Ventricular Tachycardia

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Nicholas Von Bergen and Craig J. Byrum

Introduction

Physicians caring for the young, when faced with a wide QRS complex tachycardia on electrocardiogram, are often hesitant to assign a diagnosis of ventricular tachycardia (VT), especially since the patient is often minimally symptomatic. Though young patients frequently present with rate-related aberrancy associated with supraventricular tachycardia (SVT), VT accounts for an estimated ~80 % of all wide complex rhythms across all ages. The incidence of VT in the pediatric population has been estimated around 1 in 100,000, though studies have suggested that short undetected episodes of VT may occur rarely in the general population.

When confronted with a wide QRS complex tachyarrhythmia in a child, it is always important to first consider the diagnosis of VT, and then look for other causes of the wide complex tachyarrhythmias when the diagnosis of VT has been excluded. Although most VT in young patients occurs in the setting of a structurally normal heart, those with underlying heart disease are at greater risk. As many as 15 % of patients with complex congenital heart disease (native or repaired) may develop ventricular arrhythmias over a lifetime which may be associated with an increased risk sudden death or a cardiac arrest. In addition, VT may develop in the setting of acquired myocardial disease, which can be either acute (ischemic, toxic, or inflammatory) or chronic (myopathic, neoplastic) and either focal or diffuse. Examples of some of these myocardial disorders include myocarditis, Kawasaki disease, Chaga's disease, tricyclic poisoning, focal tumors, arrhythmogenic right ventricular dysplasia (AVRC, fatty replacement areas), peripartum cardiomyopathy, and dilated or hypertrophic cardiomyopathy.

As the clinical presentation is a function not only of the rate of the tachycardia but also the underlying state of the myocardium, and the resultant blood pressure, young individuals may tolerate even sustained VT for lengthy periods of time. Therefore, the clinical response to VT should be tailored to the individual presentation and may range from conservative management to emergent cardioversion and invasive management.

Ventricular tachycardia implies an arrhythmia that arises from the working ventricular myocardium. Some forms may arise from the distal branches and the Purkinje network. Thus, junctional ectopic tachycardia will be excluded from the present discussion as it originates from the AV node—His bundle area.

N. Von Bergen, M.D. (✉)
Department of Pediatrics, University of Wisconsin—Madison, 1675 Highland Ave., Madison, WI 53792, USA
e-mail: vonbergen@pediatrics.wisc.edu

C.J. Byrum, M.D.
Pediatrics–Cardiology, Syracuse, NY, USA

M. Dick, II (ed.), *Clinical Cardiac Electrophysiology in the Young*,
DOI 10.1007/978-1-4939-2739-5_13

Electrocardiographic (ECG) Diagnosis of VT

There continues to be debate among electrophysiologists regarding the most comprehensive ECG characteristics to diagnose VT. Using relatively intuitive criteria VT can be diagnosed by:

- A QRS complex greater in duration than normal for age—i.e., a wide QRS complex (Fig. 13.1).
- Atrial (A) and ventricular (V) dissociation with more ventricular complexes than atrial complexes (Fig. 13.2).
- A rate greater than 25 % above that of normal sinus rhythm, or greater than 120 bpm in adults.

However, these criteria are not necessarily applicable to the young patients. VT in infants may be associated with QRS durations of only ~80 ms, requiring comparison of the VT-QRS duration and morphology to age-appropriate norms. Although retrograde ventricular-atrial dissociation during VT is virtually diagnostic for VT, a good number of patients may display 1:1 (Fig. 13.1) or retrograde Wenckebach conduction during VT indicating that VA dissociation is not a prerequisite for VT. Because AV dissociation may be difficult to determine, a long rhythm strip may assist in its identification. Adenosine may be given to transiently block retrograde AV nodal conduction to assist in making the diagnosis (Fig. 13.2). Additionally, capture and fusion beats may be identified by a sinus-driven beat conducting antegrade through the AV node and influencing the initial deflection (fusion beat) or normalizing (capture beat) the QRS complex. When this is seen, it suggests a ventricular origin of the wide complex arrhythmia (Fig. 13.3). The likelihood of observing these beats is inversely proportional to the VT rate. In asymptomatic patients with a ventricular rate within ~125 % of the sinus rate an accelerated idioventricular rhythm (AIVR) is likely and is associated with a benign course. AIVR is somewhat more common in infants and children than adults.

In some cases, it may be difficult to distinguish VT from an atrial-driven rhythm with aberrancy of QRS conduction. To improve the diagnostic sensitivity suggested expanded criteria may be employed:

- A QRS complex with both an initial and a terminal conduction delay Fig. 13.4a
- A shift in the QRS axis by more than 40° from baseline
- Fusion beats or capture beats (Fig. 13.3)
- A QRS axis between −90 and 180°
- QRS duration greater than 0.14 s (with RBBB) or 0.16 s (LBBB)
- A taller initial (leftward) peak in the QRS complex in a right bundle branch complex
- A delayed S wave nadir with a notched S down stroke in a left bundle branch complex (Fig. 13.4b)

Though many of these criteria may be useful, they remain imperfect as the sensitivity may be

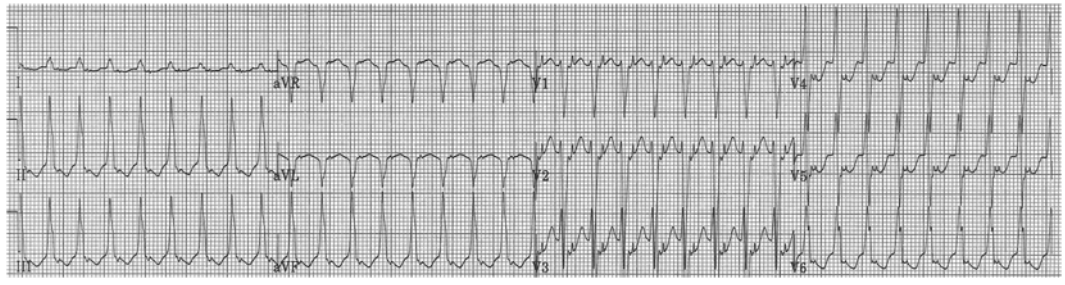

Fig. 13.1 An ECG of ventricular tachycardia presenting in a 3-day-old with mild LV dysfunction. The tachycardia exhibits a left bundle branch block with an inferior axis suggesting origin of the tachycardia from the right ventricular outflow tract. This VT has 1:1 VA conduction, with the P waves most easily seen in V1 and V2

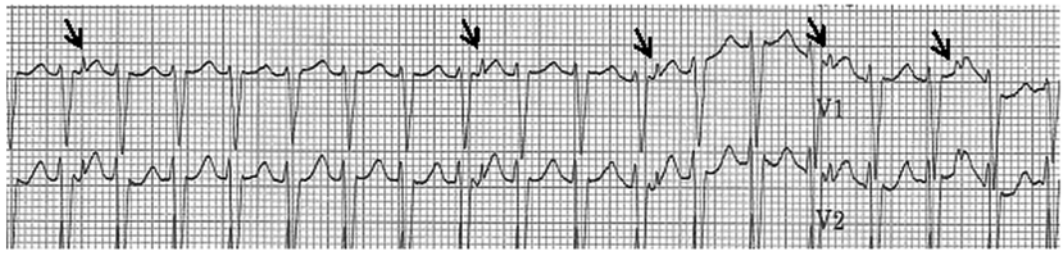

Fig. 13.2 An ECG rhythm strip on the patient from Fig. 13.1 after an adenosine bolus. Note the loss of retrograde VA conduction with adenosine administration confirming VT. Arrows indicate the retrograde P waves

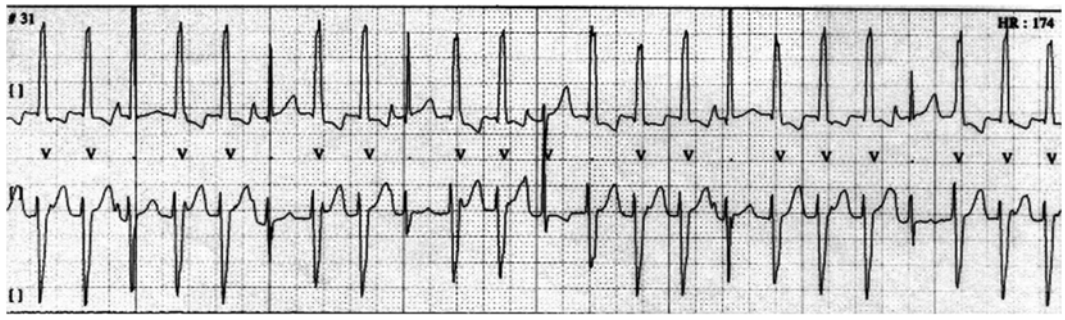

Fig. 13.3 A Holter monitor strip showing lack of VA conduction and fusion complexes (beat three and others) and sinus captures (beat six and others)

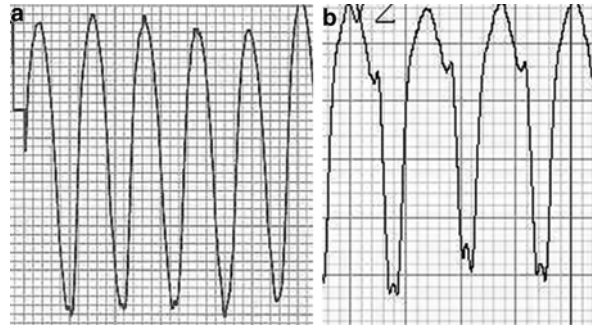

Fig. 13.4 (**a**) A 16-year-old male with VT associated with ARVC showing the initial and terminal conduction delay from V1. (**b**): A notched S negative deflection in a patient with an LBBB appearance VT from V2 from a different patient

below 50 %. Therefore, care should be used when applying them, and other potential criteria to wide QRS tachyarrhythmia diagnosis.

Often vector analysis can be employed to discern the origin of the VT. Because the wave front moves away from the point of origin, an ECG complex in the precordial leads that has predominately leftward forces (with an LBBB appearance) predicts an origin from the right ventricle and, conversely, predominately rightward forces (with an RBBB appearance) predicts an origin from the left ventricle. Further, a frontal plane (from the limb leads) superior QRS axis of the VT suggests origin from the inferior ventricular segments, and conversely, a frontal plane inferior QRS axis suggests origin of the VT from the base of the heart, usually the right ventricular outflow tract or coronary cusps.

Differential Diagnosis of Wide QRS Tachycardia

A wide QRS tachycardia in a young patient strongly suggests VT; however, other possibilities include (Fig. 13.5):

- Supraventricular arrhythmia with rate-related aberrancy (Figs. 13.5a and 13.6)
- Antidromic supraventricular tachycardia (Fig. 13.5b)
- An atrial tachycardia with preexcitation, including a Mahaim fiber (Fig. 13.5c, d)
- Ventricular tachycardia (Fig. 13.5e)
- Tachycardia with a preexisting bundle branch block
- A ventricular paced rhythm

Aberrant conduction (aberrancy) results from a delay in the recovery of the relative refractory period of the bundle branches (usually the right as it has a longer refractory period than the left) as the heart rate changes quickly producing a left or right bundle branch morphology (Figs. 13.5a and 13.6).

At times preexisting wide QRS morphology may manifest as an abnormal wide QRS tachycardia driven by an atrial tachycardia (Fig. 13.7). When possible, especially in patients with congenital heart disease, comparison of the tachycardia QRS morphology to a baseline ECG may be helpful in distinguishing a preexisting wide QRS from a true ventricular arrhythmia.

Other forms of wide QRS tachycardia include antegrade conduction during both sinus rhythm and SVT through a Mahaim fiber (Fig. 13.5d) or through an atriofascicular accessory pathway

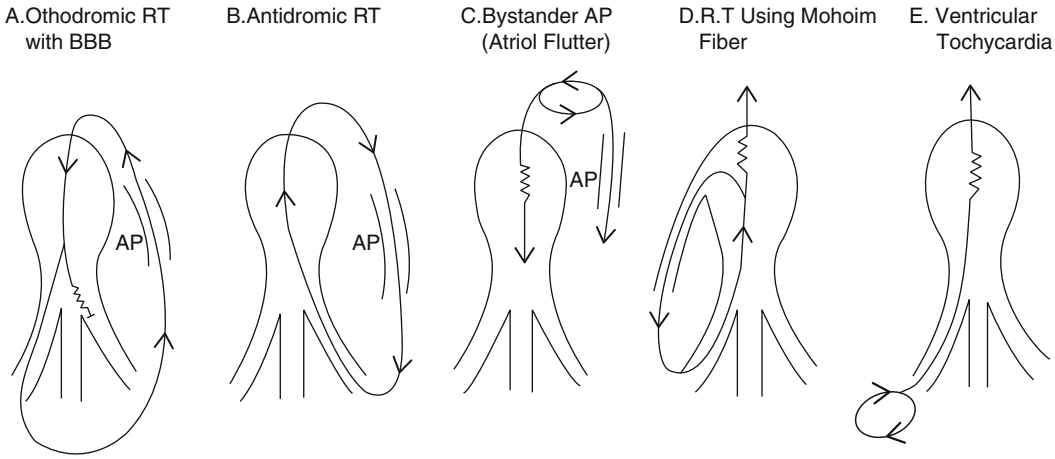

Fig. 13.5 See Differential Diagnosis [Reprinted from Benson DW, et al. Mechanisms of regular wide QRS tachycardia in infants and children. American Journal of Cardiology 1982;49(7):1778–1788. With permission from Elsevier.]

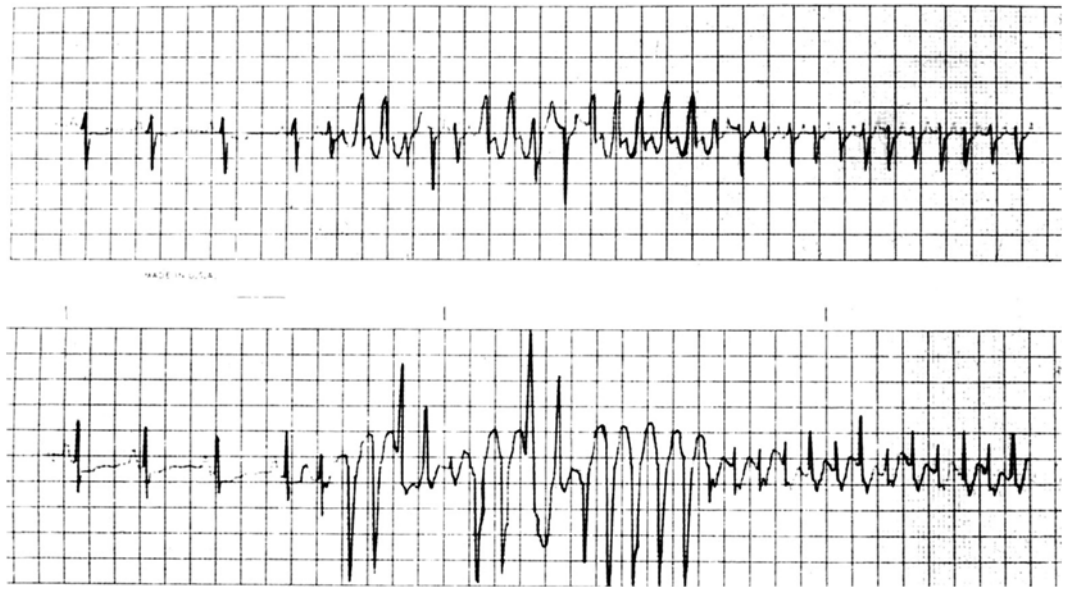

Fig. 13.6 Holter tracing of an infant with SVT and aberrancy [Figure is not a continuous ECG]—i.e., rate-related bundle branch block—both right (positive-wide QRS complexes) and left (negative-wide QRS complexes)—due to the atrial tachycardia impulses encountering relative refractoriness in the bundle branches. The refractory periods shorten in response to the shortened cycle lengths of the tachycardia, eventually normalizing the QRS complex (right-sided beats of upper and lower tracing)

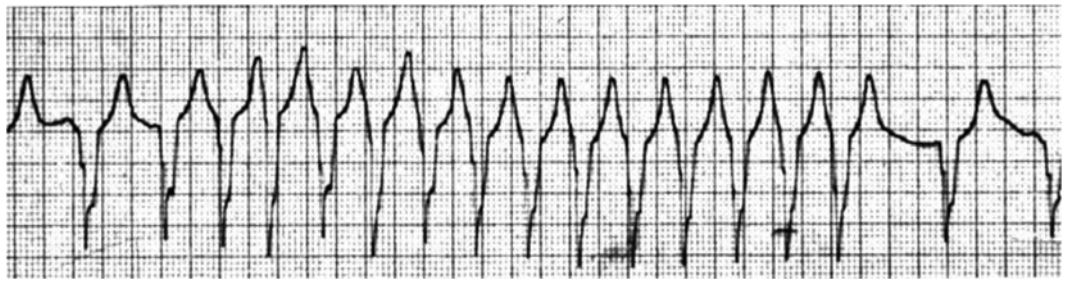

Fig. 13.7 Wide QRS tachycardia in a patient with transposition of the great arteries and ventricular septal defect following the Rastelli operation. Note the first and the last two slow wide QRS complexes are identical to the wide QRS complex tachyarrhythmia, indicating that the wide QRS complex tachycardia is due to a preexisting conduction abnormality

(not shown in Fig. 13.5); both of these anomalous connections are right sided so the wide QRS morphology in both sinus rhythm and SVT is similar and of a LBBB configuration. Rarely, atrial flutter or fibrillation (Fig. 13.5c) in the presence of preexcitation produces a wide complex rhythm due to rapid conduction through the accessory pathway. In other unusual cases, an accessory pathway may provide the atrioventricular limb of a reentrant circuit maximally activating (preexciting) the ventricular myocardium followed by retrograde (antidromic) conduction through the His–Purkinje system—AV node to "reenter" the chambers of origin (atria), resulting in an antidromic tachycardia (Fig. 13.5b); (see Chaps. 4 and 20).

Table 13.1 Factors to consider when classifying VT

Non-sustained	Sustained
Monomorphic	Polymorphic
No heart disease	Structural or function myocardial disease
Focal myocardial process	Diffuse myocardial process
Specific drug response (e.g., calcium blocker)	No specific drug response
Exercise suppressible	Exercise induced
Automatic or triggered automatic	Reentry or multiple reentrant
No underlying electrical disease	Underlying electrical disease

Table 13.2 Classification of VT by duration

Duration of VT	Description
Salvo	A few beats in a row
Non-sustained VT	Typically 3–4 beats in a row up to runs of 30s in duration
Sustained VT	An episode longer then 30s, or requiring termination due to hemodynamic compromise
Repetitive VT	Multiple salvos or non-sustained VT events in close proximity
Incessant VT	Lengthy sustained VT that often dominate the cardiac rhythm

Classification of Ventricular Tachycardia

One may classify VT from a variety of frames of reference including duration, morphology, etiology, association with underlying heart disease, relationship to physical activity and presumed origin of the tachycardia of the origin (Tables 13.1 and 13.2).

Electrocardiographic Morphology

Monomorphic VT refers to one dominant QRS form during VT whereas polymorphic refers to a changing QRS shape and beat-to-beat interval within the episode.

A form of polymorphic VT is "bidirectional tachycardia" where the QRS alters between one form and another, typically flipping its axis every other beat across the baseline. This may be seen in a portion of patients with catecholaminergic polymorphic VT (CPVT), with digoxin toxicity, or with some inherited arrhythmias. Polymorphic VT is considered more predisposed to degeneration to ventricular fibrillation.

Electrophysiologic Mechanism

Understanding of the mechanism of VT may be important in deciding on either medical management strategies or invasive management strategies. Three known arrhythmia mechanisms occur: reentry, automaticity, and triggered automaticity. Reentrant arrhythmias in pediatric and congenital heart patients are seen in the scar formation after cardiac surgery, with myocardial ischemia due to coronary artery anomalies, or with abnormal coronary perfusion related to chronic abnormal loading conditions. These the scarred areas, by virtue of their slow conduction property and their anatomic relationship to other scarred areas or to naturally occurring electrically inert borders like AV grove, may form corridors of slow electrical conduction through which electrical activity may enter, exit, and then reenter. Stretch on the myocardium may induce an extra depolarization and imitate a reentry VT in patients with abnormal loading condition.

Clinical Presentation

Ventricular tachycardia may present with a variety of clinical scenarios. It is not infrequent to encounter patients who are completely asymptomatic with the diagnosis. Often these young patients have no underlying structural heart disease and may have either repetitive non-sustained VT or sustained but rather slow VT. Those patients with non-sustained VT may be picked up by finding an irregular heartbeat during routine examination. There are some patients who have a sensation of a racing heart others, with minimal symptoms. Associated symptoms during tachycardia may include dizziness, particularly

at the onset of tachycardia, vague chest discomfort, some dyspnea, weakness and headache. Surveillance with regular ECG (Holter) monitoring may be helpful; to detect and evaluate this potential late complication.

Sustained VT over time may lead to tachycardia-mediated cardiomyopathy. In the young patient presenting with clinical congestive heart failure in the presence of a chronic, modestly increased rate, VT, in an otherwise structurally normal heart, presents a diagnostic challenge; determination of the primary cause—VT or primary myocardial disease—is required. VT secondary to a primary dilated cardiomyopathy may be best treated with medication and possibly implantable cardio-defibrillator, though the value of the defibrillator is unproven. On the other hand, tachycardia induced heart failure patients may best benefit from primary treatment of the tachycardia either through medication or catheter ablation. Often the cardiac function will return to normal with the elimination of symptoms.

The most serious but rare clinical presentation of VT in the young is syncope or sudden death. These presentations are unusual in a structurally normal heart save for those with underlying electrical disease (inheritable arrhythmia syndromes, see Chap. 19) or those with infiltrative or hypertrophic diseases of the myocardium. Rather, the number of patients with this presentation will be those who have had surgery for congenital heart disease or those who have an acutely or chronically dysfunctional myocardium after infectious or inflammatory insult.

Clinical Investigation

History

As seen in patients with supraventricular tachycardia, patients who perceive VT will typically report a sudden onset and cessation of the arrhythmia. A history of acquired or congenital heart disease and therapy, substance abuse, possible exposure to toxins or contact with infectious agents (viruses) is important. The family history should be explored vigorously, often with a trained genetic counselor, focus on syncope, seizures, sudden death, or specific inheritable cardiac arrhythmia diagnoses.

The Physical Exam

During an arrhythmia, the examination findings reflect the presence of the tachycardia as well as the inefficiency of cardiac contraction and the AV dissociation. The rate would be elevated and pulses may be weak. The observation of cannon waves in the neck reflecting episodic atrial contraction against a closed AV valve would be useful. Diastolic filling sounds may reflect underlying primary cardiomyopathic condition. Abnormal heart sounds, murmurs, rubs, and chest scars will suggest the likely presence of congenital heart disease and the related surgical palliations or corrections. Rarely will VT will respond to vagal maneuvers.

Echocardiogram

Patients with ventricular arrhythmias require an echocardiogram to rule out structural heart disease and to evaluate cardiac function. In addition for those without congenital heart disease, echocardiography (with Doppler) should focus on possible tumors, ventricular hypertrophy as well as myocardial dysfunction brought on by abnormalities of preload or afterload or by conditions primarily affecting contractility.

Cardiac Magnetic Resonance (CMR)

MRI appears to offer the advantage of viewing the ventricles in three dimensions and it may give a better view of both anatomy and function. In addition, it provides a useful sensitivity for detecting myocardial abnormalities such as areas of fatty replacement in the patient with arrhythmogenic right ventricular dysplasia. Delayed enhancement and the presence of fibrosis on CMR have also been suggested as a marker of risk for a patient to develop arrhythmias in hypertrophic cardiomyopathy and other congenital heart populations. In selected cases with scar-related VT, CMR may also assist in the planning process for scar/fibrosis mapping in patients who may undergo an electrophysiologic study and ablation. In selected cases, CMR images may be imported into and merged with electro-anatomical

mapping systems used during catheter electrophysiologic assessment to enhance and clarify points of intracardiac anatomy.

Holter Monitor

Holter monitoring is useful for assessing the results of ablation or medications. It can also give one a measure of the density (percentage of tachycardia beats relative to total beats) of non-sustained VT episodes that may serve to trigger sustained VT episodes.

Treadmill Exercise Testing

Treadmill exercise testing can be helpful for diagnosis of exercised-provoked VT (such as CVPT) and in ascertaining the effectiveness of antiarrhythmic therapy or ablative therapy. Some tachycardias are suppressed by exercise and increasing sinus rates but the prognostic significance of that observation is unclear.

Hemodynamic Catheterization

As part of the electrophysiologic study, hemodynamic data and angiography may be helpful to determine the current hemodynamic status of the heart and circulation, especially in patients with congenital heart disease. Since the primary goal in the management of ventricular tachyarrhythmias in this population is to first improve the hemodynamic status with medical or surgical therapy when appropriate, hemodynamic catheterization and angiography can provide directions. In addition, the coronary arteries are best imaged by the transcatheter-selective coronary approach.

Electrophysiology Study With/Without Ablation

Electrophysiologic investigation may be indicated for those patients with documented non-sustained VT, symptoms suspicious for tachyarrhythmia or a high density of premature ventricular systoles. It may also be indicated to evaluate for sustained VT in those patients with underlying structural heart disease and the effectiveness of orally administered antiarrhythmic therapy. The goal is to determine if the patient has inducible VT that places the individual at higher risk. Even so, sensitivity and specificity of an EP study remains imperfect for prognosis of an arrhythmia.

An ablation may be considered in patients presenting VT if there is substrate that may be effectively eliminated. This is often considered the first line for patients with a monomorphic VT associated with symptoms or functional/structural abnormalities. In others, without a substrate that can be reliably ablated such as channelopathies, or polymorphic VT, an ICD should be considered in place of ablation therapy.

Common Types of Ventricular Tachycardia in the Pediatric Population

Accelerated Ventricular Rhythm (AVR)

This arrhythmia simulates VT but is usually not considered a VT. It has characteristics similar to VT (Figs. 13.8 and 13.9).

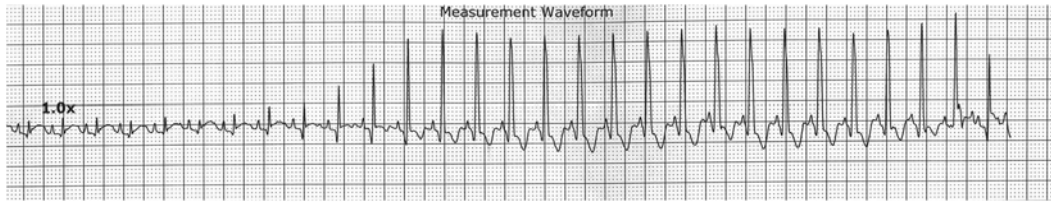

Fig. 13.8 Accelerated ventricular rhythm in 22-day-old infant at virtually the same rate as the sinus tachycardias. Note the fusion beats at the beginning of the AVR and at the end as the sinus impulse partially captures ventricular activation. Note also the more narrow QRS of the AVR that is likely age related

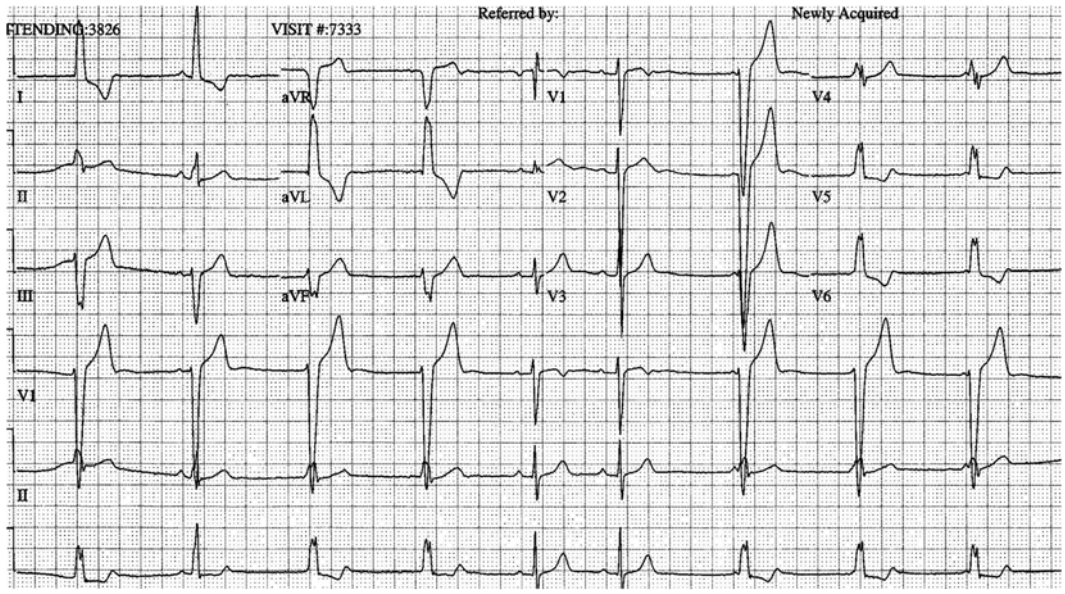

Fig. 13.9 Ten-year-old girl with a normal structural heart and a "slow" ventricular rhythm

Diagnostic ECG Features of AVR

(a) At least three beats of a ventricular rhythm which is ≤125 % of the rate of the sinus rhythm.
(b) Fusion beats often appear at the onset and termination of the arrhythmia.

Background

AVR is seen in all age groups and has been described in the presence of heart disease. In pediatrics, it segregates to the infant age group and is usually found in patients with a normal heart while in older adults it may be associated with coronary artery disease. It has also been seen with medications, digitalis intoxication, myocarditis, and other cardiomyopathies.

If the surrounding sinus rate is elevated, an accelerated idioventricular rhythm will also be proportionately elevated. Care needs to be taken when diagnosing AVR so as not to label it the more serious ventricular tachycardia.

Evaluation

The diagnosis and evaluation of AVR is similar to the evaluation for premature ventricular beats that includes evaluation for symptoms and density (frequency relative to total beats).

Treatment

Treatment, if necessary, of AVR should be done under the guidance of a pediatric cardiologist.

(a) Accelerated ventricular rhythm is benign so usually no treatment is necessary.
(b) If an etiology is identified, then it should be addressed if possible.
(c) If there is evidence of ventricular dysfunction or symptoms, then medication or electrophysiology study and ablation be may considered, but this would be the exception.

Outcome

AVR is well tolerated and often spontaneously resolves. If AVR is associated with an inciting

cause (such as a medication or myocarditis), then AVR will likely resolve once the cause is removed. In patients with structurally normal hearts and no obvious etiology, the course can range from resolution to intermittent sustained runs of AVR.

Right Ventricular Outflow Tract Ventricular Tachycardia

ECG features:

(a) Wide complex with a left bundle branch block morphology.
(b) An inferior QRS axis.

Background

Right ventricular outflow tract tachycardias are the most common form and site of origin of VT in the pediatric population. The posterior wall of the conal septum of the RVOT is in close proximity to the aortic outflow tract, valve and coronary cusps, all just posterior to it. Therefore, careful evaluation of the left-sided outflow tract and coronary cusps should be considered when approaching an ablation of RVOT-VT, especially if ambiguity is encountered in mapping the VT origin. Lead I may help distinguish if this arrhythmia is more likely to be RVOT-free wall (more upright), or RVOT septal (more isoelectric or negative; more detailed localization techniques have been described. RVOT-VTs may be either suppressed or evoked with exercise. Consideration of other causes of RV tachycardia such as Arrhythmogenic Right Ventricular Tachycardia (ARVC) needs to be entertained when evaluating patients with right-sided arrhythmias as this disorder carries a more serious prognosis (Figs. 13.10, 13.11, and 13.12).

Outcome

The decision to treat patients with RVOT tachycardia depends on patient symptoms in combination with cardiac function. If RVOT-VT is most often just a single beat or 2–3 beats and without symptoms, it is usually well tolerated. However, it may present with episodes of sustained arrhythmias. In rare cases, this arrhythmia may cause tachycardia-induced cardiomyopathy. Fortunately, RVOT-VT can often be successfully ablated, restoring cardiac function.

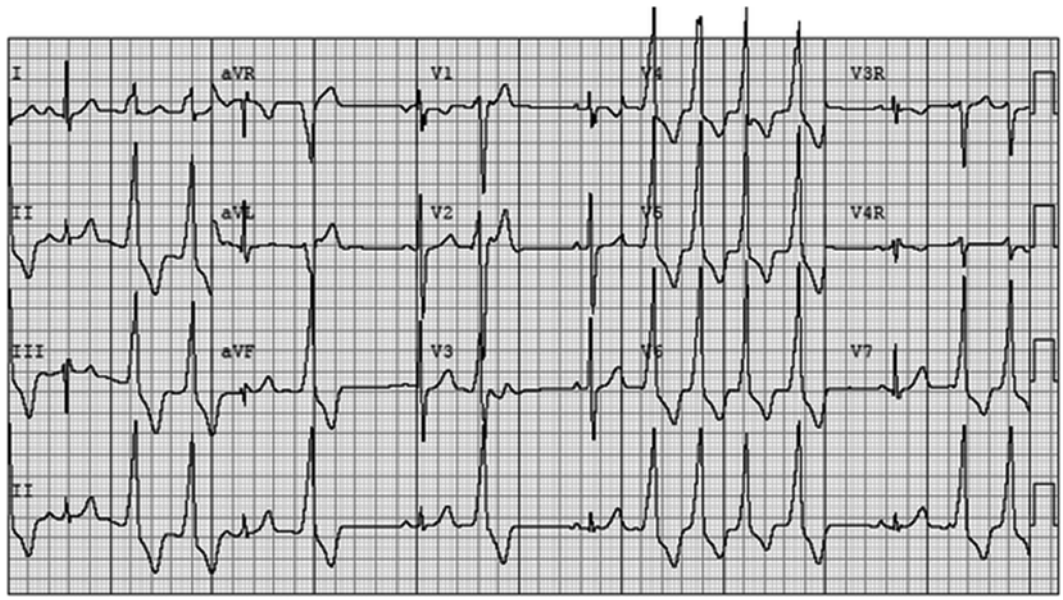

Fig. 13.10 This ECG shows ventricular ectopic activity occurring in single PVCs and repetitively as VT salvos in a 12-year-old with no symptoms but with mild LV dilation. This ECG is consistent with an RVOT-VT, however, because the origin of this RVOT-VT was mapped to the left side of the right ventricular outflow tract—i.e., high on left side of the conal or infundibular septum—it was successfully ablated in the left coronary cusp

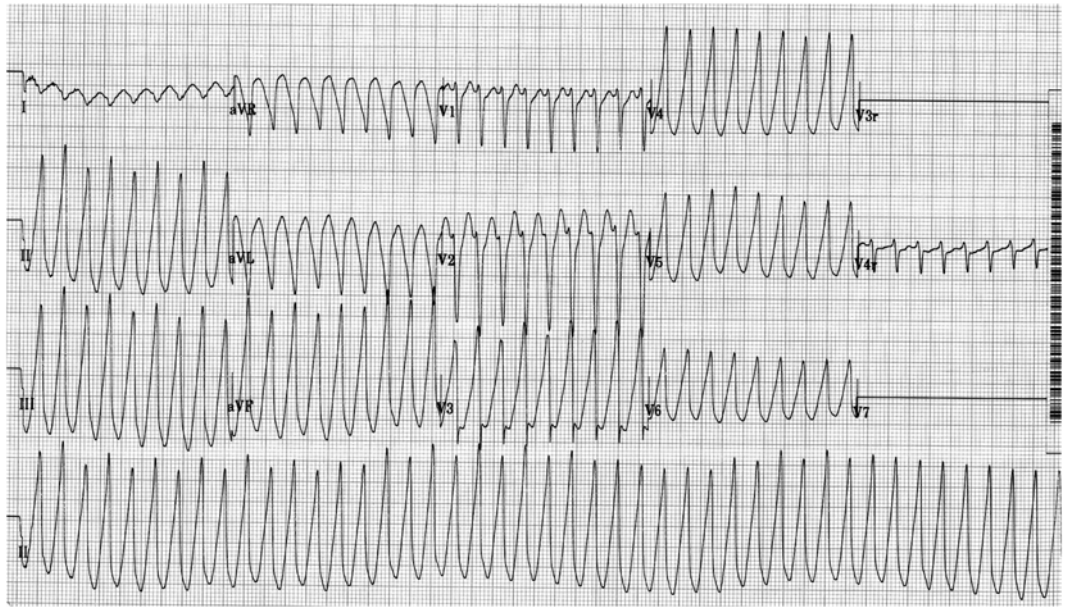

Fig. 13.11 RVOT-VT in a 16-year-old boy with an otherwise normal heart. Cryoablation was successful in the left coronary cusp

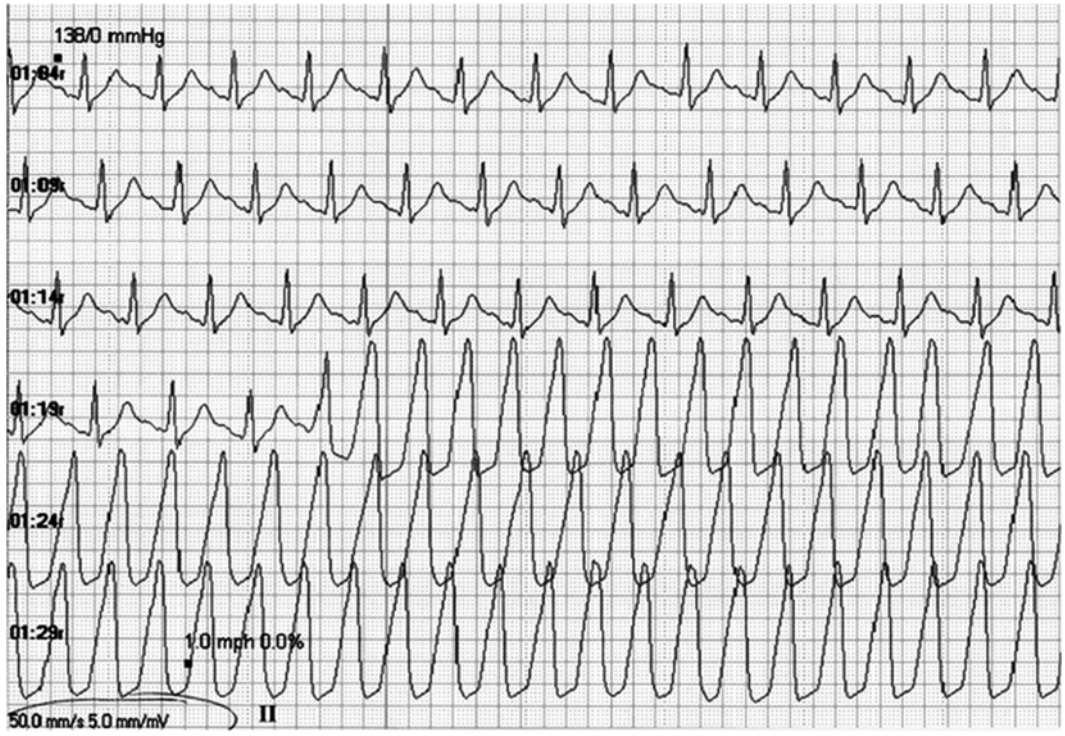

Fig. 13.12 Initiation of VT during treadmill exercise test—same patient as in Fig. 13.10

Left Posterior Fascicular Reentrant Tachycardia

ECG features:

(a) Wide complex with right bundle branch block morphology.
(b) A superior QRS axis (Figs. 13.13, 13.14, and 13.15).

Background

Although less common than RVOT tachycardia, left ventricular posterior fascicular VT is well known in pediatric patients and is most often associated with a structurally normal heart. This arrhythmia is supported by a reentry circuit within the distal fibers of the left posterior fascicle. The impulse propagates retrogradely through the left side of the His–Purkinje system to the contralateral right bundle branch slightly later as manifested by the right bundle branch block configuration, resulting in a slightly narrower QRS complex.

Outcome

Left posterior fascicular reentrant tachycardia is generally unresponsive to adenosine and

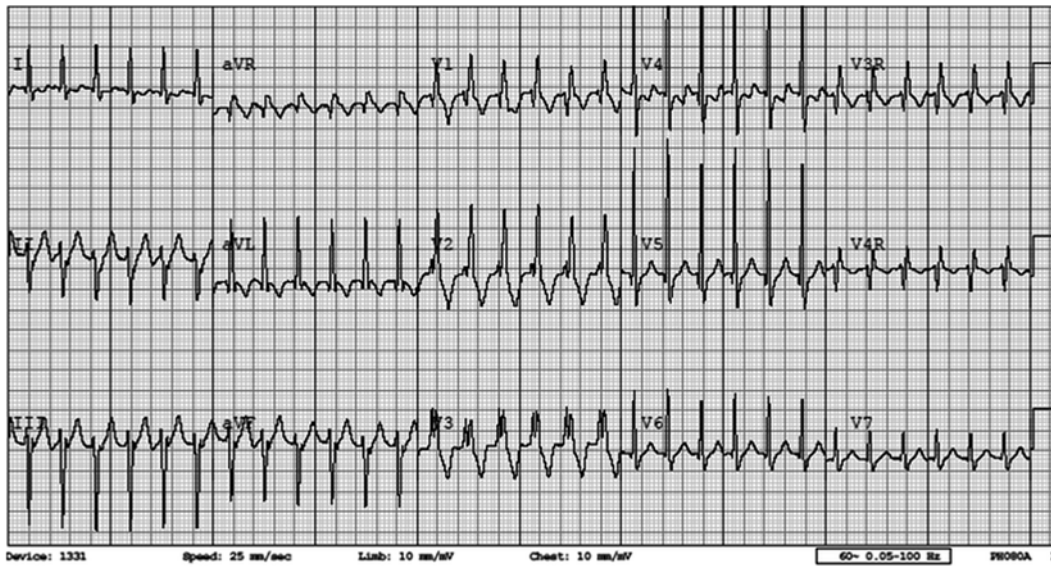

Fig. 13.13 Electrocardiogram in an 11-year-old presenting with findings stained tachycardia. This wide complex tachycardia exhibits right bundle branch block morphology and left superior axis and is narrower than the usual VT that involves mostly working myocardium. This tachycardia is highly suggestive of left posterior fascicular VT. After conversion to sinus rhythm with a calcium channel blocker this patient underwent radiofrequency ablation along the mid-low septum in the region of the left posterior fascicle permanently eliminated the VT

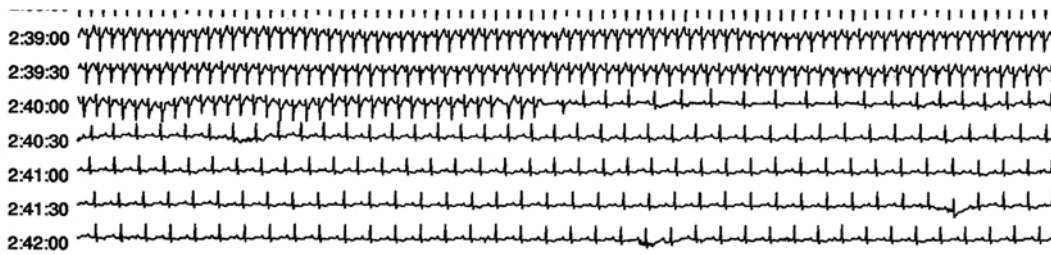

Fig. 13.14 Monitor tracing in an 8-year-old boy with left posterior fascicular ventricular tachycardia acutely and successfully treated with a verapamil bolus

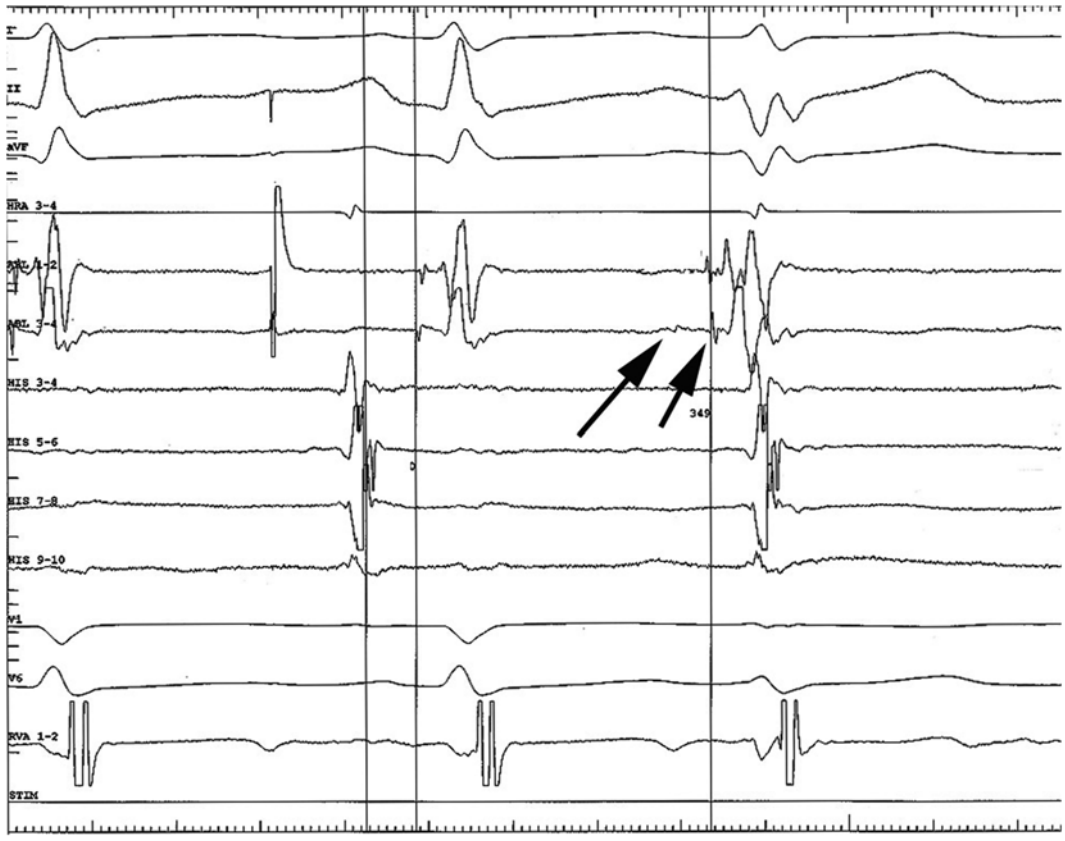

Fig. 13.15 Intracardiac electrograms demonstrating the activation sequence in the left posterior fascicle in sinus rhythm (second beat) and in the premature ventricular beat identical in morphology of the QRS complex during the left posterior fascicular tachycardia (third beat). The fifth and sixth tracings from the top are electrograms recorded through the distal (ABL 1-2) and proximal (ABL 3-4) electrode pairs on the ablation catheter. Note the proximal pair (sixth tracing) in the third beat is preceded by a prepotential (*left arrow*) before the left posterior fascicular signal (*right arrow*). Also notice the electrode pair (ABL 3-4) in the second (sinus) beat is activated before the distal electrode pair that is further (i.e., apical) along the fascicle. In contrast, the activation sequence is the reverse for the VPB indicating a change in the activation pattern for it and the VT. This VT is also amendable to successful ablation in selected patients. Entry into the left ventricle via the retrograde (larger patients) or transseptal approach provides access to the left posterior fascicle identified by a potential representing the specialized conducting left posterior fascicle

beta-blockers, but it typically exhibits a calcium channel blocker sensitivity providing both the acute and long-term treatment of this arrhythmias.

Ventricular Tachycardia in the Setting of Repaired Congenital Heart Disease

Background

Ventricular arrhythmias including VT following congenital heart disease surgery is a well known, late-term complication. Age at operation, interval from surgery, QRS duration and pulmonary regurgitation have been identified as risk factors for ventricular tachycardia and sudden cardiac death; despite these observations, the overall survival of this group remains favorable into the fourth decade and beyond. Ventricular extrastimulation at electrophysiology study may yield further risk stratification and prompt therapeutic considerations such as medication, ablation, or device therapy (Fig. 13.16).

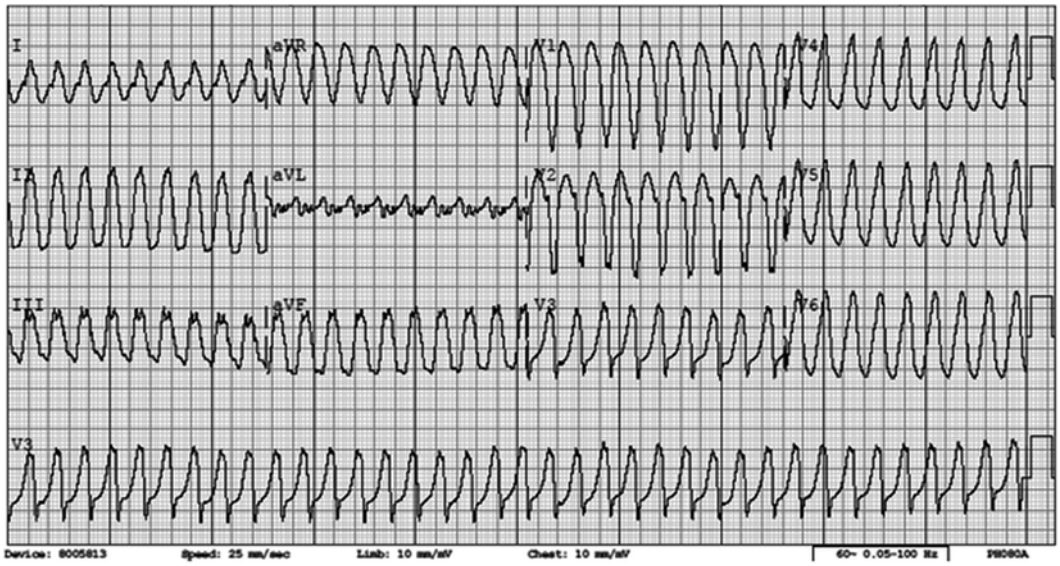

Fig. 13.16 This monomorphic VT presented in a 27-year-old late after repair of tetralogy of Fallot. The inferior axis and left bundle branch block morphology predicted origin from the right ventricle near his RVOT outflow patch. This VT was successfully ablated

Ventricular tachycardia in the setting of genetic or molecular cellular abnormalities: (see Chap. 18)

Some ventricular tachycardias are seen in the setting of genetic potassium, sodium, or calcium ion channel defects producing abnormalities in depolarization and repolarization of the sarcolemma (see Chap. 19). The most frequent is the congenital long QT syndromes due to alterations in structure and function of transmembrane potassium or sodium channels. The Brugada syndrome (sodium channel defect) has been identified as an important cause of sudden death in certain subpopulations. Some of these ventricular tachycardias are catecholamine provoked, such as in catecholaminergic polymorphic ventricular tachycardia.

Other entities are more likely to show ventricular arrhythmias after pauses in cardiac rhythm as in the pause-dependent type torsade de pointes arrhythmia or rarely, in the presence of complete heart block and profound bradycardia. However, even with advances in genetic testing there remains a number of young patients presenting with VT or sudden death in which no underlying structural, functional, electrical, or genetic abnormality has been identified.

Treatment of VT

Immediate visual and physical inspection of the patient is critical assessing heart rate, reparations, color, and perfusion. Acute treatment should follow Pediatric Advanced Life Support and Advanced Cardiovascular Life Support (PALS, ACLS algorithm: (https://www.acls.net/images/algo-arrest.pdf). In particular, if the patient has evidence of hemodynamic compromise during the VT, then cardiac pulmonary resuscitation (CPR) with compressions should be initiated followed by direct current cardioversion/defibrillation with an automatic external defibrillator (AED) if a "shockable" rhythm (VT or Ventricular fibrillation), identified by the device, is present.

Intravenous amiodarone may also be used in most patients, though has a wide side effect profile (see Chap. 21). Other medications such as esmolol or procainamide may also be considered

during an event. A procainamide slow push over 20 min (10–15 mg/kg) may be effective for VT and would also be expected to convert many other non-VT tachycardias that are dependent on the presence of an accessory pathway. Esmolol has less of a proarrhythmia effect and can also be easily titrated. Blood pressure monitoring is critical as the blood pressure will be in this group of patients and all three of these medications may further lower the blood pressure. Polypharmacy should be avoided.

If patients present without hemodynamic compromise the caregiver, often a bystander and first responder, may tailor the treatment to the history (symptoms), clinical status of the patient, and electrocardiographic findings. The ECG may reveal the more common forms of VT seen in pediatric patients without known heart disease that may pose a life-threatening risk. Left posterior fascicular VT (RBBB morphology with a superior QRS axis) often responds to calcium channel blockers; therefore, a slow verapamil push may be considered. With this arrhythmia and verapamil, caution is advised for use in neonates <6 weeks old. Though not typically used for the treatment of VT, adenosine may occasionally be useful in patients with RVOT-VT.

Arrhythmia Prevention

After initial management and conversion to sinus rhythm, chronic therapy must be considered. In patients with congenital heart disease, one should consider addressing any underlying functional hemodynamic abnormality of the heart as an improvement in function may considerably reduce the VT or reduce symptoms associated with episodes. An example would be reduction by pulmonary valve replacement of right ventricular volume overload due to unrestricted pulmonic insufficiency and subsequent progressive RV dilation in a patient with a transannular patch repair of tetralogy of Fallot.

In patients with minimal symptoms and no underlying structural or functional myocardial disease, conservative management may be appropriate while monitoring for spontaneous resolution of the arrhythmia. It has been suggested that the spontaneous resolution rate of VT may be between 30 and 70 % depending the VT location. In older patients without heart disease, conservative management is also generally appropriate for those with repetitive non-sustained VT in the absence of symptoms and in the absence of significant ventricular dysfunction.

Pharmacological therapy (see Chap. 21) can be guided by the morphology of the tachycardia and the clinical situation. Beta-blockers are one of the most commonly used medications as an initial medication for the suppression of VT, and are generally well tolerated. Beta-blockers may be very effective for patients who have no cardiac malformations, for example, patients with the long QT syndrome. Similarly, oral verapamil therapy is well tolerated and effective for patients with calcium-sensitive left posterior fascicular VT. Digitalis has no direct role in the treatment of ventricular arrhythmias but as a medication given to improve myocardial function it may have indirect benefit along with the use of low dose beta-blockers or afterload reduction therapy.

Type IA antiarrhythmic agents such as procainamide, quinidine, and disopyramide have had a diminishing role over time in the prophylactic treatment of VT primarily due to their high side effect and frequent dosing profiles and the availability of more effective antiarrhythmic medications in other classes. Likewise Type IB agents such as diphenylhydantoin and tocainide are rarely used. Thus, the use of Type IA or Type IB agents should be individualized to situations where a patient cannot benefit from catheter ablative therapy and potentially when the individual medication has been shown at electrophysiologic induction study and clinical follow-up to be beneficial in suppressing the tachycardia.

Type IC agents have been shown to be nearly equally effective in the treatment of both supraventricular and ventricular arrhythmias in the young. However, flecainide should be avoided in patients with congenital heart disease (especially in patients with slow ventricular conduction—long QRS complex, and these are not indicated for PVC suppression. Patients with ischemia-induced arrhythmias may be at increased risk for

ventricular arrhythmias soon after initiating medication; therefore, one should start these medications in a well-monitored setting if there is any underlying structural heart disease.

Type III agents may also be highly effective in the suppression of VT. Sotalol has a significant component of beta-blockade effect in addition to its Type III (block the potassium channel) action. Recently dofetilide, which has been approved for atrial arrhythmias, has been shown to be potentially effective for the treatment of ventricular tachycardia though there remains limited data on its use for ventricular arrhythmias in the pediatric population. Amiodarone can be highly effective; however, the benefit needs to be weighed against the risk of long-term side effects when a young patient is placed on chronic therapy. Amiodarone should be avoided if an electrophysiologic study is contemplated because of the long half-life.

Procedural Management

Surgical management

Transcatheter ablation has virtually replaced the surgical approach to the treatment of ventricular tachycardia. Effective antiarrhythmic medication and implantable defibrillation devices may supplant thoracotomy. The exception may be when the surgical approach may be required for management of hemodynamic lesions as well as ablation of arrhythmias. For lesions such as cardiac tumors, aneurysms, or large ventricular patches, surgical excision may be needed.

Catheter Ablation

Current guidelines for catheter ablation of VT in adults with structural heart disease, prior myocardial infarction or cardiomyopathy include patients with symptomatic monomorphic VT which is drug refractory or for the patient who does not desire medications; for patients having VT storms not due to a reversible cause; for frequent arrhythmias which are presumed to cause ventricular dysfunction; for bundle branch reentry; or for recurrent sustained polymorphic VT/VF which is drug refectory. It also can be considered in patients with VT despite antiarrhythmic therapy and in some patients post-myocardial infarction as an alternative to antiarrhythmic medication. In patients with a structurally normal heart, the procedure is indicated for patients who have symptomatic sustained or repetitive non-sustained type monomorphic VT; for patients with VT not responsive to medication therapy; or for recurrent sustained polymorphic VT/VF where it is felt the trigger can be targeted. In the pediatric population, the most successful experiences in catheter ablation of VT certainly come from patients with a structurally normal heart and a focal origin of their tachycardia.

In patients felt to have sustained monomorphic VT from the right ventricular outflow tract experience confirms that some of these tachycardias will originate from the left side of the outflow septum or within the aortic coronary cusp so the operator needs to be prepared for a possible retrograde aortic and/or transseptal-trans-mitral approach (Fig. 13.17).

In these patients, careful evaluation of the coronary arteries is important to avoid collateral damage during the ablation. Radiofrequency lesions should be at least 5 mm from the coronary arteries; alternatively, cryotherapy may be considered and has less risk of permanent damage if the lesion is halted at the first (electrocardiographic) sign of coronary injury. Another group of patients where catheter ablation can be contemplated are those with arrhythmogenic right ventricular cardiomyopathy. Caution must be exercised in this disease however since the disease process is often progressive and effect of the ablation may be short lived. In these patients, recent studies suggest ablation may be more effective if approached from the epicardium. Other disorders such as Chagas disease or a presumed epicardial VT focus has resulted in an increase in the number of epicardial ablations in adult patients with VT.

Patients with underlying structural heart disease are much more likely to have reentry circuits as the mechanism for their arrhythmia. These reentry circuits often develop around areas of scar, such as that seen after a right ventriculotomy for tetralogy of Fallot repair. In these cases, the superior aspect of the scar may define one boundary of

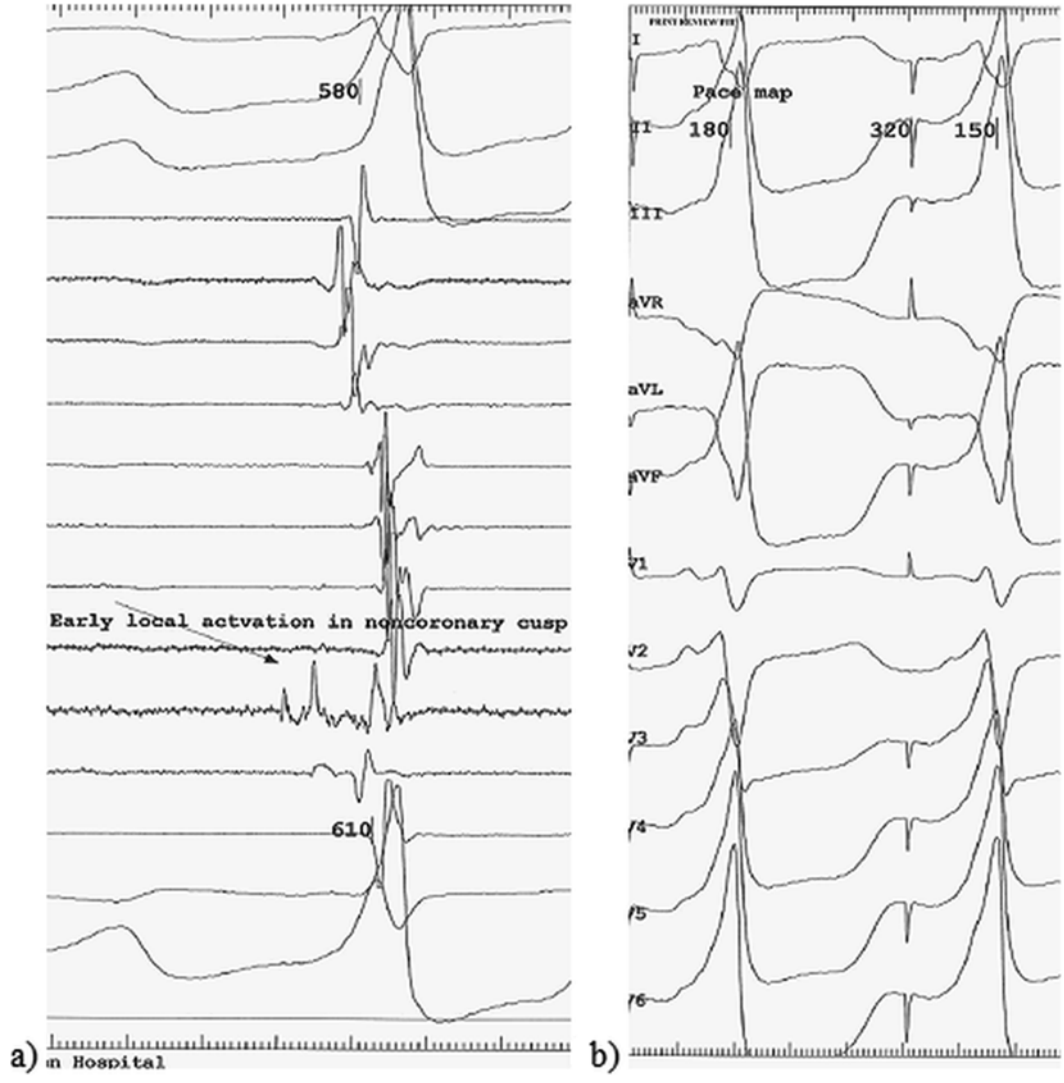

Fig. 13.17 (**a**) Intracardiac electrogram showing the earliest local activation of a focal ventricular arrhythmia within the noncoronary cusp. (**b**) Pace mapping showed 12/12 match between the ventricular arrhythmia (first beat) and the paced ventricular beat (second beat)

a portion of the circuit and the pulmonary valve annulus may define the other boundary. Having such a simple reentry circuit invites ablation across the corridor between the inert electrical barriers (such as from the scar to the pulmonary valve area), thus interrupting the possibility of reentry through the corridor in either direction. Other potential well-defined inert electrical barriers setting up potential corridors in the heart would include AV valve annuli, septal patches, or other scars. Difficulty in assessing and treating reentry VT arises when there are multiple reentrant circuits between and around a number of inert electrical barriers. Therefore, when considering catheter treatment of VT, a careful and very complete review of the underlying cardiac structure, the prior surgical techniques along with extensive mapping of the electrical circuits using computer-based electro-anatomic mapping systems are essential to detect possible reentry circuits.

Several approaches are available for mapping and ablation of VTs. Single point catheter mapping can be accomplished for focal origin tachycardia by isolating the earliest site of ventricular activation. Once found, pacing at the suspected site can be used to compare the "pace map" to the 12-lead EKG of the VT looking for a match to the clinical tachycardia. If the ECG during the VT is identical in all 12 leads to the ventricular depolarization pattern in all 12 leads of the ECG during pacing with the ablation catheter, one can infer that the ablation catheter tip electrode is located at the origin of the VT. Pace mapping techniques are useful when the inducible tachycardia is not hemodynamically stable enough to allow activation mapping during the VT. In addition, if the VT mechanism is triggered activity or automaticity at a focal site, then ablation at the best pace map site has a high likelihood of ablating the arrhythmia. Anesthesia may suppress VT, especially RVOT arrhythmias, therefore lightening or transitioning off of inhaled anesthetic may be necessary during the procedure if the arrhythmia is non-inducible.

If the tachycardia is due to a reentry circuit, an exact pace map may be found anywhere along the slowly conducting corridor (as well as a side branch of the circuit) and a focal ablation site is less likely to be successful since complete block across the slow conduction corridor of the reentry circuit is required. Therefore, lesions are typically placed across an isthmus of conductive tissue between nonconductive barriers to create block across the reentry loop. In addition, the elegant technique of concealed entrainment to establish the location of the critical slow conduction zone by observing VT behavior during and just after pacing is helpful in identifying appropriate ablation targets in reentry VT; the technique can be applied in the patient with congenital heart disease and the usual reentry arrhythmia. The advent of electro-anatomical mapping and non-contact mapping has allowed more precise delineation of early excitation sites from focal tachycardias and has allowed better definition by virtue of three-dimensional representations of the slow conduction paths between low voltage inert zones. These mapping techniques allow improved catheter localization and can reduce the need to use radiation for the procedure (Fig. 13.18). Additionally, some of these systems have the potential to delineate the reentry circuit or origin of tachycardia in a single beat so that sustained tachycardia is not required, an especially important benefit in those with hemodynamically unstable VT.

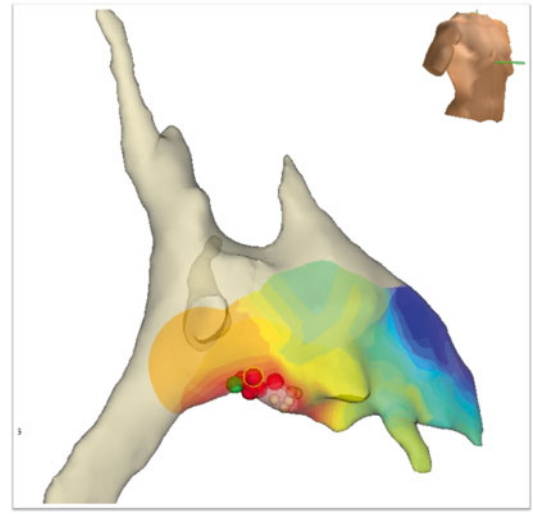

Fig. 13.18 An RAO view of the 3-D geometry (IVC, SVC, RA, and RV) and activation map in a 12-year-old female with ventricular tachycardia who underwent a radiation-free procedure. The spherical marking near the inferior base of the RV indicate the site of successful ablation of a focal tachycardia confirmed with electro-anatomic 3-D mapping and pace mapping

The ability of ablative lesions in the ventricular myocardium to be effective is still limited by the ability to create transmural lesions. Therefore, various catheter technologies have been developed to assist with transmural lesions delivery.

Device Therapy

An Implantable cardioverter-defibrillator is potentially lifesaving for patients with arrhythmias that are not amendable to ablative therapy or who remain at risk on chronic oral antiarrhythmia medication. Patients without underlying structural heart disease who may need device therapy are those who have the long QT syndrome,

CPVT, Brugada, or those who have suffered a cardiac arrest due to an unknown cause. Although less common in childhood, those with arrhythmogenic right ventricular cardiomyopathy may benefit from an ICD, as might those with dilated or hypertrophic cardiomyopathies. Among patients with congenital heart disease, the most widely represented group are those late after repair of tetralogy of Fallot. As the number of adults with congenital heart disease continues to increase, now more in number than children with heart disease, there is expected to be an emerging number of patients with more complex and evolving heart disease who may require ICD placement. Many of these, such as those who have completed the surgical staged therapy for single ventricle anatomy may present unique challenges due to concerns with vascular access, complex arrhythmias, variable anatomy, and intolerance of the arrhythmias.

Incorporation of brady and antitachycardia pacing into ICD units and combining pace/sense function into defibrillating leads has also benefited those needing ICD therapy. While the availability of transvenous ICD systems appears to be a benefit, the risk of having to use a transvenous system in a young patient incurs not only the risk of venous thrombosis limiting later access but also the need and risk of lead extraction expected over longer life span in the growing patient. Due to these concerns, epicardial leads and even leadless pacemakers and ICDs are being developed. With such varied cardiac anatomy in the pediatric and congenital heart population (especially the single ventricle group) the approaches for device placement call for creativity.

In all children with VT who require devices, the significant problem of inappropriate shock delivery due to over sensing, lead damage, sinus tachycardia entering the tachycardia detection zones, and other causes has not been fully surmounted. Also, the psychological burden of living with an ICD needs to be carefully and consistently monitored and addressed. The presence of the ICD in one's body is certainly challenging to a developing individual, but the anticipation that the device may cause a shock, or that it may not work in the intended way potentially could create a chronic psychological stress in any individual. Because of this many physicians also recommend close affiliation with a psychologist to assist with the emotional support needed by patients and their families.

Summary

Ventricular tachycardias in the young are a diverse group of arrhythmias generally considerably different in etiology and presentation than those seen in the adult with ischemic heart disease. There are a wide variety of ventricular substrates and a superimposed variety of electrophysiologic mechanisms that challenges diagnosis and treatment. The ability of many young patients to tolerate some forms of VT may allow the clinician to approach the arrhythmia in a more deliberate and conservative fashion. In many young patients with idiopathic LV or RV VT, the arrhythmia may spontaneously remit. Nonetheless VT still remains an important cause of sudden death in the young. This compels the understanding and utilization of a variety of diagnostic and treatment strategies. In patients after surgery for congenital heart disease, VT needs a comprehensive approach based on a complete analysis of the arrhythmia and cardiac malformation. For patients with cardiomyopathies and electrical heart disorders, exciting work in genetics offers the hope that further individualization of therapy and understanding of prognosis may soon be at hand. Finally, heightened awareness and increased use of newer myocardial imaging and ablation techniques offer hope for an improved ability to risk stratify and treat this important arrhythmia.

Suggested Reading

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