

PACES/HRS Expert Consensus Statement on the Evaluation and Management of Ventricular Arrhythmias in the Child With a Structurally Normal Heart



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Preamble

The purpose of this consensus statement is to provide up-to-date recommendations on the evaluation and treatment of

ventricular tachycardia (VT) in children with structurally normal hearts (idiopathic VT [IVT]). IVT is usually benign and often resolves spontaneously without treatment; however, it is essential to distinguish this problem from potentially life-threatening conditions that can occur with absent or minimal structural heart disease (long QT syndrome [LQTS], arrhythmogenic right ventricular dysplasia/cardiomyopathy [ARVC], myocarditis, and cardiac tumors). As concluded in a recent review, because of the rare nature of this condition, the often small case series that describes it, and confusion with potentially lethal mimicking disease processes, "Currently no standard diagnostic approach exists, and management is heterogeneous."¹

The Pediatric and Congenital Electrophysiology Society (PACES) in conjunction with the Heart Rhythm Society (HRS) formed a writing committee to address this lack and propose expert consensus guidelines. Selected members from within PACES and HRS have reviewed and analyzed the published scientific literature, carefully assessing the absolute and relative risks of diagnostic and therapeutic procedures so as to provide a practical approach to optimize patient care. This consensus statement is directed at all health care professionals who treat young patients with idiopathic monomorphic VT, broadly considered to include premature ventricular complexes (PVCs), nonsustained VT, and sustained VT. Accelerated idioventricular rhythm will also be discussed in this document. Polymorphic VT, as seen in LQTS and catecholaminergic polymorphic VT (CPVT), is thoroughly discussed in the consensus document with regard to patients with arrhythmias secondary to genetic ion channelopathies and thus will be discussed only briefly.² For the purposes of this document, we are referring to patients from the neonatal period through adolescence, up to 18 years of age, who would be cared for primarily by pediatricians and pediatric cardiologists. We use the terms *infants* for those younger than 3 years, including toddlers in this group because of similar issues of ability to cooperate with tests, and recommendations against ablation; and *children* for those who are 3–18 years old. This is a diverse group in terms of symptoms, signs, and ability to endure various diagnostic and therapeutic options; age-related distinctions in care will be discussed. This document is to be considered as expert consensus-based guidance. A specific care plan for a particular patient must be made by the health care provider, the patient, and his or her parents after careful consideration and a thorough discussion of patient characteristics that impact risks and benefits.

Methods and evidence

For the purposes of this document, we defined *consensus* as 75% or greater agreement by the writing members. Writing

committee members were selected by PACES or HRS on the basis of their expertise in the field. The 11 pediatric electrophysiologists and 2 adult electrophysiologists on the writing committee were tasked with performing a formal literature review and then weighing the strength of the evidence on various aspects of diagnosis and treatment of young patients with IVT. It is acknowledged that the published evidence for most of the recommendations made herein is limited, but the depth of knowledge and experience of the writing group is believed to provide justification for consensus recommendations based on expert opinion. In some situations, the writing committee had difficulty arriving at consensus, largely owing to the lack of sufficient evidence and/or experience. In some situations, recommendations were based partially on additional data from the treatment of adult patients with similar disorders, as the number of pediatric patients treated for some of these conditions is insufficient to make definitive judgments as to the efficacy of treatment. In other situations, such as the utility of exercise testing in the evaluation of ventricular arrhythmias, a consensus on recommendations for specific clinical situations could not be reached. It was also difficult to establish complete consensus on the relative benefits of antiarrhythmic drug therapy versus ablation in patients who require treatment for ventricular arrhythmias. Writing committee members felt that this decision tree held multiple acceptable options and critically depends on local expertise, which is variable in pediatric centers, particularly in non–right ventricular outflow tract ventricular arrhythmias.

The framework for the construction of the recommendations depended on the following variables: (1) age of the patient, with regard to the ability to comply with diagnostic procedures and risk of therapy, expressed in the document by the distinction between infants and children; (2) severity of symptoms; (3) presence of ventricular dysfunction suspected to be caused by frequent ectopy; and (4) type of ventricular arrhythmia (e.g., uniform vs polymorphic, frequent ectopy vs sustained VT, and right ventricular outflow tract vs other sites). The committee was divided into subgroups to best review key aspects of the evaluation and management of IVT. These sections included detailed reviews and assessments of the following topics: (1) overview of the condition, (2) clinical presentations, (3) evaluation and exclusion of more dangerous conditions, and (4) therapeutic options. All committee members have reviewed the entire document and have agreed with its contents by consensus vote as described above.

The committee reviewed, ranked evidence, and made recommendations based on the standard process previously described in the *Methodology Manual and Policies from the American College of Cardiology and American Heart Association Task Force on Practice Guidelines* June 2010; these are summarized below.

A. Classification of Recommendations

- Class I: There is evidence and/or general agreement that a procedure or treatment plan is beneficial, useful, and effective (“is recommended”)
- Class II: There is conflicting evidence and/or divergence of opinion about the usefulness and/or efficacy of the procedure or treatment plan
 - Class IIa: Weight of evidence/opinion is in favor of usefulness/efficacy (“may be useful”)
 - Class IIb: Weight of evidence/opinion is less well established by evidence/opinion (“may be reasonable”)
- Class III: There is conflicting evidence and/or general agreement that a procedure or treatment plan is not useful/effective, and in some cases may be harmful (“not recommended”)

B. Level of evidence

- Level of evidence A: Data available from multiple randomized clinical trials or meta-analyses
- Level of evidence B: Data available from a single randomized trial or nonrandomized trials
- Level of evidence C: Only consensus opinion of experts, case studies, or standard of care

Document review and approval

This document was reviewed by the PACES executive committee and through the HRS review process. All writing members approved the final version. The writing committee thanks all reviewers for their suggestions and sponsoring organizations for their support. Author and reviewer disclosures are given in [Appendices 1](#) and [2](#), respectively.

1. Introduction

There have been multiple small series describing sustained VT in the pediatric patient with a normal heart, but there are very limited data regarding the incidence of this entity in the general population.^{3–7} In a school-based heart disease screening program in Japan, the incidence of nonsustained or sustained VT was estimated to be between 0.2 and 0.8 per 10,000 children.⁸ The majority of these (54%) disappeared on follow-up. Roggen et al⁹ found a sustained VT frequency of 1.1 episodes per 100,000 children when studying a single center over a 10-year period. Half of these patients had structural heart disease, while the rest were divided between neonatal idiopathic left and idiopathic right VTs, with the majority (53%) originating from the right ventricle. Mortality occurred only in patients with underlying heart disease. Thus, VT in the pediatric patient with a structurally normal heart is rare and carries a good prognosis.

2. Clinical presentations

There are a number of possible presentations of the child with idiopathic ventricular arrhythmias, ranging from infrequent ectopy to incessant VT; these will be considered individually.

2.1. Ventricular ectopy

PVCs are frequent in neonates, infants, and children. When these are rare and isolated, they rarely need further evaluation. However, when ectopy becomes more frequent, herein defined as more than 10% of beats in a 24-hour period, it should be followed longitudinally. The initial decision as to when to

proceed with evaluation, such as echocardiography and ambulatory monitoring, to define the 10% burden is made when the clinician recognizes multiple PVCs during electrocardiography (ECG) or frequent ectopy during physical examination, with ECG confirmation of PVCs as the etiology. The choice of 10% ectopy as a definition of “frequent” is acknowledged as being lower than that commonly associated with ventricular dysfunction but seems a reasonable cutoff for monitoring purposes given the day-to-day variability in frequency.

The prevalence of PVCs in healthy children varies with age. Nearly 20% of the neonates have uncomplicated ventricular ectopy consisting of uniform PVCs or couplets.¹⁰ This decreases to 10% of toddlers and school-age children and increases to 20%–30% of the normal adolescents.^{11–14} The ectopy burden and grade are important.¹³ In otherwise normal adolescent boys, although some ventricular ectopy is common, less than 5% will have more than 50 beats per 24 hours and less than 2% will have multiform PVCs, couplets, or nonsustained VT on 24-hour monitoring.^{11,12,15}

The origin of the PVCs and the response to exercise should be analyzed. Some reports suggest that the suppression of PVCs with exercise indicates a more benign condition, but suppression with exercise is so common that it is difficult to use this criterion diagnostically.¹⁶ There is evidence that PVCs that originate from the left ventricle (right bundle branch block [RBBB] morphology) are more likely to regress overtime.¹⁶ PVCs that originate from the right ventricular outflow tract (RVOT) are typically benign. However, they may be an early presentation of ARVC. Thus, when PVC burden exceeds age-based normal ranges, it is important to evaluate patients for possible underlying pathology. The new task force for ARVC diagnosis lowered the criteria for ectopy to 500 ectopic beats in a 24-hour period in patients who have other features concerning for ARVC.¹⁷

Ventricular ectopy may present as isolated beats or as nonsustained VT. The rate of nonsustained VT may be an important characteristic, as will be discussed in the following section. The asymptomatic patient with frequent ectopy should be monitored for decline in cardiac function. On rare

occasions, if the burden of ventricular beats is substantial, children can develop cardiac dysfunction, likely related to dyssynchrony. There is very little data on this in children, but studies in adults suggest a burden of at least 10% ectopy, and generally 20%–30% is needed to increase the risk of ventricular dysfunction.^{4,18,19}

Complex ventricular ectopy has been defined as bigeminy, multiform ectopy, couplets, or nonsustained VT.²⁰ These arrhythmias may identify a subset of patients in populations with cardiovascular disease who are at increased risk of death and sudden cardiac death.²¹ The longitudinal data of Biffi et al,²² looking at the largest prospectively studied cohort of apparently healthy persons (all trained athletes) identified with frequent and/or complex ventricular ectopy, confirm that the presence of frequent and/or complex ventricular ectopy does not confer an ominous prognosis in the absence of structural heart disease. A careful evaluation of such patients is warranted, as several patients with cardiomyopathy were identified during evaluation for ectopy, and the presence of bidirectional ventricular ectopy or tachycardia can indicate a channelopathic condition. A detailed family history for sudden death or aborted sudden death should be sought and patients assessed for the clinical features of CPVT, Andersen-Tawil syndrome, and other channelopathies.²³

2.2. Accelerated idioventricular rhythm

In adults, numerical criteria differentiate VT from accelerated idioventricular rhythm, but in children, the age-based variability of sinus rhythm precludes purely numerical classification; however, it is the relationship to the expected sinus rate that is used. Ventricular escape rhythms are defined as slower than sinus rhythm; idioventricular rhythms are similar to sinus rhythm; and accelerated idioventricular rhythms are slightly faster than sinus rhythm, defined as within 10% of the underlying sinus rate. VT is defined as being at least 10%–15% faster than the expected sinus rate or greater than 120 beats/min in older teens and young adults at rest.²⁴ Accelerated idioventricular rhythm appears to be due to enhanced automaticity of myocardium or His-Purkinje fibers. Although typically benign, in the setting of metabolic disarray, ischemia or myocardial disease may be a harbinger of more malignant ventricular arrhythmias.

Accelerated idioventricular rhythm of the newborn may present in the first hours of life, typically discovered because of a slightly irregular rhythm, or in a child who is being monitored for another reason. In the asymptomatic infant, once structural heart disease has been ruled out, as have temperature and metabolic and electrolyte abnormalities, this is considered a benign phenomenon. Newborns with benign accelerated idioventricular rhythm do not require treatment, but should be observed longitudinally to ensure they remain asymptomatic and the rhythm resolves as anticipated, usually within the first year of life.^{25,26}

Idioventricular rhythm, accelerated or not, can be seen in older children as well and appears to be similarly quite

benign and generally self-resolving.²⁷ The asymptomatic patient should be monitored for potential decline in cardiac function, as in frequent ventricular ectopy. If the burden of ventricular beats is substantial, on rare occasions children can develop cardiac dysfunction, which may be dyssynchrony induced. Ablation of the ventricular focus may be indicated in these cases to restore cardiac synchrony and has been demonstrated to restore normal cardiac function.²⁸

2.3. Monomorphic ventricular tachycardias

2.3.1. Right ventricular outflow tract tachycardia

Definition. RVOT tachycardia is one of the most common ventricular arrhythmias seen in young patients, accounting for 60%–80% of all IVTs.²⁹ The tachycardia originates from an area cephalad to the tricuspid valve and caudal to the pulmonary valve, most commonly from the posteroseptal region or the right ventricular free wall just below the pulmonary valve. Less commonly, tachycardia foci can originate from sites above the pulmonary valve or near the bundle of His.³⁰ Similar morphologies can be seen from VT originating from the left ventricular outflow tract (LVOT) or aortic cusps. The tachycardia is monomorphic with left bundle branch block (LBBB) QRS morphology and an inferior axis (Figure 1). There is often a late transition ($>V_3$) in the precordial leads.³¹

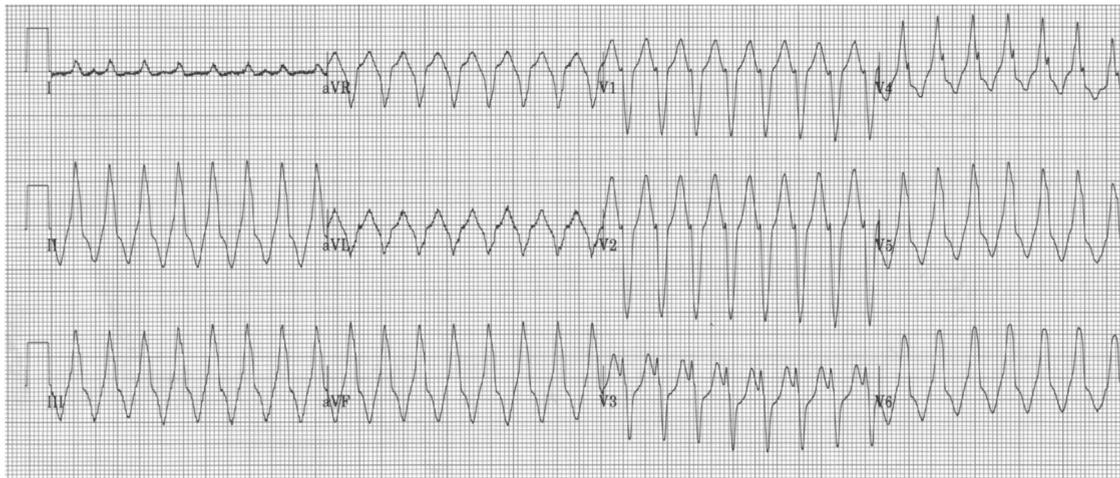
Mechanism. The most common mechanism of RVOT tachycardia is triggered automaticity due to cyclic adenosine monophosphate (AMP)–mediated activity.^{32,33} Thus, these tachycardias are responsive to adenosine, calcium-channel blockers, and β -blockers and often respond to maneuvers that decrease cyclic AMP levels, including vagal maneuvers. Rarely, RVOT tachycardia may be due to automaticity or reentry that can be either sensitive or nonsensitive to verapamil.³⁴

Clinical characteristics. Two clinical variants of RVOT tachycardia have been described and may be a spectrum of the same disease. The most common variant consists of frequent PVCs or nonsustained monomorphic VT occurring at rest or in the recovery period after exercise.^{35–37} The amount of ventricular ectopy usually decreases during exercise. The less common variant manifests as longer runs of monomorphic VT triggered by exercise or stress.³⁸

The typical mean age at presentation is 8 years, with rare forms of tachycardia occurring in infancy.³ RVOT tachycardia occurs more commonly in women, and there is an increased occurrence of this arrhythmia associated with the menstrual cycle.^{39,40}

Symptoms of palpitations or near-syncope occur in approximately 50%–67% of the patients.^{3,41} Syncope is uncommon and should raise the suspicion of an alternative diagnosis or an associated cardiomyopathy.⁴¹ RVOT tachycardia can be reproduced approximately 25%–68% of the time during exercise stress testing.^{3,34}

While RVOT tachycardia usually occurs in an otherwise normal heart, there have been reports of structural abnormalities in the RVOT detected by computed tomography or



Important ECG characteristics of this site of origin are brisk upstrokes of the initial 40 msec of the QRS (signifying normal myocardium), left bundle branch morphology and a strongly positive inferior axis. Lead I is positive in this example, but may also be negative.

Figure 1 A 12-lead electrocardiogram recorded during sustained idiopathic ventricular tachycardia arising from the right ventricular outflow tract. Important electrocardiographic characteristics of this site of origin are brisk upstrokes of the initial 40 ms of QRS morphology (signifying normal myocardium), left bundle branch morphology, and a strongly positive inferior axis. Lead I is positive in this example, but may also be negative.

magnetic resonance imaging (MRI).^{42–44} These include focal thinning of the right ventricular wall, segmental abnormalities, and fatty infiltration in up to 25% of those studied. The significance of these findings must be interpreted in the context of rigorous criteria for the diagnosis of ARVC, as in the past, many patients with these minor abnormalities have been inappropriately diagnosed with ARVC.¹⁷

The differential diagnosis of RVOT tachycardia includes myocarditis, tumors, CPVT, ARVC, Uhl anomaly, or coronary artery disease.^{45,46} One report has shown that up to 14% of these tachycardias may be associated with the finding of focal myocarditis.⁴⁷ The diagnosis of ARVC is important to consider and may warrant further evaluation with signal-averaged ECG (SAECG), echocardiography, MRI, and/or right ventricular angiography. This is particularly true in patients with exertional syncope, a high burden of PVCs, an abnormal baseline ECG, or a worrisome family history of sudden death in the young. Unlike RVOT tachycardia, VT associated with ARVC is usually due to reentry and is not typically responsive to adenosine or vagal maneuvers.⁴⁸ Furthermore, ventricular arrhythmias associated with ARVC may have multiple QRS morphologies, indicating a more diffuse disease of the right ventricle.⁴⁹ Occasionally, VT with LBBB QRS morphology and an inferior axis may arise from the LVOT.^{39,50} LVOT tachycardias may originate from the aortic root, aortic-mitral valve continuity, superior base of the left ventricular septum, mitral valve annulus, or the epicardial surface along the course of the coronary vein. The mechanism of these tachycardias appears to be due to cyclic AMP-mediated triggered automaticity, similar to RVOT tachycardias, although a majority of these may be due to automaticity in younger patients.³⁴ Aortic cusp tachycardias arise from the right coronary cusp more often than the left and rarely from the noncoronary cusp (Figure 2).^{51,52} Tachycardias arising from

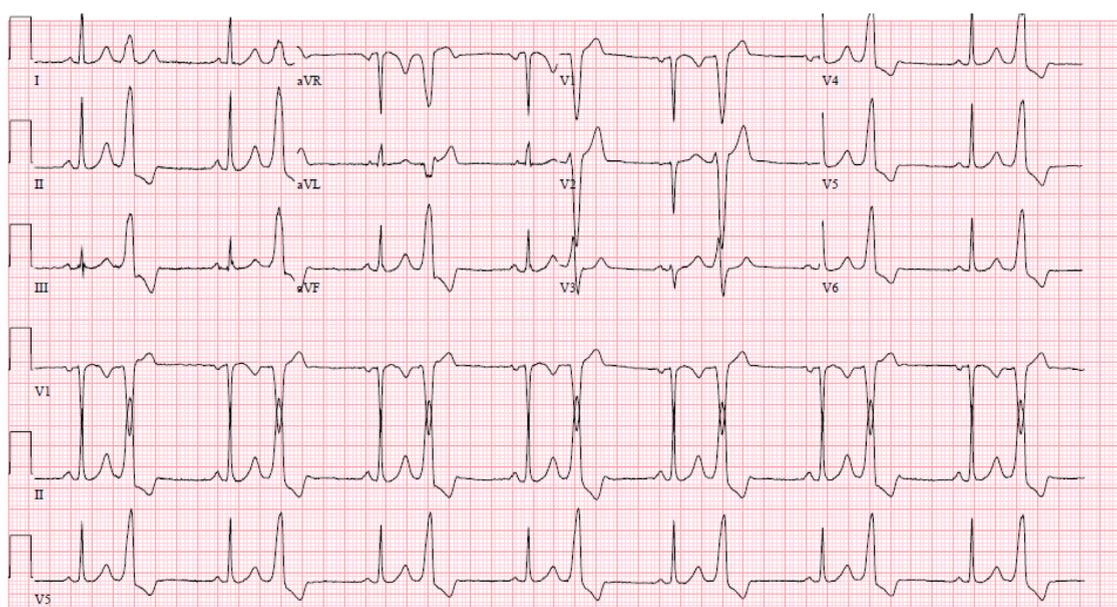
the aortic-mitral valve continuity can be distinguished by RBBB QRS morphology, while epicardial VTs usually have a characteristic slurred late peaking QRS complex.^{53,54}

VTs with LBBB QRS morphology and superior axis are less common in young patients than in adults. Focal arrhythmias arising from the body of the right ventricle, including the anterior free wall and the tricuspid valve annulus, have been described in patients with structurally normal hearts.^{38,55} The mechanisms of these tachycardias have been attributed to enhanced automaticity.⁵⁵ Some studies have suggested that tachycardias with LBBB QRS morphology and superior axis are more often associated with occult structural right ventricular abnormalities, and thus a detailed evaluation for ARVC should be pursued.^{56,57}

Natural history. The natural history of RVOT tachycardia is usually benign with a good long-term prognosis. Spontaneous remission may occur in 5%–65% of the patients, while the arrhythmia burden usually declines over time in those that persist.^{1,3,5,8,27,58–60} The wide range of reported remission rates appear to be related to the age at presentation, with virtually all infants having spontaneous resolution and older children more likely to have persistence. There are rare reported cases of sudden death with possible idiopathic RVOT tachycardia. All these reports precede our understanding of ARVC and of more precise tachycardia mechanisms; an analysis of these papers suggest that these cases were most likely related to unrecognized cardiomyopathy.^{7,61–64} Recent studies have shown no mortality with up to 80-month follow-up in children with structurally normal hearts.^{3,27}

2.3.2. Incessant ventricular tachycardia in infancy

Definition. This rare form of VT has been described to occur in infancy. It is usually monomorphic and most commonly arises from the left ventricle.



Idiopathic ventricular arrhythmias can also arise from the left ventricular outflow tract. The most common sites are the right and left coronary cusps. Compared to the RV outflow tract, the LV outflow tract at the level of the coronary cusps is posterior and slightly inferior. The major effect of this VT location on the ECG is an earlier precordial R wave transition (as the site of origin is more posterior). In this example, the PVCs have a left bundle branch block morphology and an inferior axis, but the transition lead is V₃ (earlier than is typical in RV outflow tract tachycardia); there is also a broad R wave in lead V₂.

Figure 2 An electrocardiogram recorded during frequent ectopy from the right coronary cusp. Idiopathic ventricular arrhythmias can also arise from the left ventricular outflow tract. The most common sites are the right and left coronary cusps. Compared to the right ventricular outflow tract, the left ventricular outflow tract at the level of the coronary cusps is posterior and slightly inferior. The major effect of this ventricular tachycardia location on the electrocardiogram is an earlier precordial R-wave transition (as the site of origin is more posterior). In this example, premature ventricular complexes have a left bundle branch block morphology and an inferior axis, but the transition lead is V₃ (earlier than is typical in right ventricular outflow tract tachycardia); there is also a broad R wave in lead V₂.

Mechanism. The mechanism of this tachycardia is thought to be automaticity. It has been associated with ventricular tumors, either isolated hamartomas (Purkinje cell tumors) or more diffuse histiocytoid or lymphocytoid tumors.^{65–67} It may also be associated with acute or chronic myocarditis in very young patients. A specific cause is not found in 50% of the patients.⁶

Clinical characteristics. The clinical course is characterized by incessant tachycardia with heart rates often greater than 200 beats/min. Older children are more likely to have symptoms of heart failure.⁶ Incessant VT greater than 80% of the day may be associated with tachycardia-induced cardiomyopathy.⁶

Natural history. Infants with incessant VT may have spontaneous resolution over a 1–2-year time period, but mortality rates of up to 15% have been reported.^{6,65}

2.3.3. Intrafascicular verapamil-sensitive reentrant tachycardia

Definition. Intrafascicular verapamil-sensitive reentrant tachycardia or idiopathic left VT is a ventricular arrhythmia arising from the mid to the apical portion of the left ventricular septum.⁶⁸ It accounts for 10%–15% of IVT. The tachycardia is monomorphic with RBBB QRS morphology and a superior axis (Figure 3). Right axis deviation is seen in 5%–10%.⁶⁹

Mechanism. This tachycardia is thought to be due to a reentry circuit in the vicinity of the left posterior fascicle and has commonly been referred to as *fascicular VT*.^{68,70,71} It had been thought that the circuit includes antegrade conduction down the left fascicle and then moves upward along an adjacent branch of conductive tissue.⁷² This tachycardia is characteristically sensitive to verapamil and occasionally may respond to adenosine, but not to Valsalva maneuvers.⁷³

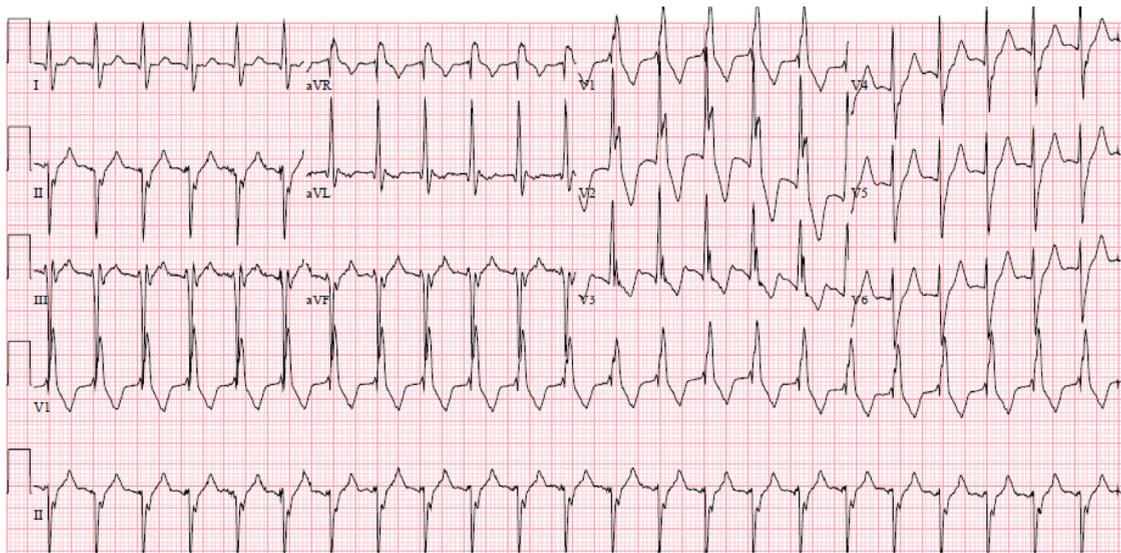
Clinical characteristics. The most common clinical course consists of sustained or nonsustained episodes of monomorphic VT triggered by stress or exercise. Tachycardia rates range from 120 to 250 beats/min, usually from 150 to 200 beats/min. The tachycardia is generally well tolerated, especially at the slower rates.

The tachycardia occurs in boys more frequently than in girls, with the age of onset often in the teen years.⁷⁴ Most patients have mild symptoms of palpitations or dizziness and are rarely limited from activity because of exercise-induced tachycardia.

Natural history. The natural history of idiopathic left VT is usually benign with reports of spontaneous remission.^{75–77} There are rare reports of sudden cardiac death owing to tachycardia-induced cardiomyopathy.^{27,78}

2.3.4. Bundle branch reentry tachycardia

Definition. Bundle branch reentry tachycardia is a reentry tachycardia using the His-Purkinje system. This tachycardia



The morphology of this tachycardia is essentially identical (and often confused with) SVT with right bundle branch block and left anterior hemiblock, due to its origin in the vicinity of the left posterior fascicle.

Figure 3 An electrocardiogram recorded during sustained idiopathic left ventricular tachycardia (intrafascicular verapamil-sensitive reentrant tachycardia). The morphology of this tachycardia is essentially identical to (and often confused with) supraventricular tachycardia with right bundle branch block and left anterior hemiblock because of its origin in the vicinity of the left posterior fascicle.

often occurs as a result of His-Purkinje disease associated with left ventricular enlargement and heart failure.⁷⁹ It may be observed in young patients with myotonic dystrophy, in whom delayed conduction in the His-Purkinje system is common despite preserved left ventricular function.^{79,80} The baseline ECG usually shows evidence of His-Purkinje disease such as a nonspecific interventricular conduction delay or PR prolongation. VT is monomorphic with LBBB QRS morphology and a superior axis. It is less common to have RBBB QRS morphology with an inferior axis.

Mechanism. The usual reentry circuit involves movement down the right bundle branch, crossing the septum near the apex and traveling upward toward the base via the left bundle branch. The His bundle is activated in a retrograde fashion.⁸¹ The less common reentry circuit moves in the opposite direction, traveling up the right bundle branch across the septum at the base and down the left bundle branch. Rare cases have described the reentry circuit occurring within the anterior and posterior fascicles of the left bundle branch.

Clinical characteristics. Bundle branch reentry tachycardia is rare even in the adult population. This arrhythmia is unique in that it is dependent exclusively on the specialized conduction systems and is usually limited to those with advanced structural heart disease.⁸² This tachycardia has not been described in young patients with structurally normal hearts but has been described in those with His-Purkinje disease related to myotonic dystrophy.⁸⁰ Symptoms include palpitations or dizziness when left ventricular function is preserved but syncope or cardiovascular collapse is common in the setting of diminished cardiac function.

Natural history. The natural history of bundle branch reentry tachycardia is mostly related to the underlying disease. Progressive His-Purkinje disease and late development of heart block have been reported in patients with myotonic dystrophy.

2.4. Complex ventricular ectopy and polymorphic ventricular tachycardia

Definition and recognition

Polymorphic VT is characterized by beat-to-beat variations in QRS morphology and/or axis. It is uncommon in the pediatric population and carries a less favorable prognosis than does monomorphic VT.⁸³ As it is rare in patients with a structurally normal heart and no channelopathy, discussion will be limited to its recognition and differential diagnosis. This may be a hemodynamically unstable arrhythmia, and although events may terminate spontaneously, the potential for degeneration into ventricular fibrillation exists. It can be divided into bidirectional VT and true polymorphic VT, a classic example of which is torsades de pointes (TdP), as seen in LQTS.⁸⁴

Bidirectional VT is characterized by an alternating beat-to-beat QRS axis on the 12-lead ECG and has been described in the setting of digoxin toxicity and in two channelopathic conditions—Andersen-Tawil syndrome and CPVT.^{85,86} Both channelopathies can result in polymorphic VT and are well described in a previous consensus document.⁸⁴

Clinical characteristics and differential diagnoses

Polymorphic VT in a young person without structural heart disease can occur in the setting of QT prolongation, either at baseline or precipitated by specific drugs, hypokalemia, or hypomagnesemia. These arrhythmias are usually self-limited

but may degenerate into ventricular fibrillation and result in sudden cardiac death. Although provocation of polymorphic VT by a noncardiac medication is less common than by antiarrhythmic medications, a number of noncardiovascular drugs have been withdrawn from the market because of unexpected sudden cardiac death associated with prolongation of the QT interval and TdP.⁸⁷ The hallmark mechanism of drug-induced QT prolongation and TdP is the blockade of the cardiac delayed rectifier potassium channel.⁸⁸ Incidence is difficult to estimate, and the occurrence of acquired QT prolongation and TdP is unpredictable. Often multiple risk factors are present in a given individual, for example, drug exposure coupled with electrolyte imbalance. A female preponderance has been born out in multiple studies.^{89,90} Cocaine, amphetamines, and the weight-reducing substances phentermine and chlorpheniramine have been associated with the occurrence of polymorphic VT; multiple agents are often present and interacting.^{91,92} Recently, methadone alone or in combination with other agents has come under scrutiny as a cause of polymorphic VT.⁹³ Methadone is mainly metabolized by the CYP3A4 isoenzyme of the hepatic cytochrome P450 system, which is used by numerous other QT-prolonging agents. Since drug addicts are prone to concomitant medical conditions, they are at high risk of developing this complication from methadone. When polymorphic VT is suspected or confirmed, a careful review of possible drug exposure is required.

While it is arguable whether the heart is truly normal in myocarditis, "occult" myocarditis implies no demonstrable structural pathology. Complex ventricular arrhythmias have been documented in this setting. A series of 17 patients with otherwise clinically silent lymphocytic myocarditis presented with potentially life-threatening ventricular arrhythmias.⁹⁴ Bidirectional VT, although rare, has been seen in the setting of myocarditis.⁹⁵ A study of children with frequent ectopy and a structurally normal heart as evaluated by noninvasive imaging revealed that 9% had myocardial biopsy changes consistent with lymphocytic myocarditis, and all 50% had biopsy evidence of subclinical disease related to cardiomyopathy or myocarditis.⁹⁶ In a pediatric study of myocarditis, 29% of the patients had arrhythmias including ventricular arrhythmias.⁹⁷ In a retrospective series by Friedman et al,⁹⁸ 12 patients with biopsy findings of myocarditis all had associated monomorphic and/or polymorphic VT and ectopy. This cohort had a normal mean shortening fraction on echocardiography at presentation. Resolution of the inflammation was not always associated with complete arrhythmia resolution, and some patients were maintained on antiarrhythmic medications.

Polymorphic VT and bidirectional VT are rare in pediatric patients in the setting of a structurally normal heart. This arrhythmia should alert one to the potential for underlying pathology such as a channelopathy, drug toxicity including digoxin toxicity, and myocarditis. Therapy for hemodynamically significant events includes magnesium, β -blocking agents, and possibly lidocaine. The use of other antiarrhythmic agents with their tendency to prolong the QT interval

owing to I_{Kr} (delayed rectifier potassium current) blockade should be approached cautiously. Correcting the underlying cause including normalizing electrolyte abnormalities is imperative. Pacing at a relatively rapid rate may suppress the arrhythmia if it does not respond to magnesium.

3. Evaluation

There are several important considerations regarding the evaluation of young patients with idiopathic ventricular arrhythmias. First and foremost is the need to evaluate for malignant causes of arrhythmias. As previously noted, the management of VT owing to genetic mutations resulting in altered function of cardiac ion channels, cardiac myocytes, or intracellular matrix is directed toward the specific underlying disorder and is not included in the scope of this document. As such, a very thorough evaluation, including careful scrutiny of the baseline ECG and echocardiographic characteristics, must be undertaken to exclude the presence of these recognized disorders (LQTS, short QT syndrome, Brugada syndrome, CPVT, ARVC, cardiomyopathy, etc.); if identified, disease-specific management can then be initiated. Second, it is important to distinguish the following characteristics: degree of symptoms; grade of ventricular arrhythmias (uniform/multiple morphology and PVCs/non-sustained VT/sustained VT); presumed VT site of origin (RVOT, idiopathic left ventricular VT, and others); and presence or absence of hemodynamic effects of the arrhythmia. Finally, the age of the patient is important, particularly with regard to tolerating various diagnostic tests. Younger children cannot cooperate with SAECG and exercise testing; the requirement for general anesthesia to acquire cardiac MRI in infants and younger children should discourage its use except in very specific situations.

3.1. History and physical examination

Between 0% and 50% of the patients with a structurally normal heart and ventricular arrhythmias report symptoms at presentation or during follow-up. Symptoms range from nonspecific discomfort to rare cases of syncope.^{3,4,6,58,83} The extremely rare cases of aborted cardiac death are mostly attributable to tachycardia-induced cardiomyopathy. Palpitations are most frequently described in older children, but aborted sudden death, heart failure, and syncope have no difference in frequency across age groups.⁸³ The absence of a prodrome before a significant symptom such as syncope is concerning for a more malignant form of ventricular arrhythmia, but the presence of a prodrome may be falsely reassuring.⁹⁹ In a study of 35 patients with a cardiac channelopathy, more than half of the patients with syncope reported some type of prodrome. Exercise-related syncope should be thoroughly investigated, with a high index of suspicion for ventricular arrhythmias related to a channelopathy or structural heart disease. A thorough drug history must be included in the evaluation of patients suspected of ventricular arrhythmias, including prescribed, illicit, and recreational drugs and supplements (e.g., energy drinks and

body building products). A complete family history is necessary, as some patients with inherited channelopathies can present with undifferentiated VT. In a study of 87 families with a child who suffered a sudden cardiac arrest, 23 (27%) reported that a family member had suffered sudden death before the age of 50 years secondary to a “heart condition.”¹⁰⁰ Physical examination is often unrevealing in these patients, who generally have structurally normal hearts, but it is important to identify those with structural disease. VT may also be identified incidentally on routine examination, screening ECG, or testing for another purpose; symptoms may be absent or relatively mild. Here too, careful symptom and family history are important and may impact management.

3.2. Electrocardiography

The baseline ECG is critical and informative in the patient with VT. LQTS, Brugada syndrome, ARVC, and short QT syndrome, as well as the cardiomyopathies, all have characteristic findings on ECG that may be important for evaluation and diagnosis. The resting ECG may also have abnormalities consistent with an electrolyte abnormality, myocarditis, or hypertrophy. A conduction abnormality such as preexcitation or bundle branch block may lend weight to the diagnosis of supraventricular tachycardia in a patient with documented wide complex tachycardia. Conduction delay may also be a marker of an underlying pathologic condition (e.g., sarcoid and ARVC) predisposing to ventricular arrhythmia.

3.3. Ambulatory monitoring (Holter monitoring)

Ambulatory ECG, or Holter monitoring, has been used extensively when evaluating the patient with IVT and a structurally normal heart.¹⁰¹ Home telemetry monitoring is being increasingly used for arrhythmia surveillance, but as of yet, there is no literature on its use in this pediatric population. As approximately 50% of the patients may be asymptomatic, especially patients with an accelerated idioventricular rhythm, Holter monitors can be quite useful to determine arrhythmia burden, which has been associated with the development of cardiomyopathy in adult patients.^{28,102–107} No studies have been performed in the pediatric population to support or refute this finding. Even a significant burden of monomorphic ectopy may be asymptomatic and warrant observation only. The distinction of monomorphic from polymorphic ventricular ectopy is also critical. The finding of exertional bidirectional VT, even asymptomatic, may portend a more serious diagnosis. Holter monitor may also be useful for assessing efficacy of therapy such as degree of β -blockade or reduction of PVC burden. Its role in the diagnosis of LQTS has been questioned.¹⁰⁸

More prolonged monitoring with event monitors has been useful in evaluating sporadic episodes and correlating them with symptoms. The implantable loop recorder has been shown to be efficacious in children, especially when a serious arrhythmia is suspected.¹⁰⁹ A retrospective multicenter study found symptom-rhythm correlation possible in

100% of the patients.¹⁰⁹ However, the automatic detection algorithm may not be optimal in children. Kothari et al¹¹⁰ reported missed detection of polymorphic VT and a high false-positive rate. In practice, long-term ambulatory monitoring can, in most cases, provide data similar to those offered by an implantable loop recorder.

3.4. Exercise testing

Exercise testing can be very useful in elucidating adrenergic-sensitive ventricular arrhythmias. During exercise testing in a wide range of pediatric patients, VT was induced in just less than 1%.¹¹¹ The majority of these patients had either LQTS or structural heart disease. Among patients with known VT, more than 50% had inducible arrhythmia with exercise.^{112,113} In a study addressing diagnosis-specific characteristics of VT, exercise-induced arrhythmia was seen in 20%–40% of the patients with IVT (either right or left sided) and in 100% of the patients with CPVT.⁸³ Thus, this test is especially useful when trying to distinguish patients with CPVT or LQTS from others with apparent structurally normal hearts. Ambulatory ECG or event monitoring may be unrevealing in these disease states, especially if the patient is sedentary.¹¹⁴

3.5. Special electrocardiographic techniques

SAECG is a technique that improves the signal-to-noise ratio on the surface ECG, allowing for identification of low-amplitude signals at the end of the QRS complex, called “late potentials.” These late potentials indicate regions of abnormal myocardium, demonstrating slow conduction. There have been limited studies using SAECG in the pediatric population, mainly in the patient with postoperative congenital heart disease and patient with ARVC.^{115–122} This technique has not been investigated in the pediatric patient with IVT. The finding of an abnormal SAECG should prompt the clinician to investigate the possibility of ARVC more thoroughly. Microvolt T-wave alternans is a fluctuation in the amplitude or morphology of the T wave on every other beat. It is assessed during exercise or with atrial pacing and has been shown to be useful in assessing risk of life-threatening arrhythmia in patients who have had myocardial infarction.¹²³ Alexander et al¹²⁴ studied T-wave alternans in more than 300 pediatric and congenital heart disease patients and found an eight-fold increased risk of cardiac arrest with an abnormal T-wave alternans pattern. Importantly, T-wave alternans identified only 26% of the patients who had suffered cardiac arrest. No studies of children with structurally normal hearts and VT have been performed.

Heart rate variability, a marker of cardiac autonomic control, is a measure of the beat-to-beat variation in the cardiac cycle length. Standardized analyses of frequency-domain and time-domain RR interval variability are widely available from ambulatory cardiac recordings. Decreased heart rate variability is correlated with the risk of sudden cardiac death in patients after myocardial infarction.¹²⁵ Several small studies of adult patients with IVT originating

from the outflow tract suggest a predominance of sympathetic activation immediately before the initiation of tachycardia.^{126–128} A single small study of children with either IVT or frequent ventricular ectopy found diminished time-domain variables as compared to those in normal children.¹²⁹ These studies may lead to important clues regarding the mechanism of these arrhythmias, but there is a lack of adequate sensitivity and specificity to predict risk of arrhythmia or sudden death.

3.6. Cardiac imaging

An echocardiogram is an important test when assessing the patient thought to have IVT in order to rule out structural heart disease. Evaluation should include wall thickness assessment, quantitation of systolic function, measurement of indices of diastolic function, and exclusion of valvular lesions, coronary artery anomalies, and cardiac tumors. The present document addresses patients in whom significant congenital anomalies are absent and in whom echocardiogram excludes the diagnosis of any form of cardiomyopathy or overt ARVC. Serial echocardiography is also useful in children with a high burden of ventricular arrhythmias, as some patients have developed cardiomyopathy owing to a high burden of frequent ventricular arrhythmias over follow-up.

Cardiac MRI allows for the assessment of structure, function, and presence of fibrosis. This test may be most important when assessing a patient for ARVC, as abnormal MRI findings constitute major criteria for the diagnosis of this disease.¹¹⁵ However, considerable concern has been raised about the false-positive rate for the diagnosis of ARVC by cardiac MRI, especially in nonexpert hands. It is hoped that the updated, more quantitative diagnostic criteria will reduce this problem.¹⁷

MRI may also identify tissue abnormalities not appreciable by other means; late gadolinium enhancement (delayed enhancement) may suggest areas of scarring or fibrosis that may be due to myocarditis and can be the substrate for the development of ventricular arrhythmias.¹³⁰ MRI may also be useful when the echocardiogram suggests or cannot exclude coronary anomalies or tumors.

3.7. Electrophysiological testing

Intracardiac electrophysiological testing (EPS) is performed in conjunction with catheter ablation once the decision has been made to eliminate a documented VT. The role of programmed stimulation as a purely diagnostic test in young patients has not been well studied, but would be expected to be minimal. The use of EPS to confirm a VT mechanism, guide medical therapy, or stratify risk rarely exceeds non-invasive means, and false-positive results of aggressive stimulation may be misleading. EPS is clearly not useful in the investigation of unexplained syncope; the fairly low pretest probability of VT as the cause of syncope in this population, along with the test's low specificity, compromises its positive predictive value.^{131–135} However, EPS

may be useful in rare circumstances, such as to evaluate patients with nonsustained polymorphic VT, to assess for the inducibility of sustained arrhythmia, to look for low-voltage areas consistent with scar, and/or to help determine how aggressive to be with medical or device therapy.

3.8. Laboratory testing

Laboratory evaluation is warranted in all patients with complex, multiform ectopy or polymorphic VT to include assessment for acute inflammation as seen in myocarditis and to exclude drug toxicity and metabolic or electrolyte disturbance. Similar evaluation should be carried out as part of the initial evaluation in any patient with acute presentation of VT and especially in those in whom there is a clinical suspicion of myocarditis.

3.9. Genetic testing

Performed with cardiac evaluation, genetic testing may be used to evaluate a molecular diagnosis of LQTS, short QT syndrome, CPVT, and Brugada syndrome.⁸⁴ In addition, phenotypically negative relatives of the affected patients, capable of passing on an abnormal gene, can be identified through techniques such as cascade screening.¹³⁶ The details of genetic testing as a strategy have been considered comprehensively in a consensus document regarding patients with arrhythmias secondary to genetic ion channelopathies.¹³⁷

4. Therapy

4.1. General considerations

The decision to initiate therapy for the management of frequent ventricular arrhythmias in infants, children, and adolescents is dictated by the age, symptomatology, specific diagnosis, and electrical and hemodynamic impact of the arrhythmia. As such, a wide range of possible appropriate therapies exists, anywhere from reassurance and discharge to aggressive antiarrhythmic medication or catheter ablation. Given the often benign nature of IVT in children, it is anticipated that following a thorough diagnostic investigation, the majority of patients will not require therapy. While considering whether to treat, in patients too young to voice a complaint, clinicians should be alert to signs of possible hemodynamic compromise. These may include overt signs of decreased cardiac output, such as changes in perfusion or measurable markers of metabolic acidosis. Any event that resembles syncope or aborted sudden death should be considered significant. There are few obvious signs and symptoms in these youngest patients; thus, subtle signs such as excessive irritability or poor feeding should be considered important. Older patients who have proven arrhythmia-associated symptoms that may be markers of hemodynamic compromise, such as dizziness, syncope, shortness of breath, easy fatigability, or chest pain, should also be considered for therapy. When the impact of ventricular arrhythmia is significant enough to warrant more than observation, therapies should be instituted in a stepwise fashion, with continuous reevaluation of the selected course to ensure that

the possibility/probability of side effects from the treatment does not exceed that of the disease. Likewise, the goals of the selected therapies should be clearly defined and communicated with the family before implementation, as complete elimination of the arrhythmia is often not necessary and an attempt to do so may result in an unacceptable risk of side effects.

There have been a number of descriptive publications detailing the natural and unnatural history of IVT.^{3,4,9,60,68,73,83,138–140} In each, there are patients who received medical therapy and others that were followed without intervention. No randomized controlled trials exist, and in these case series, there has been variation in criteria for a successful result. Studies of response to various medications have often been grouped by arrhythmia site of the origin/mechanism on the basis of ECG characterization and will be discussed further below. Song et al⁸³ reported 37 patients between 0 and 19 years who presented with IVT between 1999 and 2009. Of these, there were 13 patients with LBBB morphology and 24 with RBBB morphology. Twenty were treated with antiarrhythmic medications, with complete resolution in 14 and improvement in 5. There was 1 death in this group, but the details are not presented. Prognosis for complete resolution of tachycardia was especially high in infants, with or without treatment. Pfammatter et al,⁴ of the European Working Group, reported 98 pediatric patients with IVT followed for an average of 47 months, of which 25 did not receive antiarrhythmic medications. No patient died. In 40 patients, medical therapy was successfully withdrawn at an average of 23 months after presentation, with only 3 having a relapse. At study completion, 23 patients remained on therapy. These patients included only 7% of those presenting in infancy versus 30% of those presenting later, once again demonstrating a high rate of spontaneous resolution, especially in those presenting in infancy.

In the Toronto study by Wang et al,³ there were 72 patients with idiopathic ventricular arrhythmias, including 19 with accelerated ventricular rhythm, 30 with right ventricular VT, and 23 with left ventricular VT. No deaths were reported. Of the 32% of the patients with accelerated ventricular rhythm, all responded to therapy, with β -blocker being the most common medication used. In the right ventricular VT group, two-thirds were treated with medications, with an overall success rate of 60%. Calcium-channel blockers were effective in 12 of 13 or 92% of patients treated in the left ventricular VT group, although they are generally not recommended in children younger than 1 year.

The above studies and others present somewhat heterogeneous approaches to the medical management of the pediatric patient with VT. As seen frequently in medicine, the decision to treat must be individualized. When medical therapy is indicated, it is often appropriate to start with β -blockers or calcium-channel blockers, given that they are generally very well tolerated with few side effects in children and adolescents. β -Blockers are almost always the first-line therapy chosen for infants, although caregivers should be

counseled regarding signs and symptoms of hypoglycemia in this age group. Likewise, calcium-channel blockers have proved quite efficacious as first-line therapy for ventricular arrhythmias, although they generally are not recommended in children younger than 12 months because of reported incidences of profound hemodynamic compromise.^{141–145}

The choice of therapies beyond these first-line medications must take into consideration not only the patient's age and the arrhythmic substrate but also the experience and expertise of the institution at which the patient is undergoing treatment. Whereas in some institutions initiating a more aggressive (often class III) antiarrhythmic medication for a refractory ventricular arrhythmia would be standard practice, referral for ablation of the arrhythmic substrate may be an equally appropriate option in higher-volume centers with experience in a pediatric population.

4.2. Special considerations and exclusions

As previously noted, the management of VT due to genetic mutations resulting in altered function of cardiac ion channels, cardiac myocytes, or intracellular matrix is directed toward the specific underlying disorder and is not included in the scope of this document. Recommended therapies and guidelines for the acute management of polymorphic VT in infants and children have been established and likewise are beyond the scope of this document.¹⁴⁶

4.3. Ventricular ectopy/tachycardia in infancy

Isolated ventricular ectopy or tachycardia presenting in infants is often discovered as an incidental finding in the course of evaluating an unrelated condition. Management is based on a thorough review of the functional and symptomatic impact on the infant's growth and development. The few large cohort studies that have been performed demonstrate that the majority of infants with isolated VT are successfully managed with conservative observation alone. A large multicenter review by Pfammatter et al⁴ reported that as compared with children older than 1 year, infants with VT are less likely to experience symptoms (22% vs 38%; $P < .05$) and more likely to have complete resolution (89% vs 56%; $P < .01$).⁴ These findings were confirmed in a single-center report by Levin et al,⁶⁰ which also demonstrated no statistical difference in time to resolution of VT between infants who received outpatient antiarrhythmic medications and those who did not.

4.4. Substrate-based management

Ventricular ectopy

The prevalence of ventricular ectopy in school-age children has been shown to increase with age.¹⁴⁷ The long-term prognosis is favorable, particularly in those experiencing VT of an outflow tract origin. A majority of patients have a decrease in arrhythmia burden with time, and many have complete resolution.^{6,14,27,83} In addition, even patients with very frequent PVCs are rarely symptomatic, with the ectopy most often discovered during routine medical evaluation.

Routine treatment of patients with antiarrhythmic medications has not been shown to decrease arrhythmia burden nor hasten resolution of ventricular ectopy and is not recommended in the absence of rhythm-correlated symptoms.^{1,27}

Ventricular ectopy-induced cardiomyopathy

Patients with very frequent ectopy, defined herein as greater than 10% ectopic beats on 24-hour ambulatory monitoring, should be monitored for the development of PVC-related ventricular dysfunction. Numerous reports describing the development and reversibility of this form of cardiomyopathy in children have been published.^{4,148–150} It is important to realize that no large study addresses the risks of developing this dysfunction in the pediatric population and that the burden of PVCs required to produce this effect is unclear and variable.

Several studies in an adult population have attempted to define the incidence of and identify risk factors for the development of cardiomyopathy in the face of frequent ventricular ectopy. These have demonstrated that cardiomyopathy in this population is relatively uncommon. One study by Hasdemir et al²⁸ found an incidence of cardiomyopathy of 6.8% in a group of 249 patients referred for evaluation owing to frequent monomorphic PVCs or VT over a 6-year period. Work by Niwano et al¹⁰³ in which 239 patients presenting with frequent ventricular ectopy but normal left ventricular ejection fraction followed for 5 years revealed that no one developed cardiomyopathy. Other studies have reported an incidence of up to 30% but are hampered by selection bias, as they were performed in populations referred for ablation. In patients with frequent PVCs presenting with tachycardia-induced cardiomyopathy and referred for ablation therapy, several risk factors have consistently been identified.^{28,151–153}

These include male gender, high PVC burden, and asymptomatic status. These studies have independently shown an association between cardiomyopathy and PVC QRS duration or epicardial origin, persistence of PVCs or frequent monomorphic VT, and a longer duration of palpitations (in symptomatic patients). Most studies have found that it is rare to develop dysfunction with a PVC burden less than 20%–30%, but reversible myopathy in patients with as little as 5% PVCs has been reported.^{28,152,153}

In pediatric patients presenting with evidence of diminished left ventricular function in the face of frequent PVCs, management should include medical or ablative options to diminish the arrhythmia burden. Medical management should be initiated with the medication least likely to result in significant side effects. β -Blockers generally are the first choice if ventricular function has not severely deteriorated. Calcium-channel blockers must be used cautiously in infants and in the setting of ventricular dysfunction, and class III agents such as amiodarone may be necessary. Ablative therapy may be highly effective, with cure rates up to 95% shown in adult populations,^{148,149} and is recommended if medical therapy is unsuccessful in controlling ectopy and reversing dysfunction.

Predicting which pediatric patients may develop a tachycardia-induced cardiomyopathy is obviously difficult. Thus, routine surveillance of ventricular function should be performed in patients with persistent ectopy.

Accelerated idioventricular rhythm

An accelerated idioventricular rhythm is nearly always benign.^{3,25,138,154} Accelerated idioventricular rhythm may be observed in well-trained athletes, where it is simply felt to represent increased vagal tone at rest and immediately resolves with the initiation of activity. Rarely, observation of this rhythm may be a harbinger of a more severe and symptomatic form of VT and therefore ongoing surveillance until resolution has been recommended.^{155,156} When medications have been used, response to a wide variety of agents has been very favorable.⁸³ In the absence of symptoms, ventricular dysfunction, or evidence of an additional underlying arrhythmogenic condition, there is no indication for intervention.

Ventricular tachycardia originating from the outflow tracts

Treatment of infants and children with outflow tract VT should be reserved for those with symptoms and/or frequent, prolonged, or rapid episodes. When treatment is indicated, the choice of medical therapy versus catheter ablation should be driven by institutional expertise. Once again, initial therapy with lower-risk medication is recommended, with class I and III agents reserved for failure to control VT with β -blocker or calcium-channel blocker in the older child. As discussed in a separate section, catheter ablation is effective and can be performed at a relatively low risk, but it should be noted that perforation can be a complication of outflow tract ablation.

Intrafascicular verapamil-sensitive reentrant tachycardia

This less common form of VT has a relatively narrow QRS owing to its origin adjacent to the normal conduction system and is often initially misdiagnosed as supraventricular tachycardia in children. As with many other forms of arrhythmia, infants with this VT frequently experience resolution with time; such resolution is much less common in older children and adolescents.¹⁵⁷ Episodes are often paroxysmal, frequently brought on by stress or exercise and may last for hours before spontaneously abating. VT is generally well tolerated, although patients are usually symptomatic during the events. Prolonged episodes can lead to tachycardia-induced ventricular dysfunction.¹⁴⁹

Conservative observation alone of patients with this VT usually is not adequate owing to the symptomatic nature of the arrhythmia, but may be useful in children with infrequent, self-terminating episodes. While this form of VT is highly responsive to intravenous verapamil during acute events, the use of oral verapamil to prevent subsequent episodes has been shown to be ineffective in more than 20%.¹⁵⁸ In such instances, another pharmacologic agent, such as a β -blocker or class III antiarrhythmic agent, or catheter ablation is warranted.

Implantable cardioverter-defibrillators

The incidence of sudden cardiac death in pediatric patients with ventricular ectopy/VT in the absence of structural heart disease, myopathies, and channelopathy is very low. As such, the need for the placement of an implantable cardioverter-defibrillator (ICD) in this population is exceedingly rare. The highest incidence of sudden death among pediatric patients with VT (13%) was reported by Deal et al⁷ in a 1986 description of 24 patients treated between 1974 and 1986 at a single institution. Importantly, in each of the 3 deaths in these 24 patients, there were findings suggestive of underlying myopathy or channelopathy, conditions less well understood in that era.

Collins et al¹⁵⁸ recently reported 3 deaths, 2 of which could be reasonably attributed to arrhythmia among a cohort of 152 pediatric patients treated for left VT at 22 centers across North America, South America, and Europe. This report describes another 3 patients who underwent placement of an ICD; all 3 had diminished ventricular function, and 2 had polymorphic VT at presentation. The third child was subsequently found to have evidence of myocarditis. Only the patient with myocarditis had appropriate ICD therapies during follow-up. One patient underwent ICD removal 2 years after presentation after successful ablation of his VT, having received no appropriate ICD therapies. Given the very low rate of sudden death in this population, ICD implantation is not recommended in pediatric patients with VT in whom careful evaluation has not revealed any evidence of underlying myopathy, channelopathy, or structural heart disease, unless the tachycardia cannot be adequately controlled, and in the judgment of the specialist the patient has a risk of sudden death higher than expected in this population.

Lifestyle modifications: exercise restrictions

There is a paucity of data with regard to the risk of sports participation and exercise in individuals with IVT. Suppression of ventricular ectopy during exercise stress testing, and the known low risk of sudden death in this population, would suggest that the risk of sudden death during exercise is minimal. Frequent and complex ventricular arrhythmias in the structurally normal heart are not uncommon in trained athletes and do not appear to convey risk for sudden death.²²

The consensus statements for sports participation from the Bethesda Conference #36 and the European Society of Cardiology (ESC) provide some insight into recommendations for sports participation in those with IVT.^{159,160} The Bethesda Conference #36 states that the asymptomatic athlete without structural heart disease and short (<10 beats) bursts of monomorphic VT at rates less than 150 beats/min that suppress or do not worsen during exercise is eligible to participate in all competitive sports. Similarly, the ESC consensus statement allows full participation in competitive sports for asymptomatic athletes without structural cardiac disease if nonsustained VT is rare, is not triggered by exercise, presents without short RR interval, and occurs in the absence of a family history of sudden death.

Athletes with frequent or sustained IVT, particularly induced by exercise, have the option of ablation of the arrhythmogenic focus. The Bethesda Conference #36 allows for full participation in athletics after successful ablation of VT, but recommendations for those not choosing ablation are unclear. The ESC consensus statement gives recommendations specific to slow VT, intrafascicular verapamil-sensitive reentrant tachycardia, and RVOT tachycardia. In the absence of cardiac disease, arrhythmogenic conditions (channelopathies and cardiomyopathy), and a family history of sudden death and symptoms (presyncope, lightheadedness, and exertional fatigue), the ESC allows for all sports participation except in those with high risk of syncope.

In light of the paucity of outcomes data for patients with IVT who wish to participate in activities, ablation of the arrhythmogenic focus should be considered in patients with symptomatic, frequent, and/or exercise-induced sustained or nonsustained IVT before full participation in competitive athletics. Athletes with infrequent, asymptomatic, sustained, or nonsustained IVT that suppresses with exercise, and in whom a thorough evaluation to exclude more malignant causes of IVT has been performed, may participate fully in competitive athletics.

As to the possible use or restriction of use of stimulant medications in this group of patients, the committee did not feel that there was sufficient data on which to make recommendations or that this topic was within the scope of our statement. For discussion of this topic, please review the American Heart Association scientific statement published in 2008.¹⁶¹

Catheter ablation

Catheter ablation is effective for many ventricular arrhythmias.¹⁶² Efficacy and risks are determined by the associated heart disease, location of the arrhythmia origin, and patient size.¹⁶³ Idiopathic ventricular arrhythmias are often well tolerated, minimally symptomatic, and not associated with a risk of sudden death.³ After consideration of risks and benefits, catheter ablation is a reasonable option when treatment is required. In patients younger than 2 years, ablation has been used successfully for the treatment of life-threatening, usually incessant VTs.^{164,165} Ablation of incessant VT during extracorporeal support has also been reported.¹⁶⁵ Ablation in infants is generally a last resort and should be reserved for arrhythmias that are incessant or sufficiently frequent to contribute to ventricular dysfunction and cannot be controlled medically. Otherwise, ablation should be deferred until the child is larger, especially since many resolve spontaneously.⁶⁰ For older children, the decision to proceed to ablation is determined by assessment of the risks and benefits as they relate to other therapeutic options.

Ablation procedure considerations in children

Although ablation of VT has been performed effectively in infants, risks are likely greater for children weighing less than 15 kg.^{163,166} The risks of ablation lesion size, injury to adjacent coronary arteries, and from fluoroscopy exposure are concerns.

Fluoroscopy exposure. Fluoroscopy for imaging during ablation exposes patients to a risk of radiation. For children, long-term risks of neoplasm may be greater than those for adults who have a shorter postprocedure average life expectancy. The accuracy of estimated long-term risks of radiation exposure is not clear.¹⁶⁷ The lifetime mortality risk from cancer has been estimated to be 13%/Gy for male patients and 16%/Gy for female patients aged 10 years in a study by Clay et al¹⁶⁸ in children with a mean weight of 52 kg at ablation. This group measured radiation exposure at selected sites during biplane fluoroscopy, with attention to measures to minimize exposure, including pulsed fluoroscopy at 15 frames/s, low energy output settings, columnated field images, lead shielding, and avoiding magnification modes. During a median fluoroscopy time of 18.3 minutes, the greatest exposure was 43 mGy measured in the right scapular region. The estimated lifetime risk of fatal malignancy was 0.02%. Mean fluoroscopy times in experienced centers are often in the range of 20 ± 8 minutes. The duration of exposure is not an adequate indication of patient exposure, as frame rates, energy, and columnation have a major effect. Some centers have reduced fluoroscopy to less than 5 minutes with the use of electroanatomic mapping systems.^{165–169} The use of fluoroscopy should be as limited as possible and employ appropriate settings and equipment to minimize exposure.^{169–171}

Coronary artery injury. The coronary arteries are at risk for injury from ablation performed along the atrioventricular annuli, in the sinuses of Valsalva, and in the epicardium, including within the coronary sinus and cardiac veins.^{172,173} Coronary injury has been reported after catheter ablation of accessory pathways in children, can appear late after ablation, and can be asymptomatic.^{172,173} In a study of piglets, Paul et al¹⁷⁴ observed that adjacent atrial radiofrequency (RF) lesions produced medial injury that resulted in stenosis evident 6 months later. It appears to be more difficult to injure coronary arteries with catheter cryoablation.^{175,176} There is limited experience with the use of cryoablation for VT, and recurrences may be greater than those with RF ablation.^{165,177,178}

Effect of growth on ablation lesions. In infants, the size of an ablation lesion in the ventricle may increase with time. RF and cryoablation lesions have been studied in infant animals.^{179,180} Ventricular lesions increased in time, essentially doubling in volume at 1 year. Lesions at the atrioventricular groove appeared to remain relatively stable in size, although depth appears to increase with time. The mechanism is uncertain, but may involve proliferation of fibroblasts and matrix in the lesion borders. These findings further support avoiding catheter ablation in infants.

Catheter ablation of specific ventricular arrhythmias

The approach to ablation and the risks and efficacy are related to the site of origin of the arrhythmia. The ventricular outflow tract regions are the most common origin for

ventricular arrhythmias in the absence of structural heart disease.¹⁸¹ The RVOT is most common followed by the LVOT/aortic sinuses of Valsalva and then the mitral and tricuspid annuli.^{181,182} These arrhythmias typically have a focal origin that can be targeted for ablation guided by activation mapping.¹⁶² When the arrhythmia is infrequent, pace mapping can be used. Failure of ablation can be caused by quiescence of the arrhythmia in the electrophysiology laboratory that may be aggravated by anesthesia. Administration of β -adrenergic agonists is often necessary for arrhythmia induction.

In the RVOT, the focus is often found close to the pulmonary valve annulus but can originate anywhere in the region, including sites adjacent to the membranous septum or in sleeves of muscle extending above the pulmonary valve.¹⁶² Risks are, to some extent, related to the focus location. In the free wall of the outflow tract, perforation and tamponade are concerns. In the para-Hisian region, there is a risk of heart block. In its leftward posterior aspect, the RVOT is in close proximity to the left main coronary artery, raising the theoretical possibility of injury that is likely greater in children than in adults.¹⁸³

The LVOT and aortic sinuses VTs have not been well characterized in children.¹⁸⁴ Arrhythmias with a prominent R wave in lead V_1 suggest a left ventricular origin, but localization can only be reliably established by mapping. Those that originate from the right aortic sinus of Valsalva often cannot be distinguished electrocardiographically from those that can be ablated from the RVOT. Ablation from the left or right sinus of Valsalva, and rarely the noncoronary sinus, is required in some.¹⁸⁵

The major ablation concern in this area is the proximity of the ablation site, which is often in the base of the cusp, to the coronary artery ostia. Great care with assessment of this distance is mandatory to avoid the risk of acute coronary injury. A distance of greater than 5–8 mm between the ablation site and the coronary artery has been suggested, and this is less likely to be achieved in small hearts.¹⁸⁵ Damage to the aortic root or aortic valve is also possible.¹⁸⁵ The effect of ablation lesions on these structures in a growing heart is unknown. Some foci are located in an inaccessible area between the aortic annulus and the great cardiac vein with the proximal left anterior descending and circumflex coronary arteries overlying the region, precluding ablation.^{172,186} In adults, some foci have been successfully ablated within the great cardiac vein but proximity to coronary arteries and small size of this vein will likely limit this approach in smaller hearts.¹⁵¹ Successful ablation of LVOT PVC foci and foci near the aortic annulus has been reported in children, but limited data are available.^{187,188} Children considered for ablation often have sustained or very frequent repetitive nonsustained VT or ventricular ectopy, sometimes associated with reduced ventricular function, and have usually failed antiarrhythmic drug therapy.^{3,187,189} Small case series have demonstrated successful abolition of VT in the majority of cases.^{1,187,190,191}

In intraventricular verapamil-sensitive reentrant VT, ablation targeting either presystolic Purkinje potentials during

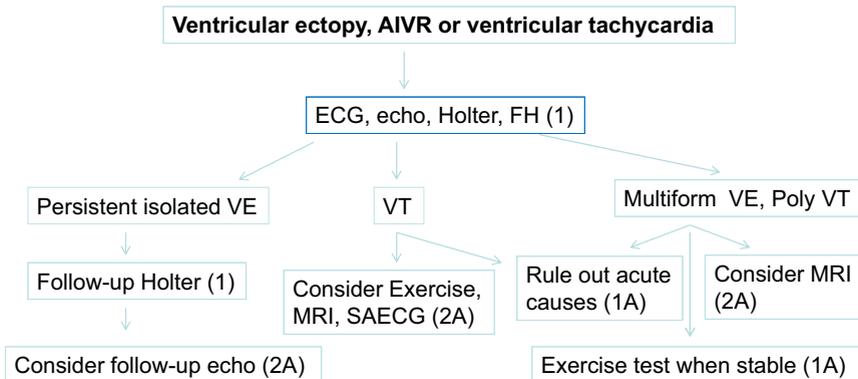
tachycardia or diastolic potentials that may be markers for the retrograde pathway of the circuit is effective in more than 80% of the patients.¹⁹² The latter approach is useful when VT is not inducible or is terminated by mechanical pressure.

Potential complications relate to arterial catheterization and need to access the left ventricle. Damage to the Purkinje system sufficient to change the QRS morphology is rare in adults. Experience in children is limited.^{192,193}

5. Recommendations

A. Evaluation of children with ventricular arrhythmias and a structurally normal heart (summary in Figure 4)

- | | |
|-----------------|---|
| <i>Class 1</i> | <ol style="list-style-type: none"> 1. Infants and children suspected of having ventricular arrhythmias should have a 12-lead ECG, echocardiography, 24-hour ambulatory ECG monitoring, and a detailed personal and family history (Level of evidence: C). 2. Infants and children presenting acutely with multiform or complex ventricular ectopy or polymorphic VT should have laboratory evaluation that includes a metabolic panel and toxicology screen (Level of evidence: C). 3. Exercise stress testing is recommended in children with multiform or complex ventricular ectopy felt to be medically stable when the child is felt to be able to cooperate with such testing and otherwise meets established criteria for exercise stress testing (Level of evidence: C). 4. For infants and children with previously documented frequent ventricular ectopy, and when continued ectopy is confirmed or strongly suspected, follow-up 24-hour ambulatory ECG monitoring is recommended (Level of evidence: C). |
| <i>Class 2a</i> | <ol style="list-style-type: none"> 1. Exercise stress testing may be useful in children with persistent frequent ventricular ectopy or outflow tract tachycardia when the child is felt to be able to cooperate with such testing and otherwise meets established criteria for exercise stress testing (Level of evidence: C). 2. MRI may be useful in infants and children with incessant or complex forms of ventricular ectopy or tachycardia as part of the evaluation of possible myocarditis in patients who are considered stable enough to undergo testing safely (Level of evidence: B). 3. MRI may be useful in children with ventricular arrhythmias in whom there is clinical suspicion of ARVC (Level of evidence: B). |
| <i>Class 2b</i> | <ol style="list-style-type: none"> 1. MRI may be reasonable in older infants with ventricular arrhythmias in whom there is strong clinical suspicion of ARVC (Level of evidence: C). 2. SAECG may be reasonable in children with ventricular arrhythmias in whom there is clinical suspicion of ARVC (Level of evidence: B). |
| <i>Class 3</i> | <ol style="list-style-type: none"> 1. Diagnostic EPS with no intention for catheter ablation is not recommended for pediatric patients with ventricular arrhythmias and presumed structurally normal hearts, except under special circumstances discussed in the text (Level of evidence: C). 2. MRI is not recommended in infants with accelerated ventricular rhythm (Level of evidence: C). |



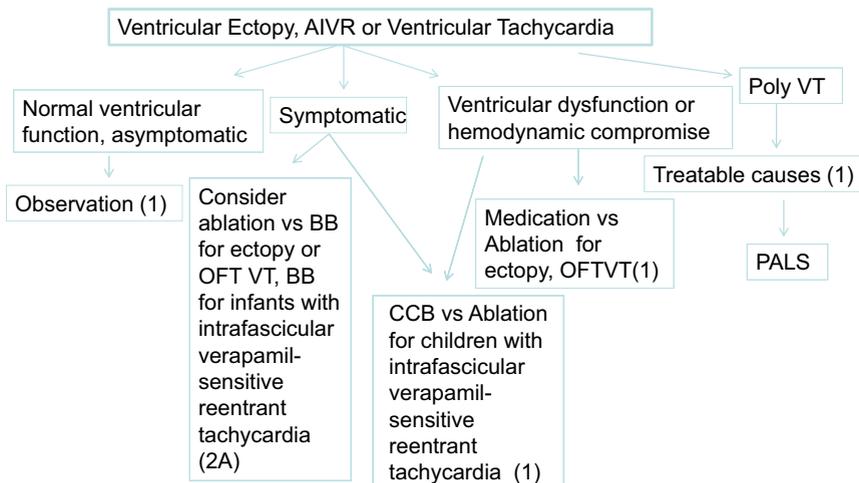
See text for details. Numbers in parentheses refer to level of recommendation. Abbreviations not in text: AAD= antiarrhythmic drug; BB= beta blockers; CCB= calcium channel blocker; OFT VT= outflow tract tachycardia; PALS= pediatric advanced life support guidelines.

Figure 4 Evaluation algorithm. See text for details. Numbers in parentheses refer to the level of recommendation. AIVR = accelerated idioventricular rhythm; ECG = electrocardiogram; FH = family history; MRI = magnetic resonance imaging; SAECG = signal-averaged electrocardiogram; VE = ventricular ectopy; VT = ventricular tachycardia.

B. Treatment of children with ventricular arrhythmias and a structurally normal heart (summary in Figure 5)

- Class 1*
1. Asymptomatic infants and children with normal ventricular function and frequent but isolated ventricular ectopy or accelerated ventricular rhythm should be observed, with no medical or ablative therapy (Level of evidence: B).
 2. Infants and children with well-tolerated idiopathic outflow tract tachycardia that is infrequent, slow, and self-terminating should be monitored, with no medical or ablative therapy (Level of evidence: B).
 3. Children with VT or frequent ventricular ectopy thought to be causative of documented ventricular dysfunction should be treated either medically or with catheter ablation* (Level of evidence: C).
 4. Children who experience hemodynamic compromise due to presumed idiopathic outflow tract tachycardia should be treated either medically or with catheter ablation* (Level of evidence: C).
 5. Symptomatic children and infants older than 1 year with presumed intrafascicular verapamil-sensitive reentrant tachycardia should have initial medical management with a calcium-channel blocking agent or catheter ablation* (Level of evidence: B).
 6. Infants and children with an acute presentation of polymorphic VT should have prompt correction of treatable causes such as electrolyte abnormalities or drug toxicity (Level of evidence: C).
- Class 2a*
1. In asymptomatic infants and children with frequent complex or multiform ventricular ectopy, β -blocker therapy can be useful. If this does not control the arrhythmia, suppressive therapy with calcium-channel blockers can also be useful. If this arrhythmia is very well tolerated and infrequent, only observation can be useful (Level of evidence: C).
 2. In symptomatic children with presumed idiopathic outflow tract tachycardia, or with rhythm correlated symptoms due to ventricular ectopy or accelerated idioventricular rhythm, suppressive therapy with a β -blocker or catheter ablation can be useful (Level of evidence: C).
 3. In infants younger than 1 year with presumed intrafascicular verapamil-sensitive reentrant tachycardia, medical therapy with β -blocker therapy can be useful (Level of evidence: C).
- Class 2b*
1. In infants and children with frequent complex or multiform ventricular ectopy, treatment with other agents (class I or III) after failure of β -blockers and/or calcium-channel blockers may be reasonable (Level of evidence: C).
 2. Catheter ablation* may be reasonable in children with complex ventricular arrhythmias where one morphology dominates or when there is a suspected trigger that can be targeted (Level of evidence: C).
 3. ICD implantation may be reasonable in children or older infants with polymorphic VT when the arrhythmia persists after acute treatable causes have been ruled out if sudden death risk persists (Level of evidence: C).
- Class 3*
1. Catheter ablation in infants and toddlers is not recommended, except in the case of VT that cannot be adequately controlled medically *and* is not tolerated hemodynamically (Level of evidence: C).
 2. Exercise restrictions are not recommended in children with normal ventricular function, no or minimal symptoms, and well-tolerated and/or well-controlled monomorphic ventricular arrhythmias (Level of evidence: C).
 3. ICD implantation is not recommended in patients with IVT, regardless of symptoms, unless the tachycardia cannot be adequately controlled with medication and/or catheter ablation *and* in the judgment of the specialist the patient has a risk of sudden death higher than expected in this population (Level of evidence: C).

*Catheter ablation for ventricular arrhythmias in children should be performed only by centers and physicians with expertise in ablation therapy in pediatric patients



See text for details. Numbers in parentheses refer to level of recommendation. Abbreviations not in text: AAD= antiarrhythmic drug; BB= beta blockers; CCB= calcium channel blocker; OFT VT= outflow tract tachycardia; PALS= pediatric advanced life support guidelines.

Figure 5 Treatment algorithm. See text for details. Numbers in parentheses refer to the level of recommendation. AIVR = accelerated idioventricular rhythm; BB = β-blocker; CCB = calcium-channel blocker; OFT VT = outflow tract tachycardia; PALS = Pediatric Advanced Life Support; VT = ventricular tachycardia.

C. Indications for catheter ablation in children with idiopathic ventricular arrhythmias*

- Class 1** Catheter ablation is recommended in children with:
1. Ventricular dysfunction or hemodynamic compromise presumed to be due to ventricular ectopy or tachycardia, either as primary therapy or in patients not controlled medically (Level of evidence: C).
 2. Intrafascicular verapamil-sensitive reentrant tachycardia, either as primary therapy or if not controlled by calcium-channel blockers (Level of evidence: C).
- Class 2a** Catheter ablation can be useful in:
1. Symptomatic children with presumed idiopathic outflow tract tachycardia (Level of evidence: C).
 2. Symptomatic children with rhythm-correlated symptoms due to frequent ventricular ectopy or accelerated idioventricular rhythm (Level of evidence: C).
- Class 2b** Catheter ablation may be reasonable to consider in children with polymorphic ventricular arrhythmia where one morphology dominates or when there is a suspected trigger that can be targeted (Level of evidence: C).
- Class 3** Catheter ablation is not recommended in:
1. Infants and toddlers, except in the case of VT that cannot be adequately controlled medically and is not tolerated hemodynamically (Level of evidence: C).
 2. Asymptomatic ventricular ectopy or tachycardia that is not suspected of causing ventricular dysfunction (Level of evidence: C).
 3. Ventricular arrhythmias due to transient reversible causes, such as acute myocarditis or drug toxicity (Level of evidence: C).

*Catheter ablation for ventricular arrhythmias in children should be performed only by centers and physicians with expertise in ablation therapy in pediatric patients

Appendix 1

See Tables A1 and A2

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Appendix 1 PACES/HRS VT writing group disclosures

Writing group	Employment	Consultant/ advisory board	Speakers' bureau/ honoraria	Research grant	Fellowship support	Board Mbs/ stock options/ partner	Others
David J. Bradley, MD, FACC, FAAP	University of Michigan	None	None	None	None	None	None
David J. Callans, MD, FHRS, FACC, FAHA	Hospital of the University of Pennsylvania	1; Biosense Webster Biotronik, Medtronic, St. Jude Medical	None	None	None	None	None
Jane E. Crosson, MD, FACC	Johns Hopkins University	None	None	None	None	None	None
Anne Dubin, MD, FHRS, FAHA, FACC	Stanford University, Pediatric Cardiology	None	None	None	2: Medtronic	None	None
Susan Etheridge, MD, FHRS, FACC, FAAP, CEPS	University of Utah, Department Pediatric Cardiology	None	None	None	None	0: SADS board member/Vice President, PACES Treasurer, ACC member, AAP member, AHA members	None
Andrew Papez, MD, FHRS, CCDS	Arizona Pediatric Cardiology Consultants	1: St. Jude Medical	None	None	None	None	None
John R. Phillips, MD, FAAP, FHRS, FACC	West Virginia University Children's Hospital	None	None	None	None	None	None
Larry A. Rhodes, MD, FAAP, FHRS, FACC	West Virginia University School of Medicine	None	None	None	None	None	None
Elizabeth Stephenson, MD, MSc, FHRS, CCDS, CEPS	The Hospital for Sick Children-Cardiology	None	1: Glaxo-SmithKline, Medtronic Minimed, Johnson & Johnson	4: Medtronic Minimed, Boehringer Ingleheim	None	None	2: Neurometrix
William Stevenson, MD, FAHA, FHRS, FACC	Brigham and Women's Hospital Cardiac Arrhythmia Service	None	None	None	None	None	0: Johnson and Johnson-intellectual property rights 3: Editor, Circulation Arrhythmia and Electrophysiology-AHA Journal
Frank Zimmerman, MD, FAHA, FHRS	The Heart Institute for Children	None	None	None	None	None	None

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

Appendix 2 PACES/HRS VT peer reviewer disclosures

Peer review	Employment	Consultant/advisory board	Speakers' bureau/honoraria	Research grant	Fellowship support	Board	
						Mbs to members	Others
Victoria Vetter, MD, FHRS	Children's Hospital of Philadelphia	None	None	None	None	None	None
Bradley Knight, MD, FHRS	Northwestern University, Feinberg School of Medicine	2; Boston Scientific, Medtronic, St. Jude Medical, Biosense Webster, Estech	2; Biotronik, Biosense Webster, Boston Scientific, Medtronic	None	3: Boston Scientific, Medtronic, St. Jude Medical, Biotronik, Biosense Webster	None	Royalty income, 2: Up to date
Peter Aziz, MD, CEPS	Cleveland Clinic Foundation	None	None	None	None	None	None
Mina Chung, MD, FHRS	Cleveland Clinic	0; Boston Scientific, Medtronic, St. Jude Medical, Zoll Medical Corp, Amarin, Biotronik 1; National Institutes of Health, Japanese Society of Electrophysiology, Japanese Circulation Society, Axio Research	1: American College of Cardiology Foundation	5: National Institutes of Health	None	None	Royalty income; 1; <i>UpToDate</i> , Jones & Bartlett Publishers

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