PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease: Executive Summary

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 more than 90% of children with CHD in developed countries today are expected to survive to 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 more than 1 million adults with CHD in the United States alone, more than 100,000 in Canada, and 1.8 million in Europe.[3–5] Rhythm disorders, which span the entire spectrum of bradyarrhythmias 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

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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

Table 1 Classification of congenital heart disease complexity in adults

Complexity	Type of congenital heart disease in adult patients
Simple	*Native disease*
	Isolated congenital aortic valve disease
	Isolated congenital mitral valve disease (except parachute valve and 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, and ventricular inversion)

Adapted from Warnes et al.[8]

Table 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

electrophysiology, adult congenital heart disease (ACHD), and CHD surgery. The writing committee included representation from 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). 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 no sizable 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. The writing committee retained the nomenclature for complexity of CHD (i.e., simple, moderate, and complex/severe) proposed by the ACC/AHA task force on practice guidelines for adults with CHD,[8] which are summarized in Table 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, and III) and its accompanying level of evidence (levels A, B, and C) as summarized in Table 2.

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 the ACC, AHA, EHRA, CHRS, and ISACHD. All writing members approved this final version.

The following Executive Summary represents the complete set of recommendations for the recognition and management of arrhythmias in adults with CHD. The reader is referred to the supporting text in the full document for further information.

Although detailed recommendations regarding training and skills required to qualify as an electrophysiologist with expertise in ACHD are beyond the scope of this consensus document, basic competencies are summarized in Table 3.[18–21]

Figure 1 summarizes the recommended approach to rhythm control in adults with CHD and IART or atrial fibrillation.

Recommendations for thromboprophylaxis in adults with CHD and IART are summarized in Figure 2.

An overview of recommendations is summarized in Figure 3.

3. Delivery of care and ensuring access to care

3.1. 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 ACHD centers of excellence that serve the surrounding medical community as a resource for consultation and referral *(Level of evidence: C)*.[10]
	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)*.[8,11,12]
	3. Diagnostic and interventional catheter- and device-based electrophysiological 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)*.[7,13,14]

3.2. Recommendations for adults with CHD requiring invasive electrophysiological interventions

Recommendations	
Class I	1. Consultation with an ACHD specialist should be sought before invasive electrophysiological interventions in adults with CHD *(Level of evidence: C)*.[8,11,12]
	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)*.[6,7,15]
	3. Invasive electrophysiological 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)*.[16]
	4. The electrophysiology laboratory and postprocedure recovery unit should be suitable for the care of adults with CHD, which includes the following:
	a. Adult appropriate equipment *(Level of evidence: C)*;
	b. Nursing and technical staff certified in adult cardiac life support and trained in basic CHD anatomy *(Level of evidence: C)*;
	c. ACHD cardiothoracic surgical backup and operating room access *(Level of evidence: C)*.[17]

Table 3 Basic requirements for electrophysiologists with expertise in adult congenital heart disease

Completion of specialized fellowship training in adult or pediatric electrophysiology with demonstrated acquisition of required clinical competencies[18–21]
Formal affiliation with an established adult congenital heart disease center[8,20]
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[6,7,15]
Experience and skills in managing adults with congenital heart disease and arrhythmias,[20] including
• Noninvasive testing
• Electrophysiology studies
• Catheter ablation, including with three-dimensional electroanatomic mapping systems and large-tip/irrigated catheters
• Intraoperative procedures
• Cardiac rhythm management devices

4. Evaluation and diagnosis of arrhythmias

4.1. 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, and syncope), documented new-onset or worsening arrhythmias, or resuscitated sudden cardiac death *(Level of evidence: C)*.[22] 2. A resting 12-lead electrocardiogram (ECG) is indicated in adults with CHD who are evaluated for arrhythmias *(Level of evidence: C)*.[23] 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)*.[24–27] 4. Cardiac event loop recorders are indicated to establish whether sporadic symptoms are caused by transient arrhythmias *(Level of evidence: C)*.[28,29] 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)*.[27,30,31] 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 electrophysiological testing *(Level of evidence: B)*.[28,32]
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)*.[33,34]
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)*.[33]
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)*.[8,32,34] 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)*.[14,35] 3. Coronary artery evaluation is indicated in assessing life-threatening ventricular arrhythmias or resuscitated sudden cardiac death in adults with CHD older than 40 years 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)*.[36,37]
c. Electrophysiological testing	
Class I	Electrophysiological 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)*.[27,38,39]
Class IIa	Electrophysiological 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 electrophysiological procedure *(Level of evidence: B)*.[33,38,40–42]
Class IIb	Electrophysiological 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)*.[33]

4.2. 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, at periodic intervals for CHD of moderate complexity, and occasionally for simple forms of CHD *(Level of evidence: C)*.[8,11,12] 2. Surveillance for adults with moderate or severe CHD should include a standard 12-lead ECG at least once a year *(Level of evidence: C)*.[23] 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)*.[30]
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)*.[25,26] 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)*.[27,30,31,38,43]
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)*.[38,44] 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)*.[41]

5. Medical therapy

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

Recommendations	
Class I	In adults with CHD, the choice of pharmacological therapy for arrhythmia management should consider factors such as coexisting sinus node dysfunction, impaired atrioventricular (AV) nodal conduction, systemic or subpulmonary ventricular dysfunction, associated therapies, child-bearing potential, and acquired comorbidities *(Level of evidence: B)*.[45,46]
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)*.[47,48] 3. In the pharmacological 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*).[47,48] 4. In adults with CHD and frequent recurrent symptomatic IART, an ablation strategy is preferable to long-term pharmacological therapy *(Level of evidence: B)*.[49–55] 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 pathological hypertrophy of the systemic ventricle, systemic or subpulmonary ventricular dysfunction, or coronary artery disease *(Level of evidence: C)*.[56] 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 or $\geq$500 ms in the presence of ventricular conduction delay (*Level of evidence: B*).[45,57,58] 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*).[45,57,58] 7. Subject to standard precautions and barring any contraindication (e.g., creatinine clearance $<$20 mL/min, hypokalemia, corrected QT interval $>$440 or $\geq$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*).[59,60]

Class IIb	1. It may be reasonable to liberalize the use of β-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*).[41,61] 2. Subject to standard precautions (e.g., renal insufficiency, hypokalemia, severe sinus node dysfunction or AV nodal disease, uncorrected QT interval >460 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*).[45,46,62]
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*).[63–67] 2. Dronedarone is not recommended in patients with a history of heart failure, moderate or severe systolic ventricular dysfunction, or moderate or complex CHD due to potential concerns over worsening heart failure and increased mortality (*Level of evidence: B*).[68,69]

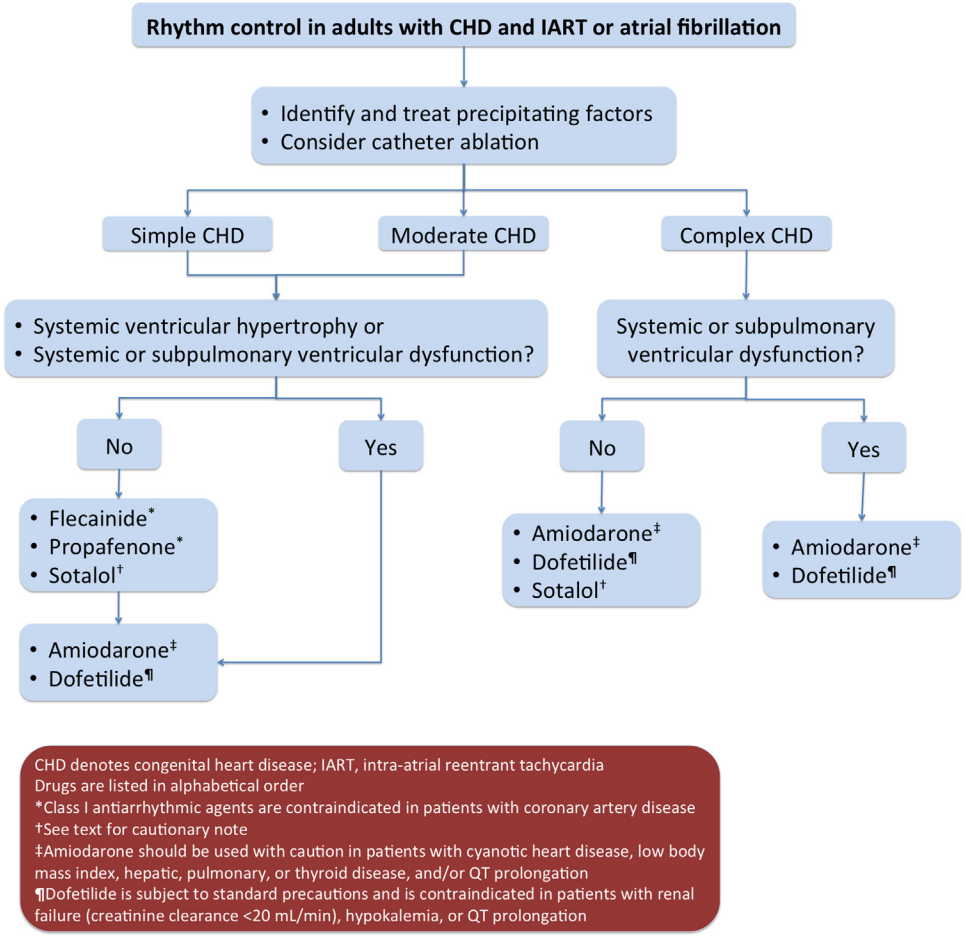
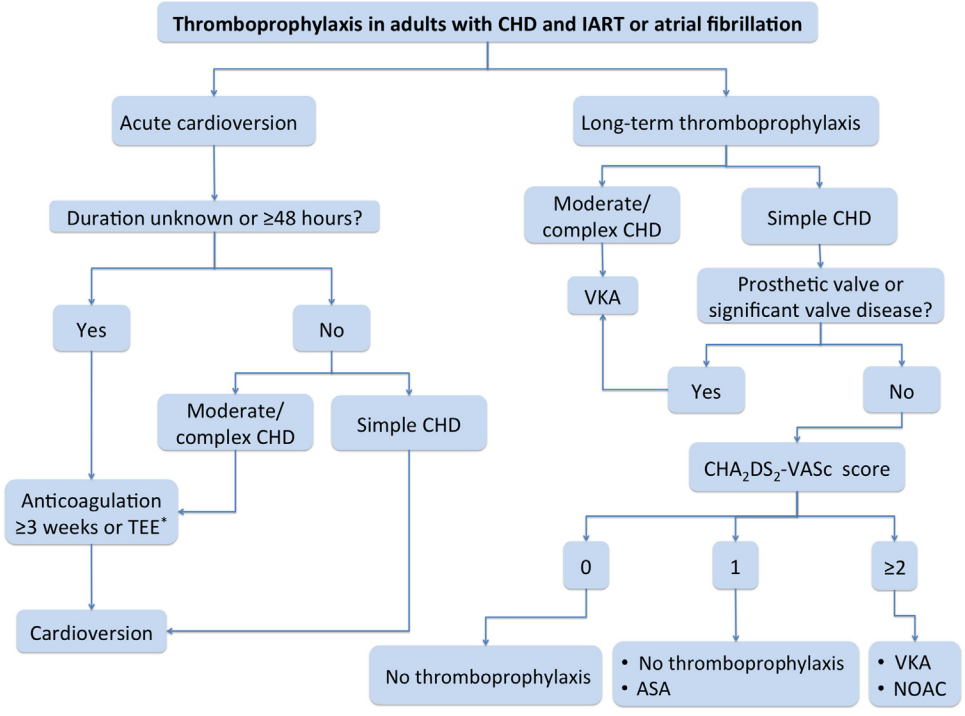


Figure 1 Rhythm control in adults with congenital heart disease and intra-atrial reentrant tachycardia or atrial fibrillation

5.2. Recommendations for thromboprophylaxis

Recommendations	
Class I	1. For adults with simple forms of CHD and hemodynamically stable IART or atrial fibrillation of unknown or ≥48-hour duration, therapeutic anticoagulation is recommended for at least 3 weeks before cardioversion or, alternatively, a transesophageal echocardiogram may be performed to rule out intracardiac thrombus *(Level of evidence: B)*.[48,56,70–72]
	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)*.[73–77]
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 before cardioversion or perform transesophageal echocardiography to rule out thrombus, regardless of arrhythmia duration *(Level of evidence: B)*.[42,78]
	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)*.[73–77]
	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)*.[74–77,79]
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)*.[80,81]
	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)*.[82–85]
Class III	1. Pending further studies, there is 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)*.



CHD denotes congenital heart disease; IART, intra-atrial reentrant tachycardia; TEE, transesophageal echocardiography; VKA, vitamin K antagonist; NOAC, newer oral anticoagulant; CHA_2DS_2-VASc: Congestive heart failure; Hypertension; Age (≥75 years, 2 points; 65-74 years, 1 point); Diabetes; Stroke, transient ischemic attack, or thromboembolism (2 points); VAscular disease; Sex category (female); ASA, aspirin

*Patients with Fontan palliation are at particularly high risk of thromboembolic complications such that TEE may be prudent prior to cardioversion even if therapeutic anticoagulation is received for ≥3 weeks

Figure 2 Thromboprophylaxis in adults with CHD and IART or atrial fibrillation.

6. Catheter ablation

6.1. Recommendations for catheter ablation of atrial tachyarrhythmias 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)*.[86–89] 2. Catheter ablation is useful for adults with CHD and symptomatic and/or drug refractory IART or focal atrial tachycardia *(Level of evidence: B)*.[86,90–101] 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)*.[102] 4. A three-dimensional electroanatomic mapping system is indicated for guiding ablation of postoperative atrial tachyarrhythmias in adults with CHD *(Level of evidence: B)*.[90,91,103–105]
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)*.[105–107] 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)*.[89,97,108–110] 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)*.[111]
Class IIb	1. It may be reasonable to perform invasive diagnostic electrophysiology studies in patients with Ebstein anomaly before anticipated cardiac surgery *(Level of evidence: B)*.[112] 2. In adults with CHD and symptomatic atrial tachyarrhythmia refractory to pharmacological and standard ablation therapy, it may be reasonable to consider AV node ablation and pacing as third-line therapy *(Level of evidence: C)*.[113,114]

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

Recommendations	
Class I	Catheter ablation is indicated as adjunctive therapy to an implantable cardioverter-defibrillator (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)*.[33,115]
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)*.[116,117]
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)*.[117] 2. Catheter ablation may be reasonable in adults with CHD and frequent ventricular ectopy associated with deteriorating ventricular function *(Level of evidence: C)*.[33]
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)*.[33] 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)*.[33]

7. Bradyarrhythmias and pacemakers

7.1. 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)*.[39,118–120] Devices that minimize ventricular pacing are preferred *(Level of evidence: B)*.[121–125]
	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)*.[39,126–130]
	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)*.[39,131–133]
	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)*.[39,134–136]
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)*.[39,137]
	2. Permanent pacing is reasonable for adults with CHD and sinus or junctional bradycardia for the prevention of recurrent IART *(Level of evidence: C)*.[39,138–140] Devices with atrial antitachycardia pacing properties are preferred in this subpopulation of patients *(Level of evidence: B)*.[139–142]
	3. Permanent pacing is reasonable for adults with congenital complete AV block and an average daytime resting heart rate <50 beats/min *(Level of evidence: B)*.[39,143,144]
	4. Permanent pacing is reasonable for adults with complex CHD and an awake resting heart rate (sinus or junctional) <40 beats/min or ventricular pauses >3 seconds *(Level of evidence: C)*.[39] 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)*.[139–142]
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 beats/min or ventricular pauses >3 seconds *(Level of evidence: C)*.[39,118,119,145] 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)*.[139–142]
	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)*.[39,146]
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)*.[39]
	2. Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure before endocardial lead placement, or alternative approaches for lead access should be individualized *(Level of evidence: B)*.[8,11,147]

8. Sudden cardiac death and ICDs

8.1. 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)*.[41,43,148–150]
	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)*.[39,41,43,148,149,151] Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD therapy in carefully selected patients *(Level of evidence: C)*.[152–154]
	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)*.[39,155–159]
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 ≥ 180 ms, extensive right ventricular scarring, or inducible sustained ventricular tachycardia at an electrophysiology study *(Level of evidence: B)*.[38,43,44,160–165]
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 ≥ 140 ms, or severe systemic AV valve regurgitation *(Level of evidence: C)*.[41,159,166–169]
	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: C)*.[39,169,170]
	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 an electrophysiology study *(Level of evidence: B)*.[38,39,171]

	4. ICD therapy may be considered for nonhospitalized adults with CHD *awaiting heart transplantation (Level of evidence: C)*.[39,172] 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)*.[39,173]
Class III	1. All class III recommendations listed in the current ACC/AHA/HRS guidelines apply to adults with CHD *(Level of evidence: C)*.[39] These include the following: 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 (CRT). 2. Adults with CHD and advanced pulmonary vascular disease (Eisenmenger syndrome) are generally not considered candidates for ICD therapy *(Level of evidence: B)*.[174,175] 3. Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure before endocardial lead placement, or alternative approaches for lead access should be individualized *(Level of Evidence: B)*.[8,11,147]

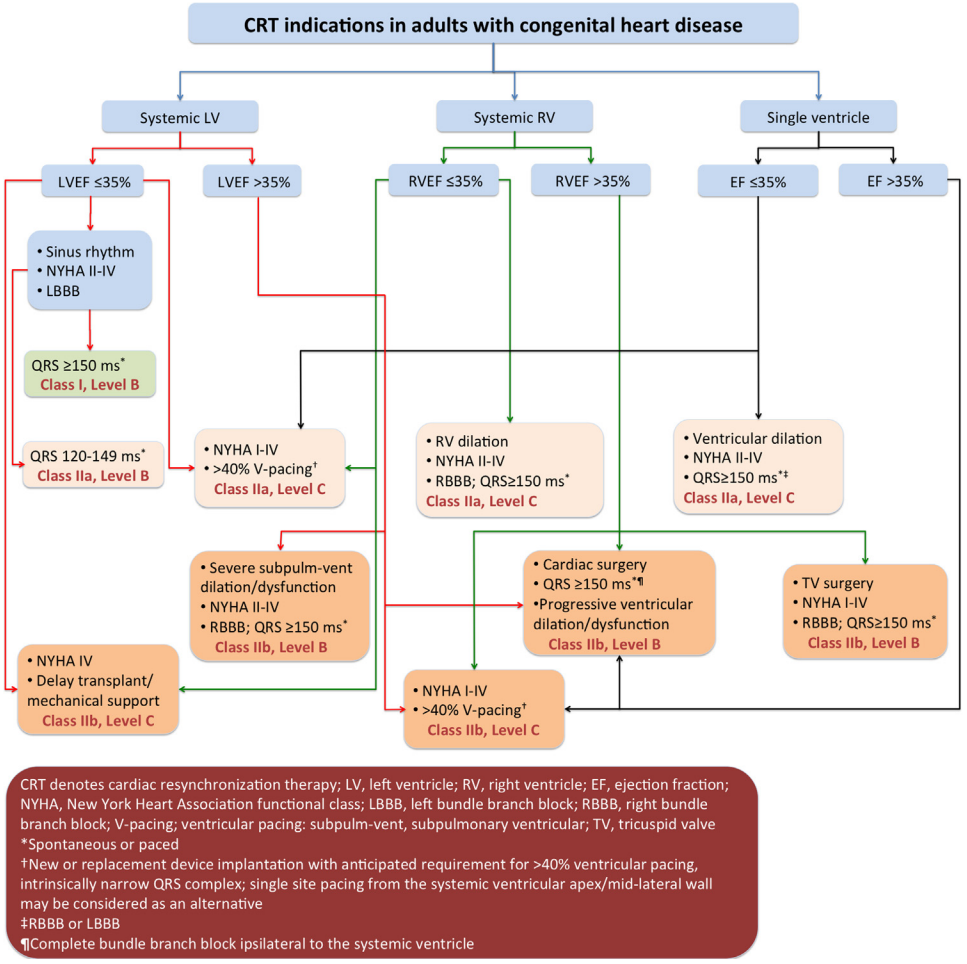


Figure 3 Overview of recommendations for CRT in adults with CHD. Please refer to the text for additional information.

9. Cardiac resynchronization therapy

9.1. Recommendations for cardiac resynchronization therapy

Recommendations	
Class I	CRT is indicated in adults with CHD, a systemic left ventricular ejection fraction $\leq$35%, sinus rhythm, complete left bundle branch block (LBBB) with a QRS complex $\geq$150 ms (spontaneous or paced), and NYHA class II to IV (ambulatory) symptoms *(Level of evidence: B)*.[39,176–179]
Class IIa	1. CRT can be useful for adults with CHD, a systemic left ventricular ejection fraction $\leq$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)*.[39,176–185] 2. CRT can be useful for adults with a systemic right ventricular ejection fraction $\leq$35%, right ventricular dilation, NYHA class II to IV (ambulatory) symptoms, and complete right bundle branch block (RBBB) with a QRS complex $\geq$150 ms (spontaneous or paced) *(Level of evidence: C)*.[176–179,186–189] 3. CRT can be useful in adults with CHD, a systemic ventricular ejection fraction $\leq$35%, an intrinsically narrow QRS complex, and NYHA class I to IV (ambulatory) symptoms who are undergoing new or replacement device implantation with anticipated requirement for significant ($>$40%) ventricular pacing *(Level of evidence: C)*.[39,190–201] Single-site pacing from the systemic ventricular apex/mid-lateral wall may be considered as an alternative *(Level of evidence: C)*.[202–207] 4. CRT can be useful for adults with a single ventricle ejection fraction $\leq$35%, ventricular dilatation, NYHA class II to IV (ambulatory) symptoms, and a QRS complex $\geq$150 ms due to intraventricular conduction delay that produces a complete RBBB or LBBB morphology (spontaneous or paced) *(Level of evidence: C)*.[177]
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)*.[191–196,198–208] 2. CRT may be considered in adults with CHD undergoing cardiac surgery with an intrinsic or paced QRS duration $\geq$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)*.[176–179] 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 $\geq$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)*.[178,186] 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 $\geq$150 ms, and NYHA class II to IV (ambulatory) symptoms *(Level of evidence: C)*.[209–212] 5. CRT may be considered in selected adults with CHD, NYHA class IV symptoms, and severe systemic ventricular dysfunction in an attempt to delay or avert cardiac transplantation or mechanical support *(Level of evidence: C)*.[178]
Class III	1. CRT is not indicated in adults with CHD and a narrow QRS complex ($<$120 ms) *(Level of evidence: B)*.[213] 2. CRT is not indicated in adults with CHD whose comorbidities and/or frailty limit survival with good functional capacity to $<$1 year *(Level of evidence: C)*.[39,214]

10. Surgical options

10.1. Recommendations for an electrophysiology study before ACHD surgery

Recommendations	
Class IIa	A preoperative electrophysiology 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)*;[38,152,215–219] 2. Documented sustained supraventricular tachycardia, excluding atrial fibrillation *(Level of evidence: C)*;[215,216,220] 3. Ventricular preexcitation (*Level of evidence: B/C*).[87,221,222]
Class IIb	A preoperative electrophysiology 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)*;[15,216] 2. Moderate or complex CHD known to be at high risk for atrial arrhythmia development but without

	documented sustained arrhythmia *(Level of evidence: C)*;[223] 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)*.[224]
Class III	1. A preoperative electrophysiology 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 electrophysiology 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)*.

Principles of arrhythmia interventions at the time of surgery for CHD are outlined in Table 4. These include (1) inferomedial right atrial (cavotricuspid isthmus) ablation for classic atrial flutter, (2) modified right atrial Maze procedure for multiple IART circuits, and (3) left atrial Cox-Maze III procedure for permanent or long-standing atrial fibrillation.[225] The need for permanent atrial pacing may be required for bradycardia or as an antitachycardia device.

Table 4 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 dependent	Cavotricuspid isthmus ablation
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 and/or right atrial Maze and/or 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

10.2. 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)*.[215,226–229] 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)*.[215,226,228]
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)*.[225,230–236] 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)*.[216,237]
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)*.[216,237]

10.3. 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 is reasonable in adults with CHD and clinical sustained monomorphic ventricular tachycardia *(Level of evidence: B)*.[117,238,239]
Class IIb	1. Surgical ventricular tachycardia ablation guided by electrophysiologic mapping may be considered in adults with CHD, no clinical sustained ventricular tachycardia, and inducible sustained monomorphic ventricular tachycardia with an identified critical isthmus *(Level of evidence: C)*.[117] 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)*.[238]

10.4. 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)*.[215,226–229,240] 2. Concomitant atrial arrhythmia surgery should be considered in adults with Ebstein anomaly undergoing cardiac surgery *(Level of evidence: B)*.[237,241,242]
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)*.[242,243] 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)*.[243] 3. Closure of the left atrial appendage may be considered in adults with CHD undergoing atrial arrhythmia surgery *(Level of evidence: C)*.[244]
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 affect 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)*.[245]

Appendix 1. PACES/HRS ACHD writing group disclosures

See Table A1

Appendix 2. PACES/HRS ACHD peer reviewer disclosures

See Table A2

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Table A1

Writing group	Institution	Consultant/Advisory board	Speakers' bureau/ Honoraria	Research grant	Fellowship support	Board members/stock options/partner	Others
Anne M. Dubin, MD, FHRS	Standford University	None	None	None	3; Medtronic	None	None
Barbara J. Deal, MD, FACC	Children's Memorial Hospital	None	None	None	None	None	None
Carole A. Warnes, MD, FRCP, FACC, FAHA	Mayo Clinic						
Charles I. Berul, MD, FHRS	Children's National Medical Center	1; Johnson and Johnson	None	None	None	2; American Heart Association	None
Curt J. Daniels, MD, FACC							
Edward P. Walsh, FACC, MD, FHRS	Boston Children's Hospital	None	None	None	None	None	None
Frank Cecchin, MD, FACC	Children's Hospital Boston	1; St. Jude Medical	None	None	None	None	None
George F. Van Hare, MD, FACC, FHRS (Co-Chair)	St. Louis Children's Hospital	None	None	None	None	0; Heart Rhythm Society, HRS Consulting, Heart Rhythm Foundation	None
James C. Perry, MD, FACC, FHRS	UCSD/Rady Children's Hospital	1; Medtronic, U.S. Department of Justice	None	2; Medtronic	None	None	None
Jan Janousek, MD, PhD	Kardiocentrum and Cardiovascular Research Center	None	None	None	None	None	None
John K. Triedman, MD, FACC, FHRS	Children's Hospital Boston	1; Bionsense Webster	None	None	None	2; *UpToDate*	None
Joseph A. Dearani, MD, FACC	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 J. Silka, MD, FACC, FAHA	Children's Hospital of Los Angeles	None	None	None	None	None	None
Mitchell I. Cohen (SCDC Rep), MD, FACC, FHRS	Arizona Pediatric Cardiology Consultants	1; Medtronic	None	None	None	None	None
Natasja de Groot, MD, PhD	Erasmus University Center	None	None	None	None	None	None
Paul Khairy, MD, PhD, FRCPC (Chair)	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

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Ronald K. Kanter, MD, FHRS	Duke University Medical Center	None	None	None	None	None	None
Seshadri Balaji, MBBS, PhD	Oregon Health and Science University	None	None	None	None	None	None
Stephan P. Seslar, MD, PhD	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

Peer reviewer	Employment	Consultant/Advisory board	Speakers' bureau/ Honoraria	Research grant	Fellowship support	Board members/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 Heart 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