

PACES/HRS expert consensus statement on the use of catheter ablation in children and patients with congenital heart disease



Developed in partnership with the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American Academy of Pediatrics (AAP), the American Heart Association (AHA), and the Association for European Pediatric and Congenital Cardiology (AEPC)

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ablation, and high-powered ablation catheters using passive or active tip cooling. In addition, the uses of catheter ablation in pediatrics have expanded to include the management of virtually every arrhythmia, including atrial fibrillation (AF). The Pediatric and Congenital Electrophysiology Society (PACES), in conjunction with the Heart Rhythm Society (HRS), has recently published related consensus guidelines and documents that include the following: the uses of catheter ablation for the management of asymptomatic Wolff-Parkinson-White (WPW) pattern found on the ECG (2012)²; adult patients with congenital heart disease (2014)³; and ventricular arrhythmias in children with a structurally normal heart (2014).⁴ This consensus statement provides up-to-date clinical practice guidelines for the use of catheter ablation in children 0–18 years of age with any arrhythmia, and in all patients with congenital heart disease. This statement attempts to also be consistent, where possible, with the 2012 and 2014 documents noted above. In case of conflict, the more recent publication of this document would indicate it supersedes the prior recommendations.

Methods and Evidence

As with the 2012 and 2014 documents, the writing group for this document was formed by PACES in conjunction with HRS. The goal of the writing committee was to focus primarily on the methodology, utility, and safety of catheter ablation in various clinical settings, and not on the overall management of particular arrhythmias. By necessity, some aspects of management are included, particularly when incorporating the history of symptoms and treatments that might lead to a choice of ablation versus other therapeutic options. In general, the guidelines assume that the specific arrhythmia diagnosis will be identified before the final decision for ablation takes place; the document does not typically specify what testing is or should be performed to obtain the diagnosis. The consensus committee has reviewed and ranked the evidence, and has made recommendations based on the standard process described in a 2014 summary document on the ACC/AHA Practice Guidelines,⁵ which are summarized here in Table 1. This statement is directed to all health care professionals who treat pediatric patients or who treat those with congenital heart disease and arrhythmias.

PREAMBLE

In 2002, an expert consensus statement was published on catheter ablation in children and adults with congenital heart disease (CHD), based on a conference held at the North American Society of Pacing and Electrophysiology in the year 2000.¹ Since publication in 2002, numerous technological improvements have been introduced, including catheter cryoablation, 3-dimensional (3D) mapping, percutaneous epicardial

Table 1 Classification of recommendations and levels of evidence⁵

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

For the purposes of this document, we have attempted to divide the recommendations into three categories: *Safety*—those related to general issues, such as substrate location, the risk of AV block, and coronary risk; *Procedure*—issues related to laboratory equipment, personnel, fluoroscopy use, ablation energy, catheter choice, sedation/anesthesia, and pre- and post-ablation procedure management; and *Arrhythmia*—issues related to the specific condition being managed. If the recommended guidelines are the same or largely overlapping for particular tachyarrhythmias, such as for atrioventricular (AV) node reentry tachycardia (AVNRT) and AV reentry tachycardia in the absence of pre-excitation (AVRT), the guidelines might be combined with specific references to the safety or procedure sections for issues involving items such as location or energy choice.

The 2002 consensus document had multiple recommendations that depended on patient age, typically expressed as “>4 years” or “<5 years”. This committee reviewed the available evidence related to patient age and weight, which is detailed below (see Section 3.2), and provided its own expert opinions. The committee concluded that, in general, age was a less important factor than patient weight in determining the risk to benefit ratio for ablation. The committee chose a cutoff of “approximately 15 kg” to be used for multiple ablation indications, recognizing that a precise cutoff for weight or age should not be specified to cover every circumstance. The neonate or very small infant is the one exception, for whom age is also taken into account in some circumstances.

The 24 members of the writing committee were selected for their expertise within their fields by PACES and/or HRS, and include 19 pediatric electrophysiologists, two adult electrophysiologists, two anesthesiologists, and an electrophysiology nurse practitioner. Because the evidence for the management of many pediatric arrhythmias is frequently limited to retrospective series or prospective adult series, the committee members were asked to consider the available evidence and their own approaches to patient management when deriving their expert opinions and suggested recommendations. Recommendations were then considered by the entire group. For the purposes of this document, “consensus” was defined as a 75% or greater agreement by the writing members, as was defined for the recently published ventricular arrhythmia consensus document.⁴

Document Review and Approval

This document was reviewed and approved by the PACES executive committee and the Scientific and Clinical Documents Committee of HRS. All the writing members approved the final version. Author and reviewer disclosures are provided in [Appendices 1](#) and [2](#), respectively.

1. Overall Results, Risks, Safety, and Complications According to Pediatric Catheter Ablation Registries and Multicenter Reports

Ablation Efficacy

Multicenter data from the Pediatric Electrophysiology Society were first reported in 1994 from the Pediatric

Electrophysiology Catheter Ablation Registry.⁶ Data from within the same registry were subsequently compared from two separate eras of radiofrequency (RF) ablation only, 1991–1995 (*Early Era*, $n = 4193$ procedures) and 1996–1999 (*Late Era*, $n = 3407$ procedures). The results from all substrates showed an improvement in the success rate from 90.4% in the Early Era to 95.2% in the Late Era.⁷

Beginning in 2000, data were prospectively collected as part of the Prospective Assessment after Pediatric Cardiac Ablation (PAPCA) study, again only for RF ablation.⁸ A total of 2761 ablation patients from 41 centers were prospectively enrolled, of whom 481 followed a standard protocol for a period of two years.⁹ The initial success rate for all the supraventricular tachycardia (SVT) substrates was 93%. Accessory pathway (AP) ablation success was 94% (left freewall 98%; right freewall 90%; left septal 88%; right septal 89%), AVNRT 99%, atrial ectopic focus 93%, junctional ectopic tachycardia (JET) 100%, and ventricular tachycardia (VT) 78%. Recurrence at 12 months was related to the substrate and was highest for right-sided APs (24.6% right septal; 15.8% right freewall) compared with left-sided pathways (9.3% left freewall; 4.8% left septal). Recurrence was lowest for AVNRT at 4.8%.¹⁰ A more recent multicenter report in 2014 from three Czech Republic centers included 708 procedures in 633 pediatric patients and showed somewhat lower acute and long-term success rates for AP ablation and AVNRT.¹¹

There are no registry or prospective trial data for the use of cryoablation in the pediatric population.

Safety and Complications

There is evidence that with increased experience and improved technology, the number of acute procedural complications can be reduced (e.g., from 4.2% in 1991–1995 to 3% in 1996–1999); however, there are no studies systematically capturing all short- and long-term risks of catheter ablation.⁷ Death and major complications are rare, but are more frequent when there is underlying heart disease, lower patient weight, greater number of RF applications, and left-sided procedures.¹²

The three most common serious complications, defined as requiring emergency or ongoing treatment, were AV block (second and/or third degree), catheter perforation or pericardial effusion, and thrombi or emboli. When analyzed for all ages from the Early to the Late Era, rates were lower in the Late Era, but no *significant* differences were found for any complication.⁷ Early Era vs Late Era: AV block, 36 of 4050 (0.89%) vs 18 of 3187 (0.56%), $P = .14$; perforation or effusion: 28 of 4050 (0.69%) vs 17 of 3187 (0.53%), $P = .40$; thrombi or emboli: 15 of 4050 (0.37%) vs 6 of 3187 (0.19%), $P = .15$. In the PAPCA registry,⁹ the complication of AV block persisted, with a rate of 1.2% compared with the 0.56% in the preceding Late Era registry.⁷ The highest rates were with AVNRT at 2.0% and septal APs at 3.0%. Although not included in any registries, the increased use of cryoablation for substrate targets in the septal areas (slow AV node pathway for AVNRT; anterior and mid-septal APs) appears to be associated with much lower AV nodal injury rates (see the cryo discussion in Section 3.3).

Three procedural deaths occurred (two APs; one atrial ectopic focus) in the Early Era compared with one procedural death (AP) in the Late Era ($P = .31$).¹² No deaths were recorded during the PAPCA registry years or from the recent Czech multicenter retrospective study.¹¹

Minor complications such as pseudoaneurysms, arteriovenous fistula, and minor bleeding are typically not reported in randomized or prospective studies; however, some reports on the pediatric population mention a 1%–3% incidence of minor vascular complications,^{11,13,14} and studies on adult patients note minor vascular complications of up to 5%.¹⁵ Although there are some data that congenital heart disease might increase the risk of a vascular complication,¹⁶ there are no controlled studies directly supporting this observation.

Thrombosis and embolism are always a concern during invasive procedures, but reports are anecdotal. Severe complications, such as acute coronary sinus thrombosis, have been reported.¹⁷ Recent findings from MRIs routinely performed on adults after atrial ablation raise concerns about the risk of microembolisms that could cause brain lesions, but there are no reports on children to date. High energy delivery and poor electrode tissue contact appear to be risk factors in adults.¹⁸

Direct injury due to ablation energy can cause damage to all adjacent structures, such as the esophagus, the coronary arteries, and the phrenic nerve. Again, there are no systematic reviews, but studies on adults and animals suggest a relationship to certain regions, higher energy levels, and multiple applications.¹⁹ Coronary artery spasm, stenosis, and occlusion have been reported in almost all AV groove locations except the anterior, where the coronary arteries are larger and further from the groove.^{20–23} Details from pre- and post-ablation coronary angiography in a pediatric SVT ablation series are reviewed in Section 3.2;²⁴ however, angiography is typically not routinely performed after ablation, thus the risk of asymptomatic stenosis might be underestimated. Reports of echocardiogram findings have not shown any obvious wall motion or significant valve abnormalities.²⁵ Cryoablation appears to have a lower risk of coronary injury²⁶ (see Section 3.3). There is general concern based on animal studies that both RF and cryoablation lesions in young children might grow after the procedure^{27,28}; however, there have been no reports of similar findings in humans. The effects of ionizing radiation are discussed in the *Fluoroscopy* section (Section 3.4).

Summary

From prior registries, recent multicenter retrospective reports, and a wide spectrum of non-registry, single-center reports, including case reports, the following points can be made regarding the pediatric population:

- Catheter ablation for pediatric patients with structurally normal hearts has an overall high acute success rate, varying from 87%–98%, depending on the mechanism and target site.
- Death is rare, and in most recent reports the rate appears to have decreased.

- Using RF energy, the risk of AV block for AVNRT ablation is approximately 1%, and for anterior and mid-septal pathways the risk is approximately 1%–3%.
- With cryoablation, permanent complete AV block has not been reported in the literature regarding children and adults (see the cryo discussion in Section 3.3).
- Serious complications continue to occur at a low rate (approximately 1%).
- There are no long-term outcome studies examining patients in adulthood after pediatric ablation; thus, concerns remain regarding scarring, late AV block development, latent coronary injury, and a small risk of malignancy secondary to radiation exposure (see Section 3.4 for a complete discussion).
- Recurrence continues to be a problem that might not have changed over time despite data showing improvements in acute procedural success rates over time.

2. Clinical Presentations

2.1 AV Node Reentry Tachycardia

AVNRT accounts for approximately a quarter of the ablation substrates in a pediatric EP laboratory.⁷ The mean clinical AVNRT age of onset has been reported to be a decade later than AVRT, and is likely due to changes in AV node physiology with age.^{29,30} As a result, the proportion of ablation procedures related to AVNRT increases with advancing pediatric age.³¹ Although AVNRT is generally considered to have a benign course, symptoms can be troublesome for children, and the family might prefer to proceed with definitive catheter ablation therapy as a curative measure.¹

AVNRT Subtypes

The issues relevant to catheter ablation are discussed here, with the general characteristics of AVNRT discussed extensively in the literature. *Typical* AVNRT (slow-fast) is by far the most common form in pediatrics, with *atypical* AVNRT (fast-slow and slow-slow) accounting for less than 10% of cases. Atypical AVNRT can be confused with ectopic atrial tachycardia (AT) or special AP-mediated tachycardias, but the summation of EP findings is usually diagnostic.³²

Dual AV Node Physiology

Dual AV node physiology is defined as the presence of both a fast and slow pathway (SP) for AV conduction, and is generally considered necessary for the development of AVNRT. The mere presence of dual AV node physiology, however, does not establish a tachycardia diagnosis because this finding is frequently present in healthy individuals.³³ The classical definition of dual AV node physiology (an atrio-His [AH] jump greater than 50 ms with a 10-ms decrement in the A1A2) is only met in approximately half of pediatric patients with AVNRT,³⁴ leading to the suggestion for using a shorter AH jump in pediatrics, and the need for clinician judgment.^{34,35} The presence of sustained SP conduction can be suggested by a PR interval greater than or equal to the RR interval during atrial decremental pacing, which is further evidence for dual AV node

physiology. Although a PR interval greater than or equal to RR was observed in only 60% of pediatric AVNRT patients, it was far more frequent in the AVNRT patients compared with the controls (13%, $P < .001$).³⁴

Diagnosis

Because dual AV nodal physiology can be a normal finding, diagnosing AVNRT for any given patient generally requires additional evidence of AVNRT. Even when there have been multiple clinical events, however, AVNRT can be difficult to induce in the EP lab in pediatric patients, who are typically under anesthesia or sedation. Consequently, the diagnosis can be made from an accumulation of findings. Inducibility or observation of SVT with the relevant characteristics in the EP lab provides the best confirmation. Other supportive data include dual AV nodal physiology, one or more echo beats consistent with AVNRT, and a history of documented SVT consistent with AVNRT. Isoproterenol testing can assist with SVT induction. In three smaller studies of pediatric patients with a history of documented SVT, but without inducible AVNRT and in some cases without dual AV nodal physiology, there was significant improvement in symptoms after empiric SP ablations, both with RF and cryoablation.^{36–38} As a result, when SVT has been documented, but is not inducible, and dual AV nodal physiology is present at electrophysiological testing, the committee considers this a Class IIa recommendation for ablation. When there are clear symptoms consistent with SVT, but the arrhythmia has not been documented and is not inducible, and dual AV nodal physiology is present at electrophysiological testing, the committee considers this a Class IIb recommendation, strongly suggesting that every effort should be made to document the clinical arrhythmia prior to a catheter procedure. Although the three quoted references for ablation without SVT induction also support ablation in patients with documented SVT who do not meet the criteria for dual AV nodal physiology, the committee did not consider this scenario to fit any specific class of recommendation. However, the language used in the indications is “evidence for dual AV nodal physiology,” allowing for the observation that not all children meet the specific adult criteria.

Ablation Technique

Once a diagnosis of AVNRT has been established, an SP modification is typically performed. Fast pathway ablation techniques are not recommended due to the high risk of inadvertent heart block (8%–23%).^{39,40} Catheter ablation techniques for SP modification vary from center to center and depend on whether RF energy or cryoenergy is used. Although RF and cryoablation share some common features, there are important differences in how these procedures are approached and in the expected responses to ablation.

Typically, a combination of anatomy and the electrogram characteristics of the SP is used to identify the optimal locations for ablation applications. With RF energy, the presence of mild to moderate junctional acceleration suggests a positive effect, but junctional acceleration does not occur with cryoablation

applications; thus, during cryoenergy delivery, EP testing is typically performed and the cryo application is terminated prematurely if SP conduction or AVNRT persists. Alternatively, multiple applications or an ablation line are placed, with testing only performed thereafter.⁴¹ Most operators using cryoenergy perform additional applications at successful locations to improve success rates and reduce recurrences,^{41–46} either in a linear pattern,⁴¹ a “freeze-thaw-freeze” cycle,^{26,42,43} or a triple freeze-thaw cycle.⁴⁵ Longer applications have also been used to improve success.⁴⁶ Data from adult and pediatric patients suggest lower recurrence rates with a 6-mm tip cryoablation catheter rather than a 4-mm tip,^{47,48} and in practice most pediatric operators use a 6-mm or larger tip.

Determining the optimal endpoint of ablation varies depending on whether RF or cryoablation is used. Various studies using RF ablation have demonstrated that single AV nodal echo beats are an appropriate end point with low recurrence rates. Because AV block has been reported in up to 2% of cases,⁹ more aggressive therapy to eliminate SP conduction should not be pursued with RF ablation. However, when the same endpoint was used in a large multicenter prospective trial comparing RF with cryoablation in adults, the recurrence rate for cryoablation was more than double that observed in the RF group (9.4% vs 4.4%, $P = .029$).⁴⁹ Some cryoablation studies show similar recurrence rates with and without complete SP ablation, and elimination of dual AV node physiology as a procedural endpoint⁵⁰; however, many clinicians use complete SP ablation as an endpoint to reduce recurrences.^{51,52} Eliminating *sustained* SP conduction when it is initially present also appears to reduce recurrences.⁵³ In summary, following RF or cryoablation, the presence of single AVNRT echo beats in the absence of sustained SP conduction is an acceptable endpoint. Some studies suggest that the absence of any SP conduction, which is safely achievable with cryoablation, also helps prevent recurrences.^{51,52}

When indicators of SP conduction are subtle at the start of the case, determining when to terminate the procedure becomes much more challenging. As an example, a single AVNRT echo beat at the end of a case is a much less tolerable endpoint when this was the only positive finding at the start of the procedure. Determining an optimal endpoint can be crucial to demonstrating a sufficient ablation effect and reducing recurrences.

Outcomes and Complications

The decision to perform an RF ablation versus a cryoablation remains somewhat controversial, and there are centers that use both approaches.⁵⁴ Determining which modality is optimal depends on two factors: successful outcomes and potential complications. Generally, the most serious potential complication in AVNRT ablation is permanent inadvertent AV block (frequently requiring a pacemaker). As reviewed above, multicenter studies of RF ablation continue to demonstrate a low risk of AV block requiring pacemaker placement.^{9,55–57} With cryoablation, which was not included in prior registries, permanent first-degree AV block has been described,⁵⁵ but AV block requiring pacemaker implantation has never been reported.

Although the lack of reports does not necessarily indicate that AV block requiring pacemaker implantation has never happened, the difference in AV block rates is important, and is the primary impetus for many centers choosing cryoablation as the primary ablation modality for AVNRT. Other centers continue to use RF energy to optimize success and recurrence rates; however, as discussed in Section 3.3, the outcome differences between RF and cryoablation are currently minimal. Most would agree that cryoablation success and recurrence rates depend on the techniques and catheters used, but the learning curve for either technique might be the most crucial element for predicting outcome.⁵⁸ Regardless of the initial choice for the catheter treatment of AVNRT, it is reasonable that cryotherapy be an available option at centers performing pediatric ablations for AVNRT (see Section 4.1 for safety recommendations in which cryotherapy availability is Class IIa).

2.2 Atrial Tachycardia in the Structurally Normal Heart

In patients without CHD, AT can be controlled with antiarrhythmic medications in the majority of patients younger than three years of age; however, AT is frequently resistant to medical therapy in older patients.^{59,60} When tachycardia onset is prior to three years of age and the arrhythmia is controlled with medication, more than 70% of patients will have spontaneous resolution of tachycardia^{59,60}; however, for patients in whom arrhythmia presents after three years of age, less than 50% will achieve sinus rhythm with medical therapy, and less than 25% will resolve spontaneously. Thus, when adequate control cannot be achieved with medical therapy at any age, or as an alternative to chronic medical therapy in patients over three years of age, ablation is a reasonable option. In addition, if patients present with a tachycardia-induced cardiomyopathy, ablation can be considered a first-line therapy for smaller patients (less than 15 kg), because cardiac function might normalize after ablation.^{59,61} When possible, rate control can be helpful for hemodynamic stability prior to the procedure; however, ablation has been successfully performed on patients with severely depressed function, even while receiving extracorporeal membrane oxygenation support (ECMO).⁶²

Traditionally, most AT in pediatric patients has been ascribed to an automatic focus, particularly when the tachycardia is persistent or intermittently incessant; however, adenosine-sensitive AT, thought to be due to microentry or triggered automaticity, has been reported and should be considered for the patient who presents with paroxysmal tachycardia that responds to adenosine that is not consistent with AVRT or AVNRT.⁶³ These later tachycardias are often labeled “non-automatic focal atrial tachycardia” or NAFAT. Most of these focal tachycardias can be ablated successfully.⁶³ Though commonly originating from the area of the pulmonary veins or the crista terminalis, recent reports have also demonstrated less common AT foci originating from the noncoronary cusp, the left coronary cusp, the mitral annulus-aorta junction, the coronary sinus ostium, and the AV valve annuli.⁶⁴

In the 1994 report from the Pediatric Ablation Registry,^{6,65} ablation of ectopic AT had an 86.7% acute success rate, slightly higher for left atrial foci (89.9%) than right atrial foci (84.2%). The success rate was unchanged when it was evaluated again in 2002⁷; however, the emergence of 3D electroanatomic mapping (EAM) can improve the success rate.⁶¹ Failure to ablate ATs has been attributed partially to epicardial foci and foci deep in the atrial appendages. Awareness of these issues and others should be considered when discussing ablation as a treatment option. Interestingly, although the risk of pulmonary vein stenosis has been highlighted in the AF literature when ablating within a pulmonary vein or its orifice, there are no reports in the pediatric literature of pulmonary vein stenosis during ablation of atrial ectopic tachycardia within a pulmonary vein.^{6,66,67} There are multiple possible explanations, including a lower likelihood when ablating a single focus due to focal ablation, ablation locations near the orifice where the pulmonary veins are larger, and a lack of recognition in the absence of a post-ablation angiogram. However, most pediatric patients undergo post-procedural echocardiograms within the first year after ablation, and there have been no reports of pulmonary vein stenosis to date. It seems likely that stenosis would occur occasionally, but due to the focal nature of the arrhythmia in pediatrics, ablation rarely if ever causes clinically important stenosis. Awareness of the potential for such a complication based on the literature from adults with AF might lead to the consideration for cryo-based therapy for ablation within a smaller pulmonary vein.

Atrial flutter, or macroreentrant AT in the absence of CHD is uncommon in children and most commonly occurs in neonates. In the absence of other coexisting arrhythmias, atrial flutter typically does not recur and would not require ablation in the neonatal population.⁶⁸ Atrial flutter as a lone finding can also occur uncommonly in children or adolescents,^{6,69} but is often initiated by either AVRT or AVNRT. Thus, with lone atrial flutter outside the neonatal period, a complete diagnostic electrophysiological evaluation is warranted, and management can occur as with other ATs.

Inappropriate sinus tachycardia (IST) is a rare form of AT at any age, and even more so in adolescents.⁷⁰ Although ablation to modify the sinus node is occasionally an effective therapy for IST, it can also lead to multiple complications and should be only rarely used with adolescents. Because a 2015 consensus statement including IST made “routine” use of ablation Class III, we have included that language in these recommendations as well, but we have also allowed rare Class IIb use as reflected in the text of the above referenced document.⁷¹ Ablation is not recommended for sinus tachycardia related to postural orthostatic tachycardia syndrome or other autonomic conditions.⁷¹

The notes below apply to all the recommendations in the document.

**Smaller patients* in general weigh less than approximately 15 kg, and *larger patients* weigh more than approximately 15 kg.

⁺The precise definition of “*medical therapy that is either not effective or associated with intolerable adverse effects*” is left up to the practitioner and family to decide. In general, however, the threshold for ineffectiveness and intolerability should be higher in smaller patients. For example, failure or

intolerability of a beta-blocker alone might be adequate in larger patients, but not in smaller patients, unless there are additional circumstances. In the smallest patients, failure or intolerability of drug combinations from multiple classes, including membrane stabilizing agents with adequate loading time, would be required prior to the decision for ablation.

[#]*Recurrent* is defined as arrhythmias that have more than one symptomatic or significant paroxysm of tachycardia.

[^]*Persistent* is defined as arrhythmias that are either present continuously (incessant), incessantly intermittent, or incessantly non-sustained.

[&]*Usual complex* refers to a QRS in tachycardia identical to that in sinus rhythm, regardless of QRS duration or morphology.

LOE— Level of Evidence

Indications for ablation procedures in pediatric patients with narrow (*usual*[&]) complex SVT (AVNRT/AVRT/AET/NAFAT/Atrial Flutter) without CHD

Class I

1. Ablation is recommended for documented SVT, recurrent[#] or persistent[^], associated with ventricular dysfunction in larger^{*} patients (LOE: C).
2. Ablation is recommended for documented SVT, recurrent[#] or persistent[^], when medical therapy is either not effective or is associated with intolerable adverse effects (the medical therapy used prior to ablation depends on patient weight)⁺ (LOE: C).
3. Ablation is recommended for documented SVT, recurrent[#] or persistent[^], when the family wishes to avoid chronic antiarrhythmic medications in larger^{*} patients (LOE: C).
4. Ablation is recommended for recurrent acute hemodynamic compromise (hypotension or syncope) from SVT in larger^{*} patients (LOE: E).
5. Ablation is effective for recurrent[#] SVT requiring emergency medical care or electrical cardioversion for termination in larger^{*} patients (LOE: C).

Class IIa Ablation can be useful in the following cases:

1. Recurrent[#] clinical symptoms clearly consistent with paroxysmal SVT in larger^{*} patients, and one of the following: evidence of AV accessory pathway involvement; inducible SVT (LOE: C).
2. Slow pathway modification in larger^{*} patients with a history of documented SVT, when SVT is not inducible at electrophysiological testing, but evidence for dual AV nodal physiology with or without single AV nodal echoes is demonstrated (LOE: C). Cryotherapy should be considered for slow pathway modification (LOE: B).

Class IIb Ablation can be reasonable for the following cases:

1. Slow pathway modification in larger^{*} patients with clinical symptoms clearly consistent with paroxysmal SVT but not documented, when SVT is not inducible at

electrophysiological testing but evidence for dual AV nodal physiology with or without single AV nodal echoes is demonstrated. (LOE: C). Cryotherapy should be considered for slow pathway modification (LOE: B).

2. Recurrent[#] clinical symptoms clearly consistent with paroxysmal SVT in smaller^{*} patients, and one of the following at the electrophysiology study: evidence of AV accessory pathway involvement; inducible SVT (LOE: C). Medical therapy should be considered prior to ablation (LOE: C). Cryotherapy should be considered for slow pathway modification (LOE: B).
3. Recurrent acute hemodynamic compromise (hypotension or syncope) from SVT in smaller^{*} patients (LOE: C).
4. Intermittent symptomatic SVT which is nonsustained (less than 30 seconds) in larger^{*} patients (LOE: C).

Class III Ablation is not recommended for the following cases:

1. SVT controlled with medical therapy in the absence of intolerable adverse effects in smaller^{*} patients (LOE: E).
2. Clinical symptoms consistent with SVT, but no inducible SVT, and no evidence for dual AV nodal physiology demonstrated during EP testing (LOE: C).
3. Slow pathway modification when dual AV node physiology is demonstrated after ablation of a different arrhythmia substrate (such as an accessory pathway) when there is no inducible AVNRT (LOE: C).

Indications for ablation procedures in pediatric patients with inappropriate sinus tachycardia

Class IIb Rarely, ablation can be reasonable for the following:

1. Inappropriate sinus tachycardia when autonomic causes such as postural orthostatic tachycardia syndrome have been ruled out, and medical therapy is either not effective or is associated with intolerable adverse effects (LOE: C).

Class III Ablation is not recommended for the following:

1. Sinus tachycardia due to postural orthostatic tachycardia syndrome, or as part of routine care for inappropriate sinus tachycardia (LOE: C).⁷¹

2.3 Wolff-Parkinson-White Pattern and Accessory Pathway Mediated Tachycardias

The most common cause of SVT in the younger pediatric population is AVRT using an AP.⁶⁹ In the landmark description published in 1930,⁷² WPW syndrome was used to describe a “bundle-branch pattern” with a short PR interval in healthy young people with paroxysmal tachycardia. As noted in the introduction, the distinct ECG pattern of an antegradely conducting AP will be referred to in this document as *WPW pattern*, synonymous with the term *pre-excitation*, regardless of whether SVT is present or inducible. This section addresses catheter ablation in the management of APs that conduct in the antegrade

direction only, the retrograde direction only (*concealed*), and both the ante- and retrograde directions. The acute and chronic management of usual AVRT due to a typical *concealed* or retrograde-only AP is similar to AVNRT, and is addressed as such in these guidelines (see SVT recommendations above).

The incidence of WPW pattern is 0.1% to 0.5% in the general population.⁷³⁻⁷⁵ A manifest AP can symptomatically present clinically in one of four ways: AVRT, AF/flutter, ventricular dysfunction due to asynchronous contraction through the AP, or sudden death. WPW pattern can also be incidentally discovered on an ECG. Because a significant number of incidentally discovered pediatric patients with WPW pattern will develop a symptomatic arrhythmia, the management of asymptomatic pediatric patients with WPW pattern was recently addressed in a consensus statement published in 2012.² The statement includes an in-depth discussion of the relevant risk factors for sudden cardiac death.

The most common time for patients with WPW to present with AVRT is in the neonatal period. After such a presentation, episodes of SVT decrease in frequency over the first year of life in over 90% of patients, with SVT recurring in approximately 30% of patients at an average age of seven to eight years.⁷⁶ Furthermore, there is evidence that in the first year of life, the AP loses antegrade conduction in as many as 40% of patients, with SVT becoming noninducible in a similar percentage, suggesting loss of retrograde conduction as well.⁷⁷ If pre-excitation persists, however, there is as high as a 29-fold increase in the risk of an SVT recurrence.⁷⁸ Due to the decrease in the recurrence of SVT after presentation in infancy, possible spontaneous resolution and a higher risk of ablation in smaller patients (see Section 3.2), ablation therapy in infancy should be reserved for those who fail medical therapy. Patients who present with WPW pattern and SVT after the age of one year have a greater than 90% risk of SVT recurrence⁷⁹; therefore, in larger patients who have a lower procedural risk, ablation may be considered a first line therapy.

Ventricular Dysfunction Secondary to Preexcitation. Patients with a WPW pattern on the ECG occasionally present with ventricular dilation and dysfunction. Although there are some causes of cardiomyopathy associated with WPW, primarily hypertrophic, most of these cases appear to be due to ventricular dyssynchrony from eccentric activation of the ventricle, observed most commonly in patients with more prominent preexcitation on the ECG and right anterior septal pathways,^{80,81} but also associated with pathways in other locations.⁸² It is important to note that the dysfunction resolves in most cases after AP ablation, sometimes immediately, suggesting it is a direct effect of the eccentric activation and might or might not represent dyssynchrony-induced cardiomyopathy.⁸¹⁻⁸⁵ A number of these reports demonstrating improvement after ablation have been published since the 2012 guidelines for asymptomatic WPW, leading to a change in the guidelines' recommendation for ablation in patients with dyssynchrony presumed due to ventricular preexcitation from Class IIb to Class IIa.

Atriofascicular fibers (some experts continue to call them "Mahaim fibers") are specialized APs that typically conduct only in the antegrade direction. These fibers connect the atrium

to some part of the ventricle or conduction system via a slowly conducting fiber, and its atrial end has AV node-like properties. Although atriofascicular fibers can have spontaneous automaticity and can participate in antidromic tachycardia, their antegrade conduction is decremental, and the risk of sudden death is thought to be lower than that for a non-decremental antegrade AP. Although more difficult to ablate, and with a higher recurrence rate than a typical AP, most of these pathways can be ablated successfully, and the procedure indications are the same as for other antegrade-conducting APs.⁸⁶

Fasciculoventricular pathways connect the His bundle or a bundle branch to the ventricular muscle, and can have both surface ECG and some electrophysiological features similar to antegrade-conducting accessory AV pathways; however, they have distinct electrophysiological characteristics, do not participate in tachycardia circuits, and are typically benign.⁸⁷ Attempted ablation of a fasciculoventricular pathway also carries a risk of AV block; thus, these fibers should not be targeted for ablation unless involvement in tachyarrhythmias can be definitively demonstrated.⁸⁷

Permanent junctional reciprocating tachycardia (PJRT) arises from a specialized form of retrograde-only AP, which is due to a unique fiber that has very slow and decremental conduction, resulting in a long RP tachycardia. PJRT occurs at less than one year of age approximately 60% of the time and is incessant in about half of patients; however, because of its relatively slow rate (140 to 200 bpm), it can be difficult to appreciate during routine examinations, particularly in newborns and infants, and can lead to a tachycardia-induced cardiomyopathy if untreated.¹⁰

The success, recurrence, and complication rates for AP ablation with any conduction characteristics in any location have been addressed above in Section 1. The risk of AP ablation clearly varies based on the position in the heart and should be considered in the decision to perform an ablation in a pediatric patient, also depending on the technology used. Although there have been no recent large series, the current complication rate is likely lower than in the earlier registries because the introduction of cryoablation technology has decreased the incidence of AV block as a complication. The risks and benefits of ablating a para-Hisian or mid-septal pathway, however, should be weighed prior to proceeding with an ablation. For APs located in the coronary sinus or aortic cusp, care should be taken to carefully delineate proximity to the coronary arteries to avoid inadvertent coronary damage.

Indications for ablation procedures in pediatric patients with WPW Pattern (see SVT indications for patients with WPW pattern and AVRT; see CHD indications for patients with WPW and CHD)

§Predictors of high risk for cardiac arrest in WPW pattern include the following:

- The shortest preexcited RR interval during atrial fibrillation, or during incremental atrial pacing ≤ 250 ms
- Multiple accessory pathways

Class I Ablation is recommended for the following:

1. WPW pattern following resuscitated cardiac arrest (LOE: B).
2. WPW pattern with syncope when there are predictors of high risk for cardiac arrest[§] (LOE: B).

Class IIa Ablation can be useful for the following:

1. WPW pattern with ventricular dysfunction presumed due to dyssynchrony in larger* patients, or when medical therapy is either not effective or associated with intolerable adverse effects⁺ in smaller* patients (LOE: B). (Note: This indication was Class IIb in the prior guidelines for asymptomatic WPW).²
2. WPW pattern without symptoms, in which there are predictors of high risk for cardiac arrest[§] in larger patients* (LOE: C).
3. WPW pattern with syncope, without predictors* of high risk for cardiac arrest in larger* patients (LOE: C).
4. WPW pattern without symptoms in larger* patients when the absence of WPW pattern is a prerequisite for participation in personal or professional activities (LOE: E).

Class IIb Ablation can be reasonable for the following:

1. WPW pattern without symptoms in larger* patients with predictors of low risk for cardiac arrest[§], as a patient or family choice (LOE: E).

Class III Ablation is not recommended for the following:

1. WPW pattern caused by a fasciculoventricular accessory pathway (LOE: C).
2. WPW pattern without symptoms in smaller* patients (LOE: C).

2.4 Atrial Fibrillation in the Adolescent

2.4.1 Structurally Normal Heart

AF is the most common sustained cardiac arrhythmia in adults and is associated with major complications, including ischemic cerebrovascular emboli, heart failure, and recurrent hospitalizations. AF in the pediatric population is rare, and like in adults, is typically associated with structural heart disease, myocardial dysfunction, and repaired congenital defects.⁸⁸ In addition, the association of WPW syndrome with preexcited AF has been well documented. What is known as *lone* AF (not associated with risk factors) in adolescents is rare and has been associated with a family history of AF, underlying SVT substrates, high vagal tone, and occasionally medication, alcohol intoxication, or illicit drug use.⁸⁹⁻⁹¹

A small number of retrospective series have documented the role of SVT in triggering AF.^{92,93} Successful catheter ablation of the SVT substrates has prevented AF recurrences in more than 15% of adolescent patients with AF.^{92,93} Also, in one series of nine adolescent patients with lone AF, pulmonary vein isolation or ablation of irregular, rapidly

firing foci in the left atrium or crista terminalis prevented AF recurrence in seven patients.⁹⁴ In one other series of pediatric AF, for those not undergoing catheter ablation, the AF recurrence rate was 49% at 12 months.⁹² Although there is scarce literature for the use of wide-area pulmonary vein isolation in pediatric patients,⁹² a number of centers have performed these procedures with good success in adolescent patients with recurrent AF who required repeated cardioversion and who were unresponsive to medical therapy. The lack of data and attendant procedure risks suggest the procedure should be performed only in exceptional cases.

2.4.2 Congenital Heart Disease and AF

Recent data suggest the onset of AF in patients with CHD occurs at a much younger age than in the general population.⁹⁵ Despite this earlier onset, AF remains rare in patients under the age of 18. In general, evaluation and management of AF in adolescents with CHD requires a highly individualized approach because the current understanding of the specific mechanisms of AF in CHD is insufficient to support such empiric strategies as those employed in the adult patient. Evaluation should include exclusion of specific hemodynamic lesions, assessment for the presence of macroreentrant circuits or other SVT substrates as initiators, and the potential for ablation of the initiating substrate to reduce the AF burden. If surgery is performed for hemodynamic lesions, the concomitant use of atrial arrhythmia surgery might be appropriate based on the specifics of the case.³

2.4.3 Recommendations for Patients With and Without CHD

The prior guidelines for the management of arrhythmias in adults with CHD classified pulmonary vein isolation as a Class IIa recommendation for patients with symptomatic drug-refractory AF and AV node ablation as a Class IIb recommendation when both medical therapy and ablation have failed.³ Due to the risks of pulmonary vein isolation, this committee determined that for patients 18 years and younger with or without CHD, pulmonary vein isolation can rarely be considered (Class IIb), differentiating these younger patients from adults. The procedure might be performed in selected cases where simple ablation measures and medical therapy have failed; however, it should be performed by or with an electrophysiologist experienced in AF ablation, and when relevant, familiar with the complexities of CHD. The committee determined that there was no significant role for AV node ablation for patients 18 years and younger, but that it did not meet the requirements for a Class III recommendation.

Indications for ablation procedures in pediatric patients with AF, with or without CHD

Class I

None

Class IIa Ablation can be useful for the following:

1. Any SVT substrate identified as the initiating event for AF, using either ambulatory ECG monitoring or invasive EP evaluation. Provocative SVT substrates include forms

of paroxysmal SVT, a single ectopic atrial focus unrelated to or within a larger pulmonary vein, or ectopic focus/foci initiating AF from a single pulmonary vein orifice, requiring isolation of the single vein (LOE: C).

Class IIb Ablation can rarely be considered for the following:

1. Empiric wide-area circumferential isolation of all pulmonary veins in the absence of a documented triggering focus or arrhythmia, as is performed in adults with paroxysmal AF, in adolescent pediatric patients when AF is recurrent, requires repeated DC cardioversion, and medical therapy is either not effective or is associated with intolerable adverse effects⁺. The procedure should be undertaken by or with the direct assistance of a clinician experienced in the technique (LOE: C).

Class III Ablation is not recommended for the following:

1. Empiric wide-area pulmonary vein isolation for rare episodes of paroxysmal AF not requiring cardioversion, or when well controlled on medical therapy.
2. Incidental induction of AF during an electrophysiology study.

2.5 Junctional Ectopic Tachycardia

JET is a relatively uncommon tachyarrhythmia caused by pathologically increased automaticity or triggered activity within the AV node or bundle of His. JET has been classified into postoperative, congenital, and idiopathic.^{96,97}

Postoperative JET, the most frequent subtype, is relatively common after cardiac surgery. Risk factors for development include a longer bypass time, manipulation and/or surgery around the AV node or His, greater inotropic support, electrolyte abnormalities (hypomagnesemia in particular), and genetic influences.^{98–100} Catheter ablation is typically not recommended for postoperative JET because it can generally be managed medically until spontaneous resolution is achieved.

JET unrelated to the postoperative period is uncommon. The congenital form of JET, occurring in the first six months of life, is frequently refractory to antiarrhythmic therapy and historically has relatively high morbidity and mortality.¹⁰¹ JET has been associated with congenital cardiac tumors,¹⁰² myocarditis,¹⁰³ and congenital heart block.¹⁰⁴ In rare cases, infants have been diagnosed with a familial form of JET, and although some family ancestries suggest an autosomal-dominant inheritance, the genetic basis for this condition is currently unknown.¹⁰⁵ The poor response to medical therapy and the potential for the development of tachycardia-associated cardiomyopathy have prompted catheter ablation attempts in selected high-risk patients with congenital JET. Catheter ablation was attempted in almost 50% of the patients in the largest published series.^{54,106–109}

Idiopathic JET (defined as occurring after the first six months of life and not associated with the postoperative period) has a more variable presentation, although it might also be difficult to

control with antiarrhythmic drug therapy. Transcatheter ablation for treatment of idiopathic JET has been reported to be effective, although it presents a risk for inadvertent AV block. Prior reports indicate this risk can be as high as 10% to 20% using RF energy; however, this risk is substantially decreased using cryotherapy.¹⁰⁸ After ablation, a slow junctional rhythm can be present, but this findings might not be an indication of a higher recurrence risk.¹¹⁰

Indications for ablation procedures in pediatric patients with JET

Class I

1. Ablation is recommended for persistent[^] or recurrent[#] idiopathic JET, or congenital JET associated with ventricular dysfunction, when medical therapy is either not effective or associated with intolerable adverse effects⁺ (LOE: C).
2. When ablation of JET is being performed, cryotherapy is the preferred first choice due to the high risk of AV block. RF energy should be used with extreme caution, after a detailed discussion with the family and patient concerning the high risk of AV block and the potential need for permanent pacing (LOE: C).

Class IIa Ablation can be useful for the following:

1. Persistent[^] or symptomatic recurrent[#] idiopathic JET, or congenital JET in larger^{*} patients, when medical therapy is either not effective or is associated with intolerable adverse effects⁺ (LOE: C).

Class IIb Ablation might be reasonable for the following:

1. Persistent[^] or symptomatic recurrent[#] idiopathic JET, or congenital JET in larger^{*} patients, as an alternative to chronic antiarrhythmic therapy that has been effective in controlling the arrhythmia (LOE: C).
2. Persistent[^] or symptomatic recurrent[#] idiopathic JET, or congenital JET in smaller^{*} patients, when medical therapy is either not effective or is associated with intolerable adverse effects⁺ (LOE: C).

Class III Ablation is not recommended for the following:

1. Postoperative JET, due to its potential for spontaneous resolution (LOE: B).
2. Idiopathic or congenital JET adequately controlled with antiarrhythmic medications, in smaller patients^{*} (LOE: C).

2.6 Ventricular Tachycardia with a Structurally Normal Heart

The prior guidelines document on the management of ventricular arrhythmias in pediatric patients with structurally normal hearts extensively reviews the relevant mechanisms of ventricular arrhythmia.⁴ Consequently, this document will reference the

prior document for much of the background and will focus primarily on technical issues and any differences in the recommendations related to the role of catheter ablation.

Ventricular arrhythmias are uncommon in patients who do not have structural heart disease, are often well tolerated, and are not usually associated with a risk of sudden cardiac death.^{4,111,112} Children considered for ablation are typically symptomatic and/or have reduced ventricular function—in both cases determined to be correlated with the arrhythmia. They also have frequently failed drug therapy, and have sustained or frequent repetitive nonsustained VT or ventricular ectopy.^{111,113–115} Small case series have demonstrated success in the majority of these selected cases in which ablation has been performed.^{113,116} Ablation for incessant ventricular arrhythmia that is producing hemodynamic deterioration can be life-saving and has been used even in young children and infants requiring extracorporeal support.^{117,118} In general, however, ablation in infants and small children should be deferred until the child weighs more than 15 kg. The site of arrhythmia origin is an important determinant of the risks, the likelihood of success, and the approach to ablation. There are no large series documenting outcomes and complications in children.

Outflow Tract, Periannular, and Papillary Muscle Focal Origin Arrhythmias

Idiopathic focal arrhythmias can originate from almost any site in the left or right ventricle (LV, RV), but most commonly originate from the RV outflow tract (RVOT), followed by the LV outflow tract (LVOT)/aortic sinuses of Valsalva, and then the mitral and tricuspid annuli.^{115,119,120} The prior guidelines presented an in-depth discussion of the natural history and general management of these arrhythmias, and from the standpoint of catheter ablation, the guidelines considered conditions that led to hemodynamic compromise as a Class I indication and other symptomatic conditions as Class IIa—both as an alternative to medical therapy.⁴ Differences in classification by this committee are discussed below.

Intrafascicular Verapamil-Sensitive VT (Belhassen's Tachycardia)

This sustained monomorphic VT, with its characteristic ECG findings, is thought to be due to reentry in or near the fascicles of the left bundle branch, typically the posterior.¹²¹ Its presentation and natural history were reviewed in the prior guidelines,⁴ and the condition accounts for 85% of the LV-sustained VTs encountered in children who are considered for catheter ablation.¹²⁰ The targets for ablation are either presystolic Purkinje potentials along the posterior or anterior fascicle during tachycardia, or diastolic potentials that might be markers for the retrograde pathway of the circuit. Ablation is effective in over 80% of patients.¹²² Tachycardia is susceptible to mechanical “bump” termination that might render it no longer inducible. An empiric line of applications perpendicular to the long axis of the LV and from the mid-inferior septum to the inferior wall might then be effective.¹²³ Ablation through the

region of the posterior fascicle (or anterior fascicle, depending on the axis of the VT targeted) producing a QRS axis change as evidence of interruption of a portion of the fascicle was effective in one small series of six patients ages 3.5 to 17.5 years.¹²⁴ The results of a multicenter survey found ablation was attempted in 129 of 152 patients.¹²⁰ The procedure was successful in 71% of patients. Lack of inducibility was the most common cause of failure. Potential complications in addition to those related to arterial catheterization and LV access include left bundle branch block (less than 1%) and rare AV block.^{120,124,125} Ablation might be more difficult for the less common nonposterior fascicle variants. From a series of 16 patients with a median age of 9.5 years, 10 had typical posterior fascicular QRS morphology, and ablation was successful in all; however, two required two procedures.¹²⁵ The anterior fascicular variant or multiple VTs were present in six patients, and the initial procedure was successful in only half of the patients. Two patients suffered conduction system injury with left bundle branch block, and AV block in one each, respectively. The prior guidelines committee considered ablation for this condition as a Class I indication as an alternative to medical therapy in patients over 1 year of age who fail calcium channel blockade, or in older patients, and this committee is in agreement with using the “larger” patient definition.

Ventricular Tachycardia Associated with Cardiomyopathies

Dilated, hypertrophic, restrictive, or LV non-compaction cardiomyopathies can cause polymorphic or monomorphic VT. Most sustained monomorphic VTs are due to scar-related reentry; however, bundle branch reentry and focal VTs also occur.^{112,126} Depending on the location of the scar, epicardial ablation is frequently required. Experience in children is limited.

Arrhythmogenic RV cardiomyopathy can cause monomorphic VT, polymorphic VT, and nonsustained arrhythmias that are frequently exercise induced. Sustained monomorphic VT is typically due to scar-related reentry, but focal origin VTs also occur.¹²⁷ Scarring is more extensive in the epicardium than the endocardium, and ablation from the epicardium is frequently required.^{127,128} Experience in children is limited.^{112,127} In a series of 17 children ages nine to 18 years, ablation abolished inducible VTs in 16 patients, with no recurrences during follow-ups of six to 42 months.¹²⁷ Four patients had pericardial bleeding.

Ventricular Tachycardia in Infancy

Hemodynamically significant, rapid VT in this age group is typically secondary to congenital diverticulum or aneurysm, cardiac tumor, congenital defects of fatty acid metabolism, acute metabolic disturbance, intoxication, channelopathy, or infection. There are a few reports of successful catheter ablation of VT sources from congenital cardiac tumors.^{129,130} Idiopathic VT and accelerated idioventricular rhythm of infancy (AIVR) in this age group are slower, are clinically well-tolerated, and are reported to be associated with spontaneous resolution in 89% of patients at medium-term follow-up.¹³¹ Catheter ablation plays a minimal role in this patient group.

Frequent Ventricular Ectopy without Tachycardia

In the past, even when frequent, premature ventricular contractions (PVCs) had been considered completely benign in children and deserving of treatment only when associated with bothersome symptoms that are not suppressed by simple recommendations, such as elimination of caffeine. However, uniform PVC-induced cardiomyopathy is now a well-known form of arrhythmia-induced cardiomyopathy in adults, most often when the PVC burden exceeds 20,000 in 24 hours.¹³² The anatomic distribution of foci is similar to that of VT in the normal heart, with predilection to the outflow tracts.¹³³ Because successful ablation of the focus can result in normalization of systolic function in many cases,¹³² the committee was in agreement with the prior guidelines that frequent ectopy from predominantly a single focus causing ventricular dysfunction should be a Class I recommendation, and symptomatic ventricular ectopy without dysfunction should be Class IIa. Mapping techniques are as described above and in the prior guidelines⁴ for normal heart VT. Although there are no published series on catheter ablation for this entity dedicated to children, large series have included children as young as eight years of age.¹³⁴

Percutaneous Epicardial Mapping and Ablation

VT associated with arrhythmogenic RV cardiomyopathy and other idiopathic and familial cardiomyopathies frequently requires ablation from the epicardium, which is achieved by percutaneous pericardial access.^{118,127,135} Attempted epicardial access is associated with a risk of pericardial bleeding, abdominal bleeding, and post-procedure pericarditis. Epicardial ablation has a risk of injury to adjacent coronary arteries, the phrenic nerve, and the lung.¹³⁶ Experience in children is limited.^{118,127,137,138} There are anecdotal reports of surgical and percutaneous pericardial access in infants and small children as last-resort therapies.^{118,138}

Risks of Ablation

Coronary artery injury can occur with ablation along the AV annuli, in the sinuses of Valsalva, in the conal septum, and in the epicardium.^{24,139–141} Coronary injury can present as immediate coronary occlusion, or as coronary stenosis or occlusion late after the procedure.¹⁴² Cryoablation might have lower risk of coronary injury, but there is limited experience with its use for ventricular arrhythmias, and recurrences can be greater than with RF ablation.^{118,143–146}

In growing myocardium, a ventricular ablation lesion can increase in size with time.^{27,28} In animal studies, ventricular lesions in infant animals doubled in volume by one year. Thus, minimizing ablation in normal contracting myocardium is an important consideration, particularly with small children and infants.

Guidelines—Differences from the Prior Document

After a review of the literature and multiple consensus surveys of the members, this committee determined that there were no clear reasons why outflow tract tachycardias and fascicular tachycardias would be treated differently with regard to catheter

ablation. Specifically, symptomatic VT from either cause should be treated as a Class I indication as an alternative to medical therapy in larger patients. A note is added, however, concerning the potentially higher risk of ablation in the aortic sinuses, recommending additional consideration for tachycardia origins in proximity to a coronary artery.

The prior committee included accelerated idioventricular rhythm with correlated symptoms as a Class IIa indication for ablation in the prior guidelines.⁴ However, after multiple considerations, this committee could only reach consensus as a Class IIb recommendation due to the low likelihood of this condition being clearly symptomatic.

Indications for ablation procedures in pediatric patients with ventricular arrhythmias without congenital heart disease

Class I

1. Ablation is recommended for frequent ventricular ectopy or tachycardia, predominantly from a single focus thought to be causing ventricular dysfunction, when medical therapy is either not effective or is associated with intolerable adverse effects (the medical therapy used depends on patient weight⁺), or as an alternative to medical therapy in larger* patients (LOE: C).
2. Ablation is effective for recurrent[#] or persistent[^] symptomatic intrafascicular verapamil-sensitive reentrant VT, idiopathic outflow tract VT, or VT with hemodynamic compromise, when medical therapy is either not effective or is associated with intolerable adverse effects (the medical therapy used depends on patient weight⁺), or as an alternative to medical therapy in larger* patients (LOE: C). (Notes: Additional consideration should be used for tachycardia origins in proximity to a coronary artery; e.g., aortic sinus or outflow tract VT was a Class IIa indication in the prior pediatric guidelines for ventricular arrhythmias).

Class IIa

Ablation can be useful for the following:

1. Frequent ventricular ectopy with correlated symptoms in larger* patients (LOE: C).

Class IIb

Ablation can be reasonable for the following:

1. Accelerated idioventricular rhythm with correlated symptoms in larger* patients (LOE: C). (Note: This indication was Class IIa in the prior pediatric guidelines for ventricular arrhythmias⁴).
2. Recurrent/frequent polymorphic ventricular arrhythmia when there is a suspected triggering focus, arrhythmia, or substrate that can be targeted (LOE: C).

Class III

Ablation is not recommended for the following:

1. VT in smaller* patients that either is controlled medically, or is hemodynamically well tolerated without ventricular dysfunction (LOE: C).

2. Accelerated idioventricular rhythm in smaller* patients (LOE: C).
3. Asymptomatic ventricular ectopy, VT, or accelerated idioventricular rhythm that is not suspected of causing or leading to ventricular dysfunction (LOE: C).
4. Ventricular arrhythmias due to transient reversible causes, such as acute myocarditis or drug toxicity (LOE: B).

3. Special Issues

3.1 Congenital Heart Disease—General Comments

Guidelines for the overall management of arrhythmias in adults with CHD (ACHD), including the use of antiarrhythmic drugs, surgery, and/or catheter ablation, were published in 2014.³ This section focuses on the role of catheter ablation in any age patient with CHD, including adults.

Symptomatic tachyarrhythmias occur in a substantial proportion of patients with surgically treated congenital heart defects. As a rule, tachyarrhythmias are both more prevalent and have greater clinical impact in relation to the complexity of the CHD, older age of the patient, and the degree of cardiac dysfunction. In addition, a variety of forms of CHD are associated with tachyarrhythmias prior to any surgical intervention. For example, Ebstein's anomaly of the tricuspid valve is associated with AV and atriofascicular accessory connections in approximately 20% of patients; congenitally corrected transposition of the great arteries (TGA) is associated with AV accessory connections in 2% to 5% of patients; and some forms of heterotaxy (especially in the presence of ventricular l-looping and AV septal defect) are associated with twin AV nodes.^{147,148} Acquired postoperative arrhythmia substrates can develop secondary to chronic pressure and volume overload, fibrosis, chronic hypoxemia, and surgically created and natural conduction barriers.

Multiple classification systems have been devised to reduce the myriad of CHD abnormalities and combinations to an ordinal scheme of severity level, and each system was developed for different clinical contexts, primarily surgical. The ACC/AHA task force on practice guidelines for adults with CHD proposed a classification system in 2008,¹⁴⁹ which was used in the 2014 PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease.³ Although the current guidelines have been developed for children and all patients with CHD, we included this classification system as [Table 2](#), with caveats included for certain lesions when they are clinically relevant in children. Some of the current guidelines are derived from that document and specifically refer to that severity schema. The classification system is necessarily arbitrary in that it does not take into account either anatomic variability or the spectrum of severity within some defects, such as Ebstein's anomaly of the tricuspid valve.

The majority of clinically important tachyarrhythmias in CHD are due to macroreentry, whereas a minority are due to other mechanisms. Natural and surgical barriers to conduction, such as valve annuli, venous orifices, patches, conduits, and surgical scars, play important roles. Consequently, a

priori knowledge of the specific congenital anatomy, current hemodynamic issues, and all available surgical and prior catheter-based procedures, through pre-procedural imaging and careful record review, regardless of the institution where they were performed, is a requisite component of procedural planning. For example, after a Mustard operation for D-TGA, the location of the intra-atrial baffle with respect to the coronary sinus can have important electrophysiological implications.¹⁵⁰ Preprocedural imaging using a variety of methodologies can provide an assessment of venous access to the heart, details of native anatomy and surgical reconstruction, analysis of ventricular function, and when feasible, provision of a detailed anatomic shell for image-merge with the intraprocedural electroanatomic construction. Review of all pathological tachycardias from prior ECGs and rhythm strips is critical to procedural planning. Finally, if there are concomitant hemodynamic issues that might be contributing to or exacerbating the arrhythmia, consideration should be given to how and when they will be addressed when planning an ablation procedure.

Procedural Considerations

Planning for catheter access to the chambers of interest is important. When inferior vena cava access is not available, transhepatic access can be an alternative approach.^{151–153} In patients who have undergone an atrial switch procedure for D-TGA or a Fontan operation with an intra-atrial baffle or extracardiac conduit, the tachyarrhythmia substrate can be located partially or completely in the pulmonary venous portion of the atria. If a fenestration is not present, access to that chamber can require needle perforation of pericardial material, fabric, or thick muscle,¹⁵⁴ which can be assisted by an RF puncture technique.¹⁵⁵ Although very expensive and not widely available, magnetic driven navigation systems (Stereotaxis; St. Louis, MO) enable catheter negotiation of the most complex anatomies, including retrograde approaches through the aortic valve to both the ventricle and atrium,^{156,157} with reduced fluoroscopy times and relatively high acute and chronic ablation success rates.¹⁵⁸

The use of EAM systems should be considered essential for tachyarrhythmia substrate localization in most patients with CHD.³ These technologies enable 3D spatial displays of unipolar or bipolar electrogram activation times and voltages, and identification of areas of putative scar. Other areas of interest, including valvular and venous structures, surgical patches, double potentials, His bundle electrograms, and ablation application sites can be outlined or otherwise tagged. Previously obtained cardiac renderings from cardiac MRI or CT scans can also be uploaded into the real-time geometry created by EAM to enhance anatomic accuracy^{159,160}; however, alignment of the prior and real-time images can be problematic. Real-time use of intracardiac echocardiography (ICE) has been used to help achieve accurate anatomic modeling during the actual tachyarrhythmia,¹⁶¹ can assist with transbaffle or transseptal needle puncture, and can help assess the stability of the ablation catheter¹⁶² during mapping and energy delivery. Finally, real-

Table 2 Classification of congenital heart disease complexity

| Complexity | Type of congenital heart defect in the adult patient | Caveats in children |
|--|---|---|
| Simple | <i>Native disease</i> | |
| | Isolated congenital aortic valve disease | Great variability and can be in “moderate” or “severe/complex” category |
| | Isolated congenital mitral valve disease (except parachute valve, cleft leaflet) | Great variability and can be in “moderate” or “severe/complex” category |
| | Small ASD | Moderate and most large ASDs |
| | Isolated small ventricular septal defect (no associated lesions) | |
| | Mild pulmonary stenosis | |
| | Small patent ductus arteriosus | |
| | <i>Repaired conditions</i> | |
| | Previously ligated or occluded ductus arteriosus | |
| | Repaired secundum or venosus atrial septal defect without residua | |
| Moderate | Repaired ventricular septal defect without residua | |
| | Aorto-left ventricular fistulas | |
| | Anomalous pulmonary venous drainage, partial or total | “Severe/complex” category if obstructed or mixed |
| | Atrioventricular septal defects, partial or complete | “Severe/complex” category if significantly unbalanced |
| | Coarctation of the aorta | |
| | Ebstein’s anomaly | Great variability and can be in “severe/complex” category |
| | Infundibular right ventricular outflow tract obstruction of significance | |
| | Ostium primum atrial septal defect | |
| | Patent ductus arteriosus, not closed | Great variability and can be in “simple” category |
| | Pulmonary valve regurgitation, moderate to severe | |
| | Pulmonary valve stenosis, moderate to severe | |
| | Sinus of Valsalva fistula/aneurysm | |
| | Sinus venosus atrial septal defect | |
| | Sub-valvar or supra-valvar aortic stenosis | |
| | Tetralogy of Fallot | |
| | Ventricular septal defect, with: | “Severe/complex” if single ventricle |
| | -Absent valve or valves | |
| | -Aortic regurgitation | |
| -Coarctation of the aorta | | |
| -Mitral disease | | |
| -Right ventricular outflow tract obstruction | | |
| -Straddling mitral or tricuspid valve | | |
| -Subaortic stenosis | | |
| Severe/complex | Conduits, valved or nonvalved | |
| | Cyanotic congenital heart disease, all forms | |
| | Double-outlet ventricle | “Moderate” if uncomplicated subaortic VSD, or if tetralogy of Fallot-type |
| | 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 | Great variability of heterotaxy would place some in “moderate” category |
| | not included above (e.g., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion) | |

Modified from Warnes et al.¹⁴⁹

time visualization of electrode catheters helps reduce exposure to ionizing radiation. Most importantly, acute procedural success appears to be enhanced by use of these systems.¹⁶³

All relevant energy sources are applicable to patients with CHD; however, for macroreentrant mechanisms in atrial or ventricular muscle, transmural lesions might be necessary, sometimes at depths greater than 1 cm. In such cases, RF

energy delivered through actively or passively cooled tips is generally useful, resulting in higher power delivery, larger lesion volume, and greater acute and long-term success rates.^{164,165} Tissue contact force monitoring can also be useful, but has not been reported in patients with CHD. In addition, a variety of techniques are under development to improve the prediction of lesion size and/or to increase the lesion volume.

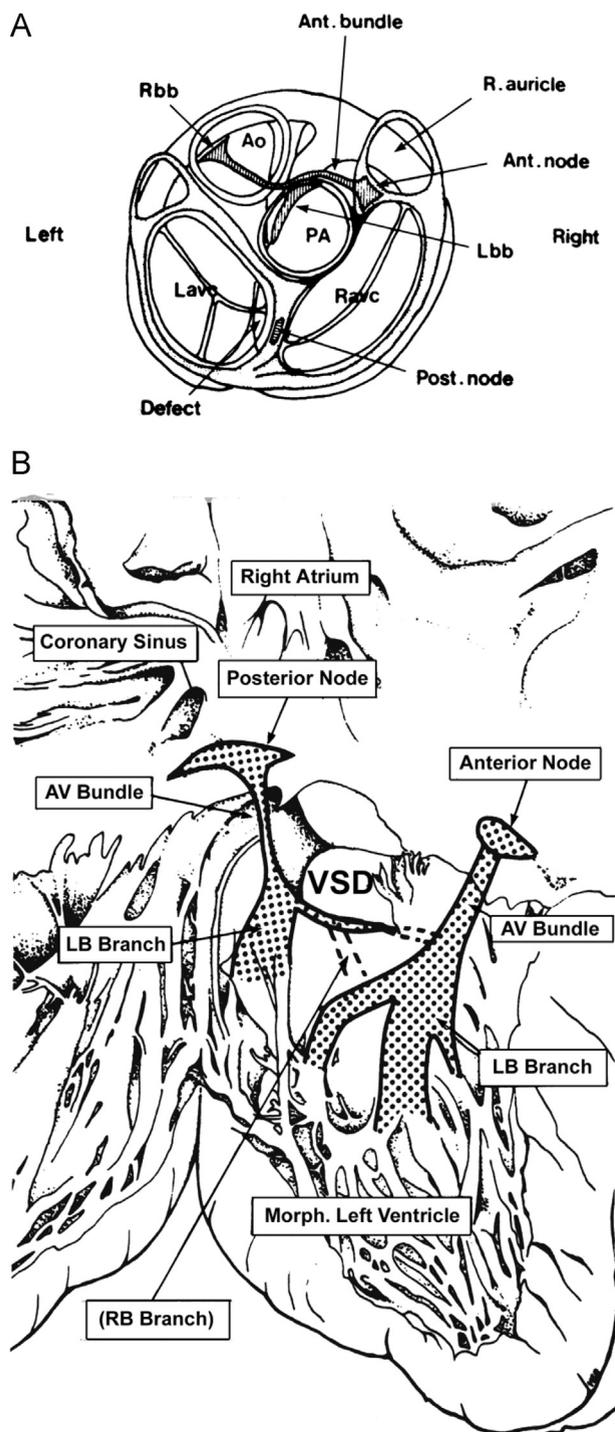


Figure 1 **A:** Diagram of the base of the heart and the AV conduction system in L-transposition of the great arteries (corrected) as seen from above. Note the bicommissural mitral valve on the right and the tricommisural tricuspid valve on the left. The AV node can either lie posteriorly (Post. node) in the septum in a somewhat normal location, anteriorly on the right-sided mitral valve (Ant. bundle), or in both places. (Reproduced with permission from Anderson et al.¹⁶⁶) **B:** Diagram of the conduction system of a patient with corrected transposition in a crisscross heart, with AV valve anatomy similar to that shown in A. Note the dual-conduction system with both the posterior and anterior AV nodes penetrating into the ventricles, and near connection of the conduction systems within the ventricle. (Reproduced with permission from Symons et al.¹⁶⁷)

There are specific considerations for mapping and ablation in patients with twin AV nodes and complex CHD

(Figure 1).^{166,167} Identification of anterior and posterior His bundle electrograms related to the (usually single) AV annulus requires careful electroanatomic rendering. Mapping during induced tachycardia enables determination of which AV node/s conduct in an antegrade manner and which conducts in a retrograde fashion. It is the latter that is usually targeted for ablation.

Outcomes

Acute and chronic success rates after catheter ablation in patients with atrial or ventricular tachycardias related to CHD will be discussed in the sections devoted to those entities. In patients with CHD undergoing catheter ablation of APs, the acute and long-term success rates of 80% and 68%, respectively,¹⁶⁸ are substantially lower than in children with structurally normal hearts: 96% and 85%, respectively.^{9,10} Moreover, catheter ablation in children with CHD is associated with higher complication rates and hospitalization costs.¹⁶⁹ Due to the frequent occurrence of both congenital and acquired tachyarrhythmia substrates in patients with Ebstein's anomaly, there is evidence that these patients should routinely undergo electrophysiological testing with the option of catheter ablation prior to tricuspid valve surgery, irrespective of symptom burden.¹⁷⁰

Catheter ablation of tachyarrhythmia substrates in patients with CHD can also be viewed in the broader context of their surgical care. Although beyond the scope of these guidelines, the role of intraoperative substrate modification using various energy forms following tachyarrhythmia substrate elucidation in the electrophysiology laboratory, or less commonly in the operating room, is evolving.¹⁷¹ Likewise, there is growing interest in the provision of prophylactic intraoperative ablation at the time of congenital heart surgery to prevent subsequent macroreentrant tachycardias.¹⁷² The past 20 years of tachyarrhythmia substrate mapping in the electrophysiology laboratory have advanced our knowledge of the most common anatomical regions that promote these tachyarrhythmias.

3.1.1 Atrial Tachycardias in Patients with Congenital Heart Disease (Not Including Fibrillation)

Atrial arrhythmias represent a major source of long-term morbidity and are a significant contributor to mortality after repair or palliation of CHD. In a population-based study of patients with CHD, the 20-year risk of developing an atrial arrhythmia was 7% in a patient aged 20 years and 38% in a patient aged 50 years.⁹⁵ Although automatic AT and NAFAT are well recognized, intra-atrial reentry tachycardia (IART) is the most common form of postoperative AT, especially in patients younger than 40 years of age.

The incidence of late atrial tachyarrhythmia has been reported to be 29% to 60% after an atriopulmonary-type Fontan procedure, 14% to 48% after atrial switch (Mustard and Senning) procedures for D-TGA, and 20% after tetralogy of Fallot repair.¹⁷³ Although ASD is considered a simple lesion with an excellent long-term prognosis, these patients

are at risk of late development of atrial flutter and fibrillation, especially if undergoing surgery at older ages.^{174,175}

The newer modifications of the Fontan procedure have been shown to be less prone to the development of atrial arrhythmias, with the incidence reduced by 50% to 70% compared with the atriopulmonary Fontan procedure.^{176–181} In general, the emergence of AT correlates highly with the presence of heart failure, and is associated with thromboembolic events and sudden cardiac death, especially in patients after Fontan or atrial switch operations.^{176,182,183}

Intra-Atrial Reentry Tachycardia

Mapping and Ablation. Given the highly significant correlation of IART with morbidity and mortality in patients with CHD and the disappointing results of pharmacologic therapy, catheter ablation was used early in the history of the technology. Mapping techniques have been based on activation mapping and entrainment with concealed fusion.¹⁸⁴ Successful targets are regions of slow conduction between combinations of natural conduction barriers (vein orifices, AV valve annuli, and crista terminalis) and surgically created barriers (patches, conduits, and dense scar). With increased experience, it has become clear that certain areas in the right atrium represent “hot spots” for potentially successful ablation lesions.¹⁸⁵ The tricuspid valve-inferior vena cava (TV-IVC) isthmus has been found to be a critical area.^{150,187} For patients after the Mustard or Senning procedure, this isthmus is bisected by the atrial baffle. To achieve isthmus block, the portion of the isthmus connected to the tricuspid valve must frequently be reached by either a retrograde transaortic or an antegrade transbaffle approach.

Macroreentrant circuits in Fontan patients are anatomically distinct, with the lateral right atrial wall the most common area for successful RF applications.¹⁸⁵ Pericaval circuits have been identified specifically in Fontan patients and successfully ablated.¹⁸⁶ A dual loop *figure of 8* reentry has been identified in 20% to over 50% of patients with CHD and IART,^{187–189} and in the great majority of those with surgically repaired ASDs.¹⁸⁸

Outcomes. Several studies are available to assess procedure efficacy and safety. From 9 case series, a total of 580 patients underwent 772 procedures.^{158,160,163,164,190–193} The patient groups consist of those with single-ventricle physiology, primarily after some form of Fontan operation, D-TGA after a Mustard or Senning operation, and other types of CHD after biventricular repair, mostly tetralogy of Fallot and ASDs. Acute success varied from 66% to 97%, recurrences from 10% to 59%, and longer-term success from 53% to 92%. Major factors related to outcome included the following: (1) severity of CHD (single ventricle and D-TGA patients have poorer outcomes); (2) older age at repair (associated with worse outcomes); and (3) use of EAM and irrigated tip catheters (associated with higher success rates). The outcome of patients with two-ventricle repairs (excluding Mustard and Senning patients), such as ASDs and tetralogy of Fallot, has been much more encouraging, with

acute success rates of 98% to 100%; and long-term sinus rhythm, with or without antiarrhythmic medications, in 84% to 96%.^{19,194,195} Many apparent recurrences in patients with CHD have been due to the appearance of new arrhythmias, including AF.

Non-Automatic Focal Atrial Tachycardia

In addition to IART, patients with repaired CHD can present NAFAT, defined as focal atrial tachycardias (FATs) that are inducible and terminable with programmed atrial stimulation. Mapping and ablation of NAFAT has been described in patients with all forms of CHD. Clustering of the NAFAT foci in the right atrium and in the ostia of the pulmonary veins has been described.¹⁵⁰ We note that the ECG features of this arrhythmia can be difficult to distinguish from IART; however, the tachycardia tends to be paroxysmal and self-limited, unlike IART, which is usually persistent.¹⁹⁵ The success rate of NAFAT ablation has varied from 77% to 100%.^{150,195,196}

Catheter ablation in the postoperative atrium, especially using high-output RF energy, is associated with complications previously referenced in this document for surgically naïve patients. The risk of phrenic nerve damage is probably higher in the patient undergoing surgery due to distortion of the nerve’s epicardial course and the potential for fibrous adherence to the epicardial surface. If the substrate to be ablated is potentially near the normal course of a phrenic nerve, careful mapping of its course is advisable, using high-output pacing.

3.1.2 Ventricular Tachycardias in Patients with Congenital Heart Disease

Ventricular tachyarrhythmias are well-known late sequelae after surgical repair of a variety of forms of CHD. Most experience on this topic has been accumulated with tetralogy of Fallot.³ Sustained ventricular arrhythmias are frequently associated with a risk of sudden death, and ICD implantation is an important consideration for many patients.³ The arrhythmia substrate can be related to morphological variants of the heart defect itself or ventricular incisions, scar, and patches. Monomorphic VT is frequently due to a stable macroreentrant circuit that can be targeted for ablation.¹⁹⁷ Rapid polymorphic VT is typically seen with systemic or pulmonary ventricle failure, in either a two- or single-ventricle repair.³ Catheter ablation is typically not an option unless polymorphic VT is repeatedly induced by an ectopic beat that can be targeted for ablation. Catheter ablation of sustained monomorphic VT is recommended for recurrent VT that is not adequately suppressed by antiarrhythmic drug therapy, and can be considered following the initial presentation of sustained monomorphic VT. In patients with postoperative VT after ICD implantation, ablation can be beneficial by reducing the arrhythmia burden and the number of ICD shocks.

Prevalence

Although a publication in 2000 reported sustained monomorphic VT in 4.5% of a population who underwent surgery at a mean age of eight years, a more recent document underscores the importance of age in this population. The prevalence was less than 10% before 25 years and reaches greater than 30% by 35 years of age.

Age at repair and at follow-up appear to be important factors in the prevalence of VT in this population. In a multicenter study published in 2000 of 793 patients who had undergone tetralogy of Fallot repair at a mean age of eight years, sustained monomorphic VT occurred in 4.5% of the patients during a mean postoperative follow-up of 21 years. Sudden cardiac death occurred in another 2% of the patients.¹⁹⁸ However, in a more recent multicenter study of 556 adults (mean age 37 years) with repaired tetralogy of Fallot, ventricular arrhythmias had occurred in 14.6% of the patients.¹⁷³ The prevalence was less than 10% before the age of 25 years, and increased markedly after the age of 45 years. With current routine surgical correction performed early in infancy via a transatrial-transpulmonary approach that avoids right ventriculotomy, it is hoped that the prevalence of VT will decrease.¹⁹⁹

Anatomy and Pathophysiology

After surgery for tetralogy of Fallot, sustained monomorphic VT is typically due to macroentry through anatomically defined isthmuses in the RV.^{197,200,201} Several potential isthmuses have been identified (see Figure 3 of Kapel et al¹⁹⁷): (1) between the superior aspect of the tricuspid valve annulus and an unexcitable scar/patch in the free wall of the RVOT; (2) between the pulmonary trunk and the RVOT patch (in patients without a transannular patch); (3) between the tricuspid valve annulus and the VSD patch; and (4) between the VSD patch and the pulmonary trunk.^{197,200–204} Focal origin VTs, possibly due to microentry, have also been reported.²⁰⁵ In patients who have had sustained VT, multiple morphologies of inducible VT are common and can be due to use of the same isthmus in different directions, or to VTs using different isthmuses.¹⁹⁷ VT is occasionally related to a scar in an LV location or in other diseased RV myocardium.²⁰⁶

Mapping and Ablation

Initiation of VT with programmed stimulation allows assessment of the QRS morphology of the VT, helps confirm the diagnosis, and provides a potential assessment of the procedural endpoint of inducible VT absence. In early reports, contact mapping combined with pace mapping and entrainment mapping was performed with satisfactory results.²⁰² This approach has limitations, particularly in patients with fast and hemodynamically unstable tachycardias. Based upon results from EAM systems, areas containing fibrosis with either surviving myocardium or dense fibrosis are typically characterized by electrograms with a bipolar voltage of less than 0.5 mV. The aim of endocardial

mapping is to identify anatomical isthmuses (i.e., viable myocardium) between unexcitable areas, as described above. Once the potential anatomic isthmuses are defined, the isthmus involved in the VT is sought. The QRS morphology often suggests the exit from an isthmus. If VT is hemodynamically tolerated, activation mapping or entrainment mapping can be used. If the VT is not tolerated, an isthmus can be selected for ablation based on pace mapping, or all the potential isthmuses can be targeted for ablation. Pacing near the exit of the isthmus usually produces a QRS morphology similar to that of the VT.^{197,207} A delay between the stimulus and the QRS indicates the conduction time from the pacing site to the exit from the isthmus. At entrance sites to the isthmus, however, the paced QRS can be different from that of the VT even if that isthmus is in the circuit. The isthmus involved in the VT is then targeted for ablation.^{197,200–202,204} One of the most common isthmuses is between the ventricular septal patch and the pulmonary valve annulus (conal septum), and it occasionally requires ablation from the LV side of the septum.¹⁹⁷ The isthmus across the free wall between an incision or patch and the tricuspid annulus is also common, tends to be broad and thick, and can be difficult to interrupt.^{197,199} The isthmus between a VSD patch and the tricuspid annulus might contain the conduction system, with a risk of heart block from ablation. A line of ablation applications is placed to transect the respective isthmus during sinus rhythm or stable VT. The endpoint of ablation directed at transecting an isthmus should ideally be the demonstration of complete conduction block along the ablation line. This outcome can be assessed by pacing on one side of the ablation line and demonstrating late and inverted activation on the opposite side of the line in the appropriate sequence.^{200,201,203} Double potentials can be present along the line, as can absence of capture during pacing. VT inducibility should be assessed as well. The continued presence of inducible VT might be due to failure to achieve conduction block, or VT arising from a different isthmus or other site, such as a more left-sided structure.^{197,206}

Ablation Outcomes

Experience with ablation of VT in postoperative CHD patients is still limited; however, several case series have reported satisfactory results,^{197,200–202,204,208} with success rates ranging from 80% to 100%. Significant complications were infrequent, but AV block can occur. Femoral access complications and low cardiac output with delayed recovery from hypotensive VT or frequent pacing can occur when ventricular function is poor.¹⁹⁷ Earlier reports found recurrence rates up to 40% during mid-term follow-up,²⁰⁸ probably in part due to insufficient lesion formation. Recently, Kapel et al¹⁹⁷ reported ablation of 61 VTs in 34 adults with repaired CHD, of whom 28 had tetralogy of Fallot. An anatomic isthmus was identified for all but two, and noninducibility of VT and demonstration of isthmus block were achieved in 25 patients. At 46-months follow-up, none of those 25 had recurrent monomorphic VT, but one who also had poor ventricular function had an episode of ventricular fibrillation. Ablation failure was attributed to the

inability to achieve conduction block in an isthmus, or the inability to identify a critical isthmus in nine patients; four of these patients subsequently had recurrent VT.

Due to the risk of recurrence and sudden death, ICD implantation is advised in patients with failed ablation and in those with depressed ventricular function even if ablation is successful.²⁰¹

Indications for ablation procedures in patients with CHD (for atrial fibrillation, see Section 2.4.2 and accompanying indications)

Class I

1. Ablation is recommended for recurrent[#] or persistent[^] SVT related to accessory AV connections or twin AV nodes in patients with CHD when medical therapy is either not effective or associated with intolerable adverse effects (the medical therapy used depends on patient weight⁺). Ablation is also recommended as an alternative to medical therapy for larger* patients (LOE: B).
2. Ablation is effective for patients with WPW pattern and high-risk or multiple accessory pathways, as commonly encountered in Ebstein's anomaly, in larger* patients with CHD (LOE: C).
3. Ablation is effective for recurrent symptomatic atrial tachycardia in patients with CHD occurring outside the early postoperative phase (less than three to six months) when medical therapy is either not effective or associated with intolerable adverse effects (the medical therapy used depends on patient weight⁺). Ablation is also recommended as an alternative to medical therapy for larger* patients (LOE: B).
4. Ablation is recommended for adjunctive therapy to an ICD in patients with CHD and recurrent monomorphic VT, a VT storm, or multiple appropriate shocks that are not manageable by device reprogramming or drug therapy (LOE: C).

Class IIa Ablation can be useful for the following:

1. Sustained monomorphic VT in patients with CHD causing symptoms or hypotension, when medical therapy is either not effective or associated with intolerable adverse effects (the medical therapy used depends on patient weight⁺). Ablation is also recommended as an alternative to medical therapy in larger* patients. The decision whether to have an ICD as adjunctive therapy should follow the ACHD guidelines³ (LOE: B).
2. Recurrent[#] or persistent[^] AVNRT when medical therapy is either not effective or associated with intolerable adverse effects in larger* patients with moderate or complex CHD (mild CHD can be managed as narrow [usual[#]] complex SVT) (LOE: C).
3. Substrates that have a reasonable likelihood of contributing to tachyarrhythmias in the postoperative period, in the absence of other indications, when impending

- congenital heart surgery will result in restriction of vascular or chamber access following surgery (LOE: C).
4. Substrates that have a reasonable likelihood of contributing to tachyarrhythmias in the postoperative period, in the absence of other indications in larger* patients with Ebstein's anomaly prior to anticipated cardiac surgery (LOE: C). (Note: This was a Class IIb indication in the prior ACHD guidelines³).
 5. Recurrent asymptomatic atrial tachycardia occurring outside the early postoperative phase (less than three to six months) in larger* patients with CHD who are at increased risk of thromboembolic events or worsening heart failure, or in smaller* patients when medical therapy is either not effective or associated with intolerable adverse effects (LOE: C).
 6. Frequent ventricular ectopy, predominantly from a single focus, thought to be contributing to deteriorating ventricular function in larger* patients with CHD, or in smaller* patients when medical therapy is either not effective or associated with intolerable adverse effects (LOE: C). (Note: This was a Class IIb indication in the prior ACHD guidelines³).

Class IIb Ablation can be reasonable for the following:

1. SVT with acute hemodynamic compromise in smaller* patients with CHD (LOE: E).
2. Creation of complete AV block with permanent pacing in patients with atrial tachyarrhythmias refractory to all medications and substrate-targeted catheter ablation, who are not candidates for surgical therapy (LOE: B).

Class III Ablation is not recommended for the following:

1. Atrial tachyarrhythmias that can be managed medically in the early postoperative period (less than three to six months postoperatively) (LOE: C).
2. Asymptomatic ventricular ectopy and stable ventricular function in patients with CHD (LOE: C).
3. Prophylactic therapy of ventricular arrhythmias in patients with CHD deemed to be at increased risk for sudden cardiac death, and in whom an ICD is otherwise indicated (LOE: C).

3.2 Infants and Small Children

Transcatheter ablation of cardiac arrhythmias in infants (1 year of age and younger) and small children (15 kg and less) is controversial.^{7,209} Studies assessing ablation in small children have not limited their analysis to children under this weight or to one specific age group, and those studies that address ablation in "infants" have not always defined this group in the same terms.^{210–217} Therefore, where data exist, issues related to subsets of children such as infants or those specifically weighing less than 15 kg are the focus of this section.

Ablation Substrates

AVRT, AVNRT, and FAT account for 80%, 5%, and 15% of SVT in children younger than 1 year of age, respectively, and

approximately 65% to 75%, 10% to 25%, and 10% of SVT, respectively, in children who are between 1 and 5 years of age.⁶⁹ Atrial flutter, JET, and VT in small children are much less common.^{3–10}

Clinical Presentation and Medical Therapy

Although the majority of tachycardias occurring in small children are well tolerated, a number of infants might present with congestive heart failure. Nevertheless, the tachycardia can typically be managed conservatively with anti-arrhythmic medication,^{218,219,220} with the knowledge that 60% to 90% of patients who present with WPW or AVRT in infancy and 20% to 50% with less common tachycardias will not have a recurrence of tachycardia before the age of 5 years after medication is discontinued at six to 12 months of age.^{76,209,218} Despite the success of medical therapy in most infants, the following circumstances can lead to the need for an attempt at definitive therapy with catheter ablation in small children:

- Failure of medical therapy after escalation to Class I and/or Class III anti-arrhythmic agents for common forms of SVT.²²¹
- Complex and life-threatening arrhythmias with inadequate response to medical therapy.^{131,222}
- Intolerability or high risk of malignant proarrhythmia from drug therapy, particularly when Class I/III agents are used alone and in combination.²²³

Cardiac arrest secondary to rapid ventricular conduction over an AP during AF in patients with WPW appears to be very uncommon in young children; thus, high-risk AP conduction characteristics alone are not an indication for ablation in small children.²²³

Catheter Ablation

Nearly all the data regarding ablation of cardiac arrhythmias in small children have involved the use of RF energy; however, because of the risks detailed below, some practitioners now use cryoenergy as their first choice for ablation in small children.

Outcomes. The success rate of RF ablation in small children in terms of eliminating any particular substrate or arrhythmias on the whole is similar to that of older children,^{6,65,210} perhaps because ablations in infants have been performed by more experienced pediatric electrophysiologists—a factor associated with successful pediatric ablation procedures in general.^{210,224} The presence of multiple APs has been found to be a risk for ablation failure, whereas pathway location and CHD have not been related to ablation failure in small patients with APs.^{65,210}

Complications

Children weighing less than 15 kg or at ages younger than 5 years have been shown to be at increased risk for complications during RF ablation,^{6,7,65} both when compared directly with larger or older children, and when compared

with reports on large pediatric groups undergoing ablation.^{211–215,217} Although these studies lacked statistical power, reported complications in smaller and younger children have been consistently more severe than those reported in older children and adolescents, including death, heart block, cardiac perforation and effusion, and coronary injury.^{210–215,217} Most practitioners consider the smallest infants (less than 5 kg) to be at higher risk from catheter ablation and use a higher threshold for transitioning from medical- to catheter-based therapy. However, there are no data other than anecdotal reports to support differences in outcomes or complications between these smallest patients and those up to approximately 15 kg. Consequently, the committee chose a weight of 15 kg as the threshold for changes in the recommendations, with occasional reference to minor modifications for the smallest infants.

Death. The Pediatric Ablation Registry reported an overall mortality rate of 0.12% for patients with a structurally normal heart and 0.89% for patients with CHD.¹² A review of 111 infant procedures (younger than 18 months of age) in the Registry reported two deaths (1.8%): one patient without and one with CHD.²¹⁰ From 1991–1995, 281 of 3912 SVT ablations (7%) were performed on children younger than five years of age; however, three of the seven deaths (43%) that occurred were in this younger age group, which was disproportionately high.^{12,65} Although subsequent case series of small children at various ages and weights have not reported deaths related to ablation, it is not possible to know if this represents selection bias, better patient selection, or improved techniques for the recognition of higher risk.^{211–217}

AV Block. Considering the relative sizes of RF lesions and the triangle of Koch in children, it is not surprising that RF ablation for septal APs in infants is associated with a greater likelihood of heart block than ablation in other locations.²¹⁰ Although weight less than 15 kg was not associated with AV block during ablation of AP mediated tachycardias in early registry studies, it is likely that there were too few patients with septal pathways in this weight class to have enough power to show a significant association.²⁰⁹

Cardiac Perforation and Pericardial Effusion. The incidence of cardiac perforation and/or pericardial effusion after ablation in the pediatric population has been reported to be 0.1%,⁹ whereas in infants and small children, the reported risk has ranged from 1.3% to 2.2%.^{210–214,217}

Coronary Injury. All the reports of coronary artery injury following ablation have involved RF energy. They most frequently occurred following AP ablation as single case reports, or in larger series in the absence of coronary angiography prior to ablation.^{9,20,225} One report of 112 pediatric patients describes a series of patients who underwent coronary angiography before and after attempted AP ablation: two (1.8%) had post-ablation coronary stenosis.²⁴ The small number of infants or small children in that study, however, precluded any subgroup analysis. Because the mechanism of coronary injury is likely to be a combination of direct thermal injury and subsequent inflammatory response, the proximity of the coronary arteries to the lesion location is likely to be an

important factor in coronary damage. In a study on normal heart specimens, the distance from various locations along both AV valves to the closest coronary artery correlated with age, and in children three years of age and younger, most potential ablation locations were within a distance equal to or less than the depth of a typical RF lesion.²²⁶

Potential Procedural Modifications

An increase in complications has been related to a greater number of RF applications over 20 seconds indexed to patient weight, suggesting that fewer and shorter applications might help prevent complications in smaller patients.²²⁷ Limiting the number of RF applications has been associated with decreased mortality; thus, avoiding “insurance” applications and limiting the number of applications overall, even if it means the procedure is unsuccessful, might be reasonable for this age group.^{210,227}

Cryotherapy can offer important advantages for small children. Cryo lesions are smaller than RF lesions, contributing to the ability to safely create cryo lesions anywhere—even adjacent to the His bundle.²²⁸ In addition, cryoablation allows for reversible loss of tissue function.^{229,230} In the largest cryoablation clinical study in small children, defined as children less than 15 kg or younger than five years of age, cryoablation was found to have a success rate of 74% and a recurrence rate of 20%,²¹⁷ whereas RF—which was used after failed cryoablation—had a success rate of 81% and a recurrence rate of 30%. Regardless of technique, these outcomes are not as good as in older children; however, importantly, no complications were found in the cryoablation-alone group, which was statistically lower than the 12.5% major complication rate for the RF after failed cryoablation group.

Prior Indications for Ablation in Small Children

In the 2002 guidelines, the cutoff for age in a variety of the recommendations was either younger than four years or younger than five years of age.¹ The only ablation indications for the younger group were Class II and were limited to tachycardia associated with severe ventricular dysfunction refractory to complex drug therapy, or when effective medications caused intolerable adverse effects.¹

Recommendations

Based on the data reviewed above, the task force chose an approximate cutoff of 15 kg to differentiate indications for ablation, recognizing that an exact size or age should not be specified for individual patients. The nomenclature used refers to children less than approximately 15 kg as “smaller children” and over 15 kg as “larger children.” In general, catheter ablation in the very smallest children (3 to 7 kg and younger than 6 months of age) should be reserved for life-threatening arrhythmias, or refractory arrhythmias after multiple failed attempts at medical management, which can include various combination therapies. Ablation should be performed by an experienced pediatric electrophysiologist who

undertakes various strategies to reduce risk, including limiting power and temperature as well as the number and duration of applications. The use of alternate sources of energy prior to using RF, such as cryoablation, can be considered.

The specific guidelines that emanate from the discussions in this section are necessarily distributed throughout this document because they apply to numerous sections, including every arrhythmia-based indication set, and the safety and procedural guidelines in Section 4 below. However, the significant differences between managing this patient group and older larger patients were important enough to collate the relevant indications and guidelines for smaller patients in a single location, in addition to their being included in other sections.

Indications for Ablation and Safety Guidelines for Infants and Smaller Patients (<15 kg)

Relevant Footnotes. *Smaller patients are in general less than approximately 15 kg, and larger patients greater than approximately 15 kg.

⁺The precise definition of “medical therapy that is either not effective or associated with intolerable adverse effects” is left up to the practitioner and family to decide. In general, however, the threshold for ineffectiveness and intolerability should be higher in smaller patients. For example, failure or intolerability of a beta-blocker alone might be adequate in the larger patient, but not in the smaller patient, unless there were additional circumstances. In the smallest patients, failure or intolerability of drug combinations from multiple classes, including membrane stabilizing agents with adequate loading time, would be required prior to the decision for ablation.

Class I Ablation is recommended for the following:

1. Documented SVT, recurrent[#] or persistent[^], when medical therapy is either not effective or associated with intolerable adverse effects (LOE: C).
2. WPW pattern following resuscitated cardiac arrest (LOE: B).
3. WPW pattern with syncope when there are predictors of high risk for cardiac arrest[§] (LOE: B).
4. Persistent[^] or recurrent[#] idiopathic JET, or congenital JET associated with ventricular dysfunction, when medical therapy is either not effective or associated with intolerable adverse effects⁺ (LOE: C).

When ablation of JET is being performed, cryotherapy is the preferred first choice due to the high risk of AV block. RF energy should be used with extreme caution, after a detailed discussion with the family or patient concerning the high risk of AV block and the potential need for permanent pacing (LOE: C).

5. Ventricular ectopy or tachycardia with ventricular dysfunction, when medical therapy is either not effective or associated with intolerable adverse effects (LOE: C).
6. Recurrent[#] or persistent[^] SVT related to accessory AV connections or twin AV nodes in patients with CHD when medical therapy is either not effective or associated with intolerable adverse effects (LOE: B).

7. Ablation is effective for recurrent symptomatic atrial tachycardia occurring outside the early postoperative phase (less than three to six months) in patients with CHD, when medical therapy is either not effective or associated with intolerable adverse effects (LOE: B).
8. Pediatric cardiovascular surgical support should be available in-house during ablation procedures for smaller patients* (LOE: E).

Class IIIa Ablation can be useful for the following:

1. WPW pattern with ventricular dysfunction presumed due to dyssynchrony when medical therapy is either not effective or associated with intolerable adverse effects⁺ (LOE: B).
2. Recurrent sustained monomorphic VT causing symptoms or hypotension despite antiarrhythmic drug therapy in patients with CHD (LOE: B).
3. Recurrent asymptomatic atrial tachycardia occurring outside the early postoperative phase (more than three to six months) in patients with CHD who are at increased risk of thromboembolic events or worsening heart failure, when medical therapy is either not effective or associated with intolerable adverse effects (LOE: C).
4. Substrates that have a reasonable likelihood of contributing to tachyarrhythmias in the postoperative period in the absence of other indications, when impending congenital heart surgery will result in restriction of vascular or chamber access following surgery (LOE: C).
5. Frequent ventricular ectopy thought to be contributing to deteriorating ventricular function in patients with CHD, when medical therapy is either not effective or associated with intolerable adverse effects (LOE: C).

Class IIb

1. Recurrent clinical symptoms clearly consistent with paroxysmal SVT, and one of the following at electrophysiology study: evidence for involvement of an AV accessory pathway; inducible SVT (LOE: C). Medical therapy should be considered prior to ablation (LOE: C). Cryotherapy should be considered for slow pathway modification (LOE: B).
2. When ablation is indicated, cryoablation can be useful to avoid the higher risk of complications with RF ablation (LOE: C).
3. Recurrent acute hemodynamic compromise (hypotension or syncope) from SVT (LOE: C).
4. Persistent[^] or symptomatic recurrent[#] idiopathic JET, or congenital JET, when medical therapy is either not effective or associated with intolerable adverse effects⁺ (LOE: C).
5. Polymorphic ventricular arrhythmia when there is a suspected triggering focus, arrhythmia, or substrate that can be targeted (LOE: C).
6. Creation of complete AV block with permanent pacing in patients with atrial tachyarrhythmias refractory to all medications and substrate-targeted catheter ablation who

are not candidates for surgical therapy (LOE: B).

Class III Ablation is not recommended for the following:

1. SVT controlled with medical therapy in the absence of intolerable adverse effects (LOE: E).
2. WPW pattern without symptoms (LOE: C).
3. Postoperative JET, due to its potential for spontaneous resolution (LOE: B).
4. Idiopathic or congenital JET adequately controlled with antiarrhythmic medications (LOE: C).
5. VT or ventricular ectopy that is either controlled medically, or is hemodynamically well tolerated without ventricular dysfunction (LOE: C).
6. Accelerated idioventricular rhythm (LOE: C).
7. Atrial tachyarrhythmias that can be managed medically in the early postoperative period (less than three to six months postoperatively), while postoperative remodeling is still occurring (LOE: C).

3.3 Cryotherapy

The 2002 NASPE expert consensus document on catheter ablation in pediatrics cites a single reference on cryoablation and refers to it as an investigational technology.¹ Since that publication, cryoablation has evolved into a widely used and frequently preferred ablation technique. Conflicting opinions are strong regarding the efficacy and role of cryoablation relative to RF ablation. A 2010 survey of pediatric electrophysiologists reported that 80% used cryoablation for less than half of their ablations, and 10% did not use it at all, whereas 40% used cryoablation as a first-line therapy for AVNRT.⁵⁴ The most common reasons reported for avoiding cryoablation were high recurrence rates, poor catheter handling, and no personally experienced complications with RF-induced AV block. Nonetheless, the literature supports an exceptionally high safety profile using cryoablation, with no reports of AV block,²³¹ even in small children²¹⁷ or in the presence of a His potential.²³² Thus, cryoablation has been generally recognized as a safer alternative to RF ablation in proximity to the normal conduction system, a feature that has made the technology particularly desirable in the pediatric population, in whom a complication can lead to a lifetime of morbidity, since most children with SVT are otherwise well.

Technology

The cryoablation catheter was introduced in the 1990s.²³³ There are currently three ablation tip sizes commercially available: 4 mm, 6 mm, and 8 mm; and three curve variations (Medtronic/CryoCath, Minneapolis, MN), which require a dedicated ablation system. The catheter tip is chilled to -80° C, producing an ice-ball at the catheter tip and freezing contacted tissue. A single application should last at least four minutes, and most clinicians employ at least one extra four-minute cycle at successful locations after a brief thaw, frequently referred to as “freeze-thaw-freeze,” which has been shown to extend the lesion’s size.^{42,44,45} In piglets, both

cryoablation and RF lesions have been shown to grow over time. Atrial and ventricular lesions reached two to three times their original size, whereas lesions at the AV groove increased only in depth, not in volume.²⁸

Benefits

Cryoablation has several potential safety advantages over RF ablation, including (1) reversible cryomapping prior to the production of a permanent lesion⁴²; (2) adherence of the catheter tip to the endocardium upon freezing; (3) well-defined lesion margins; (4) minimal effects on adjacent coronary arteries^{24,44,143}; (5) a lower incidence of thrombus formation^{44,228}; and (6) painless application,²³⁴ as well as being associated with lower fluoroscopy use in meta-analyses.^{235,236} The first four of these issues are particularly relevant to small children because of the close proximity of a variety of critical cardiac structures to the ablation target. Some of these benefits follow from the biologic effects. First, cardiac tissue can tolerate a transient period of cooling during which cellular function and conductivity are inhibited, but no permanent damage is induced. This means cryoenergy can be applied, and if there is no desired effect, or an undesired effect, the application can be terminated and the tissue will recover full functionality. This is the intended purpose of the cryomapping mode, although brief applications at full ablation temperature are also typically reversible; this feature of cryoenergy application has contributed most significantly to its safety. Application reversibility allows for cryoenergy applications very near the conduction system with little risk of permanent AV block. Despite this benefit, cryoablation near the AV node should be applied with caution, and the assumption should be made that AV block is still a possible effect.

The risk of coronary artery injury also appears to be decreased with cryoablation compared with RF ablation; the latter causing coronary artery injury in 1.8% of pediatric patients in a prospective study.²⁴ Warming coronary blood flow limits the effect of cryoablation on the coronary arteries, and the elastic tissue shrinkage observed from RF application heating is not a concern. An animal model showed occasional adventitia and medial necrosis, but preservation of the intima without coronary artery stenosis from annular cryoablation applications.⁴⁴

Limitations

The primary limitation of cryoablation is the higher recurrence risk compared with RF in most studies, including meta-analyses^{235,236}; however, most recent reports have found low recurrence rates, comparable to RF, for AV node modification.^{45,47,51,58,235,237} The catheters are stiff and the deflection mechanics are inconsistent, in many cases making the catheter difficult to manipulate. In addition, due to the freeze, the electrograms from the catheter are lost during cryo applications, and the cryo “dose” cannot be adjusted by changing the temperature setpoint on all the commercially available catheters. Because applications last four minutes or

more, and a freeze-thaw-freeze technique is optimal, the ablation time and case duration might be increased.

Cryotherapy for AVNRT

Cryoablation for AVNRT has been the subject of at least 12 pediatric studies, all retrospective.^{41,45,47,55,57,58,238-242} In addition, two meta-analyses incorporating pediatric studies^{235,236} and many adult studies have been published.^{48,49,51,229,237,243-250} Acute success for most studies is greater than 90%, and two meta-analyses found no difference in the acute success of cryoablation compared with RF.^{235,236} Recurrence after cryoablation has been higher in most studies than after RF, as confirmed in meta-analyses (9% vs 3.5%).^{235,236}

The 4-mm tip catheter was used in the earlier studies, whereas later studies reported the use of 6- and 8-mm tips. Several studies have shown lower recurrence with the 6-mm tip compared with the 4-mm tip; however, meta-analyses do not show a significant difference.^{47,48,235,236,244} The use of the freeze-thaw-freeze technique is inconsistent across studies. Studies reporting the use of this technique^{45,51,58,237,247,248} generally report lower recurrence rates than those that do not. Procedural endpoints are important, and complete elimination of the SP improved outcomes in several studies.^{51,243,245} A meta-analysis showed the most important endpoint is non-inducibility on isoproterenol.²⁵¹ As noted previously, no study has reported permanent second-degree or higher AV block resulting from AVNRT cryoablation, with over 5000 patients represented, including a recent large adult study with over 1300 patients.²⁵² Clearly, the lack of prior reports should inspire caution when ablating near the AV conducting system.

Cryotherapy for Accessory AV Pathways

The evolution of the cryoablation technique has also been evident in studies on AP ablations. There are no randomized trials comparing RF with cryoablation for AP ablation. Acute success rates are widely variable, between 62% and 97%, with the most recent studies being greater than 90%—similar to RF ablation.²⁵³⁻²⁵⁵ With rare exceptions,^{255,256} recurrence risk after cryoablation for APs has been high: 12.5% to 45%, but appears to be reduced when AP conduction blocks earlier in the application.²⁵⁷ As with AVNRT, there have been no reported occurrences of permanent second-degree or higher heart block using cryoablation for APs, regardless of location.

Cryoablation for Other Substrates

A few reports on cryoablation use for pediatric JET have demonstrated its feasibility for this substrate,^{110,258} and a survey of 70 pediatric electrophysiologists reported that 83% would choose cryoablation for this arrhythmia substrate.⁵⁴ Early studies of FAT substrates showed poor outcomes using cryoablation^{145,254}; however, cryoablation may be a useful technique to avoid phrenic nerve injury when ablating FAT, or for a focus within a pulmonary vein.²⁵⁹

3.4 Fluoroscopy Exposure

Risks of Ionizing Radiation

Radiation risks include both deterministic and stochastic effects. Deterministic effects are dose dependent and predictable, including skin erythema, epilation, cataracts, retarded bone growth, sterility, decreased white blood cell count, organ atrophy, and fibrosis. The threshold level for deterministic effects to occur is 2 Gy. Stochastic effects are not dose dependent, but rather follow a linear, no threshold model. They include malignancies and hereditary defects. These risks accrue both to the patient and to the staff within the lab.

Using dosimeters to measure radiation exposure in both adults²⁶⁰ and pediatric patients,²⁶¹ the lifetime increased risk of fatal malignancy from a catheter ablation procedure using fluoroscopy has been estimated to be 0.02% to 0.03%, with the lung at greatest risk, followed by the bone marrow, breast, and thyroid. However, based upon measured DNA breaks in white blood cells in pediatric patients undergoing diagnostic or interventional catheterization,²⁶² a fourfold greater risk of malignancy was determined compared with the standard linear no threshold estimate. Given that children have a longer time span in which to express potential stochastic effects, these findings might have even greater importance.

Techniques to Reduce Procedure-Based Radiation

Efforts to reduce radiation exposure without compromising procedure outcomes should benefit the patient and personnel in the room. The general standard for radiation exposure should be *As Low as Reasonably Achievable (ALARA)*. Whether fluoroscopy is used as the primary imaging technique or as an adjunct to a nonfluoroscopic system, there are a variety of techniques to achieve ALARA, which have been outlined in a 2014 scientific statement from the American Heart Association on enhancing radiation safety (see Table 6 from Papagiannis et al^{272, 263}). The first step is for the medical team and institutional radiation safety personnel to collaborate on optimizing equipment settings and tracking radiation exposure to personnel and patients. Techniques shown to reduce radiation exposure include pulsed fluoroscopy, lower frame rate, adjusting collimators to decrease field view, limiting the use of magnification, employing the “store fluoro” function instead of cineangiography, and alternating between two views rather than a single imaging view to minimize site exposure.^{264, 265}

Over the last 15 years, ease and use of the nonfluoroscopic EAM systems has expanded, along with access to the technology in most contemporary EP laboratories. These systems have led to significant decreases in radiation exposure, and in many less complex cases, procedures can be performed with zero radiation exposure.²⁶⁶⁻²⁷³ Transesophageal or intracardiac echocardiography (ICE) can also be employed simultaneously with these 3-D imaging systems to improve visualization and identification of the anatomy

and catheters, with or without integration of the imaging modalities.^{266, 271}

Nonfluoroscopic Imaging Use in the Structurally Normal Heart

The efficacy and safety of nonfluoroscopic imaging for uncomplicated SVT ablation in pediatrics has been assessed in a number of retrospective studies^{265, 266, 268-270, 273} and two small prospective randomized controlled studies.^{267, 271} When comparing adjunct use of nonfluoroscopic imaging systems with fluoroscopy alone, the reported success rates, long term recurrence rates, and complication rates are all similar to the use of fluoroscopy alone, and total radiation exposure is significantly reduced, regardless of the type of mapping system used. To summarize, despite the absence of a large prospective trial, the data are convincing that nonfluoroscopic imaging systems reduce radiation exposure, and can be safely and effectively used for most SVT ablations in pediatric patients. Ultrasound is generally the only nonfluoroscopic imaging technology demonstrated to safely reduce or mitigate the need for fluoroscopy during transeptal puncture.

Experience and Learning Curve

The first report of catheter ablation being performed without fluoroscopy was by Drago et al²⁷⁰ in 2002; however, the imaging tools available at that time were relatively primitive, and most investigators used EAM as an adjunct. With time and improvements in technology, a number of investigators have reported complete elimination of fluoroscopy for less complex procedures, with the adjunct of either echocardiographic or brief fluoroscopy exposure during a transeptal procedure.^{266, 267, 271, 273-275} With experience, we have learned that the geometry can shift over time due to patient movement, reference electrode movement, and patient fluid status changes (particularly in the impedance-based system: NavX, St. Jude Medical, St. Paul, MN). Therefore, reference points within the body, such as the coronary sinus catheter or an esophageal catheter, help ensure the most stable images.

There are few data regarding the learning curve necessary to become comfortable with 3D mapping systems as the primary catheter visualization tool. The possibility of not using lead aprons initially might incentivize the elimination of fluoroscopy, but to date this strategy has not been evaluated in any reports. One report focuses on the experience of multiple operators at a single center using 3D mapping.²⁶⁷ Only a few procedures were completed with zero fluoroscopy, but overall exposure times were decreased by 59%, while total procedure time increased by 31 minutes. This experience might be representative of what most centers can expect until further experience is gained and the technologies continue to improve.

Limitations of Nonfluoroscopic Imaging

There remain several limitations to 3D mapping systems. These systems are a virtual estimation of the time-space reality, which can be altered by a variety of technical factors,

whereas fluoroscopy is the gold standard for temporospatial data. Three-dimensional mapping systems can only visualize the electrodes on the catheter, and must use a *cartoon* of the catheter shaft, which can be problematic when the manipulation of one catheter is being impeded by another within the heart. Furthermore, sheaths, dilators, and wires are not currently visible, which affects the performance of procedures such as a transseptal puncture, as discussed above. Permanent transvenous implantable device leads are also not shown, leading to a risk of entanglement, dislodging, or damage if no other tools for visualization are used. Lastly, artificial valves cannot be visualized, as with fluoroscopy.

Summary

ALARA should be a widely accepted principle among all laboratories conducting ablation procedures. Both retrospective and prospective studies of pediatric patients have demonstrated significant reductions or even the elimination of radiation exposure when the procedures are performed employing non-fluoroscopic imaging devices. This reduction has been demonstrated regardless of the type of system used. In addition, acute and long-term success and complication rates are not different when compared with fluoroscopy alone. Thus, in accordance with the ALARA principle, it is recommended that fluoroscopic settings be optimized for all procedures; it is reasonable to use 3D nonfluoroscopic imaging as an adjunct for ablation of SVT; and it is recommended that if available, 3D nonfluoroscopic imaging be used for all complex ablation procedures. However, efforts to minimize radiation exposure should not compromise patient safety or procedural success.

3.5 Laboratory Standards

In 2014, HRS published a consensus statement on Electrophysiology Laboratory Standards, addressing process, protocols, equipment, personnel, and safety guidelines, and including a section differentiating issues for pediatric patients.²⁷⁶ This document differentiated the pediatric-specific recommendations based on patient age, with 12 years and under being defined as a special category, as well as patients of any age with CHD. In general, the current committee supports the pediatric-specific guidelines for the laboratory issues and adopts the recommendation for participation in quality initiatives, but extends them to the defined pediatric age for this document: 18 years of age. The prior guidelines also specifically recommended “pediatric cardiac surgical availability” for patients prior to the teenage years. For teenagers and adults with CHD, the guidelines recommended a formalized cardiovascular surgical plan “to accommodate on-site emergencies (notably, cardiac perforation) and a rapid transfer made to a pediatric cardiovascular center should the anatomy of the patient require this expertise.”²⁷⁶ The current guidelines clarify and enhance the prior ones by recommending all of the following as Class I:

- In-house pediatric cardiovascular surgical support for smaller patients (less than 15 kg).

- A pediatric (or congenital) cardiovascular surgical program at the same institution where the ablation is performed for patients 12 years of age and younger, and any age patient with moderate or complex CHD, respectively. There is no specific recommendation that the service be in-house rather than otherwise on-call for emergencies, as per regulatory and institutional standards.
- Age-appropriate cardiovascular surgical program and back-up at the same institution where the ablation is performed for patients 12 to 18 years of age without moderate or complex CHD. There is no specific recommendation that the service be in-house rather than otherwise on-call for emergencies, as per regulatory and institutional standards.

3.6 Patient Preparation, Discharge, and Follow-Up

Preprocedure Preparation

Preparation for EP procedures in children should include a detailed history and physical exam. Because many young patients presenting for EP procedures have never had invasive procedures, particular attention should be given to factors that could place them at higher intraprocedural risk, including a family history of adverse reactions to anesthesia, bleeding, and clotting disorders. Specific issues for patients with CHD have been discussed above. In patients with noncardiac diagnoses such as diabetes, thyroid problems, or rheumatologic problems, it might be prudent to obtain recommendations from the consulting subspecialty services prior to undergoing the EP procedure. Routine laboratory tests are usually not indicated in young healthy patients prior to the EP procedure; however, pregnancy testing is recommended in females at an age dictated by institutional guidelines, typically between 8 and 12 years of age.²⁷⁷ A baseline echocardiogram and ECG should be obtained at some point prior to undergoing ablation.

It is common to discontinue antiarrhythmic medications five half-lives prior to EP procedures. During this period, patients are at risk for developing episodes of tachycardia. In young patients who are not able to report symptoms of tachycardia, additional monitoring might be required prior to the procedure. This monitoring can be accomplished by more frequent assessment by the caregivers, by employing an ambulatory heart rate monitor, or when indicated, by inpatient monitoring. The time for elimination of prior oral intake is tailored toward the age and condition of the patient.

Management of anticoagulation prior to EP procedures is variable and is based on the clinical situation and type of arrhythmia being treated. One strategy is to stop warfarin two to three days prior and bridge with heparin or low molecular weight heparin. However, recent guidelines in adults have suggested that patients undergoing chronic warfarin therapy do not necessarily need to have therapy interrupted when undergoing catheter ablation of AF.²⁷⁸ Likewise, patients receiving chronic aspirin therapy do not necessarily need therapy interruption prior to undergoing ablation.

Psychological preparation of the child/adolescent and family for an EP procedure poses unique challenges. Information regarding the procedure and risks must be complete for the caregivers, but an age-appropriate explanation in the presence of the child is also necessary. A major concern for many families is timing of when to give information about the procedure to their child. The preparation of young patients for invasive EP procedures is based on age and cognitive development.²⁷⁹ Approximately 50% to 75% of children experience severe distress and anxiety before surgery. Risk factors include younger age, temperament, baseline anxiety, past medical encounters, and parental anxiety.²⁸⁰ The presence and participation of the family in the preprocedure preparation is important to reducing the amount of anxiety in the child. Child life services have been shown to improve quality and outcomes in pediatric care and should be included in the preprocedure process.²⁷⁹

Intraprocedure Management

This section addresses a few concerns not covered extensively in the prior EP Laboratory Standards document.²⁷⁶

Anticoagulation. Guidelines for the anticoagulation of patients with AF are well established,²⁷⁸ and guidelines for pediatric ablation have also been proposed in a 2013 scientific statement from the American Heart Association.²⁸¹ The use of anticoagulation is also recommended for patients with intracardiac right to left shunting, regardless of the arrhythmia being treated or whether it is a right- or left-sided procedure. In most adult and many pediatric laboratories, when right-sided procedures are performed in patients without CHD and without AF or atrial flutter, it is common practice not to have an arterial monitoring catheter and not to use anticoagulation. The incidence of thromboembolic events following low-risk right-sided procedures is low, whereas the risk of bleeding with the use of anticoagulation is not trivial. Therefore, it is reasonable to not use heparin for low-risk right-sided procedures unless there are other risk factors for thromboembolism.²⁸²

Anticoagulation using heparin is recommended for left-sided procedures. The optimal activated clotting time (ACT) can vary and is often dependent on the arrhythmia being treated. Guidelines for the optimal ACT for patients with AF or atrial flutter recommend that heparin doses be adjusted to achieve and maintain a target ACT of 300 to 400 seconds.²⁷⁸ Although there is little scientific evidence to guide the target level for ACT during ablation of ectopic atrial foci or APs located in the left atrium, the consensus of the writing group and the previously published guidelines²⁸¹ was that heparin should be administered to achieve a target ACT of 250 to 300 during these procedures. Heparin should be administered before or immediately after transseptal puncture to avoid thrombus formation on the sheath or the catheter.²⁸³ Although infusion of the transseptal sheath with a heparin solution can prevent thrombus formation at the end of the sheath, there are limited data, and this practice is variable among laboratories for low-risk shorter-duration procedures.

Sheath infusion is typically used for longer-duration left-sided procedures.

Antibiotic Prophylaxis. Routine antimicrobial prophylaxis administered before invasive procedures is not recommended; however, it is common practice to administer antibiotic prophylaxis to higher-risk patients during long procedures or when permanent indwelling device leads are present. In general, it is recommended that potential sources of dental sepsis be eliminated at least two weeks prior to an invasive cardiac procedure.²⁸⁴

Postprocedure Management

Consideration should be given to removing sheaths while the patient remains under sedation or anesthesia. Application of local anesthesia at the catheter insertion sites might help decrease post-procedure discomfort. Current guidelines suggest that if patients have received heparin during the procedure, an ACT of less than 180 should be achieved prior to pulling sheaths if no vascular closure device is being used²⁷⁷; however, many laboratories accept longer ACTs for sheath removal.

In general, patients undergoing an uncomplicated ablation procedure can be admitted and discharged the day of the procedure. The decision for inpatient observation versus discharge on the day of the procedure should take into account factors such as patient age, procedural details, potential for complications, and the level of care that can be provided locally for families that have traveled long distances. Preparation for overnight observation or discharge to home should be discussed prior to the day of the procedure. The postprocedure recovery area should be staffed by professionals who are experienced in the care of pediatric patients and who are trained or certified in pediatric advanced life support according to national or regional guidelines (PALS or equivalent). It is important for pediatric patients to have the family present as soon as possible in the recovery phase to reduce anxiety and distress. It is also important to minimize pain, nausea, and vomiting as much as possible.

The use of postprocedural anticoagulation is dependent on the clinical situation. After left-sided procedures, it is common practice to treat with aspirin for a period of time (typically 15 to 60 days). Although there was once concern that left-sided ablations will result in disruption of the left atrial endothelium and activate the acute phase coagulation cascade, results from the 1998 NASPE prospective catheter ablation registry concluded that there is no compelling reason to use warfarin or even aspirin after ablation.²⁸⁵ Accordingly, some pediatric and adult laboratories do not currently use anticoagulation after routine ablation procedures, even on the systemic arterial side. The lone exception is following ablation for AF: anticoagulation with warfarin or a novel oral anticoagulant is recommended for at least two months following AF ablation.²⁷⁸ There is no consensus for the use of sub-acute bacterial endocarditis (SBE) prophylaxis following an ablation procedure to allow for complete endothelialization of disrupted endocardium.

Follow-Up

Several issues pertinent to the pediatric population are important during the follow-up evaluation. Persistent groin tenderness should raise concern regarding arterial pseudoaneurysm or arteriovenous fistula, which can be diagnosed by ultrasound. Intracardiac valvular damage or coronary artery stenosis occurs infrequently,^{7,25,210} but should be considered in the presence of new exertional chest pain or a new murmur. An echocardiogram can be performed, depending on the clinical situation; however, the PAPCA follow-up data showed minimal significant findings from routine early or late postprocedure echocardiography.²⁵ The risk for recurrence of tachycardia is highest in the first two months following the procedure, but late recurrences even after 1 year following ablation have been reported.¹⁰ After an apparently successful catheter ablation, recurrence of palpitations is not uncommon and deserves a noninvasive evaluation. However, many such events are due to overreaction to autonomically mediated sinus tachycardia in a patient who is overly “cardiac aware.” In a recent survey of PACES members, most respondents perform an ECG in the postablation setting prior to discharge, prescribe aspirin for a period of one to three months following ablation, and routinely schedule a follow-up at two to three months and at one year. The initial visit following ablation for APs was earlier than for AVNRT. An ECG was obtained at the follow-up visits, and additional testing was dependent on the clinical condition.

3.7 Anesthesia and Sedation

Continuum of Sedation. Appropriate and adequate sedation or anesthesia is a prerequisite for performance of EP procedures in pediatric patients, especially in young children or those who have psychological or behavioral constraints. The aims are to improve patient comfort, reduce movement, and have minimal effect on the arrhythmia substrate. The continuum of depth of sedation and general anesthesia has been clearly delineated by the American Society of Anesthesiologists.²⁸⁶ For patient safety and comfort, ablation in almost all pediatric and congenital patients requires moderate to deep sedation or general anesthesia. As individual responses to the sedative effects of drugs are not predictable, practitioners need to be adequately qualified to “rescue” patients when a deeper than desired level of sedation is achieved. Particular consideration should be given to the immediate availability of qualified personnel with airway management skills.

Personnel. Current guidelines (Practice Guidelines for Sedation and Analgesia by Non-Anesthesiologists) provide basic recommendations for the safe provision of sedation or analgesia by nonanesthesiologists.²⁸⁶ The Heart Rhythm Society Expert Statement on Electrophysiology Laboratory Standards provides further recommendations pertinent to the Pediatric Electrophysiology Laboratory.²⁷⁶ These recommend a trained nurse credentialed in procedural sedation providing anxiolysis or mild to moderate sedation for

pediatric electrophysiology procedures in older children (older than 13 years) without confounding variables, under the supervision of the cardiac electrophysiologist, or the presence of a certified registered nurse anesthetist (CRNA) administering anesthesia or monitored anesthesia care under the supervision of an anesthesiologist for deep sedation or general anesthesia.²⁷⁶ Credentials should be periodically renewed and practitioners should maintain current certification in Advanced Cardiac Life Support (ACLS) and PALS, as appropriate (or their country/regional equivalent).

Monitoring. All patients requiring sedation or anesthesia for EP studies should undergo basic monitoring of cardiorespiratory parameters based on the ASA standards for basic monitoring. This process should include continuous monitoring of oxygenation (peripheral oxygen saturation SPO₂), visualization of patient color, ventilation assessment (chest excursion and/or ETco₂), continuous ECG, noninvasive arterial blood pressure measurement every five minutes, and body temperature. Because these procedures can be prolonged, especially in children or adults with CHD, monitoring patency of airway, depth of sedation, adequacy of ventilation, and oxygenation is particularly important.²⁸⁶ For prolonged procedures, a urinary catheter is recommended. It is critical to have immediate access to emergency equipment, including basic and advanced airway management equipment, vascular access devices, resuscitative medications, inotropic agents, antiarrhythmic drugs, pharmacologic antagonists, and a defibrillator.²⁸⁶ The depth of sedation should be continuously monitored using any of several validated sedation scales (modified Ramsay scales).

Anesthesia vs Sedation. To help assure both patient safety and comfort, in many pediatric electrophysiology laboratories general anesthesia with endotracheal intubation is the methodology of choice for almost all ablation procedures. It is particularly recommended for a variety of patient or procedural characteristics, including but not limited to the following: age 12 years and younger; significant CHD; systemic ventricular dysfunction; myocardial ischemia; pulmonary hypertension; hemodynamic instability; respiratory comorbidities; other significant systemic comorbidities; anticipation of a prolonged procedure; the need for complete immobility (e.g., a procedure near the coronary orifices or AV conduction system, percutaneous epicardial approach); and patient or parent choice.

Conscious and Deep Sedation. Conscious sedation can be used in some adolescents and young adults. A cooperative and motivated patient will be able to undergo the length of the EP procedure with incremental dosing of midazolam and fentanyl. Further incremental intravenous procedural sedation should be based on institutional sedation guidelines, provided by credentialed nurses, using narcotics, benzodiazepines, barbiturates, ketamine, and propofol. Vascular access in the groin or the neck should be obtained after providing adequate local anesthetic infiltration. A popular combination includes buffered procaine 2% and bupivacaine 0.25%.

Monitored Anesthesia Care (MAC) and General Anesthesia. The need for endotracheal intubation or laryngeal

mask anesthesia depends on the complexity of the procedure, the need for holding ventilation, or when venous access is achieved through the neck. Disadvantages of intubation are the occurrence of a sore throat after the procedure, the physiologic perturbations of emergence, and the potential for bleeding from the vascular access sites from straining during emergence. Considerations for the selection of technique or drugs include but are not limited to arrhythmia mechanism, intrinsic autonomic tone, and coexisting heart disease. Based on the complexity and duration of the ablation procedure, the child's clinical status, co-morbidities, and hemodynamic or respiratory instability, postprocedural recovery can occur in a recovery unit, general cardiology floor, or the intensive care unit. Continuous telemetry and regular monitoring of hemodynamic, oxygenation, and ventilation parameters should be performed. Staff in this area should be trained in the postprocedural and postsedation care of pediatric EP patients. Discharge criteria include the return of baseline mental status, stable and acceptable vital signs, an absence of relevant complications, the ability to tolerate oral intake, demonstration of spontaneous urination and adequate pain control. Patients should be discharged in the presence of an adult caregiver with clear written instructions, including postprocedure diet, medications, activities, and an emergency contact number.

Electrophysiologic Considerations. Commonly used anesthetic agents and sedatives manifest effects on the electrophysiology of the heart through interactions with receptors and ion channels in the myocardium and conduction tissue, and have been reviewed in detail elsewhere.²⁸⁷ Sedative and anesthetic drugs and agents can affect automaticity and triggered activity. They can also affect intrinsic sympatho-vagal tone. These factors must be balanced against the anesthetic characteristics to provide the optimal approach. Automatic tachyarrhythmias and "normal heart VT" are particularly vulnerable to depth of sedation and autonomic nervous system manipulation. Agents least likely to have significant autonomic or EP effects include opioids, benzodiazepines, volatile inhalational agents, nitrous oxide, and propofol. At high doses, opioids have vagotonic effects with attendant sinus bradycardia. All inhalational agents, even the popular sevoflurane, can modestly increase the QTc interval. They also notoriously enhance automaticity in ectopic atrial pacemakers. Ketamine has inhibitory effects on norepinephrine reuptake, resulting in modest sympathomimetic effects. The nondepolarizing muscle relaxing agent, pancuronium, results in a brief period of sinus tachycardia related to its weak vagolytic property and norepinephrine-releasing effect. Of these agents, rocuronium is most free of autonomic nervous system effects. A number of agents should be specifically avoided unless prior experience in the patient has demonstrated minimal effects on the target arrhythmia. Dexmedetomidine, a centrally acting alpha₂ agonist, suppresses sinoatrial and AV node conduction and automaticity. Phenylephrine, frequently used to support systemic blood pressure, can result in significant vagotonia due to baroreceptor unloading. Reversible acetylcholinesterase inhibitors, such as neostigmine, are used to reverse the

effect of nondepolarizing muscle relaxing drugs, and have parasympathomimetic effects.

4. Recommendations—General Safety and Procedures

Based on the discussions above of the prior data from registries, arrhythmia backgrounds and outcomes, and the special issues of ablation in infants and patients with moderate or complex CHD, the safety of cryotherapy, radiation exposure, laboratory standards, and sedation/analgesia, the following guidelines were established by the committee.

4.1 Safety Recommendations

Class I

1. Pediatric cardiovascular surgical support should be available in-house during ablation procedures for smaller patients* (LOE: E).
2. There should be a pediatric cardiovascular surgical program at the same institution where the ablation is performed for patients 12 years of age and younger (LOE: E).^{\$}
3. There should be a congenital cardiovascular surgical program at the same institution where the ablation is performed for patients with moderate or complex CHD (LOE: E).^{\$}
4. There should be a cardiovascular surgical program at the same institution where the ablation is performed for patients 13 to 18 years of age without moderate or complex CHD (LOE: C).^{\$}
5. For patients 12 years of age and younger and/or with moderate or complex CHD (Table 2), the procedure should be performed in collaboration with an anesthesiologist who has expertise with pediatric and/or CHD patients (LOE: C).
6. Fluoroscopy use should be as low as reasonably achievable (ALARA) (see Table 6 in Fazel et al²⁶³) for all ablation procedures in pediatric and congenital cases (LOE: B).
7. When the ablation procedure will take place in the systemic arterial atrium or ventricle, or there is a known or potential right-to-left shunt, the risk of systemic embolization should be managed with systemic anticoagulation with unfractionated heparin (or an alternative in patients with heparin-induced thrombocytopenia)²⁸¹ (LOE: B).
8. When ablation takes place in proximity to a phrenic nerve, maneuvers should be undertaken to identify the location of the phrenic nerve and avoid injury, if possible (LOE: C).

Class IIa

1. It is useful to have cryotherapy available for septal substrates and proximity to smaller coronary arteries,

and to discuss the safety and efficacy of the alternative approaches of RF energy and cryotherapy (LOE: A).

Class IIb

1. When ablation is indicated for smaller children*, cryoablation can be useful to avoid the higher risks of complications from RF ablation (LOE: C).

Class III

1. Ablation is not recommended for patients with an intracardiac thrombus identified by transthoracic or transesophageal echocardiography, which is relevant to an anticipated catheter course (LOE: C).

*There is no specific recommendation that the service be in-house rather than otherwise on-call for emergencies, as per regulatory and institutional standards.

4.2 Procedural Recommendations

Class I

1. The electrophysiology laboratory and postprocedure recovery unit should be suitable for the care of pediatric and CHD patients, as indicated²⁷⁶ (LOE: C).
2. Catheter-based ablation procedures for patients with moderate or complex CHD (Table 2) or complex arrhythmias should be performed by a cardiac electrophysiologist with the appropriate expertise, and in a laboratory with appropriate personnel and equipment, for the relevant patient population (age and/or CHD) (LOE: C).
3. Preprocedural planning for patients with CHD should include a detailed review of operative notes pertaining to all previous cardiac and vascular surgeries; imaging studies assessing cardiac anatomy and recent cardiac function; vascular and intracardiac access issues; prior interventions; and all documented arrhythmias. (LOE: C).
4. A 3D nonfluoroscopic, electroanatomic mapping system should be available and strongly considered for guiding or assisting mapping and ablation of postoperative tachyarrhythmias in patients with moderate or complex CHD (LOE: B).

Class IIa

1. Participation in Quality Assurance activities, including an ablation quality outcomes registry, is reasonable and can be useful (LOE: B).
2. Irrigated or large electrode-tip RF catheters can be useful for the ablation of postoperative tachyarrhythmias in patients with CHD (LOE: B).
3. Nonfluoroscopic imaging can be useful as a primary tool or as an adjunct to fluoroscopy during ablation procedures for all pediatric patients to reduce radiation exposure in accordance with the ALARA principle and to provide

electroanatomic mapping for more complex procedures (LOE: B).

4. Cryoablation can be useful for slow pathway modification in pediatric patients with AVNRT (LOE: B); any substrate in close proximity to the AV conduction system; when catheter stability is a barrier to successful ablation (LOE: C); or where low blood flow limits the utility of RF ablation (LOE: C).
5. Catheter-based hemodynamic evaluation can be useful prior to ablation for patients with repaired moderate or complex CHD (Table 2) who are being considered for catheter ablation (LOE: C).

Class IIb

1. Integration of anatomic imaging using CT or MRI with 3D electroanatomic mapping can be considered for guiding the ablation of tachyarrhythmias in patients with complex CHD (LOE: B).
2. A remote magnetic navigation system might be reasonable for catheter ablation of tachyarrhythmias in complex CHD with difficult intracardiac anatomy or vascular access (LOE: B).

5. Summary

In the twelve years since publication of the last ablation guidelines for children and for all patients with CHD, advancements in imaging technologies and ablation energy sources have dominated the field and disproportionately influenced clinical practice. The general availability of EAM systems has led to a reduction in reliance on ionizing radiation for catheter manipulation and has helped refine the identification of arrhythmia substrates in abnormal anatomies. More robust use of higher-energy RF sources now results in greater ablation efficacy in patients with CHD; and, conversely, ablation using cryoenergy permits safer ablation of substrates in the most vulnerable parts of the heart. Together, these experiences have helped inform the recommendations in this document. In the coming years, it is expected that recently initiated registry-based projects will enable benchmarking of outcomes that will inspire the next iteration of guidelines.

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Appendix 1 Author Disclosure Table

| Writing Group Member | Institution | Consultant/ Advisory Board/ Honoraria | Speakers' Bureau | Research Grant | Fellowship Support | Stock Options/ Partner | Board Mbs/ Other |
|--|---|---|------------------|----------------|--------------------|------------------------|--|
| J. Philip Saul, MD, FHRS, FACC, FAHA, FAAP | JP Saul Consultingm, Mount Pleasant, SC | 1; Novartis Pharmaceuticals | None | None | None | None | None |
| Ronald J. Kanter, MD, FHRS, FACC | Miami Childrens Hospital, Coral Gables, FL | None | None | None | None | None | None |
| Dominic Abrams, MD | Royal Brompton & Harefield NHS Trust, London, UK | None | None | None | None | None | Equity Interests: 1; Johnson and Johnson |
| Samuel Asirvatham, MD | Mayo Clinic, Rochester, MN | 1; Biotronik, Abiomed; Boston Scientific Corp; Medtronic, Inc.; Sanofi Aventis; St. Jude Medical; Stereotaxis, Inc.; Spectranetics Corporation; Mediasphere Medical; Biosense Webster, Inc.; AtriCure, Inc.; Wolters Kluver; Elsevier | None | None | None | None | None |
| Yaniv Bar-Cohen, MD, FAAP, FACC | Children's Hospital of Los Angeles, Los Angeles, CA | None | None | None | None | None | None |
| Andrew D. Blaurock, MD, FHRS, FACC | Alexandra & Steven Cohen Children's Medical Center of NY, New Hyde Park, NY | 1; Boehringer Ingelheim | None | None | None | None | None |
| Bryan Cannon, MD | Mayo School of Graduate Medical Education, Rochester, MN | 1; St. Jude Medical; Medtronic, Inc. | None | None | None | None | None |
| John Clark, MD | Akron Children's Hospital, Akron, OH | 1; St. Jude Medical | None | None | None | None | None |
| Macdonald Dick, MD, FHRS, FACC | Univ of Michigan Health Systems, Ann Arbor, MI | 1; Pfizer, Inc. | None | None | None | None | None |
| Anne Freter, MSN, FHRS | Advocate Medical Group, Wheaton, IL | None | None | None | None | None | None |
| | | 1; Medtronic, Inc. | None | None | None | None | None |

Appendix 1 (continued)

| Writing Group Member | Institution | Consultant/ Advisory Board/ Honoraria | Speakers' Bureau | Research Grant | Fellowship Support | Stock Options/ Partner | Board Mbs/ Other |
|---|---|--|------------------|--|--------------------------------------|------------------------|------------------|
| Naomi J. Kertesz, MD, FACC, FHRS | Nationwide Children's Hospital, Columbus, OH | | | | | | |
| Joel A. Kirsh, MD, FRCP, FHRS, CEP | Hospital for Sick Children, Toronto, ON | None | None | None | None | None | None |
| John Kugler, MD, FAAC, FAAP, FAHA | UNMC/CU Children's Hospital, Omaha, NE | None | None | None | None | None | None |
| Martin LaPage, MD, MS, FAAP, FHRS, CCDS, CEPS | University of Michigan, Ann Arbor, MI | None | None | None | None | None | None |
| Francis X. McGowan, MD, FAAP | The Children's Hospital of Philadelphia, Philadelphia, PA | | | | | | |
| Christina Y. Miyake, MD | Stanford University, Redwood City, CA | None | None | None | None | None | None |
| Aruna Nathan, MBBS, FRCA | The Children's Hospital of Philadelphia, Philadelphia, PA | | | | | | |
| John Papagiannis, MD, FHRS, CEPS | Children's Mercy Hospital, Kansas City, MO | None | None | None | None | None | None |
| Thomas Paul, MD, FHRS, FACC | Georg-August-Univ Medical Univ Hospital, Gottingen, Germany | None | None | None | None | None | None |
| Andreas Pflaumer, MD, FRACP, FCSANZ, CEPS | Royal Children's Hospital Melbourne, AU | None | None | None | 2; St. Jude Medical; Medtronic, Inc. | None | None |
| Allan C. Skanes, MD, FRCPC | London Health Sciences Center, London, ON | 1; Boehringer Ingelheim; Biosense Webster, Inc.; Medtronic, Inc. | None | 2; Boehringer Ingelheim; 3; Biosense Webster, Inc. | 5; Medtronic, Inc. | None | None |
| | | None | None | None | None | None | |

Appendix 1 (continued)

| Writing Group Member | Institution | Consultant/ Advisory Board/ Honoraria | Speakers' Bureau | Research Grant | Fellowship Support | Stock Options/ Partner | Board Mbs/ Other |
|--|---|---------------------------------------|------------------|----------------|--------------------|------------------------|---|
| William G. Stevenson, MD, FAHA, FHRS, FACC | Brigham and Women's Hospital, Boston, MA | | | | | | Officer, trustee, director, or other fiduciary role: 3; American Heart Association; Intellectual Property Rights: Brigham and Women's; Biosense Webster, Inc. |
| Nicholas Von Bergen, MD | The Children's Hospital of Iowa, Iowa City, IA | None | None | None | None | None | None |
| Frank Zimmerman, MD, FAHA, FHRS | The Heart Institute for Children, Western Springs, IL | None | None | None | None | None | None |

Number Value: 0 = \$0; 1 = < \$10,000; 2 = > \$10,000 to < \$25,000; 3 = > \$25,000 to < \$50,000; 4 = > \$50,000 to < \$100,000; 5 = > \$100,000

Appendix 2 Peer-Reviewers Disclosure Table

| Peer Reviewer | Institution | Consultant/ Advisory Board/ Honoraria | Speakers' Bureau | Research Grant | Fellowship Support | Stock Options/ Partner | Board Mbs/ Other |
|---------------------|--|---|----------------------------|---|--------------------------------------|------------------------|------------------|
| James Perry | UCSD Rady Children's Hospital, San Diego, CA | 1; Medical Legal Consulting | None | None | None | None | None |
| Susan Etheridge | University of Utah, Department of Pediatric Cardiology, Salt Lake City, UT | None | None | None | None | None | None |
| Dipen Shah | Hopital Cantonal De Geneve, Thonex, Switzerland | 1; Biosense Webster, Inc. Biotronik; 2; Endosense | 1; Endosense; 2; Endosense | 3; Bard Electrophysiology; Biotronik; St. Jude Medical; 4; Biosense Webster, Inc. | None | 1; Endosense | None |
| Charles Berul | Children's National Medical Center, Washington DC | None | None | 4; Medtronic, Inc. | None | None | None |
| Barbara Deal | Children's Hospital of Chicago | None | None | None | None | None | None |
| Andrew M. Davis | The Royal Children's Hospital, Melbourne | None | None | None | 3; Medtronic, Inc.; St. Jude Medical | None | None |
| Jonathan Skinner | Starship Hospital, Paediatric Cardiology, Auckland, NZ | 0; Medtronic, Inc. | None | None | None | None | None |
| Louise Harris | University of Toronto, Peter Munk Cardiac Centre, Toronto, ON | 1; St. Jude Medical | None | None | None | None | None |
| Juha-Matti Happonen | Helsinki University Central Hospital, Helsinki, Uusimaa, Finland | 1; St. Jude Medical | None | None | None | None | None |
| Fabrizio Drago | Bambino Gesù Children's Hospital, Rome, Italy | None | None | None | None | None | None |
| Thomas Kriebel | Georg-August-University Göttingen, Göttingen, Germany | 1; AOP Orphan | None | None | None | None | None |
| Laszlo Kornyei | Gottsegen Gyorgy Hungarian Institute of Cardiology, Budapest, Hungary | None | None | None | None | None | None |

Number Value: 0 = \$0; 1 = < \$10,000; 2 = > \$10,000 to < \$25,000; 3 = > \$25,000 to < \$50,000; 4 = > \$50,000 to < \$100,000; 5 = > \$100,000