surgical approaches to managing arrhythmias,[66] recanalization of obstructed baffles or conduits to allow catheter or lead access,[67] presurgical electrophysiologic mapping, epicardial lead implantation,[7] and arrhythmia surgery (see Section 11).

4.2. Recommendations for adults with CHD requiring invasive electrophysiologic interventions

Recommendations	
Class I	1. Consultation with an ACHD specialist should be sought prior to invasive electrophysiologic interventions in adults with CHD *(Level of evidence: C)*.[8,54,55] 2. Preprocedural planning should include a detailed review of operative notes pertaining to all previous cardiac and vascular surgeries, patient anatomy, vascular and intracardiac access issues, prior interventions, and all documented arrhythmias *(Level of evidence: C)*.[6,7,68] 3. Invasive electrophysiologic interventions in adults with moderate or complex CHD that require conscious sedation or general anesthesia should be performed in collaboration with an anesthesiologist familiar with CHD *(Level of evidence: C)*.[69] 4. The electrophysiology laboratory and postprocedure recovery unit should be suitable for the care of adults with CHD, including: 1. Adult appropriate equipment *(Level of evidence: C)*; 2. Nursing and technical staff certified in adult cardiac life support (ACLS) and trained in basic CHD anatomy *(Level of evidence: C)*; 3. ACHD cardiothoracic surgical backup and operating room access *(Level of evidence: C)*.[70]

Invasive electrophysiologic procedures in adults with CHD should be performed by electrophysiologists with expertise in adults with CHD and within an environment suitable for CHD and adult patient care.[59] The technical and nursing staff involved with preprocedural, procedural, and postprocedural care must be ACLS certified and familiar with basic CHD anatomy and physiology. Preprocedural evaluation may include risk analysis that considers associated comorbidities, additional subspecialty consultation (e.g., pulmonary medicine, nephrology, infectious disease), anesthesia assessment, and supplementary preprocedural imaging or functional analysis, as needed.

5. Evaluation and diagnosis of arrhythmias

5.1. Introduction

Arrhythmias and their attendant clinical consequences become increasingly prevalent in adults with CHD as they age. Ramifications are often dependent as much on the clinical context in which the arrhythmia occurs as the arrhythmia itself. For that reason, this section stresses the importance not only of electrophysiologic testing from which one might elucidate the correct "electrical" diagnosis but also of a broader evaluation that allows care providers to understand the arrhythmia within the context of the patient's cardiovascular status. Although it is possible to make some generic recommendations regarding adults with CHD as a collective entity or within defined subgroups, in many cases, the natural and modified history of specific anatomic forms of CHD and/or associated palliative corrections dictate more precise targeting of recommendations to specific substrates. Finally, although the presence of symptoms will drive the majority of arrhythmia evaluations, it is recognized that surveillance testing in select circumstances may alert the provider to an impending or unrecognized arrhythmia.

5.2. General rhythm assessment based on cardiac history and symptom status

Arrhythmia risks in adults with CHD vary according to the underlying heart lesion, hemodynamics, and features in a patient's clinical history. Certain rhythm disorders are well known to be lesion-specific, such as accessory pathway–mediated tachycardia in Ebstein anomaly[71] and AV block in the setting of L-looped transposition of the great arteries.[72] The tendency for atrial tachycardias and sinus node dysfunction to develop in patients who have undergone extensive atrial baffling procedures is also clearly established,[32,73] as is the association of monomorphic ventricular tachycardia with such lesions as surgically repaired tetralogy of Fallot,[31] and polymorphic ventricular tachycardia or ventricular fibrillation in patients with advanced degrees of ventricular dysfunction.[40,74] Moreover, late age at time of complete surgical repair and incomplete or imperfect repair with residual cyanosis or pressure/volume loads are among the many historical items that have been implicated as general risk factors for both atrial and ventricular arrhythmias. Individual patient anatomy, surgical history, and hemodynamic status must, therefore, be ascertained fully whenever arrhythmia risks are being estimated.

The presence or absence of symptoms is a practical starting point for evaluating adults with CHD for arrhythmias. In patients with symptoms, the primary task involves determining whether the complaint is rhythm-related and, if so, documenting or replicating the rhythm disturbance so that appropriate treatment can be instituted. In asymptomatic patients, the task is to detect or predict arrhythmias and institute therapy in advance of serious symptoms through a process of surveillance testing and risk assessment, which is in many ways a far more challenging and imperfect exercise.[6]

5.3. Approach to the symptomatic patient

Careful history and physical examination reveal a mix of electrophysiologic and hemodynamic data that are vital to determining the pace and setting of the subsequent evaluation. Attributes of the symptoms, including timing, duration, context, and severity, are helpful in guiding the subsequent selection of tests. In patients with an in situ cardiac rhythm management device, device interrogation may provide the required information to clinch a diagnosis.[75] In the absence of such a device or in the event of an unrevealing interrogation, subsequent workup is determined based on level of clinician suspicion and severity of symptoms.

In patients with aborted sudden cardiac death or unexplained syncope, consideration should be given to performing a diagnostic electrophysiologic study with programmed atrial and ventricular stimulation.[75,76] For milder symptoms, some form of ambulatory monitoring is usually indicated. Frequent or incessant symptoms may be well suited for 24-hour ambulatory ECG (Holter) evaluation. In contrast, infrequent, brief symptoms are better evaluated using an event recorder or longer-duration Holter monitoring. While pursuing the electrophysiologic evaluation, the clinician must also assess the patient's anatomy and hemodynamic/functional status. Critical parameters include ventricular size and function, AV valve function, and vessel or baffle patency, which can often be elucidated by transthoracic echocardiography. When relevant, additional functional and anatomic data can be obtained through 3-dimentional imaging such as cardiac computed tomography (CT) or magnetic resonance imaging (MRI), or more invasive means such as transesophageal echocardiography or cardiac catheterization. Like history and physical examination, ECG and cardiopulmonary exercise testing provide a valuable mix of both electrophysiologic and hemodynamic/functional data.

5.3.1. Rhythm testing for symptomatic patients

5.3.1.1. Electrocardiogram. Documentation of an active arrhythmia by 12-lead ECG at the time of symptoms is a cornerstone of diagnosis, but this luxury is not always available. Observations such as marked bradycardia, AV and intraventricular conduction disturbances, QRS duration, repolarization pattern, and varied degrees of atrial and ventricular ectopic activity may all prove useful in deciphering a patient's complaint. Typical ECG features in adults with common forms of CHD are summarized in Table 5.1.[77]

Table 5.1 Typical ECG features in adults with common forms of CHD

Congenital diagnosis	Rhythm	PR interval	QRS axis	QRS configuration	Atrial enlargement	Ventricular hypertrophy	Particularities
Secundum atrial septal defect	NSR; ↑IART/AF with age	1° AVB 6%–19%	0°–180°; RAD; LAD in Holt-Oran or LAHB	rSr′ or rsR′ with RBBBi > RBBBc	RAE 35%	Uncommon	"Crochetage" pattern
Ventricular septal defect	NSR; PVCs	Normal or mild ↑; 1° AVB 10%	RAD with BVH; LAD 3%–15%	Normal or rsr′; possible RBBB	Possible RAE ± LAE	BVH 23%–61%; RVH with Eisenmenger	Katz-Wachtel phenomenon
AV canal defect	NSR; PVCs 30%	1° AVB >50%	Mod to extreme LAD; Normal with atypical	rSr′ or rsR′	Possible LAE	Uncommon in partial; BVH in complete; RVH with Eisenmenger	Inferoposteriorly displaced AVN
Patent ductus arteriosus	NSR; ↑IART/AF with age	↑PR 10%–20%	Normal	Deep S V_1, tall R V_5 and V_6	LAE with moderate PDA	Uncommon	Often either clinically silent or Eisenmenger
Pulmonary stenosis	NSR	Normal	Normal if mild; RAD with moderate/severe	Normal; severity	Possible RAE	RVH; severity correlates with R: S in V_1 and V_6	Axis deviation correlates with RVP
Aortic coarctation	NSR	Normal	Normal or LAD	Normal	Possible LAE	LVH, especially by voltage criteria	Persistent RVH rare beyond infancy
Ebstein anomaly	NSR; possible EAR, SVT; AF/IART 40%	1° AVB common; short if WPW	Normal or LAD	Low-amplitude multiphasic atypical RBBB	RAE with Himalayan P waves	Diminutive RV	Accessory pathway common; Q II, III, aVF and V_1–V_4
Surgically repaired TOF	NSR; PVCs; IART 10%; VT 12%	Normal or mild ↑	Normal or RAD; LAD 5%–10%	RBBB 90%	Peaked P waves; RAE possible	RVH possible if RVOT obstruction or PHT	QRS duration ± QTd predictive of VT/SCD
L-TGA	NSR	1° AVB >50%; AVB 2%/year	LAD	Absence septal q; Q in III, avF and right precordium	Not if no associated defects	Not if no associated defects	Anterior AVN; Positive T precordial; WPW with Ebstein
D-TGA/intra-atrial baffle	Sinus brady 60%; EAR; junctional; IART 25%	Normal	RAD	Absence of q, small r, deep S in left precordium	Possible RAE	RVH; diminutive LV	Possible AVB if VSD or TV surgery
UVH with Fontan	Sinus brady 15%; EAR; junctional; IART >50%	Normal in TA; 1° AVB in DILV	LAD in single RV, TA, single LV with noninverted outlet	Variable; ↑↑ R and S amplitudes in limb and precordial leads	RAE in TA	RVH with single RV; possible LVH with single LV	Absent sinus node in LAI; AV block with L-loop or AVCD
Dextrocardia	NSR; P-wave axis 105°–165° with situs inversus	Normal	RAD	Inverse depolarization and repolarization	Not with situs inversus	LVH: tall R V_1–V_2; RVH: deep Q, small R V_1 and tall R right lateral	Situs solitus: normal P wave axis and severe CHD
ALCAPA	NSR	Normal	Possible LAD	Pathologic ant-lat Q waves; possible ant-sept Q waves	Possible LAE	Selective hypertrophy of posterobasal LV	Possible ischemia

AF = atrial fibrillation; ALCAPA = anomalous left coronary artery from the pulmonary artery; AVB = atrioventricular block; AVCD = atrioventricular canal defect; AVN = AV node; BVH = biventricular hypertrophy; CHD = congenital heart disease; DILV = double-inlet left ventricle; EAR = ectoptic atrial rhythm; IART = intra-atrial reentrant tachycardia; LAD = left-axis deviation; LAE = left atrial enlargement; LAHB = left anterior hemiblock; LAI = left atrial isomerism; LV = left ventricle; LVH = left ventricular hypertrophy; NSR = normal sinus rhythm; PDA = patent ductus arteriosus; PHT = pulmonary hypertension; RAD = right-axis deviation; RAE = right atrial enlargement;, PVC = premature ventricular contraction; RBBB = right bundle branch block (c = complete; i = incomplete); RV = right ventricle; RVH = right ventricular hypertrophy; RVOT = right ventricular outflow tract; RVP = right ventricular pressure; SCD = sudden cardiac death; SVT = supraventricular tachycardia; TA = tricuspid atresia; TOF = tetralogy of Fallot; TV = tricuspid valve; VSD = ventricular septal defect; VT = ventricular tachycardia; WPW = Wolff-Parkinson-White syndrome.

Reproduced with permission from Khairy P, Marelli AJ. Clinical use of electrocardiography in adults with congenital heart disease. Circulation 2007;116:2734–2746.[77]

5.3.1.2. Ambulatory ECG (Holter monitoring and event recorders). Indications for ambulatory monitoring and selection of recording technique in symptomatic patients with CHD are similar to those for the general population.[78] Standard Holter monitoring is best suited for the evaluation of daily symptoms or arrhythmias. The yield in evaluating sporadic symptoms such as syncope is generally low.[79,80] More recently, devices capable of longer-duration continuous recordings (typically 2–4 weeks) have become available, combining the best features of event recorders with the best attributes of Holter monitors. Event recorders come in 2 basic forms: noninvasive and implantable. The former are most commonly used. Although data are limited, in select cases where the index of suspicion for a malignant arrhythmia is high but noninvasive monitoring in not feasible or has been unrevealing, an implantable loop recorder may prove valuable.[75,81]

5.3.1.3. Cardiopulmonary exercise testing. Exercise testing has the advantage of providing data regarding rhythm in combination with functional status and may be useful for evaluation of patients with exertional symptoms. Although exercise testing does not typically result in reliable replication of sustained clinical tachyarrhythmias, it can provide information regarding sinus node behavior, AV conduction, and nonsustained tachyarrhythmias that may reflect the underlying cause of symptoms.[82]

5.3.1.4. Data from cardiac rhythm management devices. Modern pacemakers and implantable cardioverter-defibrillators (ICDs) have the ability to function like event recorders. Programmable parameters can allow the automatic recording of atrial, ventricular, or summed electrograms that meet specified criteria. In many devices, patients can actuate a recording using a programmable magnet response. Because of the long-term nature of these recordings, these devices can provide excellent information on arrhythmia burden. In a recent single-center retrospective series, 71% of patients underwent treatment modification as a result of device telemetry.[83]

5.3.1.5. Electrophysiologic study. Providing the clinical arrhythmia is present or can be induced during the procedure, diagnostic electrophysiologic studies offer the most definitive means of characterizing the essential components (location, mechanism, and other attributes) of the rhythm disturbance. The use of this test as a screening tool to assess risk of sudden cardiac death and ventricular arrhythmias is discussed in the evaluation of the asymptomatic patient.

5.3.2. Hemodynamic testing for symptomatic patients

Hemodynamic testing in the symptomatic adult with CHD may alter the pretest probability of finding a heart rhythm abnormality at the root of the patient's symptoms and determine the potential clinical impact of such a rhythm problem.[84] The first objective is to clearly define anatomy. The modified natural history of a given CHD lesion often depends extensively on the type of palliative or corrective interventions.[85] Review of operative reports in conjunction with some form of imaging study, if not recently performed, is generally indicated. The focus of the evaluation then shifts to the patient's hemodynamic and functional status. Identification of arrhythmogenic substrates, such as ventricular dysfunction, an enlarged or hypertrophied cardiac chamber related to valve dysfunction, or other hemodynamic derangement, may provide important diagnostic and prognostic clues.

5.3.2.1. Echocardiography. Widely available and noninvasive, transthoracic echocardiography is generally the initial imaging method of choice.[8] Although usually quite sensitive and accurate in assessing semilunar and AV valve dysfunction, as well as left ventricular size and function, the accuracy of echocardiography for quantitative assessment of right ventricular size and function has been questioned.[86] Echocardiography may also fall short in evaluation of systemic venous baffles where small hemodynamic gradients may be difficult to assess but nonetheless have important clinical implications. In these and other settings, additional testing may be indicated. Transesophageal echocardiography can be helpful if transthoracic echocardiographic windows are inadequate, if a prosthetic valve or material is present, and to better assess baffle function or complex CHD anatomy.[8]

5.3.2.2. Cardiac MRI. Cardiac MRI has become an increasingly important tool in evaluation of CHD patients with arrhythmias.[87] It provides data to supplement echocardiographic assessment of anatomy, valve performance, and ventricular function. In addition, images can be imported into arrhythmia mapping systems to provide 3-dimensional representations of the endocardial surface that can be adapted for substrate mapping and activation mapping of both atrial and ventricular tachycardias.[57,88] In circumstances where importing 3-dimensional image data to facilitate arrhythmia mapping is desired but MRI cannot be performed because of implanted cardiac rhythm management devices, CT imaging can be substituted for this purpose, although radiation exposure probably mandates that it only be performed when it will directly impact management. Given the expanding clinical indications for MRI in adults with CHD, when indicated, MRI conditional implantable cardiac arrhythmia devices should be considered.

5.3.2.3. Cardiac catheterization/angiography. Cardiac catheterization allows direct pressure measurements under controlled conditions. This may be particularly important in situations where small gradients can have an important clinical impact (e.g., Fontan, Mustard baffles). Coronary angiography should be considered in patients undergoing evaluation for ventricular arrhythmias who are over 40 years of age or those with additional cardiovascular risk factors such as congenital anomalies of the coronary arteries, coronary arteriovenous fistulae, a history of coronary surgery, or the potential for coronary compression by vascular conduits or stents.[89]

5.3.3. Recommendations for the evaluation and diagnosis of arrhythmias in symptomatic adults with CHD

Recommendations	
a. Noninvasive evaluation	
Class I	1. A thorough clinical history and physical examination should be conducted in adults with CHD and symptoms suggestive of arrhythmias (e.g., palpitations, presyncope, syncope), documented new-onset or worsening arrhythmias, or resuscitated sudden cardiac death *(Level of evidence: C)*.[90] 2. A resting 12-lead ECG is indicated in adults with CHD who are evaluated for arrhythmias *(Level of evidence: C)*.[77] 3. Ambulatory ECG monitoring is indicated when there is a need to clarify or exclude an arrhythmia diagnosis, correlate arrhythmias with symptoms, evaluate risk, or determine appropriate therapy *(Level of evidence: B)*.[78–80,91] 4. Cardiac event loop recorders are indicated to establish whether or not sporadic symptoms are caused by transient arrhythmias *(Level of evidence: C)*.[75,81] 5. Patients with suspected arrhythmias and implanted cardiac rhythm management devices should undergo device interrogation to retrieve diagnostic information provided by arrhythmia detection algorithms, trended data, histograms, and/or intracardiac electrogram recordings *(Level of evidence: B)*.[83,91,92] 6. Implantable loop recorders are useful in cases where the index of suspicion for a malignant arrhythmia is high (e.g., syncope) but a symptom–rhythm correlation cannot be established by conventional noninvasive techniques or invasive electrophysiologic testing *(Level of evidence: B)*.[81,93]
Class IIa	Cardiopulmonary exercise testing can be useful in adults with CHD and known or suspected exercise-induced arrhythmias in order to provoke the arrhythmia, establish a diagnosis, or assess response to therapy *(Level of evidence: C)*.[94,95]
Class IIb	Cardiopulmonary exercise testing may be useful in selected adults with CHD and arrhythmias as part of a broader workup to exclude triggering factors such as exercise-induced oxygen desaturation or myocardial ischemia *(Level of evidence: C)*.[94]
b. Hemodynamic workup	
Class I	1. Adults with CHD and new-onset arrhythmias, worsening arrhythmias, or resuscitated sudden cardiac death should undergo hemodynamic assessment, including transthoracic or transesophageal echocardiography, to rule out potentially contributory conditions such as regurgitant or obstructive lesions, shunts, ischemia, and ventricular dysfunction *(Level of evidence: B)*.[8,93,95] 2. Magnetic resonance imaging or cardiac computed tomography is useful in assessing adults with CHD and arrhythmias when cardiac structures or function cannot be reliably assessed by echocardiography or supplementary information is required *(Level of evidence: B)*.[57,88] 3. Coronary artery evaluation is indicated in assessing life-threatening ventricular arrhythmias or resuscitated sudden cardiac death in adults with CHD over 40 years of age and in those with CHD associated with a higher risk of coronary ischemia, such as congenital anomalies of the coronary arteries, coronary arteriovenous fistulae, a history of coronary surgery, or the potential for coronary compression by vascular conduits or stents *(Level of evidence: B)*.[89,96]
c. Electrophysiologic testing	
Class I	Electrophysiologic testing is indicated in adults with unexplained syncope and "high-risk" CHD substrates associated with primary ventricular arrhythmias or poorly tolerated atrial tachyarrhythmias, such as tetralogy of Fallot, transposition of the great arteries with atrial switch surgery, or significant systemic or single ventricular dysfunction *(Level of evidence: C)*.[76,91,97]
Class IIa	Electrophysiologic testing with programmed atrial and ventricular stimulation can be useful in adults with CHD and life-threatening arrhythmias or resuscitated sudden cardiac death when the proximate cause for the event is unknown or there is potential for therapeutic intervention at the time of the electrophysiologic procedure *(Level of evidence: B)*.[33,46,76,94,98]
Class IIb	Electrophysiologic testing may be considered in adults with CHD and palpitations suggestive of sustained arrhythmia when the conventional diagnostic workup is unrevealing *(Level of evidence: C)*.[94]

5.4. Approach to the asymptomatic patient

In adults with CHD, the high prevalence of arrhythmias, progressive functional deterioration, and risk of sudden death in the absence of overt premonitory clinical symptoms has led to the practice of surveillance monitoring and, in some cases, preemptive treatment. Evidence to base recommendations regarding which patients should be screened and which screening tests should be performed is limited but growing. In addition, it is not always clear that the detection of asymptomatic arrhythmias leads to or is an indication for a change in management. Still, some common sense and data-supported recommendations can be made.

5.4.1. Rhythm testing for asymptomatic patients

5.4.1.1. ECG. Even in the absence of symptoms, the ECG can provide important information about a patient's potential for certain arrhythmias.[77] In patients with Ebstein anomaly, for

example, the prevalence of Wolff-Parkinson-White syndrome is considerably higher than in the general population. Left untreated, the presence of an accessory pathway could have important implications.[99] The routine ECG can also provide useful information about the status of the sinus node and AV conduction, and abnormalities may prompt performance of longer-term recordings to determine whether the patient meets criteria for pacemaker implant.[97] It is also clear in many forms of CHD that sinus node dysfunction is a risk factor for development of atrial tachycardias,[100] and its presence should alert the clinician to monitor more carefully for this potential. In some cases, the ECG can provide electroanatomic data. For example, in patients with tetralogy of Fallot, there appears to be a mechanoelectrical interaction whereby the QRS duration on resting ECG and rate of QRS duration change over serial assessments correlates with right ventricular size and propensity for ventricular tachycardia and sudden death.[101,102] Other attributes such as QT dispersion have also demonstrated prognostic value for sudden cardiac death in adults with CHD.[103] Finally, surveillance ECG testing plays a role in therapeutic drug monitoring in patients on antiarrhythmic medications and other QT-prolonging drugs.

5.4.1.2. Ambulatory ECG (Holter monitoring and event recording). Holter monitors are perhaps the best studied arrhythmia surveillance test. In one recent single-center retrospective review, arrhythmias were found in 31% of Holters performed on a cohort of adults with CHD followed in an outpatient setting.[80] In this series, 80% of detected arrhythmias were asymptomatic. It is important to emphasize, however, that although the prevalence of asymptomatic arrhythmias in adults with CHD may be high, it is not clear that the detection of these arrhythmias modifies therapy. In another single-center retrospective series, for example, authors found that only 4% of surveillance Holters yielded findings that resulted in a change in clinical management, at a cost per clinically significant study of $12,732.[79] The yield on surveillance monitoring was better on older patients and those with transposition of the great arteries after Mustard/Senning or Fontan palliation.

In some cases, the implications of clinically silent arrhythmias are controversial. For example, nonsustained ventricular tachycardia in patients with tetralogy of Fallot appears to correlate strongly with inducible ventricular tachycardia[76] and risk of appropriate ICD discharge,[40] but the association of asymptomatic nonsustained ventricular tachycardia with sudden death is less clear.[101,104] Holter monitoring can also be used to assess autonomic nervous system function in adults with CHD.[75] For example, in patients with tetralogy of Fallot, abnormalities in heart rate variability have been correlated with age, right ventricular pressure, and end-diastolic dimension.[105] In a separate study, heart rate turbulence correlated with right and left ventricular function and peak VO_2 during exercise testing.[106] Finally, in a prospective study of 43 patients with a variety of congenital heart defects, heart rate variability and turbulence were found to be potent risk predictors for sudden cardiac death.[107] Despite the potential value of these data, the writing group felt that the studies to date are too preliminary to support broad-scale autonomic nervous system surveillance testing recommendations in adults with CHD.

Although some event recorders allow parameters to be set to trigger auto recordings, event recorders are typically used to identify arrhythmias associated with symptoms. For this reason, event recorders are not generally used for surveillance monitoring. Devices capable of long-duration continuous monitoring have proved useful in adults with atrial fibrillation.[108] It is reasonable to expect that these tools will be beneficial for adults with CHD as well, although they have not been systematically studied.

5.4.1.3. Cardiac rhythm management devices. All major pacemaker manufacturers have developed home telemonitoring systems. This provides a unique means of monitoring patients with these devices. In a recent study, the Home Monitoring TM system (HM; Biotronik, Berlin, Germany) was used in a cohort of patients with CHD.[83] The authors found that such systems are useful not only in monitoring device performance but also in detecting asymptomatic arrhythmias.

5.4.1.4. Electrophysiologic study. Although uncommonly used as a surveillance tool, electrophysiologic studies play a role in risk assessing patients with some forms of CHD. For example, in a multivariate analysis of a multicenter cohort of 252 patients with repaired tetralogy of Fallot, inducible sustained ventricular tachycardia was an independent risk factor for clinical ventricular tachycardia and sudden cardiac death.[76] However, programmed ventricular stimulation is insufficiently predictive to recommend as a screening tool for all patients with repaired tetralogy of Fallot.[84] It should rather be reserved for patients with additional risk factors, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration $\geq$180 ms, and extensive right ventricular scarring.[76] In other forms of CHD, such as transposition with intra-atrial baffles, programmed ventricular stimulation appears to be of little prognostic value.[46]

5.4.2. Hemodynamic testing for asymptomatic patients

Surveillance hemodynamic testing in asymptomatic adults with CHD has been addressed in existing guidelines.[8,54,55] Echocardiography and cardiac MRI are used routinely to monitor valve function and ventricular size/performance in conditions such as aortic stenosis, transposition with atrial switch procedures, and tetralogy of Fallot. Hemodynamic deterioration of various sorts has been correlated with an increased risk of both atrial and ventricular arrhythmias in many CHD lesions.[31,109–111] Firm guidelines for how often such testing should be performed and the exact threshold for primary prevention rhythm interventions are not yet clearly established for the CHD population. However, when periodic hemodymanic data are viewed in combination with surveillance rhythm testing, a more complete picture of an individual patient's risk can be developed that is modestly predictive of longer-term outcome.[84,101,112]

5.4.3. Recommendations for surveillance testing for arrhythmias in asymptomatic adults with CHD

Recommendations	
Class I	1. Surveillance for asymptomatic adults with CHD should follow established guidelines, including visits at regional ACHD centers at regular intervals for complex CHD, periodic intervals for CHD of moderate complexity, and occasionally for simple forms of CHD *(Level of evidence: C)*.[54,55,111]
	2. Surveillance for adults with moderate or severe CHD should include a standard 12-lead ECG at least once per year *(Level of evidence: C)*.[77]
	3. In adults with CHD and implanted cardiac rhythm management devices, routine follow-up should include device interrogation and review of stored diagnostic information *(Level of evidence: C)*.[83]
Class IIa	1. Periodic Holter monitoring can be beneficial as part of routine follow-up in adults with transposition of the great arteries and atrial switch surgery, Fontan palliation, and in patients with tetralogy of Fallot over 35 years of age *(Level of evidence: B)*.[79,80]
	2. Programmed ventricular stimulation can be useful in risk stratifying adults with tetralogy of Fallot who have additional risk factors for sudden cardiac death, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration $\geq$180 ms, and extensive right ventricular scarring *(Level of evidence: B)*.[40,76,83,91,92]
Class III	1. Programmed ventricular stimulation is not indicated as a screening tool to routinely risk stratify patients with tetralogy of Fallot at large *(Level of evidence: B)*.[76,84]
	2. Programmed ventricular stimulation does not appear to be of value for risk-stratifying adults with transposition of the great arteries with prior atrial switch surgery, in the absence of symptoms *(Level of evidence: B)*.[46]

6. Medical therapy

In this section, therapeutic options for the pharmacologic management of arrhythmias in adults with CHD are discussed, including acute termination of atrial and ventricular tachyarrhythmias, rate control and maintenance of sinus rhythm for intra-atrial reentrant tachycardia (IART) and atrial fibrillation, and prevention of thromboembolic complications.

6.1. Atrial tachyarrhythmias

6.1.1. Acute termination

Acute termination of atrial tachyarrhythmias in adults with CHD may be achieved by synchronized direct-current shocks, overdrive pacing, or pharmacologic agents. Supraventricular tachycardias dependent on AV nodal conduction and some nonautomatic focal atrial tachycardias may be terminated by vagal maneuvers, intravenous adenosine, or nondihydropyridine calcium channel antagonists (verapamil, diltiazem).[113] Regardless of the method used for cardioversion, sustained IART or atrial fibrillation $\geq$48 hours in duration is thought to incur substantial risk for thromboembolism.[114] A predisposition to thrombus formation accompanies several moderate and complex forms of CHD such that it may be prudent to rule out intracardiac thrombus prior to cardioversion in this setting, regardless of the duration of IART or atrial fibrillation.[115–118] Naturally, urgent cardioversion is recommended in adults with CHD who become hemodynamically unstable due to IART or atrial fibrillation irrespective of arrhythmia duration or anticoagulation status.[114] Anterior–posterior pad positioning may be needed in the setting of marked atrial dilation. Although direct-current cardioversion is the most common method used to rapidly terminate atrial tachyarrhythmias, overdrive pacing of IART may be considered in patients with atrial or dual chamber pacemakers or defibrillators.[119] Due care is required to ensure that the ventricle is not rapidly paced and that ventricular pacing is maintained during rapid atrial pacing in pacemaker-dependent patients.

There is a paucity of literature regarding pharmacologic conversion of IART or atrial fibrillation in adults with CHD. General concerns include risks of proarrhythmia, such as torsades de pointes with Class III drugs, ventricular tachycardia with Class IA and 1C drugs, and severe sinus bradycardia postconversion in adults with CHD predisposed to sinus node dysfunction. Advantages over direct-current cardioversion include the lack of required sedation/anesthesia.

In a series of 19 children, 15 of whom had CHD, ibutilide successfully converted IART or atrial fibrillation in 12 patients.[120] Ibutilide was used for 74 atrial tachyarrhythmias, with a conversion rate of 71%. No patient had a bradyarrhythmia, one patient with primary pulmonary hypertension and IART developed torsades de pointes, and a Fontan patient had nonsustained ventricular tachycardia. Similarly, in 19 patients with CHD (mean age 20 years), including 9 with Fontan physiology, a single 2 mg/kg oral dose of sotalol successfully converted IART or ectopic atrial tachycardia in 84%.[121] Two required emergent pacing for severe bradycardia, and one had a fatal thromboembolic event 2 days after conversion. In a head-to-head randomized comparison of intravenous ibutilide (1 or 2 mg) versus DL-sotalol (1.5 mg/kg) in 308 patients (mean age 60 years) without CHD, both doses of ibutilide were more effective than sotalol

in converting atrial flutter, whereas only the 2-mg ibutilide dose was superior to sotalol in converting atrial fibrillation.[122] Moreover, bradycardia and hypotension were more common with sotalol. Two of 211 patients (<1%) given 2 mg of ibutilide developed polymorphic ventricular tachycardia, one of whom required direct-current cardioversion. The risk of torsades de pointes with ibutilide may be as high as 4.3%[123] and has been reported to be greater in women[124] and in African Americans.[125]

Thus, in adults with CHD presenting with IART or atrial fibrillation, 1 to 2 mg of IV ibutilide administered over 10 minutes appears to be a reasonable option for pharmacologic cardioversion when used in a monitored setting where emergency defibrillation and resuscitation facilities are immediately available. There are no efficacy and safety data regarding acute conversion of IART or atrial fibrillation with Class IA, IC, and other Class III drugs (i.e., amdiodarone, dofetilide) in patients with CHD.

6.1.2. Long-term management

Experience with chronic pharmacologic therapy for IART in adults with CHD has been discouraging, resulting in a growing preference for nonpharmacologic options in most centers. Nevertheless, long-term pharmacologic therapy is used in many instances, including for patients in whom catheter ablation is not feasible or unsuccessful. The optimal pharmacologic approach to managing IART and atrial fibrillation in adults with CHD is as yet undetermined. In those with moderate or complex forms of CHD, a rhythm control treatment strategy (i.e., maintenance of sinus rhythm) is generally preferred to rate control as the initial management approach, in the absence of prospective outcome trials. However, there remains an important role for rate control as a potential therapeutic strategy in adults with simple forms of CHD and IART or atrial fibrillation,[114] and in those with moderate or complex CHD with failed attempts at rhythm control and in whom rate control is well tolerated, recognizing that vigorous efforts to achieve AV synchrony assume greater importance in certain lesions, such as univentricular hearts or systemic right ventricles with decreased contractility. Randomized clinical trials comparing rhythm to rate control strategies in adults with and without heart failure have reported similar all-cause and cardiovascular mortality, heart failure-related hospitalizations, thromboembolic events, and quality of life.[126–131]

6.1.2.1. Rate control. It is generally assumed that uncontrolled IART or atrial fibrillation is undesirable. Indeed, sudden cardiac death has been reported as a result of rapidly conducting atrial tachyarrhythmias in patients with systemic right ventricles[46] and univentricular hearts.[33] Rate control for IART or atrial fibrillation with AV nodal blocking drugs is based on the concept that rapid ventricular rates should be prevented in order to mitigate symptoms, improve exercise capacity, and preserve cardiac function. Clinical trials in adults with and without heart failure have included targets such as a maximum heart rate of 80 bpm at rest and <110 bpm on exertion.[126,127,132] However, more lenient objectives in patients without CHD, including a mean resting heart rate >80 bpm, have been associated with similar outcomes.[133,134] As such, some management guidelines have revised the recommended resting ventricular rate target to 100 bpm.[135] The applicability of more permissive heart rate objectives to adults with CHD and IART or atrial fibrillation, particularly in the setting of the univentricular circulation, remains to be demonstrated.

Beta-blocking drugs and nondihydropyridine calcium channel antagonists (verapamil, diltiazem) can be used to achieve ventricular rate control, with insufficient evidence to recommend one agent over another.[114,135] Although the choice of medication should be individualized, digoxin is not recommended as sole therapy to control the ventricular rate response, particularly in patients with paroxysmal atrial tachyarrhythmias,[114] and controversy exists as to whether it increases mortality.[136] Beta-blockers are associated with a decreased incidence of ventricular tachyarrhythmias in patients with transposition of the great arteries and atrial switch surgery,[46] such that it may be reasonable to liberalize use of beta-blockers in this patient population if well tolerated. Nondihydropyridine calcium channel antagonists and digoxin are generally avoided in the presence of preexcitation because they may paradoxically accelerate the ventricular response rate.

6.1.2.2. Rhythm control. Before initiating antiarrhythmic therapy for IART or atrial fibrillation in adults with CHD, precipitating factors should be sought and reversible causes treated. The selection of pharmacologic agents should consider coexisting sinus node or AV node disease, heart failure, associated therapies, child-bearing potential, and comorbidities. Considering the limitations of antiarrhythmic drugs, infrequent well-tolerated recurrences of IART or atrial fibrillation is a reasonable objective.[114] Management guidelines in patients with little or no heart disease have considered flecainide, propafenone, and sotalol to be acceptable first-line antiarrhythmic agents for long-term maintenance of sinus rhythm.[114,135]

Class IC drugs have been associated with increased mortality in patients with ventricular scarring due to myocardial infarction[137,138] and in those with heart failure.[139,140] This is thought to be due, in part, to facilitation of reentrant ventricular tachycardia by decreased conduction in addition to spatially heterogeneous action potential prolongation.[141] A meta-analysis likewise found that Class IA drugs (i.e., quinidine and disopyramide) were associated with increased all-cause mortality in adults with atrial fibrillation.[142] As such, Class I agents are not recommended in patients with coronary artery disease or ventricular dysfunction.[114,135] Adults with CHD frequently have residual hemodynamic disturbances, incisional scars, intracardiac baffles, conduits, and/or extensive areas of myocardial fibrosis that may predispose to potentially fatal proarrhythmic effects from Class I agents. Yet, potential proarrhythmic risk remains

ill-defined in this patient population. In 579 young patients who were administered encainide or flecainide, 24% of whom had CHD, proarrhythmic events were observed in 7.5% of patients with encainide and 7.4% with flecainide.[143] Cardiac arrest (N = 12) and deaths (N = 13) occurred predominantly among those with underlying heart disease. In a subsequent study of 121 patients with tetralogy of Fallot, Class I agents were associated with a nearly two-fold but nonsignificant increased risk of ventricular arrhythmias.[40] Pending further safety data, the writing committee deemed it prudent to discourage Class I antiarrhythmic drug use in adults with CHD and coronary artery disease or systolic dysfunction of a systemic or subpulmonary ventricle.

General guidelines for atrial fibrillation support the use of sotalol (Class IIa indication) for maintenance of sinus rhythm in patients with little or no heart disease, an uncorrected baseline QT interval <460 ms, normal serum electrolytes, creatinine clearance >40 mL/min, and absence of risk factors associated with Class III drug-related proarrhythmia.[114] Although small retrospective studies suggest that sotalol is associated with reasonable safety and efficacy in adults with CHD,[144–146] other case series in children with and without CHD reported low efficacy and high proarrhythmia rates.[147] In a meta-analysis of antiarrhythmic drugs for atrial fibrillation, which included 12 clinical trials with 3002 patients randomized to sotalol (N = 1791) versus control (N = 1211) therapy, all-cause mortality was significantly higher with sotalol [i.e., odds ratio 2.47, 95% confidence interval (CI) (1.21, 5.05), $P = .01$].[142] A second meta-analysis that used a mixed treatment comparison reported similar results.[148] Increased mortality with sotalol was even more pronounced when small studies randomizing <100 subjects were excluded. In light of these concerns, this writing committee relegated the use of sotalol to a Class IIb indication as a first-line antiarrhythmic agent for the maintenance of sinus rhythm in adults with CHD, IART or atrial fibrillation, and preserved ventricular function.

Amiodarone is the most effective antiarrhythmic agent for maintaining sinus rhythm in patients with atrial fibrillation[149] and is the drug of choice in the setting of heart failure.[126] However, long-term therapy is limited by time- and dose-dependent side effects, particularly in young adults. These include pulmonary and liver toxicity, corneal microdeposits, photosensitivity, thyroid dysfunction (hypo- or hyperthyroidism), and adverse cardiac effects (e.g., bradycardia, torsades de pointes). Amiodarone-induced thyrotoxicosis is especially common in women with CHD and cyanotic heart disease or univentricular hearts with Fontan palliation,[150] and in those with a body mass index <21 kg/m^2.[151,152] While respecting standard precautions, amiodarone may be considered a first-line antiarrhythmic agent for the long-term maintenance of sinus rhythm in adults with CHD and IART or atrial fibrillation in the presence of ventricular hypertrophy or dysfunction, or coronary artery disease.[144,150,152,153] In the absence of such coexisting conditions, it is best reserved as a second-line agent.[144,150,152] Importantly, nonpharmacologic options should be thoughtfully considered prior to committing a young adult with CHD to long-term amiodarone therapy.

Dronaderone, an amiodarone analog without the iodine moiety, is less effective at maintaining sinus rhythm.[135,142,154,155] It has been associated with increased mortality related to worsening heart failure in patients with left ventricular systolic dysfunction.[156] Moreover, in patients ≥ 65 years of age with at least a 6-month history of permanent atrial fibrillation and risk factors for vascular disease, dronedarone was associated with increased rates of heart failure, stroke, and cardiovascular mortality.[157] Rare cases of liver failure and pulmonary toxicity have also been reported. As such, dronedarone is not recommended in patients with heart failure, moderate or severe systolic ventricular dysfunction, or moderate or complex CHD.

Dofetilide is a class III antiarrhythmic agent that selectively inhibits the rapid component of the delayed rectifier potassium current.[158] Important to extrapolations for adults with CHD, it has not been associated with increased mortality in high-risk patients with recent myocardial infarction or heart failure.[159–163] Because dofetilide is excreted by the kidneys, dosing must be adjusted to the creatinine clearance level to minimize risk of torsades de pointes (0.9%–3.3%).[161,162] It should not be administered if the QTc is >440 ms or ≥ 500 ms in the presence of ventricular conduction delay. Therapy should be initiated under continuous cardiac monitoring for a minimum of 72 hours. In general, the dose should be reduced if the QTc increases by $>15\%$ after the first dose or if the QTc exceeds 500 ms or 550 ms with a ventricular conduction delay.[88] Dofetilide was associated with reasonable success in a multicenter series of 20 adults with CHD and refractory atrial arrhythmias, 14 of whom had attempted catheter ablation.[164] Two patients who received 500 μg twice daily experienced torsades de pointes, one with truncus arteriosus and the second with a single ventricle and Fontan palliation. By adhering to strict Food and Drug Administration (FDA)-mandated guidelines regarding administration, dofetilide appears to be a reasonable alternative to amiodarone as a first-line antiarrhythmic drug in adults with CHD and ventricular dysfunction, or as a second-line agent.[114,135]

6.1.3. Recommendations for pharmacologic therapy in preventing recurrent Intra-Atrial Reentrant Tachycardia (IART) or atrial fibrillation

Recommendations	
Class I	In adults with CHD, the choice of pharmacologic therapy for arrhythmia management should consider factors such as coexisting sinus node dysfunction, impaired AV nodal conduction, systemic or subpulmonary ventricular dysfunction, associated therapies, child-bearing potential, and acquired comorbidities *(Level of evidence: B)*.[142,144]
Class IIa	1. In adults with CHD and paroxysmal or persistent IART or atrial fibrillation, an initial strategy of rhythm control is reasonable, particularly in the setting of moderate or complex CHD *(Level of evidence: C)*. 2. It is reasonable to manage adults with simple forms of CHD and IART or atrial fibrillation according to previously published guidelines for antiarrhythmic therapy in adults with atrial fibrillation or flutter and no or minimal heart disease *(Level of evidence: C)*.[114,135] 3. In the pharmacologic management of adults with CHD of any complexity, IART or atrial fibrillation, and normal AV conduction, it is reasonable to include adequate AV nodal blockade to prevent a rapid ventricular response (*Level of evidence: B*).[114,135] 4. In adults with CHD and frequent recurrent symptomatic IART, an ablation strategy is preferable to long-term pharmacologic therapy *(Level of evidence: B)*.[165–171] 5. Amiodarone can be considered a first-line antiarrhythmic agent for the long-term maintenance of sinus rhythm in adults with CHD and IART or atrial fibrillation in the presence of pathologic hypertrophy of the systemic ventricle, systemic or subpulmonary ventricular dysfunction, or coronary artery disease *(Level of evidence: C)*.[153] It should be used with caution in patients with cyanotic heart disease, a low body mass index (<21 kg/m^2), concomitant hepatic, pulmonary, or thyroid disease, or an uncorrected QT interval >460 ms or ≥ 500 ms in the presence of ventricular conduction delay (*Level of evidence: B*).[144,150,152] 6. In the absence of a coexisting condition listed above and subject to the stated precautions, it is reasonable to consider amiodarone as a second-line antiarrhythmic agent for the long-term maintenance of sinus rhythm in adults with CHD and IART or atrial fibrillation (*Level of evidence: B*).[144,150,152] 7. Subject to standard precautions and barring any contraindication (e.g., creatinine clearance <20 mL/min, hypokalemia, QTc >440 ms or ≥ 500 ms in the presence of ventricular conduction delay), dofetilide is probably a reasonable alternative to amiodarone in adults with CHD and systemic ventricular dysfunction or as a second-line antiarrhythmic agent (*Level of evidence: B*).[164,172]
Class IIb	1. It may be reasonable to liberalize the use of beta-blockers in patients with transposition of the great arteries, atrial switch surgery, and IART to protect against ventricular arrhythmias and sudden cardiac death (*Level of evidence: B*).[46,173] 2. Subject to standard precautions (e.g., renal insufficiency, hypokalemia, severe sinus node dysfunction or AV nodal disease, uncorrected QT interval >460 ms or ≥ 500 ms in the presence of ventricular conduction delay), sotalol may be considered a first-line antiarrhythmic agent for the long-term maintenance of sinus rhythm in adults with CHD and IART or atrial fibrillation (*Level of evidence: B*).[142,144,146]
Class III	1. Oral class I antiarrhythmic agents are not recommended for the maintenance of sinus rhythm in adults with CHD and IART or atrial fibrillation who have coronary artery disease or moderately to severely depressed systolic dysfunction of a systemic or subpulmonary ventricle (*Level of evidence: B*).[137–140,143] 2. Dronedarone is not recommended in patients with a history of heart failure, moderate or severe systolic ventricular dysfunction, or moderate or complex CHD because of potential concerns over worsening heart failure and increased mortality (*Level of evidence: B*).[156,157]

Figure 6.1 summarizes the recommended approach to rhythm control in adults with CHD and IART or atrial fibrillation.

6.1.3.1. Thromboprophylaxis. Prevention of thromboembolism is a major objective of pharmacologic therapy in adults with CHD and IART or atrial fibrillation.[117] Few studies have explored the association between IART or atrial fibrillation and thromboembolic complications in CHD.[26,33,118] In a series of 19 patients with CHD who underwent transesophageal echocardiography prior to cardioversion of an atrial tachyarrhythmia, atrial thrombus was detected in 37%.[116] In this small series, a strategy of anticoagulation targeting international normalized ratio (INR) values ≥ 2 for at least 4 weeks prior to cardioversion, with transesophageal echocardiography reserved for high-risk patients (e.g., complex CHD, mechanical valve, prior thromboemboli, systemic hypertension, heart failure, or ventricular dysfunction), was associated with a low rate of cardioversion-induced systemic thromboemboli.[174]

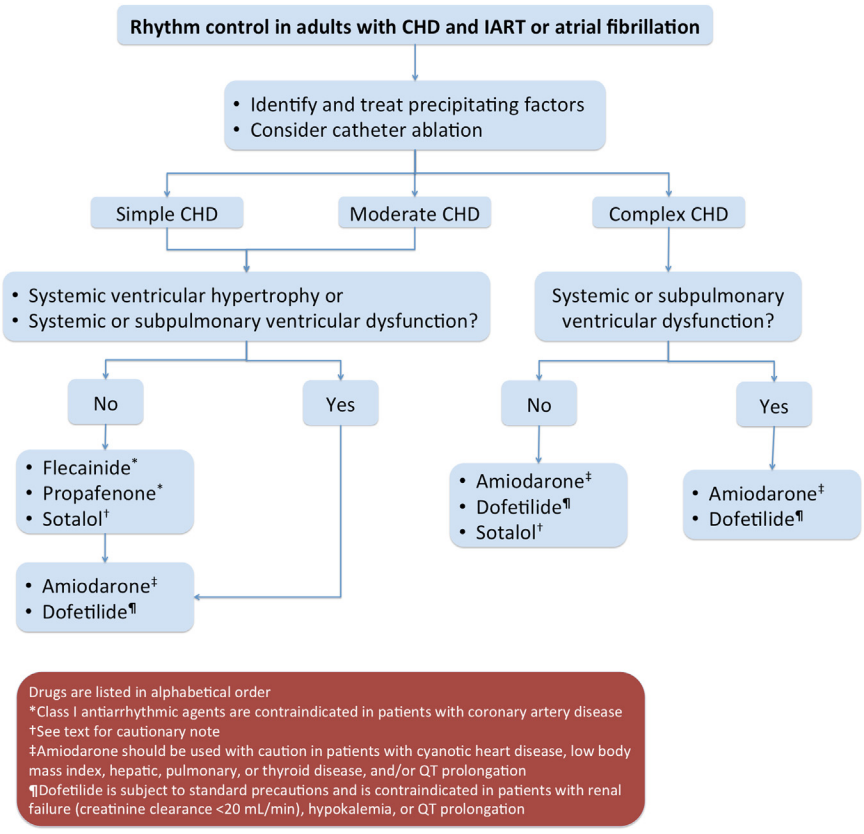

Figure 6.1 Rhythm control in adults with congenital heart disease (CHD) and intra-atrial reentrant tachycardia (IART) or atrial fibrillation.

Standard management guidelines for the prevention of thromboembolism in patients with nonvalvular atrial fibrillation or flutter recommend anticoagulation for at least 3 weeks before and 4 weeks after cardioversion for an arrhythmia of unknown or ≥48 hours' duration, regardless of the method used for cardioversion.[114,135] As an alternative to 3 weeks of anticoagulation prior to cardioversion, it is deemed reasonable to perform transesophageal echocardiography in search of intracardiac thrombus.[114,135,175,176] During the first 48 hours, the need for anticoagulation may be based on the patient's risk of thromboembolism.[114,135] Hemodynamically unstable arrhythmias should be immediately cardioverted regardless of their duration. Additionally, certain patients, such as those with a univentricular circulation, may not tolerate prolonged periods with loss of AV synchrony and may benefit hemodynamically from prompt cardioversion. With regard to long-term anticoagulation, all patients with atrial fibrillation or flutter should be stratified according to stroke and bleeding risks.[177] A combination of risk scoring systems led to the development of the $CHADS_2$ [Congestive heart failure (or systemic left ventricular systolic dysfunction), Hypertension, Age ≥75, Diabetes, Stroke (doubled)] score, which, although rather simple, is limited by poor sensitivity in identifying the lowest-risk patients.[178] The expanded CHA_2DS_2-VASC scoring system, which incorporates the additional risk factors of Vascular disease (prior myocardial infarction, peripheral artery disease, aortic plaque), younger Age (age 65–74 years), and Sex (female), appears to better identify low-risk patients in whom oral anticoagulant therapy is not beneficial.[179] Considering these scores, antiplatelet or anticoagulation therapy is recommended in most patients with atrial fibrillation or flutter.[135]

Risk scores predicting thromboembolic complications in patients with atrial fibrillation or flutter do not consider the presence, type, or severity of CHD. In the absence of large-scale prospective studies, the writing committee generally recommends pursuing a similar approach to anticoagulation/transesophageal echocardiography prior to cardioversion in adults with CHD and IART or atrial fibrillation of unknown or ≥48 hours' duration.[174] However, considering that adults with moderate or complex forms of CHD may be predisposed to thrombus formation even in the absence of atrial tachyarrhymias, it would appear prudent to pursue therapeutic anticoagulation for at least 3 weeks prior to cardioversion or perform transesophageal echocardiography to rule out thrombus in this setting, even if the IART or atrial fibrillation is <48 hours in duration.[180] The overall prevalence of thromboembolic complications in patients with CHD has been estimated to be 10- to 100-fold higher than in age-matched controls.[118] The varied pathophysiology reflects diverse predisposing substrates and includes dilated cardiac chambers with sluggish flow, intracardiac prosthetic material, pacemaker/defibrillator leads, intracardiac shunts, and associated hypercoagulable states.[117,118,181–183] Patients with Fontan palliation are at particularly high risk for

thromboembolic complications,[33,184–191] such that transesophageal echocardiography may be sensible prior to cardioversion even if therapeutic anticoagulation is received for ≥3 weeks.[116,192]

Long-term oral anticoagulation is recommended in the adult with CHD of severe complexity and IART or atrial fibrillation, and appears reasonable in those with moderate forms of CHD.[118,193–196] It is unlikely that the thromboembolic risk associated with simple nonvalvular forms of CHD is sufficiently high to justify long-term anticoagulation as a de facto approach, such that the decision to pursue antiplatelet or anticoagulation therapy in this subgroup of patients may be guided by established risk scores for stroke (e.g., CHA_2DS_2-VASc) and bleeding risk (e.g., HAS-BLED).[177,197]

Drawbacks of oral vitamin K antagonists in young patients are well known.[190,198] Several newer oral anticoagulant drugs (NOACs) have been developed to overcome some of these issues, which include fluctuations in anticoagulation effects, frequent dose adjustments, and regular serum monitoring. NOACs exert their effect by reversibly inhibiting thrombin (i.e., dabigatran) or factor Xa (i.e., rivoraxaban, apixaban, edoxaban). Several clinical trials in patients with atrial fibrillation have found NOACs to be noninferior or superior to warfarin in the prevention of stroke and systemic emboli without increasing the rate of major bleeds, while significantly reducing the incidence of intracranial bleeds.[199–202] When anticoagulation is indicated in patients with atrial fibrillation or flutter, management guidelines increasingly favor NOACs over warfarin.[114,135] In the absence of CHD-specific data, it may be reasonable to consider a NOAC as an alternative to a vitamin K antagonist in patients with simple forms of CHD and no prosthetic heart valve or hemodynamically significant valve disease.[199–201,203] In contrast, there are currently insufficient safety and efficacy data to recommend NOACs in those with moderate or complex forms of CHD. In particular, NOAC use in patients with Fontan surgery, with its associated high prevalence of hepatic impairment and altered coagulation, cannot be recommended in the absence of pharmacokinetic, pharmacodynamic, and safety data. If such data become available, the recommendations should be revised accordingly.

6.1.3.2. Recommendations for thromboprophylaxis

Class I	1. For adults with simple forms of CHD and hemodynamically stable IART or atrial fibrillation of unknown or ≥48-hours' duration, therapeutic anticoagulation is recommended for at least 3 weeks prior to cardioversion, or, alternatively, a transesophageal echocardiogram may be performed to rule out intracardiac thrombus *(Level of evidence: B)*.[114,153,174–176]
	2. Adults with complex CHD and sustained or recurrent IART or atrial fibrillation should receive long-term oral anticoagulation for the prevention of thromboembolic complications *(Level of evidence: B)*.[118,193–196]
Class IIa	1. For adults with moderate or complex CHD and hemodynamically stable IART or atrial fibrillation, it is reasonable to pursue therapeutic anticoagulation for at least 3 weeks prior to cardioversion or perform transesophageal echocardiography to rule out thrombus, regardless of arrhythmia duration *(Level of evidence: B)*.[33,180]
	2. Long-term oral anticoagulation therapy is reasonable in adults with CHD of moderate complexity and sustained or recurrent IART or atrial fibrillation *(Level of evidence: C)*.[118,193–196]
	3. Vitamin K antagonists can reasonably be considered the oral anticoagulant agent of choice in adults with moderate or complex CHD, pending safety and efficacy data on newer oral anticoagulants (NOACs; i.e., direct thrombin inhibitors and direct factor Xa inhibitors) *(Level of evidence: B)*.[193–196,202]
Class IIb	1. It may be reasonable for adults with IART or atrial fibrillation and simple nonvalvular forms of CHD to receive either an oral anticoagulant, aspirin, or no therapy for the prevention of thromboembolic complications on the basis of established scores for stroke risk (e.g., CHA_2DS_2-VASc) and bleeding risk (e.g., HAS-BLED) *(Level of evidence: B)*.[177,197]
	2. In adults with simple forms of CHD and no prosthetic heart valve or hemodynamically significant valve disease, a NOAC may be a reasonable alternative to a vitamin K antagonist when anticoagulation is indicated *(Level of evidence: C)*.[199–201,203]
`Class III	1. Pending future studies, there are currently insufficient pharmacokinetic/pharmacodynamic, safety, and efficacy data to endorse use of NOACs in adults with Fontan surgery *(Level of evidence: C)*.
	2. Anticoagulation is not indicated for the prevention of thromboembolic complications in adults with CHD and AV nodal reentrant tachycardia or accessory pathway–mediated tachycardia *(Level of evidence: C)*.

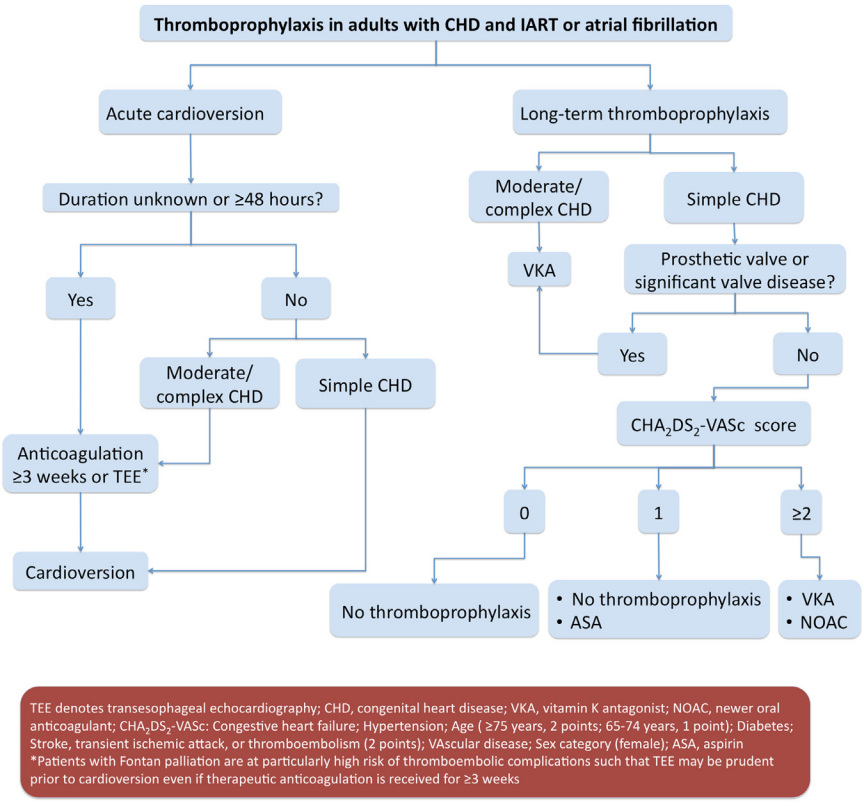

Figure 6.2 Thromboprophylaxis in adults with congenital heart disease (CHD) and intra-atrial reentrant tachycardia (IART) or atrial fibrillation

Recommendations for thromboprophylaxis in adults with CHD and IART are summarized in Figure 6.2.

6.2. Ventricular tachyarrhythmias

6.2.1. Acute termination

In adults with CHD, hemodynamically poorly tolerated ventricular tachycardia or fibrillation resulting in pulseless arrest requires management according to AHA/ACC/ESC guidelines for Adult Cardiac Life Support (ACLS).[94,204] Hemodynamically tolerated ventricular tachycardia should also be managed according to well-established adult guidelines,[94,204] while taking into consideration CHD-specific issues. For example, when direct-current cardioversion or defibrillation is required, the energy delivery vector (by chest surface paddles or patches) should take into account the cardiac location within the chest. This is important in the occasional patient with meso- or dextrocardia. Cardioversion, whether electrical or pharmacologic, should be performed expeditiously for any sustained ventricular tachyarrhythmia. Electrical cardioversion, although highly effective for reentrant ventricular tachycardia, has the disadvantage of requiring sedation. Drug therapy, although convenient, may have a delayed effect.

The mechanism for the hemodynamically tolerated monomorphic ventricular tachycardia in the young adult with surgically repaired tetralogy of Fallot is most often macroreeentry.[205,206] Intravenous preparations of amiodarone, procainamide, and lidocaine are widely available, although procainamide is more effective in rapidly terminating macroreentrant monomorphic ventricular tachycardia.[205–207] Amiodarone and procainamide can cause hypotension, requiring continuous blood pressure monitoring during administration. Lidocaine is most effective for ventricular tachycardia emanating from partially depolarized regions, as occurs in ischemic myocardium. Importantly, CHD does not render the individual immune from acquired forms of ventricular tachycardia, including ischemic and idiopathic forms. When triggered activity is the underlying mechanism, electrical cardioversion can be unhelpful, and intravenous adenosine or calcium channel antagonists may be preferred. Such agents may be harmful in the presence of scar-related macroreentry or in the presence of ischemic ventricular tachycardia.

6.2.2. Long-term management

As discussed in Section 9, the ICD is first-line therapy for the secondary prevention of sudden death in adults with CHD. Antiarrhythmic pharmacotherapy may be helpful in reducing recurrent ICD discharges. To that end, the only data available are from adults with ischemic cardiomyopathy and those with reduced ventricular function, primarily with dilated cardiomyopathy. With regard to drug efficacy, the correlation between those well-studied patient groups and adults with CHD is conjectural. Notwithstanding these caveats, sotalol has been associated with longer times to first appropriate and inappropriate ICD shocks and a reduced

frequency of shocks.[208] In a meta-analysis of 15 trials, 9 of which included patients with reduced left ventricular ejection fraction, amiodarone was associated with a 29% reduction in sudden cardiac death, with a nonsignificant effect on all-cause mortality.[209] Amiodarone combined with a beta-blocker is more effective than sotalol at preventing ICD shocks but is associated with an increased risk of drug-related adverse events.[210]

Small case series in patients with CHD from the era of serial drug testing reported favorable outcomes with mexiletine[211] and phenytoin,[212,213] but not Class I antiarrhythmic drugs.[214] Six patients with tetralogy of Fallot or double-outlet right ventricles who failed catheter ablation for ventricular tachycardia were rendered noninducible by sotalol or amiodarone.[215] In patients with drug-refractory ventricular tachycardia, a retrospective cohort study suggests that mexiletine may be added to amiodarone to reduce appropriate ICD therapies.[216] A small case series also raised the possibility that ranolazine, a drug that exerts anti-ischemic effects and also acts as an antiarrhythmic in isolation and in combination with other Class III agents, may be effective in reducing ICD shocks in refractory patients.[217]

7. Catheter ablation

7.1. General considerations for catheter ablation in adults with CHD

Decisions regarding catheter ablation for recurrent atrial, ventricular, and/or supraventricular tachycardias in adults with CHD depend, in part, on anticipated procedural success rates and associated risks, symptoms, and hemodynamic tolerance. Preprocedural evaluation should include documentation and analysis of all arrhythmias. Reports from previous surgical and catheter ablation procedures should be reviewed, and thorough knowledge of 3-dimensional cardiac anatomy obtained by echocardiography, MRI and/or CT scan.[7,218,219] Vascular access may be hampered by vascular anomalies or prior interventions such that venography can be considered. In the case of occluded veins, alternative routes such as internal jugular, subclavian, or in rare instances transhepatic access can be planned.[220] Preprocedural preparation includes insurance of a multidisciplinary team experienced with CHD (electrophysiologist, anesthesiologist, and, when deemed appropriate, cardiac surgical backup). In the event of substantial noncardiac cormorbidities or ventricular dysfunction, which predict potential postprocedural cardiorespiratory instability, need for invasive monitoring, or advanced nursing care, arrangements for an intensive care unit bed should ideally be planned beforehand.[221] In addition, the need to import MRI or CT images into an electroanotmic mapping system,[222] perform angiography of the chamber of interest, assess hemodynamics, or access the pulmonary venous atrium by a transseptal/baffle puncture[223] or retrograde via the aorta should be taken into account.

7.2. AV reciprocating tachycardia and AV nodal reentrant tachycardia

7.2.1. Epidemiology

There are well-documented associations between certain forms of CHD and AV reciprocating tachycardia, most notably the common and long-recognized co-occurrence of Ebstein anomaly and accessory pathway–mediated tachycardia.[224] Congenitally corrected transposition of the great arteries is associated with an Ebstein-like malformation of the systemic tricuspid valve and accessory pathways. A less frequently identified substrate for supraventricular tachycardia is twin AV nodes in certain heterotaxy variants.[29,225,226] Given the careful and frequent oversight of cardiac care in children with CHD and the near universal performance of a resting ECG at outpatient examinations, it is relatively uncommon for these specific problems to evade diagnosis until adulthood. Nevertheless, Ebstein anomaly and congenitally corrected transposition of the great arteries are occasionally diagnosed in adulthood on presentation of an arrhythmia, and the need or opportunity for treatment may sometimes be delayed past childhood. Additionally, there are rare instances of acquired Wolff-Parkinson-White syndrome in patients who have undergone congenital heart surgery, presumably due to an acquired functional epicardial AV connection.[227] AV nodal reentrant tachycardia has also been reported uncommonly in patients with CHD,[228–232] but little is known about its associations and natural history.

7.2.2. Mechanistic considerations

In light of the well-known association between Ebstein anomaly and accessory pathways and recent advances in surgical reconstruction of the tricuspid valve, electrophysiologic testing and catheter ablation are becoming more routine preoperative interventions.[68] It has long been recognized that the effect of atrialization of the right ventricle in Ebstein anomaly results in unusually fractionated and low-amplitude electrograms at and below the AV groove, making mapping of the accessory pathway using standard techniques challenging.[233,234] Intracoronary mapping using fine electrode wires may be useful in this setting.[235] Mahaim-type atriofascicular pathways are more common in Ebstein anomaly, and it is also frequently the case that multiple AV pathways are present.[236] This often includes coexisting concealed and manifest accessory pathways. Diagnosis and ablative management of patients with Ebstein anomaly may further be complicated by the increased prevalence of atrial and ventricular tachycardias.

7.2.3. Catheter ablation

As with most tachyarrhythmias in patients with CHD, the drivers of ablation will typically include some combination of unpredictable and poorly controlled symptoms, electrophysiologic risk (in the case of Wolff-Parkinson-White syndrome), hemodynamic vulnerability, and thromboembolic risk. Certain aspects related to co-occurrence of congenital lesions are of technical importance in planning

and performing ablations. Knowledge of vascular access limitations and exclusion of sections of the AV groove by surgical baffling are relevant to procedural planning.[237–239] With respect to planning ablation, the utility of algorithms for predicting the location of accessory pathways in patients with CHD by surface ECGs is limited.[240] Importantly, the AV node can be displaced such that its precise location may be unclear. Occasionally, identification of the His-bundle electrogram may be impossible, further complicating septal ablation and slow pathway modification in AV nodal reentrant tachycardia.

7.2.4. Ablation outcomes

Much of the outcomes literature on catheter ablation in CHD is in the form of mixed series, both in terms of anatomic and electrophysiologic diagnoses. Initial mixed case series of small numbers of patients with CHD suggested that supraventricular tachycardia ablation was feasible, but that acute clinical success rates were lower than those seen in normal anatomy.[225,241,242] In the largest series centered on ablation of pathway-mediated and AV nodal reentrant tachycardia in CHD (i.e., 105 procedures in 83 patients), an acute success rate of 80% was reported.[237] Data from the Pediatric Radiofrequency Registry suggest that patients with CHD have a higher catheter ablation procedural mortality risk than those with normal hearts.[243]

Acute ablation success rates for patients with Ebstein anomaly are lower than for patients with normal anatomies.[234,244,245] The Pediatric Radiofrequency Registry included 65 patients with Ebstein anomaly and 87 accessory pathways (including Mahaim fibers), 7 of whom had concomitant AV nodal reentrant tachycardia.[244] Other series included 21 patients with 34 pathways[234] and 32 patients with 34 pathways and 1 AV nodal reentrant tachycardia.[245] These series emphasized the occurrence of multiple arrhythmia mechanisms and the importance of accurate identification of the AV groove. Taken together, overall success rates ranged from 75%–88%, with recurrences in 27%–40% of cases.

Little has been written beyond case reports of patients who have undergone clinically successful ablation of AV nodal reentrant tachycardia in complex CHD. The site of the slow pathway has in some cases been imputed to be at nonstandard anatomical locations, based on apparent response to ablation.[231,232] In small subsets of cases reported in the context of larger series on Ebstein anomaly and transposition of the great vessels, successful slow pathway modification was successful at the expected posterior aspect of the AV septum.[237,245,246]

7.3. Atrial tachyarrhythmias

7.3.1. Epidemiology

Propensity for arrhythmias increases with time since cardiac surgery such that late postoperative atrial tachyarrhythmias are increasingly encountered in daily practice.[247] The prevalence of late postoperative atrial tachycardias varies between 4% and 30%,[31,247–250] depending, in part, on the complexity of the underlying CHD and duration of follow-up. Atrial tachyarrhythmias may cause hemodynamic deterioration, thromboembolic complications, and even sudden cardiac death,[26,33,46,248,251] and are associated with a 2-fold increased risk for mortality.[26,251] Independent predictors for mortality include poor functional class, single ventricle physiology, pulmonary hypertension, and valvular heart disease.[252]

7.3.2. Mechanistic considerations

The atria of adults with CHD are often damaged extensively by cardiac surgery and ongoing postoperative pressure and/or volume overload, resulting in impaired electrical conduction. Electrophysiologic testing in patients after Fontan surgery for single ventricles and Mustard repair for transposition of the great arteries demonstrated prolongation of atrial refractoriness and areas of intra-atrial conduction delay.[253–255] These electropathologic alterations, combined with sinus node dysfunction and frequent atrial premature beats, render adults with CHD vulnerable to developing postoperative atrial tachyarrhythmias. The most common mechanism is macroreentry within the atrial musculature, so-called IART.[6,256] Anatomic structures, areas of scar tissue, long suture lines, cannulation sites, or surgically inserted prosthetic materials often form the boundaries of these reentrant circuits.[257–260] Separation of atrial muscle bundles by fibrous tissue enhances the complexity of IART circuits as they form multiple corridors within areas of scar tissue.[257,261,262] Ectopic atrial tachycardias are less common but not infrequently observed.[260,263–266] They are typically caused by focal activity originating from low-voltage areas.[266] Their underlying mechanism is unclear, although mapping studies are suggestive of microreentry.[265,267]

7.3.3. Mapping and ablation

Late postoperative atrial tachyarrhythmias in adults with CHD are most often due to cavotricuspid isthmus-dependent (counterclockwise or clockwise) flutter or scar-based macroreentry.[44,266,268–273] Catheter ablation has proven to be safe and considerably effective.[241,246,274,275] As a curative treatment modality, it is generally preferred over long-term pharmacologic management. Reported procedural success rates range from 72% to 77%, depending in part on the complexity of the underlying defects.[260,276,277] Usage of 3-dimensional electroanatomic mapping systems for guiding ablative therapy is recommendable,[266,268,278–280] considering that atrial anatomy is often distorted. These techniques are helpful in visualizing anatomic structures, scar tissue areas, and prosthetic materials, thereby providing insight into the arrhythmogenic substrate.

Target sites for ablation are selected by combining activation and voltage mapping with entrainment maneuvers.[261,262,276,281,282] Activation and entrainment mapping aid in distinguishing reentry from focal activity; voltage mapping localizes areas of scar tissue and entrainment mapping determines whether a specific area is a crucial component of the reentrant circuit. IART is ablated by creating linear lesions within the reentrant circuit, thereby transecting critical conduction pathways.[261,262,269] When the cavotricuspid isthmus is

involved, bidirectional conduction block is sought. In the case of ectopic atrial tachycardia, the site with the earliest activation relative to the P wave is localized. This area can be directly targeted or encircled by ablation lesions. Irrigated tip catheters are associated with improved outcomes for ablation of postoperative atrial tachyarrhythmias.[280,283,284]

7.3.4. Ablation outcomes

Considering the totality of published case series (Table 7.1), the estimated acute ablation success rate for atrial tachyarrhythmias in CHD is approximately 81%.[166,168,171,260,270,277,280,283–285] Longer-term outcome studies with a follow-up period up to 5 years report recurrences in 34%–54%,[168,275,277,280] the majority of which are within the first year.[168] Compared to other subpopulations with CHD, patients who have undergone older versions of the Fontan procedure, most prevalently the atriopulmonary anastomosis, appear more likely to have acute procedural failure and arrhythmia recurrence after catheter ablation.[166,277] These "recurrences" are most often new atrial tachyarrhythmias and may be caused by different mechanisms. In addition, the location of the arrhythmogenic substrate also varies, suggesting that they more likely result from progressive atrial myopathy as opposed to arrhythmogenicity of prior ablation lesions. Despite recurrent arrhythmias, a large number of patients remain in sinus rhythm (40%–59%) after ablative therapy and have improved clinical status, as assessed by clinical scoring.[280] Multiple ablation procedures may thereby be reasonably justified.

7.4. Atrial fibrillation

7.4.1. Epidemiology

Atrial fibrillation is increasing in prevalence in the aging population with CHD. In a series of patients with CHD undergoing cardioversion over a 10-year period, 31% had atrial fibrillation, 20% as their sole presentation (i.e., without other atrial tachyarrhythmias).[286] Conditions disproportionately associated with atrial fibrillation were left-sided obstructive lesions, incompletely palliated CHD, and, to a lesser extent, Fontan surgery. In a multicenter cohort of adults with tetralogy of Fallot, atrial fibrillation surpassed IART as the most prevalent atrial tachyarrhythmia over the age of 55 years.[31] Older age, left atrial enlargement, lower left ventricular ejection fraction, and number of cardiac surgeries were independently associated with atrial fibrillation. Atrial fibrillation is a well-recognized sequela of large, unrepaired atrial septal defects in adults. Early but not late (i.e., >40 years) closure of the atrial septal defect reduces its prevalence postoperatively.[287–290] Although it is reasonable to postulate that the principles of cellular activation, wavefront propagation, and effects of myocellular hypertrophy and interstitial fibrosis are the same in patients with and without CHD, the pro- and/or antiarrhythmic effects of surgical intervention, aberrant anatomy, and chronic cyanosis on atrial fibrillation are largely unknown. Limited atrial Maze procedures may have proarrhythmic effects, particularly with respect to atypical atrial reentry circuits,[291,292] whereas extensive Maze procedures are antiarrhythmic.[293–295]

7.4.2. Catheter ablation

No specific recommendations regarding management of adults with CHD and atrial fibrillation have previously been proposed, perhaps due to lack of awareness or of data in this emerging group of patients.[296] Extrapolating from adult practice and from the literature on surgical Maze procedures in CHD, it is reasonable to infer that strategies such as catheter-based Maze procedures and AV nodal ablation with pacing are plausible treatment options. In the absence of other directive data, these interventions should be considered in conformance with recommendations for adults without CHD. Ablation (as an alternative to surgical Maze procedures) might be considered after failure of trials of cardioversion with pharmacologic rhythm control and in the context of adequate antithrombotic therapy.

With respect to catheter ablation of atrial fibrillation, operators have largely mimicked and adapted standard strategies, including isolation of pulmonary venous antra, connecting lesion sets to the left-sided AV annulus, and cavotricuspid isthmus ablation. These complex procedures require careful anatomic planning and ideally utilize techniques for real-time and/or registered volume imaging of the heart to facilitate visualization of relevant anatomy. In a series of 36 consecutive patients with predominantly simple forms of CHD (i.e., atrial septal defects in 61%) who underwent pulmonary vein

Table 7.1 Acute success rates for catheter ablation of atrial tachyarrhythmias in CHD

First Author	Year	N	Mean age (years)	Acute success
Hebe	2000	69	25 ± 18	90%
Triedman	2002	177	25 ± 12	79%
Blaufox	2002	31	18 ± 5	96%
Kannankeril	2003	47	28 ± 13	87%
Tanner	2004	36	Median 46 (9 to 67)	94%
Lukac	2005	83	Median 47 (9 to 73)	88%
Seiler	2007	40	52 ± 12 years	88%
Yap	2010	118	40 ± 13 years	69%
de Groot	2010	53	38 ± 15 years	65%
Drago	2011	31	26 ± 17 years	87%
Summary		*685*		*81% [95% CI (79%–84%)]*

CHD = congenital heart disease; CI = confidence interval.

isolation procedures for atrial fibrillation, success at 300 days was achieved in 42% compared to 53% of 355 controls without CHD.[34] By 4 years of follow-up, corresponding success rates were 27% and 36%, respectively. The value of repeat interventions and the role of pulmonary vein isolation in patients with more complex forms of CHD remain to be studied.[297] Because these procedures are currently infrequently performed, it would seem reasonable to have available for consultation the expertise of an electrophysiologist skilled in atrial fibrillation ablation.

AV nodal ablation with postablation ventricular pacing in patients with CHD has been reported as individual cases[298] and in a small series.[299] Because paced patients with complex univentricular heart disease have higher risk of mortality than those in native sinus rhythm, and because the presumed ongoing atrial arrhythmias still pose an unmitigated thromboembolic risk, this approach should only be undertaken as a last resort for symptomatic atrial tachycardia unresponsive to rate control. Location of the AV conduction system may be unpredictable.[300–302] In the absence of data specific to CHD, techniques for AV nodal ablation when elected should follow recommendations outlined for patients with normal cardiac anatomy: identification of a distinct His-bundle electrogram and application of radiofrequency energy sufficient to cause AV block within 30 seconds, preceded by an accelerated junctional rhythm. After AV nodal ablation, patients should be ventricularly paced at a lower rate of 80–90 bpm (see Section 10 for Cardiac Resynchronization Therapy recommendations), with subsequent decrements on follow-up until the desired resting heart rate is achieved.[303,304]

7.5. Recommendations for catheter ablation of atrial tachycarrhythmias in adults with CHD

Recommendations	
Class I	1. Catheter ablation is indicated for recurrent symptomatic and/or drug-refractory supraventricular tachycardia related to accessory AV connections or twin AV nodes in adults with CHD *(Level of evidence: B)*.[225,237,241,242]
	2. Catheter ablation is useful for adults with CHD and symptomatic and/or drug-refractory IART or focal atrial tachycardia *(Level of evidence: B.)*[241,246,260,266,268–272,274–277]
	3. Catheter ablation is recommended for adults with CHD, ventricular preexcitation, and high-risk or multiple accessory pathways, as commonly encountered in Ebstein anomaly *(Level of evidence: C)*.[236]
	4. A 3-dimensional electroanatomic mapping system is indicated for guiding ablation of postoperative atrial tachyarrhythmias in adults with CHD *(Level of evidence: B)*.[266,268,278–280]
Class IIa	1. Irrigated or large electrode-tip catheters can be useful for the ablation of postoperative atrial tachyarrhythmias in adults with CHD *(Level of evidence: B)*.[280,283,284]
	2. Catheter ablation can be beneficial for recurrent symptomatic and/or drug-refractory AV nodal reentrant tachycardia in adults with CHD *(Level of evidence: C)*.[231,232,237,245,246]
	3. A catheter-based procedure centered on electrically isolating pulmonary veins can be useful in adults with CHD and symptomatic drug-refractory atrial fibrillation *(Level of evidence: C)*.[34]
Class IIb	1. It may be reasonable to perform invasive diagnostic electrophysiologic studies in patients with Ebstein anomaly prior to anticipated cardiac surgery *(Level of evidence: B)*.[305]
	2. In adults with CHD and symptomatic atrial tachyarrhythmia refractory to pharmacologic and standard ablation therapy, it may be reasonable to consider AV nodal ablation and pacing as third-line therapy *(Level of evidence: C)*.[298,299]

7.6. Ventricular tachycardia

7.6.1. Epidemiology

Although ventricular ectopy and nonsustained ventricular tachycardia are relatively common, sustained monomorphic ventricular tachycardia, which is the most tractable target for catheter mapping and ablation, appears to be quite rare in adults with CHD at large. This can be inferred from the paucity of clinical cases that have been reported even in the largest series published over recent decades.[306–308] It is also evident from the efforts of several series on the epidemiology of and risk factors for sudden cardiac death in CHD.[18,40,46,76,101] Based on these observations, the incidence of sustained ventricular tachycardia in adults with CHD appears to be comparable to sudden cardiac death and is in the order of 0.1%–0.2% per year.[309]

7.6.2. Mechanistic considerations

Patients with sustained monomorphic ventricular tachycardia in the setting of CHD typically have myocardial changes related to hemodynamic loading, cyanosis, or surgical interventions that predispose to arrhythmia. Additionally, inducibility of ventricular tachycardia by programmed stimulation has been included in the many covarying risk factors for occurrence of cardiac arrest, particularly in patients with tetralogy of Fallot.[40,76] However, a clear relationship between inducible sustained monomorphic ventricular tachycardia and elevated risk of sudden death has not been established across all forms of CHD. It is important to note that in some classes of CHD, such as transposition of the great arteries with Mustard or Senning baffles, a correlation between inducible and clinical ventricular

tachycardia has not been observed despite the relatively high incidence of sudden cardiac death.[21,46]

The most common substrate for sustained ventricular tachycardia in CHD is tetralogy of Fallot. The right ventricular outflow tract area is typically heavily scarred from surgical intervention[310–313] and has anatomic features that may predispose to macroreentrant tachycardia, in a fashion similar to the cavotricuspid isthmus in atrial flutter. Mapping studies of individual cases in the 1990s postulated the importance of a critical isthmus of tissue defined by the relation of the surgical right ventriculotomy to anatomic features such as the pulmonary or tricuspid annulus, along with the observation of bidirectional use of these pathways in clinical tachycardias.[273,314–317] Subsequently, careful clinical mapping studies on larger numbers of patients identified several plausible anatomic corridors that could support right ventricular macroreentrant loops, including the conal septum and its insertion into the ventricular myocardium subjacent to the tricuspid annulus.[306]

7.6.3. Catheter ablation

Given the uncertain relationship between sustained monomorphic ventricular tachycardia and sudden death, and the relatively high risk of recurrence even after acutely successful ablation, ventricular tachycardia ablation is only rarely and under special circumstances seen as a substitute for ICD therapy, and most commonly as an adjunct. As such, catheter ablation can be helpful in reducing the risk of recurrent ICD shocks and, much more rarely, can be performed for hemodynamic risk in patients with slow but incessant tachycardias. It has also been anecdotally observed in adults with CHD, and more often in patients with normal cardiac anatomy,[318] that frequent recurrent monomorphic ventricular ectopy may sometimes be associated with decreased ventricular function. It has been suggested that this can be a reasonable indication for ablation. Although preexisting cardiomyopathy can decrease the likelihood of functional normalization postablation,[319] ablation may nonetheless be a useful adjunct in adults with CHD and frequent drug-refractory ventricular ectopy, particularly in the setting of progressive ventricular dilation or dysfunction.

The methodology for ventricular tachycardia ablation in adults with CHD is similar to that applied to atrial tachycardias, with substrate mapping using 3-dimensional electroanatomic systems generally assuming a prominent role. Careful recording of the activation sequence and application of entrainment may be used when relevant. Challenges may include the thickness of the ventricular myocardium that must be ablated to achieve anatomic block of a reentry circuit. Occasionally, the His bundle and proximal bundle branches are located in proximity to the desired ablation target. Additionally, it can be difficult to induce the clinical arrhythmia, or, once induced, it may be unstable and/or poorly tolerated hemodynamically. In such cases, it is feasible and often useful to utilize pace-mapping to identify exit sites from protected corridors of myocardium.

7.6.4. Ablation outcomes

Results of ventricular tachycardia ablation in adults with CHD have been limited to a small number of case series, many of them with mixed populations and substrates, precluding accurate estimates of long-term arrhythmia-free survival rates. The first substantial series included 16 patients with right heart lesions (predominantly tetralogy of Fallot) and demonstrated the feasibility of apparently curative ablation of circuits located on the right ventricular free wall, using a combination of sinus rhythm mapping, activation mapping, and entrainment pacing.[308] Subsequent series demonstrated the importance of inducibility and other patient factors in procedural success[307] and the plausible utility of ablation in combination with antiarrhythmic drug therapy.[215]

7.6.5. Recommendations for catheter ablation of ventricular arrhythmias in adults with CHD

Recommendations	
Class I	Catheter ablation is indicated as adjunctive therapy to an ICD in adults with CHD and recurrent monomorphic ventricular tachycardia, a ventricular tachycardia storm, or multiple appropriate shocks that are not manageable by device reprogramming or drug therapy *(Level of evidence: C)*.[94,320]
Class IIa	Catheter ablation can be considered for symptomatic sustained monomorphic ventricular tachycardia in adults with CHD and ICDs as an alternative to drug therapy *(Level of evidence: B)*.[215,306]
Class IIb	1. Catheter ablation may be reasonable in adults with postoperative CHD and nonsustained or hemodynamically poorly tolerated ventricular tachycardia by means of an empiric anatomic approach *(Level of evidence: C)*.[306] 2. Catheter ablation may be reasonable in adults with CHD and frequent ventricular ectopy associated with deteriorating ventricular function *(Level of evidence: C)*.[94]
Class III	1. Catheter ablation is not indicated for asymptomatic relatively infrequent ventricular ectopy in adults with CHD and stable ventricular function *(Level of evidence: C)*.[94] 2. Catheter ablation alone is not considered appropriate prophylactic therapy in adults with CHD deemed to be at increased risk for sudden cardiac death *(Level of evidence: C)*.[94]

8. Bradyarrhythmias and pacemakers

8.1. Introduction

In 1984, the ACC and AHA published the first clinical guidelines for permanent pacemaker implantation. This was updated in 2002 and later in 2008 in collaboration with the North American Society of Pacing and Electrophysiology (NASPE) and successor organization, HRS.[97] Although general pacemaker applications to patients with CHD based primarily on heart rate and symptoms are included in these guidelines, there are no specifics regarding anatomy, surgical repair and its consequences, implant site, or pacing mode. Implantable cardiac device therapies are increasingly indicated in adults with CHD, and physicians potentially unfamiliar with CHD are more likely to interact with these patients. There is, therefore, a need for more detailed and updated recommendations for device therapies in this growing population.

Clinical indications for pacing in adults with CHD may be inherent to the underlying anatomic substrate, occur in the immediate postoperative period secondary to injury to the conduction system, or present years later as a result of a slow but progressive deterioration in the conduction system by fibrotic encroachment. This section first discusses clinical indications surrounding pacemaker consideration (sinus node dysfunction, AV block, atrial arrhythmias) and general issues regarding permanent pacemaker implantation applicable to all adults with various forms of CHD, followed by specific structural heart defect considerations. Finally, current recommendations for pacemaker implantation are provided.

8.2. Sinus node dysfunction

Sinus node dysfunction may be observed in rare variants of heterotaxy syndrome (polysplenia, left atrial isomerism) with congenital absence of a sinoatrial node and reliance on a slower atrial or junctional escape for effective atrial depolarization. More often, pathologic sinus bradycardia or junctional rhythm, with loss of AV synchrony, is a late acquired condition following cardiac surgery. Injury to the sinus node artery, neural inputs, autonomic dysfunction, or long-standing hemodynamic perturbations may result in disordered impulse generation within the sinus node or impaired propagation of the sinus impulse to the surrounding atrial tissue.[321–325] Loss of AV synchrony can markedly worsen AV valve regurgitation, increase atrial arrhythmias, and contribute to hepatic congestion and thrombosis. In addition, a spectrum of tachyarrhythmias may occur in patients with chronic bradycardia based on reentry or automaticity that have collectively been coined bradycardia-mediated tachyarrhythmias. Table 8.1 lists common causes of sinus node dysfunction in adults with CHD.

Years of sinus node dysfunction in adults with CHD results in ineffective atrial hemodynamics that, in conjunction with scar, anatomic obstacles, and atrial hypertension, establish a milieu for atrial tachyarrhythmias. Although sinus bradycardia and sinus arrest have not been identified as risk factors for sudden cardiac death in adults with CHD, atrial arrhythmias, which may be precipitated by sinus node dysfunction,[100] are a major risk factor for sudden cardiac death.[47,326–330] Because IART tends to propagate at atrial rates of 150–250 bpm, 1:1 AV conduction in patients with healthy AV nodes is not uncommon. The risk of sudden cardiac death in patients with poorly controlled IART is 4-fold and likely related to 1:1 AV conduction degenerating to ventricular tachycardia.[23,46] Atrial tachyarrhythmias may be particularly poorly tolerated in adults with single ventricles, systemic right ventricles, ventricular dysfunction, or those having significant AV valve regurgitation. In addition, an abnormal heart rate response to exercise in adults with CHD may be associated with IART[331] and confers a higher mortality risk.[332] Any consideration for pacemaker implant among such patients must entail a determination of risk/benefit of specific devices: those with just brady- versus antitachycardia capabilities, cardiac resynchronization therapy (CRT), or an ICD.

The postoperative environment following the Senning or Mustard procedure, all varieties of the Fontan operation, Glenn shunts, or repair of Ebstein anomaly present common substrates for the gradual loss of sinus node function.[32,333–335] Loss of an atrial-derived rhythm in single-ventricle patients has been shown to result in significant pulmonary venous flow reversal, decreasing preload to the single ventricle, increasing preventricular left atrial pressures, and lower cardiac output.[18–20] Furthermore, sinus node dysfunction exposes Fontan patients to an increased risk for plastic bronchitis and protein-losing enteropathy that may resolve with atrial pacing.[21–22] However, even less complex lesions

Table 8.1 Substrates associated with a relatively high prevalence of congenital and postoperative sinus node dysfunction

Congenital sinus node dysfunction
Left-sided juxtaposition of the atrial appendages
Left atrial isomerism (polysplenia, heterotaxy syndrome)
Postoperative sinus node dysfunction
Mustard baffle
Senning baffle
Hemi-Fontan or Fontan surgery; atriopulmonary and total cavopulmonary connections
Glenn shunt
Sinus venosus atrial septal defect
Ebstein anomaly
Arterial switch operation for transposition of the great arteries (chronotropic incompetence)
Tetralogy of Fallot

such as atrial septal defects, tetralogy of Fallot, and supracardiac total anomalous pulmonary venous return are all potentially at risk for developing late sinus node dysfunction. A high prevalence of chronotropic incompetence has also been reported following the arterial switch operation for transposition of the great arteries and is thought to be mediated by sympathetic denervation.[96]

Sinus node dysfunction is typically best assessed noninvasively with ECGs, ambulatory Holter or event monitors, and exercise stress tests. Normative values for resting and peak heart rates are gender- and age-specific. An attenuated heart rate response to exercise is prevalent across the spectrum of CHD and predicts a reduction in peak oxygen consumption and increased mortality.[332,336] However, heart rate response is the limiting factor in roughly 20% of adults with CHD and chronotropic incompetence. In the majority, exercise tolerance is limited by factors such as poor heart rate O_2 uptake kinetics, depressed myocardial function, and reduced $AVDO_2$ (right-to-left shunt). Chronotropic incompetence in the absence of exercise intolerance should not be a clinical indication for a permanent pacemaker. The absolute peak heart rate value is, to a certain extent, an artificial number that should be applied with caution to patients with systemic right ventricles and univentricular hearts where ventricular filling may be compromised above a critical value, especially with aggressive rate-responsive pacing.[337,338]

A pacemaker is recommended for isolated sinus node dysfunction in adults with CHD if there are clinical symptoms related to bradycardia or loss of AV synchrony, exercise intolerance secondary to chronotropic incompetence, or bradyarrhythmia-related adverse hemodynamic effects documented by noninvasive or invasive testing. Patients with bradycardia–tachycardia syndrome are nearly always symptomatic. Even among asymptomatic adults with CHD and chronic bradycardia or junctional rhythm, noninvasive evaluation may reveal significant systolic or diastolic dysfunction, marked atrial enlargement, abnormal AV valve inflow patterns, and/or low cardiac output. Resolution of such noninvasive perturbations with temporary pacing correlated with concomitant hemodynamic evaluations in the catheterization laboratory may occasionally assist in decision-making.

Worsened AV valve regurgitation or heart failure secondary to loss of AV synchrony (junctional rhythm) should prompt consideration for atrial-based pacing to restore AV synchrony. Yet, even with maintenance of AV coupling, chronic sinus bradycardia prolongs electrical diastole and increases the interval during which a premature atrial beat may initiate a reentrant circuit. Careful noninvasive assessment of chronic bradycardia may reveal frequent premature atrial beats and/or nonsustained atrial tachycardia. The mixed nature of the bradycardia–tachycardia syndrome often requires a multimodal approach combining antiarrhythmic medication, catheter ablation, and/or antitachycardia pacemaker therapies. Although atrial antibradycardia pacing alone may result in clinical improvement and decreased tachycardia frequency, results seem somewhat equivocal.[339–342] It is currently unclear if atrial antibradycardia pacing prior to the development of IART confers prophylactic benefits. Consideration should be given to implanting pacemakers with atrial antitachycardia pacing (ATP) features in patients with sinus node dysfunction and IART, or with a high proclivity for developing IART. In those with bradycardia–tachycardia syndromes, atrial ATP is reasonably effective in terminating IART (54%)[119] and significantly reduces tachyarrhythmia-related hospitalizations.[342,343] Atrial ATP requires atrial and ventricular leads, since a $\geq$2:1 AV ratio is generally required to trigger therapy. One-to-one conduction is particularly problematic with the longer/slower circuits encountered in adults with CHD. To minimize associated risks, a concomitant AV nodal blocking agent is strongly advised.[46]

Pacing mode can be an important variable in device program decision-making. AAI or DDD pacing is preferred over isolated VVI pacing in adults with CHD and sinus node dysfunction. The deleterious effects of subpulmonary ventricular pacing are well known such that programming to reduce the percentage of ventricular pacing should be an important goal. Programming long AV delays requires the patient to have reliable AV nodal conduction. However, long AV delays may encroach on effective upper rate behavior and atrial tachycardia detection.[23–24] Although DDI(R) pacing may prevent tracking of atrial arrhythmias, patients are subject to the same limitations of long AV delays. Novel pacemaker algorithms have been developed to reduce ventricular pacing and should be considered in this population.[344–348] Also, atrial septal pacing carries the potential to improve hemodynamics and reduce unnecessary ventricular pacing when compared to appendage pacing.[349–351]

8.3. AV conduction system dysfunction

Although an improved understanding of the AV node and His-bundle conduction tissue relative to various anatomic substrates has markedly reduced the incidence of high-grade postoperative AV block, advanced AV block following CHD surgery continues to occur in 1%–3% of cases.[35–36] The highest-risk operations include closure of certain septal defects, surgery along the left ventricular outflow tract, and left-sided valve surgery. Recovery within 7–10 days can be expected in 50% of patients, with 63% recovering by 30 days.[352] For those in whom heart block is not expected to resolve, a permanent preferably dual-chamber or biventricular pacemaker is recommended. Based predominantly on earlier studies in patients with tetralogy of Fallot with transient postoperative complete heart block and residual bifascicular block, late-onset complete heart block occurs in almost 33%.[353,354] A pacemaker should, therefore, be considered in patients with postoperative transient AV block and residual bifascicular block. However, there is currently no evidence to support routine pacemaker implantation for bifascicular block in asymptomatic adults with CHD who did not have transient complete AV block.

The AV conduction tissue may be congenitally displaced and functionally rendered at risk with certain anatomic substrates, most notably AV septal defects, congenitally corrected transposition of the great arteries, and left atrial

Table 8.2 Congenital heart disease substrates associated with a relatively high prevalence of congenital and postoperative AV block

Congenital AV block
Congenitally corrected transposition of the great arteries
Atrioventricular septal defect (endocardial cushion defect)
L-Looped single ventricles
Anomalous left coronary artery arising from the pulmonary artery (ALCAPA)
Postoperative AV block
Cardiac surgery in patients with displaced AV conduction systems (congenitally corrected transposition of the great arteries, atrioventricular septal defect)
Ventricular septal defect
Valve surgery, especially mitral valve and multivalve surgery involving the tricuspid valve
Left ventricular outflow surgery, subaortic stenosis

isomerism. Table 8.2 lists common lesions associated with AV block in adults with CHD. Malalignment of the atria and ventricular septae, whether in biventricular or univentricular hearts, displaces the AV node posteriorly and inferiorly.[355,356] Caution should be exercised when operating in this vicinity or ablating in the right inferior paraseptal region. Inversion of the fast and slow components of the AV node has been reported; this knowledge is critical if considering ablation for AV nodal reentrant tachycardia in a patient with an AV septal defect.[232] In patients with congenitally corrected transposition of the great arteries, the conduction tissue is displaced anteriorly and laterally, with an elongated and fragile His bundle coursing anterior along the pulmonary valve.[43–46] This conduction system is vulnerable and at risk during surgical or catheter procedures. Heart block may also develop during pregnancy, possibly related to altered loading conditions,[357,358] and limit the ability of the systemic right ventricle to augment stroke volume as needed.[359] Patients at risk for late-onset AV block merit periodic noninvasive electrophysiologic monitoring.

Dual-chamber pacing is preferred over VVI pacing in adults with CHD and intrinsic or postoperative heart block. Concomitant echocardiographic evaluation of AV valve inflow patterns with pacemaker programming of various AV intervals may allow for identification of the longest possible diastolic filling time for maximal cardiac output.[360] Despite congenital or postoperative AV block, atrial fibrillation and IART remain an ongoing concern[72] and can complicate effective utilization of dual-chamber pacing. Pacemakers with atrial ATP features may be considered in adults with nonpermanent IART, or with the potential anatomic substrate to develop IART, in spite of complete AV block.

8.4. Preimplant considerations

8.4.1. Know the anatomy

Prior to device implantation, it is critical that the implanting physician have a thorough and accurate understanding of the congenital heart defect and cardiothoracic surgical procedure(s) performed. Meticulous attention should be given to previous operative reports, noninvasive imaging, and angiography. Congenital structural cardiac defects such as congenitally corrected transposition or Ebstein anomaly of the tricuspid valve are associated with inherent anatomic issues that can be technically challenging to any implanter not familiar with structural variances found in certain adults with CHD. A detailed understanding of the venous drainage, baffles, conduits, and any residual shunts should be sought prior to implantation. The presence of an intracardiac shunt may expose the patient to a prohibitively high risk of thromboembolism.[182] An imaging study (e.g., Doppler echocardiography) performed and interpreted by someone familiar with CHD is recommended prior to any device implant.

8.4.2. Determine venous access prior to any incisions

Although this concept may appear intuitive, it must be remembered that many adults with CHD underwent venous cannulation during cardiopulmonary bypass at a very early age. Venous patency, therefore, can never be assumed. Also, certain forms of CHD may be associated with an absent innominate vein and persistent left superior vena cava, or an unroofed or absent coronary sinus. In addition, repaired CHD defects, such as D-transposition of the great arteries with an intra-atrial baffle (Mustard, Senning procedures), commonly have narrowing or obstruction of the superior baffle limb, often requiring prepacemaker vascular stents.[361,362] Because of the close proximity of the azygos vein acting to decompress any obstruction, transthoracic Doppler echocardiography may not be sensitive enough to identify vascular obstruction. This concept also applies to any adult CHD patient with a preexisting pacemaker or one in whom a transvenous pacemaker has previously been removed. Venograms, if available from prior catheterizations, should be reviewed before the case is initiated. In the absence of prior imaging delineating upper extremity venous drainage, a preimplant CT or MRI may be helpful. Otherwise, a venogram should be performed prior to any incision.

8.4.3. Evaluate sinus and AV nodal function

Venous cannulation in an infant can have consequences on sinus node function that may not become apparent until later in life. In addition, septal patch materials causing progressive myocardial fibrosis can impinge on AV conduction tissue. Surgical incisions commonly transect the right bundle branch resulting in bundle branch block. Because adults with CHD are likely to benefit from any atrial contribution to ventricular filling, atrial-based pacing can be anticipated to be applicable for most patients. Adults with CHD can have coexisting atrial or ventricular dysrhythmias, ventricular dyssynchrony, and/or heart failure such that a preimplantation workup is required to determine the most appropriate cardiac arrhythmia device. In some cases, a

predevice electrophysiologic study can be useful in determining whether atrial or ventricular arrhythmias are inducible and help guide decisions regarding antitachycardia and/or defibrillation capabilities. This knowledge can also inform appropriate post-implant patient-specific device programming.

8.4.4. Choose optimal lead implant site

In the current era, selecting a pacing site that merely satisfies adequate pacing and sensing thresholds is no longer considered adequate. It is now well recognized that right ventricular pacing, especially the free wall and outflow tract, can have deleterious effects on ventricular function.[363–370] Although ventricular septal pacing has been advocated as preferential to the apex, any surgical patch materials can negate septal implant. Data from pediatric patients with and without CHD suggest that systemic left ventricular function is best preserved by pacing from the left ventricular apex or mid-lateral wall.[369–375] It remains to be demonstrated whether such findings are applicable to the systemic right ventricle and univentricular heart.

"Traditional" atrial pacing from the right atrial appendage has also been questioned.[351] In addition, the atrial appendage itself may have been surgically removed during CHD repair. Alternative pacing lead implant sites should be carefully considered. This can be especially important among patients with D-transposition of the great arteries and Mustard or Senning procedures in whom pacing from the left atrial appendage in the neo-right atrium carries the potential for inadvertent phrenic nerve stimulation.[7] Preimplant "pacing site mapping" can add valuable information.[376] Active fixation leads typically offer more implant options than passive fixation designs.

8.5. Issues related to specific congenital heart defects

8.5.1. Repaired septal defects

Sinus node dysfunction can be inherently associated with atrial septal defects or with their surgical correction, particularly sinus venosus defects with anomalous pulmonary venous connections.[377] Defect closure, either by suture, patch, or device, may predispose to atrial dysrhythmias, which must be considered in device selection. Prosthetic materials placed in the interatrial septum may prevent effective pacing lead implant in the Bachmann bundle septal region. Moreover, the AV node may be inherently abnormal or displaced. Primum atrial septal defects characteristically result in a superior QRS axis with a RBBB pattern, thought to be a result of an inferiorly and posteriorly displaced AV node and hypoplastic left anterior fascicle.[77,378] AV conduction problems may occur late after surgery due to progressive fibrotic changes in the interventricular septum. Prosthetic materials may impede ventricular septal lead placement.

8.5.2. D-Transposition of great arteries

Sinus node dysfunction and IART are highly prevalent in adults with Mustard or Senning procedures for transposition of the great arteries and have been estimated to occur in approximately 60% and 25%, respectively, at 20 years of follow-up.[32,329] Narrowing of the superior limb of the baffle, which can complicate transvenous lead insertion, is observed in >40% of adults with Mustard procedures, 30% of whom have hemodynamically significant obstructions.[362] In addition, baffle leaks (i.e., interatrial shunts) are highly prevalent. In the presence of interatrial shunts, transvenous leads incur an increased risk of systemic thromboemboli.[182] Moreover, a transvenous lead may inadvertently be placed across a baffle leak and into the systemic circulation. AV block, although less prevalent than sinus node dysfunction, can complicate the postoperative course, particularly in patients with a surgically repaired tricuspid valve or associated ventricular septal defect.[379] In addition, the systemic ventricle is of right ventricular morphology and can progress to early heart failure. CRT may require a hybrid approach with epicardial and transvenous leads.[7]

8.5.3. Tetralogy of Fallot

Corrective surgery for tetralogy of Fallot involves atriotomy and/or ventricular incisions and patches, predisposing to the late development of arrhythmias.[101] Surgical repair may entail outflow prosthetic materials as well as conduits, and issues related to septal prosthetic materials may apply. Dilated right-sided chambers, patchy areas of scarring, and severe pulmonary and/or tricuspid regurgitation may complicate lead placement. Given the preponderance for ventricular tachyarrhythmias in adults with tetralogy of Fallot, a predevice implant electrophysiologic study may be warranted to better assess the need for defibrillation capabilities (see Section 9).[76]

8.5.4. Univentricular hearts

A high prevalence of sinus node dysfunction and IART is observed in adults with Fontan surgery.[50] Single ventricles have limited cardiac reserve and function decreases with increased heart rates. Programming devices to limit upper tracking rates is recommended. Older adults may have had a direct atrio-pulmonary artery connection Fontan. Often, the right atrium is extremely enlarged in such patients, with preserved venous access that permits transvenous atrial lead implantation.[380] Factors to consider prior to implanting a transvenous lead for AAIR pacing in the context of sinus node dysfunction may include ruling out intracardiac thrombus, the need for concomitant anticoagulation, and potential indications for Fontan conversion with epicardial lead placement.[381] An alternative approach to the transvenous lead entails a transmural atrial lead.[382] Ventricular pacing may be performed via the coronary sinus in some[383,384] or by an epicardial approach. More recent modifications of the Fontan procedure (i.e., total cavopulmonary connections) consist of intracardiac or external conduits.[50] Transvenous atrial pacing may be feasible in some patients with intracardiac lateral tunnels but not in those with extracardiac conduits, which prevent direct venous access to the heart.[187] Ventricular pacing typically requires an epicardial approach, although patients with intracardiac lateral tunnels may have transvenous access to a coronary sinus. Previous surgical procedures can result in extensive epicardial fibrosis, often hindering effective epicardial lead placement. A combination

transvenous-atrial/epicardial-ventricular approach ("hybrid") may be a viable alternative in selected patients.

8.6. Lead extraction

Given the finite longevity of current lead designs, lead extraction is an eventuality for a substantial subset of adults with CHD and transvenous systems. Indications for lead extractions outlined in the HRS Expert Consensus document are applicable to adults with CHD, and generally include infection, life-threatening arrhythmias secondary to a retained lead fragment, thromboembolic events caused by a retained lead, and occlusion of all usable veins with the need to implant a new pacing/ICD system.[385] Lead extraction should also be considered for nonfunctioning leads in young patients. Required personnel for lead extraction in adults with CHD include a physician with specific training in lead extraction and management of associated complications, congenital cardiothoracic surgical backup, cardiac anesthesiology, and dedicated support staff.[386] Assistance by an interventional cardiologist with expertise in CHD may be necessary for deploying stents in occluded baffles and veins and for closure of intracardiac shunts if lead reimplantation following extraction is required.

Depending, in part, on length of time that leads have been in situ, the leads can be removed by simple traction, traction devices, or specialized mechanical, telescoping, laser, electrosurgical, or rotating threaded-tip sheaths.[385,387–389] Data on the safety and efficacy of different lead extraction techniques in this specific patient population are limited.[390–392] In the first reported series, laser lead extraction was successful in 91% of adults with CHD, with comparable success and complication rates to controls despite longer procedures.[390] The most common indication was infection (44%) followed by lead dysfunction (25%). In a cohort of 144 patients, 60% of whom had structural heart disease, complex extraction techniques that primarily involved a radiofrequency-powered sheath were successful in 94% of leads.[391]

8.7. Recommendations for permanent pacing in adults with CHD

Recommendations	
Class I	1. Permanent pacing is recommended for adults with CHD and symptomatic sinus node dysfunction, including documented sinus bradycardia or chronotropic incompetence that is intrinsic or secondary to required drug therapy *(Level of evidence: C)*.[97,393–395] Devices that minimize ventricular pacing are preferred *(Level of evidence: B)*.[344–348]
	2. Permanent pacing is recommended in adults with CHD and symptomatic bradycardia in conjunction with any degree of AV block or with ventricular arrhythmias presumed to be due to AV block *(Level of evidence: B)*.[97,396–400]
	3. Permanent pacing is recommended in adults with congenital complete AV block and a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction (*Level of evidence: B)*.[97,401–403]
	4. Permanent pacing is recommended for adults with CHD and postoperative high-grade second- or third-degree AV block that is not expected to resolve *(Level of evidence: C)*.[97,404–406]
Class IIa	1. Permanent pacing is reasonable for adults with CHD and impaired hemodynamics, as assessed by noninvasive or invasive means, due to sinus bradycardia or loss of AV synchrony *(Level of evidence: C)*.[97,407]
	2. Permanent pacing is reasonable for adults with CHD and sinus or junctional bradycardia for the prevention of recurrent IART *(Level of evidence: C)*.[97,119,340,342] Devices with atrial antitachycardia pacing properties are preferred in this subpopulation of patients *(Level of evidence: B)*.[119,342,343,408]
	3. Permanent pacing is reasonable in adults with congenital complete AV block and an average daytime resting heart rate $<$50 bpm *(Level of evidence: B)*.[97,409,410]
	4. Permanent pacing is reasonable for adults with complex CHD and an awake resting heart rate (sinus or junctional) $<$40 bpm or ventricular pauses $>$3 seconds *(Level of evidence: C)*.[97] A device with antitachycardia pacing properties may be considered if the underlying anatomic substrate carries a high likelihood of developing IART *(Level of evidence: B)*.[119,342,343,408]
Class IIb	1. Permanent pacing may be reasonable in adults with CHD of moderate complexity and an awake resting heart rate (sinus or junctional) $<$40 bpm or ventricular pauses $>$3 seconds *(Level of evidence: C)*.[97,393,394,411] A device with antitachycardia pacing properties may be considered if the underlying anatomic substrate carries a high likelihood of developing IART *(Level of evidence: B)*.[119,342,343,408]
	2. Permanent pacing may be considered in adults with CHD, a history of transient postoperative complete AV block, and residual bifascicular block *(Level of evidence: C)*.[97,353]
Class III	1. Pacing is not indicated in asymptomatic adults with CHD and bifascicular block with or without first-degree AV block in the absence of a history of transient complete AV block *(Level of evidence: C)*.[97]
	2. Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure prior to endocardial lead placement, or alternative approaches for lead access should be individualized *(Level of evidence: B)*.[8,54,182]

9. Sudden cardiac death and ICDs

9.1. Introduction

The term "sudden cardiac death" refers to death due to a cardiovascular cause within 1 hour of the onset or significant worsening of symptoms, or unwitnessed death in the absence of a known noncardiac condition as the proximate cause of death. Arrhythmic sudden cardiac death encompasses death due to documented or presumed arrhythmias, that is, instantaneous death in the absence of a nonarrhythmic cause at autopsy or a pulseless abrupt loss of consciousness in the absence of a nonarrhythmic diagnosis.[103] Since the first reports of sudden cardiac death following surgical repair of CHD over 30 years ago,[412] a substantial volume of literature has been generated on this topic. Evolving perspectives regarding incidence and risk factors reflect several features, including the greater number of adults with complex forms of CHD, increased awareness of these issues, improvements in interventions and device therapies, and the availability of longer-term follow-up.[413,414] In patients with CHD, the majority of sudden cardiac deaths are of arrhythmic etiology, as indicated in Table 9.1.[18,20,103] It is important, however, to bear in mind that up to 20% of sudden cardiac deaths may be due to nonarrhythmic causes such as cerebral or pulmonary embolism, myocardial infarction, heart failure, and aortic or aneurysmal rupture.

9.2. Sudden and total late mortality

Several long-term single-center studies reported the incidence of sudden and total late mortality in patients with surgically repaired CHD (Table 9.2).[18–21,415] The data are relatively consistent, with sudden cardiac death (15%–26%) and heart failure (13%–27%) accounting for nearly half of all late deaths in mixed cohorts of children and adults. These reports were limited by their retrospective nature and mean age of follow-up through age 35 years. In studies that focused exclusively on adults with CHD and included 197[19] and 1189[103] deaths, sudden cardiac death accounted for 26% and 19% of all deaths, respectively. Therefore, based on current evidence, it can be estimated that approximately 20%–25% of late deaths in adults with CHD are due to sudden cardiac events.

The incidence of sudden cardiac death in the CHD population at large is relatively low and has been estimated to be $<0.1\%$ per year.[18–21,103,415] Identified higher-risk substrates include tetralogy of Fallot, D-transposition of the great arteries with Mustard or Senning baffles, congenitally corrected transposition of the great arteries, left-sided obstructive lesions cyanotic Eisenmenger syndrome, and Ebstein anomaly.[18,103] In the United states, a 40% reduction in annualized death rates for tetralogy of Fallot and a 71% reduction for transposition of the great arteries were reported between 1979 and 2005.[416] Similarly, in Canada, 46% and 61% reductions in adjusted mortality ratios were observed for patients with tetralogy of Fallot and D-transposition of the great arteries, respectively, between 1987–1999 and 2002–2005.[2] Studies estimating the incidence of sudden cardiac death in tetralogy of Fallot (2%–3% per decade) are summarized in Table 9.3.[18,40,101,417–419]

9.3. Arrhythmic causes of sudden cardiac death

Reflecting the variations in anatomy, circulatory physiology, and surgical techniques, diverse arrhythmic causes of sudden cardiac death have been identified in patients with CHD.

9.3.1. Heart block

Postoperative complete heart block has been recognized to be a risk factor for late death, with nonpaced postoperative

Table 9.1 Causes of sudden cardiac death following surgical repair of CHD

Authors	Year	No. events	Arrhythmic	Embolic	MI/CHF	Aneurysm
Silka et al	1998	41	30 (73.2%)	5 (12.2%)	4 (9.8%)	2 (4.9%)
Nieminen et al	2007	88	73 (83.0%)	5 (17.9%)	5 (17.9%)	5 (17.9%)
Koyak et al	2012	213	171 (80.3%)	8 (37.6%)	5 (2.3%)	19 (8.9%)
Total		*342*	*274 (80.1%)*	*18 (5.3%)*	*14 (4.1%)*	*26 (7.6%)*

CHD = congenital heart disease; CHF = congestive heart failure; MI = myocardial infarction.

Table 9.2 Causes of death following surgical repair of CHD

Authors	Years	Patients	Deaths	SCD	CHF	Other CV	Noncardiac
Oeschlin et al	1981–1996	2609	197	26%	21%	34%	18%
Silka et al	1958–1996	3589	176	23%	13%	35%	12%
Nieminen et al	1953–1998	5919	582	15%	27%	31%	8%
Verheugt et al	2001–2009	6933	197	19%	26%	32%	23%
Zomer et al	2001–2010	8595	231	22%	26%	29%	24%
Total		*27,645*	*1,383*	*19%*	*24%*	*36%*	*15%*

CHD = congenital heart disease; CHF = congestive heart failure; Noncardiac = noncardiac cause of death; Other CV = other cardiovascular cause of death; SCD = sudden cardiac death.

Table 9.3 Incidence of sudden cardiac death post surgical repair of tetralogy of Fallot

Authors	Patients	Mean follow-up	SCD	SCD incidence per decade
Murphy et al	163	30 years	6%	2.0%
Nollert et al	490	25 years	3%	1.2%
Silka et al	445	22 years	2.6%	1.8%
Norrgaard et al	125	25 years	5.6%	2.2%
Gatzoulis et al	793	21 years	6%	3.0%

SCD = sudden cardiac death.

AV block associated with 28-100% annual mortality.[420] Despite improvements in pacemaker technologies, the increased risk of late sudden cardiac death has been reduced but not entirely eliminated. Recent reports indicate a much higher postoperative mortality risk for defect-matched patients who either had transient AV block for more than 3 days or were pacemaker dependent.[421] Several authors have proposed that sudden cardiac death may be precipitated by late onset AV block, late device or lead failure, or systemic ventricular dysfunction associated with right ventricular pacing.[422]

9.3.2. Atrial arrhythmias

Atrial arrhythmias frequently complicate postoperative repairs in various forms of CHD and may be poorly tolerated, particularly in those with cyanotic heart disease, systemic right ventricles, univentricular hearts, or pulmonary hypertension.[252] Atrial tachyarrhythmias have been identified as a risk factor for sudden cardiac death in multiple studies of adults with CHD. The mechanism of sudden cardiac death has been attributed to rapid AV conduction, most notably at times of exertion, with hemodynamic instability caused by the atrial tachyarrhythmia itself[33] or by its degeneration into a secondary ventricular tachyarrhythmia.[46] In patients with atrial switch palliation for D-transposition of the great arteries, IART and atrial fibrillation have been associated with increased risk for sudden cardiac death in several studies[45,47] and are a common trigger for ventricular tachyarrhythmias in those with primary prevention ICDs.[46]

Focal atrial tachycardias and less common supraventricular tachyarrhythmias, such as twin AV node reentry, are not thought to be major contributors to sudden cardiac death. However, rapidly conducting or multiple AV accessory pathways, as commonly occur in Ebstein anomaly, are a well-established substrate for sudden cardiac death.[236] In general, ICDs are not indicated to terminate high-risk atrial arrhythmias (for which the shock vector is suboptimal).[423] Rather, effective treatment may be achieved with pharmacologic therapy or, as is preferable in most, by more definitive catheter or surgical ablation.[424]

9.3.3. Ventricular arrhythmias

As in diverse populations with assorted forms of heart disease, ICDs are indicated in adults with CHD resuscitated from sudden cardiac death and in those with spontaneous sustained ventricular tachycardia, after a careful workup has failed to identify a clear reversible cause.[97] Observational studies support a high rate of appropriate shocks in adults

Table 9.4 Appropriate and inappropriate ICD discharges in patients with CHD

Authors	Year	No. patients	Population	Follow-up	Percent appropriate	Annual rate appropriate	Percent inappropriate	Annual rate inappropriate
Dore A et al	2004	13	Heterogeneous adult	2.4 years	53.8%	N/A	N/A	N/A
Yap SC et al	2007	64	Heterogeneous adult	2.7 years	23.4%	N/A	40.6%	N/A
Witte KK et al	2008	20	Tetralogy of Fallot	3.7 years	20.0%	N/A	20.0%	N/A
Khairy P et al	2008	121	Tetralogy of Fallot	3.7 years	30.6%	PP: 7.7% SP: 9.8%	24.8%	5.8%
Khairy P et al	2008	37	TGA/atrial switch	3.6 years	13.5%	PP: 0.5% SP: 6.0%	24.3%	6.6%
Khanna AD et al	2011	73	Heterogeneous adult	2.2 years	19.2%	N/A	15.1%	N/A
Koyak Z et al	2012	136	Heterogeneous adult	4.6 years	28.7%	N/A	30.1%	N/A
Uyeda T et al.	2012	12	Heterogeneous adult	2.9 years	25.0%	PP: 0% SP: N/A	16.7%	N/A

CHD = congenital heart disease; ICD = implantable cardioverter-defibrillator; N/A = not available; Percent appropriate = proportion of patients with appropriate ICD discharges; Percent inappropriate = proportion of patients with inappropriate ICD discharges; PP = primary prevention; SP = secondary prevention; TGA = transposition of the great arteries.

Reproduced with permission from Mondesert B, Khairy P. Implantable cardioverter-defibrillators in congenital heart disease. Curr Opin Cardiol 2014;29 (1):45–52.[414]

with varied forms of CHD and secondary prevention ICDs (Table 9.4).[40,46,414,425,426] In ICD recipients with tetralogy of Fallot, a multicenter study reported a 7.7% and 9.8% annual incidence of appropriate ICD therapies with primary and secondary prevention indications, respectively.[40] In this carefully selected high-risk population, the incidence of appropriate shocks exceeded reported rates for hypertrophic cardiomyopathy (5%/year)[427] and ischemic or nonischemic cardiomyopathy (5.1%/year),[428] and approached MADIT-II subgroups (e.g., 9.0%/year in New York Heart Association [NYHA] class I or II patients).[429,430] Importantly, appropriate ICD shocks is an imperfect surrogate marker that overestimates risk of sudden cardiac death approximately 3-fold because not all ICD shocks are life-saving.[431]

Selecting candidates for primary prevention ICDs at risk for developing fatal ventricular arrhythmias remains a major challenge.[414,432] In general, ICDs are indicated in adults with CHD who meet standard recognized criteria backed by solid clinical trial evidence, that is, biventricular physiology with a systemic left ventricular ejection fraction $\leq$35%, biventricular physiology, and NYHA class II or III symptoms.[97,111,428,433–435] It may also be reasonable to consider a primary prevention ICD in adults with CHD and syncope of unknown origin with hemodynamically significant sustained ventricular tachycardia, particularly in those with high-risk substrates.[76,97,436] Importantly, syncope in adults with CHD may have several potential etiologies, including conduction abnormalities and bradyarrhythmias, atrial and/or ventricular arrhythmias, and nonelectrophysiologic causes.

Sustained ventricular tachyarrhythmias and sudden death have been well characterized in patients with tetralogy of Fallot.[18,101,102,106,111,112,437–458] Factors such as left ventricular diastolic dysfunction, increased QRS duration, nonsustained ventricular tachycardia, prior palliative shunt, ventriculotomies, and inducible sustained ventricular tachycardia appear to have an additive effect on rates of appropriate ICD therapies in those with primary prevention defibrillators.[31,40] Nonsustained ventricular tachycardia has been associated with inducible sustained ventricular tachycardia by programmed ventricular stimulation[76] and with clinical ventricular tachyarrhythmias in ICD recipients.[40] In a multicenter study of 252 patients with tetralogy of Fallot who underwent programmed ventricular stimulation, inducible sustained ventricular tachycardia was independently associated with a nearly 5-fold higher rate of clinical ventricular tachycardia or sudden cardiac death on follow-up.[76] Bayesian analyses suggest that its prognostic value is insufficient to justify routine screening and that its discriminative potential is greatest in those deemed at moderate risk of sudden death.[84,459]

Conversely, the value of programmed ventricular stimulation in adults with CHD in the absence of a prior ventriculotomy is limited or unknown.[46] Analyses in a small subgroup of patients with transposition of the great arteries and intra-atrial baffles suggest that inducible ventricular tachycardia does not predict clinical events.[46] Electrophysiologic studies may nevertheless be helpful in determining atrial arrhythmia vulnerability and in assessing the AV conduction system. A decreased systemic right ventricular ejection fraction has been associated with ventricular arrhythmias and sudden death.[47,48] However, uncertainty remains as to the optimal cutoff value for risk stratification, with circumstantial evidence suggesting that it may be lower than the widely used 35% threshold for systemic left ventricles.[46] Additional proposed risk factors include a wide QRS duration, atrial tachyarrhythmias, and systemic AV valve (i.e., tricuspid) regurgitation.[45–48] To date, attempts to risk stratify patients with Mustard or Senning baffles have yielded discouraging results.[46]

9.4. Recommendations for ICD therapy in adults with CHD

Recommendations	
Class I	1. ICD therapy is indicated in adults with CHD who are survivors of *cardiac arrest* due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia after evaluation to define the cause of the event and exclude any completely reversible etiology *(Level of evidence: B)*.[40,46,460–462] 2. ICD therapy is indicated in adults with CHD and *spontaneous sustained ventricular tachycardia* who have undergone hemodynamic and electrophysiologic evaluation *(Level of evidence: B)*.[40,46,97,426,460,461] Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD therapy in carefully selected patients *(Level of evidence: C)*.[463–465] 3. ICD therapy is indicated in adults with CHD and a *systemic left ventricular ejection* fraction $\leq$35%, biventricular physiology, and New York Heart Association (NYHA) class II or III symptoms *(Level of evidence: B)*.[97,111,428,433–435]
Class IIa	ICD therapy is reasonable in selected adults with *tetralogy of Fallot* and multiple risk factors for sudden cardiac death, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration $\geq$180 ms, extensive right ventricular scarring, or inducible sustained ventricular tachycardia at electrophysiologic study *(Level of evidence: B)*.[31,40,76,84,101,313,439,445,466]
Class IIb	1. ICD therapy may be reasonable in adults with a *single or systemic right ventricular ejection fraction* $<$35%, particularly in the presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA functional class II or III symptoms, QRS duration $\geq$140 ms, or severe systemic AV valve regurgitation *(Level of evidence: C)*.[45–48,435,467] 2. ICD therapy may be considered in adults with CHD and a *systemic ventricular ejection fraction* $<$*35%* in the absence of overt symptoms (NYHA class I) or other known risk factors *(Level of evidence of: C)*.[36,97,467] 3. ICD therapy may be considered in adults with CHD and *syncope of unknown origin* with hemodynamically significant sustained ventricular tachycardia or fibrillation inducible at electrophysiologic study *(Level of evidence: B)*.[76,97,436] 4. ICD therapy may be considered for nonhospitalized adults with CHD *awaiting heart transplantation (Level of evidence: C)*.[97,468] 5. ICD therapy may be considered for adults with syncope and moderate or complex CHD in whom there is a high clinical suspicion of ventricular arrhythmia and in whom thorough invasive and noninvasive investigations have failed to define a cause *(Level of evidence: C)*.[97,469]
Class III	1. All Class III recommendations listed in current ACC/AHA/HRS guidelines apply to adults with CHD *(Level of evidence: C)*.[97] These include: a. Life expectancy with an acceptable functional status $<$1 year; b. Incessant ventricular tachycardia or ventricular fibrillation; c. Significant psychiatric illness that may be aggravated by ICD implantation or preclude systematic follow-up; d. Patients with drug-refractory NYHA class IV symptoms who are not candidates for cardiac transplantation or cardiac resynchronization therapy. 2. Adults with CHD and advanced pulmonary vascular disease (Eisenmenger syndrome) are generally not considered candidates for ICD therapy *(Level of evidence: B)*.[470,471] 3. Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure prior to endocardial lead placement, or alternative approaches for lead access should be individualized *(Level of Evidence: B)*.[8,54,182]

9.5. Unique considerations for ICDs

Placement of ICD systems in adults with CHD necessitates individualized preprocedural strategic planning, including consideration of customized implant techniques.[7,472,473] Young adults with CHD are particularly likely to outlive the expected longevity of current-generation devices and leads, often necessitating complex extraction and multiple replacement procedures.

Transvenous leads carry risks of venous occlusion, embolic vascular events in the presence of an intracardiac shunt,[182] endocarditis, and lead failure from subclavian crush. There can be difficulty with achieving proper endocardial lead positioning due to abnormal systemic venous pathways, impaired or lack of venous access to the ventricle, or right-sided AV valve disease.[414] Conversely, disadvantages of ICD systems that require epicardial and/or subcutaneous coils include more invasive procedures, higher lead failure rates, and a possibility of developing restrictive "pericardial" physiology related to defibrillation patches.[474–476] Lead malfunctions requiring system revisions remain unacceptably common in adults with CHD regardless of implant technique.[460,476,477]

Limitations with standard transvenous and epicardial ICD systems in adults with CHD have prompted development of novel implantation techniques. Animal models and

computerized algorithms support the feasibility of functional ICD systems without transvenous shocking coils or epicardial patches.[478,479] For example, subcutaneous array and coils originally designed for adjunctive use in order to lower the defibrillation threshold have been utilized as the sole defibrillation lead.[480,481] In addition, an entirely subcutaneous ICD is now available.[482] The current system has a large generator and does not have the capability for chronic antibradycardia pacing (other than postshock transcutaneous pacing) or ATP, which may be indicated in a substantial proportion of adults with CHD requiring ICDs. The subcutaneous ICD may be a reasonable option in adults with CHD in whom transvenous access is not possible or desirable and in whom bradycardia and ATP functions are not essential.[414,483,484]

9.6. Results and outcomes of ICD therapy

The National Cardiovascular Data Registry (NCDR) for ICDs includes 801 (0.30%) patients with atrial septal defects, 588 (0.22%) with ventricular septal defects, 444 (0.17%) with tetralogy of Fallot, 232 (0.09%) with transposition of the great arteries, 48 (0.02%) with Ebstein anomaly, and 11 (<0.01%) with single ventricles.[426] Limited data suggest that the longevity of ICD systems in adults with CHD is lower than the 60% 8-year survival rate observed in adults without CHD. The lead is the weakest link such that the higher system failure rate is driven by lower ICD lead survival in younger patients.[477]

In populations exclusively with CHD followed for 1.9 to 4.6 years, 127 of 476 patients (27%) received appropriate ICD discharges, corresponding to a rate of 7% to 9% per year (Table 9.4).[40,46,414,425,426,439,485–487] Predictably, patients with secondary compared to primary prevention indications experienced a higher rate of appropriate ICD discharges.[40,46,426] ATP appears highly effective (e.g., 88%) in terminating ventricular tachycardia in patients with CHD, thereby reducing the need for shocks.[488] As also summarized in Table 9.4, 123 of 463 patients (27%) with CHD and ICDs had inappropriate ICD discharges, suggesting that they are as common as appropriate therapies.[489] Frequent causes of inappropriate shocks include sinus tachycardia, supraventricular arrhythmias, T-wave oversensing, and lead failure.[40,46,414]

A few published studies suggest that ICDs may negatively impact quality of life in adults with CHD.[489–491] A strong association between depression and anxiety with quality of life was observed in a study of adolescents with ICDs.[492] A prospective multicenter study from the Alliance for Adult Research in Congenital Cardiology (AARCC) on 180 adults with CHD with (N = 70) and without (N = 110) ICDs reported a high level of shock-related anxiety.[493] This anxiety was associated with depressive symptoms and sexual dysfunction in both men and women. These studies should raise awareness about the importance of recognizing psychosexual issues related to ICDs in adults with CHD.[414,494]

9.7. Considerations regarding ICD programming

ICD programming has evolved considerably such that the one zone "shock box" approach may not be ideal for the complex patient with CHD. Tailored programming may considerably reduce the rates of inappropriate and avoidable shocks. Data addressing optimal ICD programming in adults with CHD to maximize therapeutic benefits and minimize adverse events are limited.[495] Programming detection time/intervals consists of a balance between delaying therapy for potentially unstable arrhythmias and overtreating otherwise self-terminating nonsustained arrhythmias.[423] Although data are not available for adults with CHD, several trials, predominantly in patients with coronary artery disease, have shown longer delays for ventricular fibrillation detection to be safe and effective in reducing the incidence of shocks.[496–498] For example, a reduction in inappropriate therapies and all-cause mortality was achieved by programming no therapies for ventricular tachycardia rates <200 bpm or by delaying therapies, that is, by 60 seconds at 170–199 bpm, by 12 seconds at 200–249 bpm, and by 2.5 seconds at ≥250 bpm.[499]

ICD recipients with CHD often have coexisting supraventricular arrhythmias. While most device manufacturers offer several types of algorithms and discriminators using criteria such as QRS morphology, PR logic, onset and stability to minimize inappropriate shocks for atrial tachycardias, adjunctive pharmacologic treatment, or catheter ablation may be helpful. The frequent occurrence of bundle branch block and intraventricular conduction delay can complicate device programming and discrimination of arrhythmias. Adults with CHD and IART are particularly susceptible to having longer atrial tachycardia cycle lengths that favor 1:1 conduction via the AV node. These arrhythmias are prone to either going undetected or being misclassified as ventricular tachycardia.[119] In the absence of rate-dependent aberrancy, morphology discrimination algorithms may be beneficial in these circumstances. However, programming according to prior history of supraventricular tachycardia is not always reliable because adults with CHD may have multiple supraventricular substrates with differing rates and conduction characteristics.[495] Nevertheless, programming discriminators in slower zones appears justified to avoid inappropriate shocks. If discriminators are not activated, they should be programmed to a passive mode to assist in defining future cutoff values.[495]

For patients with secondary prevention ICDs, a safety margin of 30–60 ms between the slowest spontaneous or induced ventricular tachycardia and the cutoff rate may be reasonable for the ventricular tachycardia zone, the upper limit being more appropriate in the presence of antiarrhythmic drugs.[495] A monitoring zone to detect slower ventricular tachycardia or asymptomatic atrial arrhythmias is generally recommended.[500,501] Consideration should be given to programming ATP for fast and slow ventricular tachycardia zones in patients with spontaneous or inducible ventricular tachycardia. ATP can also be delivered before or during charging. Growing evidence indicates that ATP is safe,

painless, and effective.[496,497] Limited data in adults with CHD suggest that similar outcomes should be expected in this population.[40,46] Although ATP may occasionally accelerate the ventricular tachycardia rate, algorithms providing added security can be selected.

Defibrillation is the mainstay of therapy for ventricular fibrillation and rapid ventricular tachycardia. There are no specific studies analyzing low-energy versus high-energy shocks in adults with CHD. Purported advantages of programming a low first defibrillation shock include faster charge time with its lower risk of syncope, battery preservation, and reduction of postshock myocardial depression.[502] Maximum energy shocks improve first shock success with the added advantage of carrying a higher likelihood of terminating supraventricular arrhythmias. Defibrillation testing in adults with CHD may be indicated during follow-up if there are clinically suspected changes by X-ray or measured ICD data.[503]

9.7.1. Follow-up

The goals of ICD follow-up include patient assessment, confirmation of ICD integrity and function, and ensuring optimal programming to prevent inappropriate therapies and unnecessary shocks. Remote monitoring can be helpful for routine follow-up and for early detection of device malfunction or clinical deterioration permitting prompt intervention. The initial visit should include wound assessment, with periodic follow-up thereafter. Radiography may be helpful in assessing suspected lead displacement or malfunction. Clinical situations may warrant additional ICD evaluation, including changes in antiarrhythmic medications that may affect defibrillation thresholds and/or ventricular tachyarrhythmia rates, evaluation of shocks, and symptoms suggestive of arrhythmia or device malfunction. Although routine defibrillation threshold testing is not indicated, changes in lead integrity, pacing thresholds, and chest radiographic findings have been associated with higher defibrillation thresholds on follow-up.[503]

10. Cardiac resynchronization therapy

10.1. Dyssynchronous heart failure

Electromechanical dyssynchrony causes a sequence of events that may result in pathologic ventricular remodeling leading to dyssynchronous heart failure. The pathophysiology has been documented in animal experiments[504–507] and subsequently confirmed in the clinical setting. Early electrical activation and mechanical contraction cause initial stretch of late activated segments. By the time late segments contract, early segments have initiated their relaxation phase. Local myocardial work is decreased in early contracting sites that have a low local preload and increased in late sites where preload is enhanced by preceding stretch.[505] This may lead to asymmetric myocardial hypertrophy with a reduction in regional wall thickness and volume at early contracting sites and, conversely, increases in wall thickness and volume at late contraction sites.[505] Clinical observations have confirmed these experimentally described contraction patterns and have provided evidence of inefficient myocardial work in the setting of intraventricular dyssynchrony.[367]

Intraventricular mechanical dyssynchrony begets partially asymmetric cellular remodeling,[508] which may perpetuate the initial electrical insult and further contribute to the progression of intraventricular mechanical delay. The main components of these cellular changes can be summarized as follows:

- Increased levels of mediators of fibrosis and apoptosis in late contracting myocardial segments[509];
- Decreased calcium cycling between sarcoplasmatic reticulum and cytosol, resulting in impaired excitation–contraction coupling[510];
- Reduction in beta-adrenoreceptor gene expression, leading to a blunted response to adrenergic stimulation[511];
- Connexin43 down-regulation and lateralization in late contracting myocardial segments, with a consequent reduction in myocardial conduction velocity.[512]

Electromechanical dyssynchrony with an underlying ventricular activation delay due to bundle branch block or ventricular pacing is typically characterized by clustering (spatial proximity) of early and late contracting segments. Such dyssynchrony is theoretically amenable to CRT by electrically preexciting a large late contracting area composed of several myocardial segments via a single pacing lead. Mechanical dyssynchrony may, however, also be caused by contractile disparity.[513] More vigorously contracting segments prestretch those with lesser contraction force thereby delaying their contraction peak. Segments with a low contraction force that contract later and those with a high contraction force that contract earlier may be interspersed. This form of dyssynchrony is common in the setting of ischemic or idiopathic dilated cardiomyopathy with a narrow QRS complex. It is not amenable to CRT for 2 main reasons: absence of an electrical activation delay and the inability of current technology (e.g., limited number of ventricular leads) to correct dispersed mechanical dyssynchrony. Thus, differentiation of the specific type of ventricular dyssynchrony can inform clinical decisions regarding CRT.

The prevalence of dyssynchronous heart failure in CHD is unknown. One study specifically addressed potential indications for CRT in adults with systemic right ventricles.[514] If the selection of candidates for CRT was based solely on NYHA class II or more symptoms in the presence of a QRS duration $\geq$120 ms, 9.3% of patients with Mustard or Senning procedures would qualify compared to 6.1% of those with congenitally corrected transposition of the great arteries.

10.2. Clinical studies on CRT in CHD

CRT is an established treatment modality for systolic heart failure associated with left ventricular electromechanical dyssynchrony in adults with idiopathic and ischemic cardiomyopathy. CRT leads to restoration of a normal or

near-normal electromechanical activation pattern, an increase in myocardial energy efficiency,[515] reverse structural and cellular remodeling,[508] functional improvement, and a reduction in heart failure-associated morbidity and mortality.[516–521] Despite the far more heterogeneous structural and functional substrates encountered in adults with CHD, limited evidence suggests a potential role for CRT. Studies of CRT in CHD are summarized in Table 10.1.[522–529] Series exclusively in children without CHD and case reports are excluded.

Efficacy of CRT in CHD may vary with the underlying structural and functional substrate, such as anatomy of the systemic ventricle (left, right, or single), presence and degree of structural systemic AV valve regurgitation, primary myocardial disease or scarring, and type of electrical conduction delay. Available efficacy data are derived from two multicenter surveys,[522,525] one larger retrospective single-center study,[524] and several smaller case series. None of these studies were randomized, most were retrospective, and follow-up was largely limited to a few months, precluding an analysis of the impact of CRT on long-term morbidity and mortality. Surrogate outcomes were largely limited to metrics of systemic ventricular function. No study has yet assessed the impact of CRT in a heterogeneous population exclusively limited to adults with CHD. Despite these limitations, the effects of CRT in CHD in terms of reverse ventricular remodeling appear comparable to ischemic and idiopathic dilated cardiomyopathy. Considering the totality of evidence for CRT in CHD, the following observations may be made:

- Conventional single-site ventricular pacing with systemic ventricular dyssynchrony was the most prevalent (~65%) indication for CRT[522–525];
- Presence of LBBB along with a systemic left ventricle in the absence of ventricular pacing was a minor indication for CRT (9-17%)[524,525];
- RBBB in the presence of a systemic right ventricle was an even less common indication for CRT (5-7%)[524,525];
- The majority of reported patients (58%) had NYHA class II symptoms, reflecting a more proactive approach to CRT at a time when CRT guidelines for adult ischemic and idiopathic dilated cardiomyopathy required NYHA class III or IV symptoms;
- The reported absolute increase in systemic ventricular ejection fraction following CRT ranged between 6% and 20%;
- Presence of a systemic left ventricle was an independent predictor of a greater improvement in systolic systemic ventricular function[525];
- The best responses to CRT, with near complete reverse remodeling, were observed in patients with a systemic left ventricle who were converted to CRT from conventional right ventricular pacing[523,530];
- CRT was effective in combination with corrective or palliative cardiac surgery, particularly when performed to reduce systemic AV valve regurgitation[523,525,529];
- The proportion of CRT devices with defibrillation features was low (<25%);
- Nearly 40% of heart transplant candidates referred for CRT were subsequently delisted,[530] suggesting that patients with CHD awaiting heart transplantation may benefit from screening for potentially treatable mechanical dyssynchrony;
- The proportion of nonresponders to CRT (10%–14%)[522–525] was lower than in prospective adult trials, which may reflect the retrospective nature of available studies and softer endpoints rather than greater efficacy.

Demonstration of mechanical dyssynchrony is not a prerequisite for CRT in adults with ischemic or idiopathic dilated cardiomyopathy. The only prospective trial thus far found that the predictive power and reproducibility of echocardiography were insufficient to contribute to selecting appropriate candidates for CRT.[531] However, it may be hypothesized that in adults with CHD and a diversity of structural and functional CRT substrates (e.g., presence of a systemic right ventricle, single ventricle, RBBB), QRS duration alone may be a poorer predictor of systemic ventricular dyssynchrony than in patients with a structurally normal heart. It would be premature to discount a potential role for imaging in evaluating mechanical dyssynchrony in context with other findings in this specific population.[532–535]

10.3. Technical aspects

Anatomic constraints preclude implantation of transvenous CRT systems in a sizeable proportion of adults with CHD. In the 3 largest series of CRT in children and patients with CHD, thoracotomy or hybrid lead implantation was performed in 61%.[522,524,525] Nontransvenous lead implantation is required for CHD substrates such as univentricular hearts, transposition of the great arteries with Mustard or Senning baffles, and other conditions associated with unfavorable coronary venous anatomy. A hybrid approach consisting of transvenous lead insertion in the subpulmonary left ventricle and epicardial pacing of the systemic right ventricle may be performed in patients with Mustard or Senning baffles.[7,522,524,525] In patients with single ventricles, epicardial lead placement on opposing ventricular walls has been described but is technically very demanding.[524] Although not specifically studied, some patients with univentricular hearts may benefit from pacing the late activated region in fusion with intrinsic activation using only a single ventricular lead.[536,537]

The selection of pacing site may be guided by recording the delay in local electrical activation with respect to QRS onset. Late local activation has been shown to positively correlate with the increase in ventricular maximum +dP/dt.[538] The size of the left ventricular free-wall area where a lead must be placed to achieve a given percentage of the maximum possible CRT response was shown to be 17% for at least 90% of the maximal response and 28% for 80% maximal response.[539] None of the CHD studies to date have specifically

Table 10.1 Summary of clinical studies evaluating CRT in CHD

Author	Year	No. patients	Age (years)	CHD %	Systemic RV%	Single V%	Conv pacing %	NYHA III–IV %	QRS ms	EF pre %	EF post %	Nonresp %	Design and main features
Janousek et al	2004	8	15.0* (6.9–29.2)	100	100	0.	75.0	12.5	161*	18†‡	30†‡	—	Single-center, prospective, first study on utility of CRT in systemic right ventricles
Dubin et al	2005	103	12.8† (0.3–55.4)	70.9	16.5	6.8	44.7	37.9	166*	26*	40*	10.7	Multicenter, retrospective, first large study on CRT in congenital heart disease
Khairy et al	2006	13	7.8* (0.8–15.5)	100	30.8	0	100	—	>120 in all	31*	51*	11.1	Single-center, retrospective, impaired ventricular function and conduction abnormality in all, follow-up 17 months
Moak et al	2006	6	11.3* (0.5–23.7)	33.3	0	0	100	—	204*	34*	60*	0.0	Single-center, retrospective, super-response after upgrade from conventional right ventricular pacing to CRT
Cecchin et al	2009	60	15.0† (0.5–47.0)	76.7	15.0	21.7	68.3	31.7	160†	36†	42†	10.0	Single-center, retrospective, largest reported single ventricular patient group
Jauvert et al	2009	7	24.6* (15.0–50.0)	100	100	0	71.4	100.0	160*	—	—	—	Single-center, prospective, effect of CRT in systemic systemic right ventricle
Janousk et al	2009	109	16.9 (0.3–73.8)	79.8	33.0	3.7	77.1	45.9	160†	30*	41*	13.7	Multicenter, retrospective, effects of CRT in different structural and functional substrates
Thambo et al	2013	9	36.6* (>18)	100	0	0	0	—	164*	50*	56*	—	Single-center, prospective, postoperative tetralogy of Fallot, noninvasive mapping of ventricular activation

CHD = congenital heart disease; Conv pacing = conventional pacing prior to cardiac resynchronization therapy (CRT); EF post = ejection fraction following CRT; EF pre = ejection fraction prior to CRT; Nonresp = nonresponder; NYHA = New York Heart Association; RV = right ventricle; Single V = single ventricle.

*Mean value.

†Median value.

‡Right ventricular fractional area of change.

explored the usefulness of AV and VV delay optimization during CRT follow-up. Current evidence does not support routine AV and VV optimization.[540] However, in nonresponders to CRT and in those in need of atrial pacing, evaluation of AV and VV delay may be justified to correct suboptimal device settings. No clear differences between automated electrocardiographic algorithms and CRT optimization by echocardiography have been found.[540]

10.4. Recommendations

Current North American and European heart failure and device therapy guidelines[97,540–542] are based on multiple randomized prospective trials of CRT in adults with ischemic and idiopathic cardiomyopathy.[516–521] They consistently recommend CRT by biventricular pacemakers (CRT-P) or biventricular pacemakers combined with ICDs (CRT-D) in patients with a left ventricular ejection fraction ≤35%, dilated left ventricle, wide QRS complex (≥120 ms), and NYHA class III or IV symptoms despite optimal medical therapy. CRT, preferentially by a CRT-D device, has also been recommended to reduce morbidity and/or prevent disease progression in patients with a left ventricular ejection fraction ≤35%, QRS duration ≥150 ms, sinus rhythm, and NYHA functional class II symptoms on optimal medical therapy. Growing evidence suggests that CRT is less effective in subjects with RBBB[543] and that it may be harmful (i.e., induce dyssynchrony) in the absence of QRS prolongation.[544]

Management guidelines have not previously commented on CRT indications in patients with CHD. The writing committee, therefore, adapted published CRT guidelines[97,540–542] to the adult with CHD by considering the entirety of current evidence. An overview of recommendations is summarized in Figure 10.1.

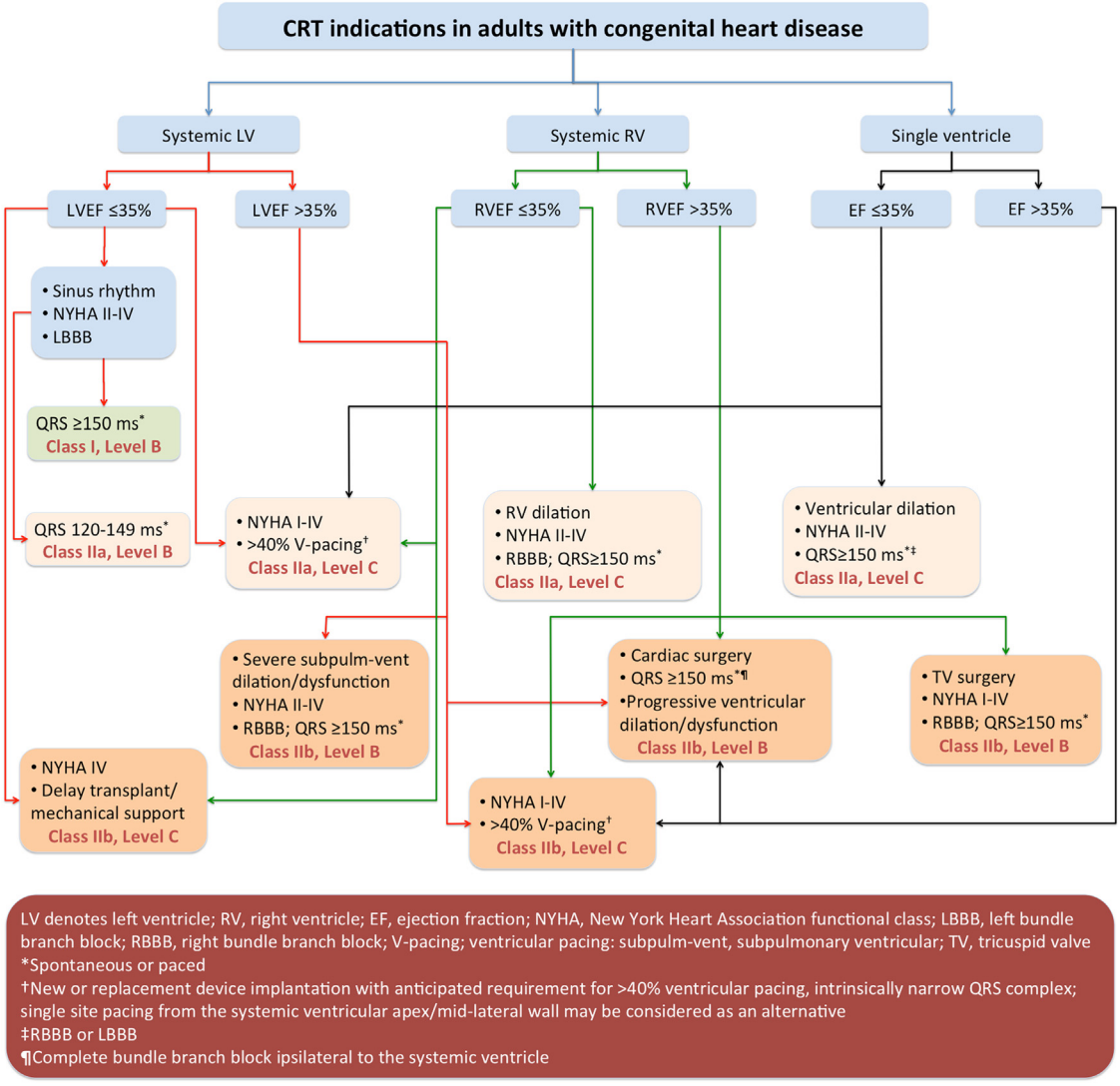

Figure 10.1 Overview of recommendations for cardiac resynchronization therapy (CRT) in adults with congenital heart disease (CHD). Please refer to the text for additional information.

Recommendations	
Class I	CRT is indicated in adults with CHD, a systemic left ventricular ejection fraction ≤35%, sinus rhythm, complete left bundle branch block (LBBB) with a QRS complex ≥150 ms (spontaneous or paced), and New York Heart Association (NYHA) class II to IV (ambulatory) symptoms *(Level of evidence: B)*.[97,522–525]
Class IIa	1. CRT can be useful for adults with CHD, a systemic left ventricular ejection fraction ≤35%, sinus rhythm, complete LBBB with a QRS complex 120–149 ms (spontaneous or paced), and NYHA class II to IV (ambulatory) symptoms *(Level of evidence: B)*.[97,516–525] 2. CRT can be useful for adults with a systemic right ventricular ejection fraction ≤35%, right ventricular dilation, NYHA class II to IV (ambulatory) symptoms, and complete right bundle branch block (RBBB) with a QRS complex ≥150 ms (spontaneous or paced) *(Level of evidence: C)*.[522–526,529,545,546] 3. CRT can be useful in adults with CHD, a systemic ventricular ejection fraction ≤35%, an intrinsically narrow QRS complex, and NYHA class I to IV (ambulatory) symptoms who are undergoing new or replacement device implantation with anticipated requirement for significant (>40%) ventricular pacing *(Level of evidence: C)*.[97,363,364,366,368,371,372,527,547–551] Single-site pacing from the systemic ventricular apex/mid-lateral wall may be considered as an alternative *(Level of evidence: C)*.[369,370,373,374,552,553] 4. CRT can be useful for adults with a single ventricle ejection fraction ≤35%, ventricular dilatation, NYHA class II to IV (ambulatory) symptoms, and a QRS complex ≥150 ms due to intraventricular conduction delay that produces a complete RBBB or LBBB morphology (spontaneous or paced) *(Level of evidence: C)*.[524]
Class IIb	1. CRT may be considered in adults with CHD, a systemic ventricular ejection fraction >35%, an intrinsically narrow QRS complex, and NYHA class I to IV (ambulatory) symptoms who are undergoing new or replacement device implantation with anticipated requirement for significant (>40%) ventricular pacing (*Level of evidence: C)*. Single-site pacing from the systemic ventricular apex/mid-lateral wall may be considered as an alternative *(Level of evidence: C)*.[363,366,368–375,547–553] 2. CRT may be considered in adults with CHD undergoing cardiac surgery with an intrinsic or paced QRS duration ≥150 ms, complete bundle branch block morphology ipsilateral to the systemic ventricular (left or right), NYHA class I to IV (ambulatory) symptoms, and progressive systolic systemic ventricular dysfunction and/or dilatation or expectation of such development regardless of the ejection fraction value, especially if epicardial access is required to implement CRT *(Level of evidence: B)*.[522–525] 3. CRT may be considered in adults with CHD and a systemic right ventricle undergoing cardiac surgery for tricuspid valve regurgitation with an intrinsic or paced QRS duration ≥150 ms, complete RBBB, and NYHA class I to IV (ambulatory) symptoms, regardless of the degree of right ventricular systolic dysfunction *(Level of evidence: B)*.[525,529] 4. CRT may be considered in adults with CHD (e.g., tetralogy of Fallot) with severe subpulmonary right ventricular dilatation and dysfunction, complete RBBB with a QRS complex ≥150 ms, and NYHA class II to IV (ambulatory) symptoms *(Level of evidence: C)*.[528,554–556] 5. CRT may be considered in selected adults with CHD, NYHA class IV symptoms, and severe systemic ventricular dysfunction in an attempt to delay or avert cardiac transplantation or mechanical support *(Level of evidence: C)*.[525]
Class III	1. CRT is not indicated in adults with CHD and a narrow QRS complex (<120 ms) *(Level of evidence: B)*.[544] 2. CRT is not indicated in adults with CHD whose comorbidities and/or frailty limit survival with good functional capacity to less than 1 year *(Level of evidence: C)*.[97,557]

11. Surgical options

11.1. Introduction

Early application of arrhythmia surgery for accessory connections associated with CHD demonstrated efficacy just as the field of catheter ablation for Wolff-Parkinson-White syndrome was developing.[558–562] Surgical interruption of accessory pathways is now largely limited to patients with failed catheter ablation attempts, particularly among those with Ebstein anomaly.[563–566] The efficacy of surgical therapy for IART in CHD is most extensively studied among patients with univentricular hearts or right heart obstructive lesions undergoing reoperations.[424,463,567] Favorable results for surgical ablation of atrial fibrillation associated with structural heart disease have been reported in large series of adults.[568,569] More modest success rates have been observed for ventricular tachycardia surgery associated with CHD, with concurrent ICD implantation often recommended.[446,463]

The majority of adults undergoing surgery for CHD are not routinely submitted to concomitant arrhythmia surgery, except at a few centers experienced with this approach. Increasing awareness of the substantial morbidity and mortality related to arrhythmias in adults with CHD, as well as the increasing numbers of patients undergoing reoperations, provides an opportunity to improve hemodynamics

and treat coexisting arrhythmias in 1 setting. Additionally, surgical interventions may potentially reduce the risk of developing de novo late arrhythmias or morbidity, by means of prophylactic lesions in the atria and pacing strategies. Stroke risk related to atrial fibrillation may be reduced by resection of the left atrial appendage, a common source of thrombi.[570]

This section of the consensus document reviews the populations of adults with CHD at highest risk for arrhythmia and reoperation, the efficacy of arrhythmia surgery, and the role of prophylactic techniques for reducing the occurrence of new onset arrhythmias.

11.2. Preoperative arrhythmia evaluation

Surgical management of arrhythmias in adults with CHD can be planned for preexisting arrhythmias or as a preemptive effort coupled with a cardiac operation. The arrhythmia intervention can usually be performed with little additional risk compared to the primary cardiac operation alone.[567] However, there is the possibility that any ablative procedure can be proarrhythmic or necessitate additional interventions, such as permanent pacing. For example, a right atrial Maze can impair sinus node function or create marked intra-atrial conduction delay.[571,572] Surgically placed lesions that are not full thickness may not eliminate the arrhythmia or perhaps even create zones of slow conduction that favor arrhythmogenesis.[573,574] Thus, prophylactic interventions should generally be reserved for patients with a definable arrhythmic substrate or high risk of further arrhythmia.

Data regarding the need for cardiac surgery has been documented in recent years via registries such as Concor, Society of Thoracic Surgeons (STS), and European Congenital Heart Surgeons Association.[575–577] These data provide information on the types of adults with CHD undergoing surgical interventions with regard to diagnosis, age, preoperative factors, and outcome. In the Concor database of 10,300 patients with a median age of 33 years, approximately 20% of patients underwent cardiac surgery during a follow-up of 15 years.[575] Reoperations constitute 16%–40% of cardiac surgeries among adults, with tetralogy of Fallot or pulmonary atresia/ventricular septal defect constituting 37% of reoperations,[575] defects associated with increased risk of sudden death and heart failure as patients age. In the absence of directed arrhythmia surgery, the impact of reoperation for hemodynamic improvement alone on risk of subsequent ventricular tachycardia and sudden death remains controversial.[463,578] Preexisting supraventricular arrhythmias generally persist in the absence of arrhythmia-specific surgery.[287]

In a series of adults with CHD undergoing multivalve surgery, concurrent arrhythmia surgery was performed in 12%.[579] The STS database that included 5265 adults with CHD operated on over 9 years had a 20% combined incidence of concurrent and primary arrhythmia surgery, including pacemaker implantation.[577] These registries have also shown that in the adult CHD surgical population, arrhythmia is the most common preoperative factor and postoperative complication, occurring in 7%–9%.[575–577] In the STS registry, the overall incidence of preoperative arrhythmia was 14%, with an additional 3% having AV block.[577] Patients undergoing Fontan revision or conversion had the highest incidence of preoperative arrhythmia, noted in 53%, followed by 16% in those having mitral vavuloplasty. Reoperation rates and prevalence of arrhythmias, as derived from numerous cohort studies, are summarized in Table 11.1.[21,26,31–33,58,76,96,236,580–583]

When open heart cardiac surgery is planned for an adult with CHD it is recommended that the individual undergo a thorough arrhythmia assessment to determine if any additional surgical interventions are required. Noninvasive evaluation, including an ECG, exercise testing, and 24-hour ambulatory cardiac rhythm monitoring, is recommended based on symptomatology. In some, an electrophysiologic study can assist in determining whether surgical management of arrhythmias is desirable. Recognizing that change in hemodynamics from surgical intervention will alter the substrate for subsequent ventricular arrhythmias, the need for preoperative invasive testing should be carefully assessed and offered to patients when there is a high probability of performing catheter or surgical ablation for the prevention of sustained ventricular tachycardia. The electrophysiologic study can help distinguish mechanisms of arrhythmias, sustainability, and hemodynamic significance. The specific arrhythmia substrate can be mapped to assist the surgeon in developing a proper ablation or incisional lesion set.

Table 11.1 Reoperation rates and estimated prevalence of arrhythmias in adults with CHD

CHD lesion	Reoperation	Atrial arrhythmias	Ventricular tachycardia
Ebstein anomaly	30%–50%	33%–60%	> 2%
Single ventricle	> 25%	40%–60%	> 5%
Tetralogy of Fallot	26%–50%	15%–25%	10%–15%
Transposition of the great arteries, atrial switch	15%–27%	26%–50%	7%–9%
Transposition of the great arteries, arterial switch	12%–20%	< 2%	1%–2%
Congenitally corrected transposition of the great arteries	25%–35%	> 30%	> 2%
Truncus arteriosus	55%–89%	> 25%	> 2%
Atrioventricular septal defect	19%–26%	5%–10%	< 2%
Atrial septal defect	< 2%	16%–28%	< 2%

CHD = congenital heart disease.

11.3. Recommendations for electrophysiologic study prior to adult CHD surgery

Recommendations	
Class IIa	A preoperative electrophysiologic study can be useful in adults with CHD and any of the following criteria, in order to identify and map arrhythmia substrates that may be addressed surgically with ablation or incisional lesion sets: 1. History of unexplained syncope or sustained ventricular tachycardia not attributed to correctable predisposing causes *(Level of evidence: B)*.[76,424,446,463,567,577,579] 2. Documented sustained supraventricular tachycardia, excluding atrial fibrillation *(Level of evidence: C)*.[424,567,575] 3. Ventricular preexcitation (*Level of evidence*: *B/C*).[225,584,585]
Class IIb	A preoperative electrophysiologic study may be considered in adults with CHD and any of the following criteria, in order to identify and map arrhythmia substrates that can be addressed surgically with ablation or incisional lesion sets: 1. Nonsustained rapid atrial or ventricular tachyarrhythmias *(Level of evidence: C)*.[68,567] 2. Moderate or complex CHD known to be at high risk for atrial arrhythmia development but without documented sustained arrhythmia *(Level of evidence: C)*[586] 3. History of palpitations or symptoms thought to be related to arrhythmia *(Level of evidence: C);* 4. Atrial fibrillation in the setting of a triggering supraventricular arrhythmia *(Level of evidence: C)*.[587]
Class III	1. A preoperative electrophysiologic study is not indicated in adults with simple forms of CHD, no history of palpitations or arrhythmia symptoms, and no significant documented arrhythmia by noninvasive testing *(Level of evidence: C)*. 2. A preoperative electrophysiologic study is not indicated in adults with CHD and permanent or persistent atrial fibrillation without evidence of a triggering supraventricular arrhythmia *(Level of evidence: C)*.

11.4. Role of surgery in treating preexisting arrhythmias

Surgical management of arrhythmias in CHD was initially performed for accessory connections[588] and subsequently for AV nodal reentrant tachycardia.[589] The treatment of other atrial tachyarrhythmias was advanced further with the introduction of the Cox-Maze procedure for atrial fibrillation and flutter.[590–592] Finally, although there has been a long history of surgical ablation in association with endocardial resection for scar-mediated ventricular tachycardia in the context of ischemic heart disease, surgical intervention is now uncommon for this indication.[593]

Catheter mapping and ablation have largely supplanted surgery for accessory conduction pathways, AV nodal reentrant tachycardia, and atrial flutter. Transcatheter approaches for paroxysmal and continuous atrial fibrillation continue to have improving success. In adults with CHD, the most common role of surgery in the treatment of tachyarrhythmias is a Maze procedure for paroxysmal or continuous atrial fibrillation or IART, performed while addressing the structural heart defect.

11.4.1. Supraventricular arrhythmias

In the early era of cardiac surgical arrhythmia treatment, >95% success was reported for Wolff-Parkinson-White syndrome.[225,234,558,561,594–596] At present, surgical treatment for AV reentrant tachycardia is reserved for patients in whom catheter ablation failed or was not feasible, particularly when surgery for structural heart disease is required.[234,558,561,594,595,597] The surgical approach to AV nodal reentrant tachycardia, now relegated to exceptional circumstances, includes a linear lesion from the posterior inferior rim of the coronary sinus ostium to the inferior vena cava and, in the setting of a right-sided AV valve, from the tricuspid valve annulus to the posterior coronary sinus os.[598,599] Surgical ablation for IART is far more common and is generally applied to patients with arrhythmias refractory to medical therapy and transcatheter procedures, or in those with associated structural heart disease that require surgery.[166,463]

Considerations in deciding to perform arrhythmia surgery include accessibility of the atria to transcatheter ablation techniques (i.e., venous access to the atrium). The right atrial lesion set described as part of the Cox-Maze III surgery was not developed for patients with CHD and was designed prior to the recognition of the importance of the cavotricuspid isthmus in perpetuating atrial reentry. Isthmus-dependent IART may be present in 30%–60% of patients with repaired CHD, and isthmus ablation alone may be adequate in the absence of multiple reentrant circuits.[586,600,601] Elimination of right-sided IART with modified right atrial Maze surgery exceeds 90% at 5–10 years of follow-up.[586] The addition of right atrial cryoablation to patients undergoing reoperation for tetralogy of Fallot reduced the incidence of late atrial tachycardia to 9%, versus 78% in patients not undergoing operative ablation.[463] In patients undergoing Fontan conversion, isthmus ablation alone was associated with higher recurrence of atrial tachycardia compared with the more extensive modified right atrial Maze.[600] Principles of arrhythmia interventions at the time of surgery for CHD are outlined in Table 11.2 These include (1) inferomedial right atrial (cavotricuspid isthmus) ablation for classic atrial flutter, (2) modified right atrial Maze for multiple IART circuits, and (3) left atrial Cox-Maze III for permanent or long-standing atrial fibrillation.[592] The need for permanent atrial pacing may be required for bradycardia or as an antitachycardia device.

Table 11.2 Operative techniques for arrhythmia surgery

Type of arrhythmia	Surgical techniques
Supraventricular	
Accessory connections	Endocardial or epicardial dissection and division, cryoablation
Focal atrial tachycardia	Map-guided resection, cryoablation
AV nodal reentrant tachycardia	Slow pathway modification with cryoablation
Right intra-atrial reentrant tachycardia	Cavotricuspid isthmus ablation
Cavotricuspid isthmus dependent	
Multiple reentrant circuits	Modified right atrial Maze
Left atrial macroreentry	Left atrial Cox Maze III lesions
Atrial fibrillation	Left atrial Cox Maze III lesions; cavotricuspid isthmus ablation ± right atrial Maze ± left atrial appendectomy
Ventricular tachycardia	
Scar related	Scar or endocardial fibrosis resection, focal ablation, lines of ablation between anatomic landmarks; map-guided resection or ablation

Atrial fibrillation in adults with CHD often occurs in the setting of left-sided heart disease, ventricular dysfunction, or unoperated septal defects.[31,602–605] Surgical ablation is usually performed at the time of valve repair in patients with atrial fibrillation that is persistent or of greater than 6 months' duration.[602,606] Importantly, right atrial Maze surgery is not effective in preventing recurrences of atrial fibrillation. In contrast, the biatrial Cox-Maze III procedure eliminates atrial fibrillation in >70% of adults,[607,608] particularly in the setting of concomitant mitral valve repair, atrial septal defect closure, or coronary bypass grafting. The surgical Maze is associated with superior freedom from recurrent atrial fibrillation when compared to catheter ablation.[609] Failure of left atrial ablation may be related to reentry via the mitral isthmus or right atrial sources.[604] Cox-Maze III lesions may be performed with a traditional "cut and sew" technique, or with cryothermy or radiofrequency ablation.[602,603,605] Use of contemporary probes/clamps/pens as an alternative to making incisions shortens operative time significantly. Efforts to minimize or "abbreviate" the left atrial lesion set are associated with higher recurrence rates of atrial fibrillation. Because the left atrium can be fully exposed during open heart surgery, performing complete pulmonary vein isolation and extending lesions to the mitral annulus and left atrial appendage, and possibly resection of the left atrial appendage, are often performed if they can be accomplished without increased morbidity or mortality from additional bypass and cross-clamp time.[607,610–613]

11.4.2. Management of the left atrial appendage

The left atrial appendage is a potential source for atrial thrombi in older patients with CHD and may predispose to thromboembolism.[117] Surgical closure techniques include external ligation or stapling, external ligation and amputation, and internal sutures. Benefits and risks related to closure of the left atrial appendage have focused on adult acquired heart disease.[614–620] To date, there have been 5 major clinical studies,[614,616–619] 1 of which was randomized.[614] Overall, no clear benefit was demonstrated, with 1 suggesting benefit, 3 reporting neutral results, and 1 demonstrating increased risk related to left atrial appendage occlusion. In adults with CHD, the majority of reoperations are valve related,[579,621] and late atrial tachyarrhythmias are the most frequent late complication. Selective closure of the left atrial appendage at the time of valve surgery can be considered, but there is insufficient evidence to support routine closure.

11.4.3. Recommendations for concomitant atrial arrhythmia surgery in adults with CHD undergoing open cardiac surgery

Recommendations	
Class I	1. A modified right atrial Maze procedure is indicated in adults undergoing Fontan conversion with symptomatic right atrial IART *(Level of evidence: B)*.[293,424,600,622,623]
	2. A modified right atrial Maze procedure in addition to a left atrial Cox Maze III procedure is indicated in patients undergoing Fontan conversion with documented atrial fibrillation *(Level of evidence: B)*.[293,424,600]
Class IIa	1. A left atrial Cox Maze III procedure with right atrial cavotricuspid isthmus ablation can be beneficial in adults with CHD and atrial fibrillation *(Level of evidence: B)*.[294,566,592,602,607,608,624,625]
	2. A (modified) right atrial Maze procedure can be useful in adults with CHD and clinical episodes of sustained typical or atypical right atrial flutter *(Level of evidence: B)*.[567,626]
Class IIb	Adults with CHD and inducible typical or atypical right atrial flutter without documented clinical sustained atrial tachycardia may be considered for (modified) right atrial Maze surgery or cavotricuspid isthmus ablation *(Level of evidence: B)*.[567,626]

11.4.4. Ventricular Arrhythmias

Ventricular arrhythmias in adults with CHD may arise from the left or right ventricle, with most occurring in the setting of a prior ventriculotomy or ventricular septal defect closure (with or without a patch) or concomitant ventricular dysfunction.[102,307,417,446] Surgical treatment ranges from cryoablation to endo- or epicardial resection and is most often applied in patients with structural heart disease requiring concomitant repair. Intraoperative map-guided ventricular tachycardia surgery has had success rates of 50%–85%.[446] Given the difficulties in adequately mapping the tachyarrhythmia substrate and the significant recurrence risks, at present, it is usually combined with ICD implantation. Historically, intraoperative empiric cryoablation of the infundibular septum between the ventricular septal defect patch and pulmonary annulus in tetralogy of Fallot was proposed but has not always been successful and potentially carries proarrhythmic risk.[578] Although correction of the hemodynamic lesion without ablation (e.g., pulmonary valve insertion/replacement for pulmonary regurgitation) may be clinically beneficial, a reduction in risk of subsequent ventricular tachycardia and sudden death has not been consistently demonstrated.[463,578,627,628]

11.5. Recommendations for concomitant ventricular arrhythmia surgery in adults with CHD undergoing open cardiac surgery

Recommendations	
Class IIa	Surgical ventricular tachycardia ablation guided by electrophysiologic mapping should be considered in adults with CHD and clinical sustained monomorphic ventricular tachycardia *(Level of evidence: B)*.[306,452,629]
Class IIb	1. Surgical ventricular tachycardia ablation guided by electrophysiologic mapping is reasonable in adults with CHD, no clinical sustained ventricular tachycardia, and inducible sustained monomorphic ventricular tachycardia with an identified critical isthmus *(Level of evidence: C)*.[306] 2. Adults with CHD and rapid ventricular tachycardia not mapped preoperatively but mapped intraoperatively may be considered for ventricular arrhythmia surgery *(Level of evidence: C)*.[452]

11.6. The role of surgery in preventing the development of arrhythmias

Prophylactic arrhythmia surgery implies that a preexisting arrhythmia has not been identified. It is, therefore, applicable to adults with CHD who have yet to have a diagnosed arrhythmia but are likely to develop one over time. Such an approach requires analysis of which populations are at highest risk for tachycardia development, which surgical lesion set to perform, and how to assess efficacy. Whereas prophylactic atrial arrhythmia surgery has been safely performed with minimal adverse consequences,[291,292] prophylactic ventricular arrhythmia surgery carries the possibility of proarrhythmia, including cardiac arrest.

Prophylactic arrhythmia surgery may be performed during primary repair of CHD or upon subsequent operations. Approximately 20% of adults undergoing CHD surgery have primary repairs, most commonly of atrial septal defects, Ebstein anomaly, and mitral or aortic valve disease. Patients undergoing primary repair of atrial septal defects beyond the age of 40 years have a high incidence of subsequent atrial arrhythmias, particularly atrial fibrillation, in 20%–35% of patients.[250,287] Lesions with the highest risk of reoperations include right heart obstructive lesions, conduits (e.g., tetralogy of Fallot, double-outlet right ventricle, and truncus arteriosus), univentricular hearts, and AV valve disease. CHD substrates associated with the highest incidence of arrhythmias over time include univentricular hearts, Ebstein anomaly, transposition of the great arteries following atrial switch, congenitally corrected transposition, atrial septal defect, and tetralogy of Fallot.[630] Patients more likely to develop atrial arrhythmias include those with significant AV valve regurgitation, greater atrial dilation, elevated pulmonary artery pressure, decreased ventricular function, a higher number of prior surgeries, and advancing age over 45 years.[31] Table 11.3 lists types of CHD that might benefit from efforts to reduce arrhythmias during surgery, and operative techniques. In asymptomatic patients with manifest accessory pathways, it is currently recommended to perform electrophysiologic study with attempted ablation prior to elective surgery whenever feasible.[8,584]

There are limited reports of prophylactic arrhythmia surgery. In patients undergoing initial Fontan surgery in whom surgical ablation in the right atrium was performed from the atriotomy to the tricuspid valve,[631] no positive impact of this intervention was demonstrated by 9 years of follow-up.[632] No arrhythmia developed in the intervention or control group.[632] A small number of patients undergoing Fontan conversion with arrhythmia surgery did not have clinical or inducible atrial tachycardia and underwent prophylactic modified right atrial Maze procedures.[424] None developed late atrial tachycardia at a median follow-up of 10 years. To assess the impact of prophylactic arrhythmia surgery, a large number of patients need to undergo a uniform lesion set. Prophylactic lesions should be reproducible by surgeons at many centers, with reliable landmarks. The lesions should carry minimal potential morbidity during surgery and should not be proarrhythmic. Electrophysiologic

Table 11.3 Prophylactic arrhythmia surgery in adults with CHD

Congenital heart substrate	Arrhythmia	Technique
Fontan revision or conversion	IART, atrial fibrillation	Modified right atrial Maze ± left atrial Cox Maze III
Ebstein anomaly	Accessory connection	Dissection and division or ablation
	IART	Modified right atrial Maze
	Atrial fibrillation	Left atrial Cox Maze III with right-sided lesion set ± left atrial appendectomy or oversew orifice
Right heart conduit revisions, tricuspid valve repair or replacement, congenital lesions with atrial dilation	IART	Cavotricuspid isthmus ablation or modified right atrial Maze
Left-sided valve repair/replacement	Atrial fibrillation	Left atrial Cox Maze III with cavotricuspid isthmus ablation, ± left atrial appendectomy or oversew orifice
Atrial septal defect closure	IART	Cavotricuspid isthmus ablation, (modified) right atrial Maze
	Atrial fibrillation	Left atrial Cox Maze III with cavotricuspid isthmus ablation ± left atrial appendectomy or oversew orifice

CHD = congenital heart disease; IART = intra-atrial reentrant tachycardia.

study prior to hospital discharge should be considered to assess the safety of prophylactic arrhythmia surgery and lack of proarrhythmic effects. Follow-up should be rigorous and long enough to assess meaningful outcomes.[633,634]

11.7. Recommendations for prophylactic atrial or ventricular arrhythmia surgery in adults with CHD

Recommendations	
Class IIa	1. A modified right atrial Maze procedure should be considered in adults undergoing Fontan conversion or revision surgery without documented atrial arrhythmias *(Level of evidence: B)*.[293,424,600,622,623,631]
	2. Concomitant atrial arrhythmia surgery should be considered in adults with Ebstein anomaly undergoing cardiac surgery *(Level of evidence: B)*.[626,635,636]
Class IIb	1. Adults with CHD undergoing surgery to correct a structural heart defect associated with atrial dilatation may be considered for prophylactic atrial arrhythmia surgery *(Level of evidence: C)*.[636,637]
	2. Adults with CHD and left-sided valvular heart disease with severe left atrial dilatation or limitations of venous access may be considered for left atrial Maze surgery in the absence of documented or inducible atrial tachycardia *(Level of evidence: C)*.[637]
	3. Closure of the left atrial appendage may be considered in adults with CHD undergoing atrial arrhythmia surgery *(Level of evidence: C)*.[614]
Class III	1. Prophylactic arrhythmia surgery is not indicated in adults with CHD at increased risk of surgical mortality from ventricular dysfunction or major comorbidities, in whom prolongation of cardiopulmonary bypass or cross-clamp times due to arrhythmia surgery might negatively impact outcomes *(Level of evidence: C)*.
	2. Empiric ventricular arrhythmia surgery is not indicated in adults with CHD and no clinical or inducible sustained ventricular tachyarrhythmia *(Level of evidence: C)*.[638]

Appendix 1

See Tables A1 and A2

References

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Table A1 PACES/HRS ACHD Writing Group Disclosures

Writing group	Institution	Consultant/ advisory board	Speakers' bureau/ honoraria	Research grant	Fellowship support	Board Mbs/stock options/partner	Others
Anne Dubin, MD, FHRS	Standford University	None	None	None	3; Medtronic, Inc.	None	None
Barbara Deal, MD	Children's Memorial Hospital	None	None	None	None	None	None
Carole Warnes	Mayo Clinic						
Charles Berul, MD, FHRS, CCDS	Children's National Medical Center	1; Johnson and Johnson	None	None	None	2; American heart Association	None
Curt Daniels							
Edward Walsh, MD, FHRS	Boston Children's Hospital	None	None	None	None	None	None
Frank Cecchin, MD	Children's Hospital Boston	1; St. Jude Medical	None	None	None	None	None
George Van Hare – HRS Chair, MD, FHRS, CCDS	St. Louis Children's Hospital	None	None	None	None	0; Heart Rhythm Society, HRS Consulting, Inc., and Heart Rhythm Foundation	None
James Perry, MD, FHRS, CEPS	UCSD/Rady Children's Hospital	1; Medtronic, Inc., U.S. Department of Justice	None	2; Medtronic, Inc.	None	None	None
Jan Janousek, MD, PhD	Kardiocentrum and Cardiovascular Research Center	None	None	None	None	None	None
John Triedman, MD, FHRS, CCDS	Children's Hospital Boston,	1; Bionsense Webster, Inc.	None	None	None	2; Up to Date	None
Joseph Dearani, MD	Mayo Clinic	None	None	None	None	None	None
Louis Harris, MBChB, FHRS	Toronto General Hospital	1; St. Jude Medical	None	None	None	None	None
Michael Silka, MD	Children's Hospital of Los Angeles	None	None	None	None	None	None
Mitchell Cohen (SCDC Rep), MD, FHRS, CCDS	Arizona Pediatric Cardiology Consultants	1; Medtronic, Inc.	None	None	None	None	None
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Table A1 (*continued*)

Writing group	Institution	Consultant/ advisory board	Speakers' bureau/ honoraria	Research grant	Fellowship support	Board Mbs/stock options/partner	Others
Paul Khairy - PACES Chair, MD, PhD	Montreal Heart Institute	1; Boehringer Ingelheim	None	5; Medtronic, St. Jude Medical, Boehringer Ingelheim, Canada Research Chair in Electrophysiology and Adult Congenital Heart Disease	4; St. Jude Medical	None	None
Peter Karpawich, MD, MS, FHRS	Children's Hospital of Michigan	None	None	None	None	None	None
Ronald J. Kanter, MD, FHRS	Duke Univ Medical Center	None	None	None	None	None	None
Seshadri Balaji, MBBS	Oregon Health and Science Univ	None	None	None	None	None	None
Stephan Seslar, MD, PhD, CCDS	Children's University Medical Group	None	None	None	None	None	None

0 =$0; 1 =≤ $10,000; 2 => $10,001 to ≤ $25,000; 3 => $25,001 to ≤ $50,000; 4 => $50,001 to ≤ 100,000; 5 => $100,001

Table A2 PACES/HRS ACHD Peer Reviewer Disclosures

Peer Reviewer	Employment	Consultant/ advisory board	Speakers' bureau/ honoraria	Research grant	Fellowship Support	BoardMbs/ stock options/partner	Others
Elizabeth Saarel, MD, FHRS, CEPS	Primary Children's Medical Center	None	None	None	None	None	None
John Sapp Jr. MD, FHRS	Queen Elizabeth II Health Sciences Center	0: Biosense Webster	None	3: Philips; 5: St. Jude Medical, Biosense Webster	None	None	None
John Rickard, MD	Johns Hopkins Medical	1: St. Jude Medical	None	None	None	None	None
Julia Indik, MD, PhD, FHRS	University of Arizona, Sarver Hearr Center	None	None	None	None	None	None

0 = $0; 1 = ≤ $10,000; 2 = > $10,001 to ≤ $25,000; 3 = > $25,001 to ≤ $50,000; 4 = > $50,001 to ≤ 100,000; 5 = > $100,001