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Evaluation of the child with an arrhythmia

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The pediatric electrophysiologist has many diagnostic and therapeutic modalities upon which to draw when caring for a child with an arrhythmia. Several recent technologic innovations will broaden and accentuate the therapeutic alternatives available to these children. Despite these advances, the management of these patients still requires a solid foundation of clinical skill and judgment. The primary care pediatric practitioner is often the first to recognize the symptoms and signs of an arrhythmia. Careful assessment of the history, physical examination, and ECG is necessary for making the correct diagnosis.

Diagnosis

History and physical examination

History taking is the essential first step in the evaluation of the child with an arrhythmia. The ability to describe symptoms accurately and to answer questions depends on the age and maturity of the child. Older children and adolescents frequently describe palpitations or a fluttering sensation in the chest. These symptoms may be accompanied by chest pain, shortness of breath, abdominal pain, or dizziness. The child may also relay a history of syncope following the onset of the palpitations. Important questions to ask these children include

1. How frequently and for how long do these episodes occur?
2. How does the episode begin and terminate (eg, sudden onset and offset or gradual warm up and cool down of the heart rate)?

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3. What, if anything, initiates the episodes?
4. What, if anything, can be done to terminate the episodes (Valsalva maneuver, standing on one's head)?

Taking a careful diet history is also important because caffeine and stimulants in over-the-counter cold medicines can precipitate certain tachycardias.

Infants and younger children, obviously, cannot provide an adequate history, and therefore the historian is often the parent or primary caregiver. Infants frequently have symptoms of irritability and poor feeding. If the arrhythmia is long standing, congestive heart failure may develop. Manifestations of heart failure in the infant include tachypnea, diaphoresis, pallor, and lethargy.

In the setting of an acute arrhythmia, the physical examination is essential for determining the degree of hemodynamic stability. From this assessment of illness severity, immediate therapeutic alternatives are determined (observation versus medical management versus electrical cardioversion). In the setting of a recurrent, paroxysmal arrhythmia, the physical examination may be helpful in evaluating any exacerbating factors, such as fever, anemia, or hyperthyroidism. In addition, the physical examination is helpful in diagnosing structural cardiac diseases that are frequently associated with arrhythmia (eg, Ebstein's anomaly and hypertrophic cardiomyopathy, among others) and infectious causes such as viral myocarditis and Lyme disease.

Adjunctive testing

Once there is a suspicion of an arrhythmia, the patient should have a 12-lead ECG taken during sinus rhythm to document baseline rhythm, conduction and repolarization intervals, axes, evidence of hypertrophy, and other considerations. Also, abnormalities on the baseline ECG can give clues to the diagnosis. For instance, a delta wave, representing ventricular preexcitation, along with a short PR interval is diagnostic of Wolf-Parkinson-White syndrome (Fig. 1). In addition, a prolonged QT interval and abnormal T-wave morphology can be seen in the long QT syndrome.

Recording the ECG during the arrhythmia is essential to making an accurate diagnosis. If the arrhythmia persists for longer than a few minutes, the patient can often be brought to the clinic or emergency room to have an ECG performed. If the arrhythmia is sporadic and short lived, capturing an ECG recording of it may be difficult. An ambulatory ECG (Holter) monitor, which records the rhythm for 24 hours, is often useful if the arrhythmia occurs on a daily basis. If the arrhythmia is more episodic, external event recorders can be useful. These monitors come in two varieties. The loop recorder is worn by the patient for a prolonged period of time (ie, 1 month) and continuously records an ECG tracing. The monitor stores a tracing after being activated by the patient or surrogate at the time of an event. These monitors are useful if the arrhythmia is of short duration or if the consciousness of the patient is altered by the arrhythmia. The nonlooping or patient-activated event monitor is placed on the patient's chest wall

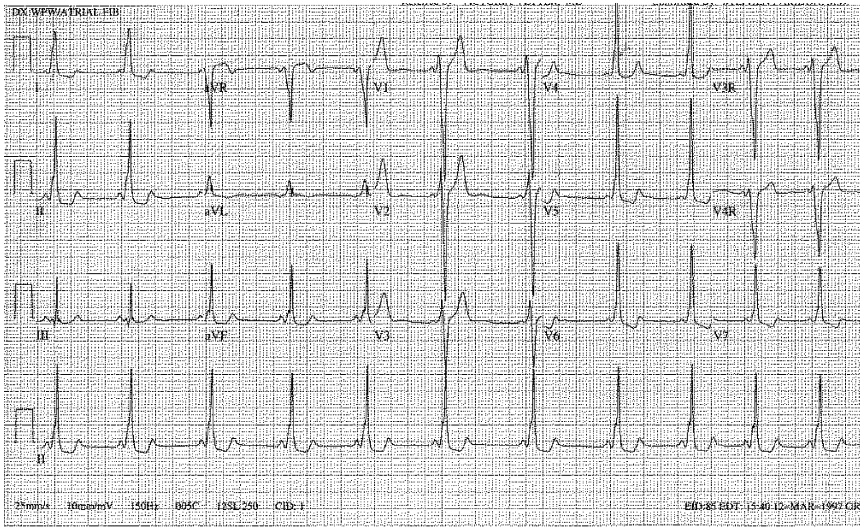


Fig. 1. ECG from infant with Wolf-Parkinson-White syndrome. Note the short PR interval and the slurred upstroke of the QRS complex (delta wave).

to record an ECG only at the time of symptoms and requires that the arrhythmia persist for at least a few minutes.

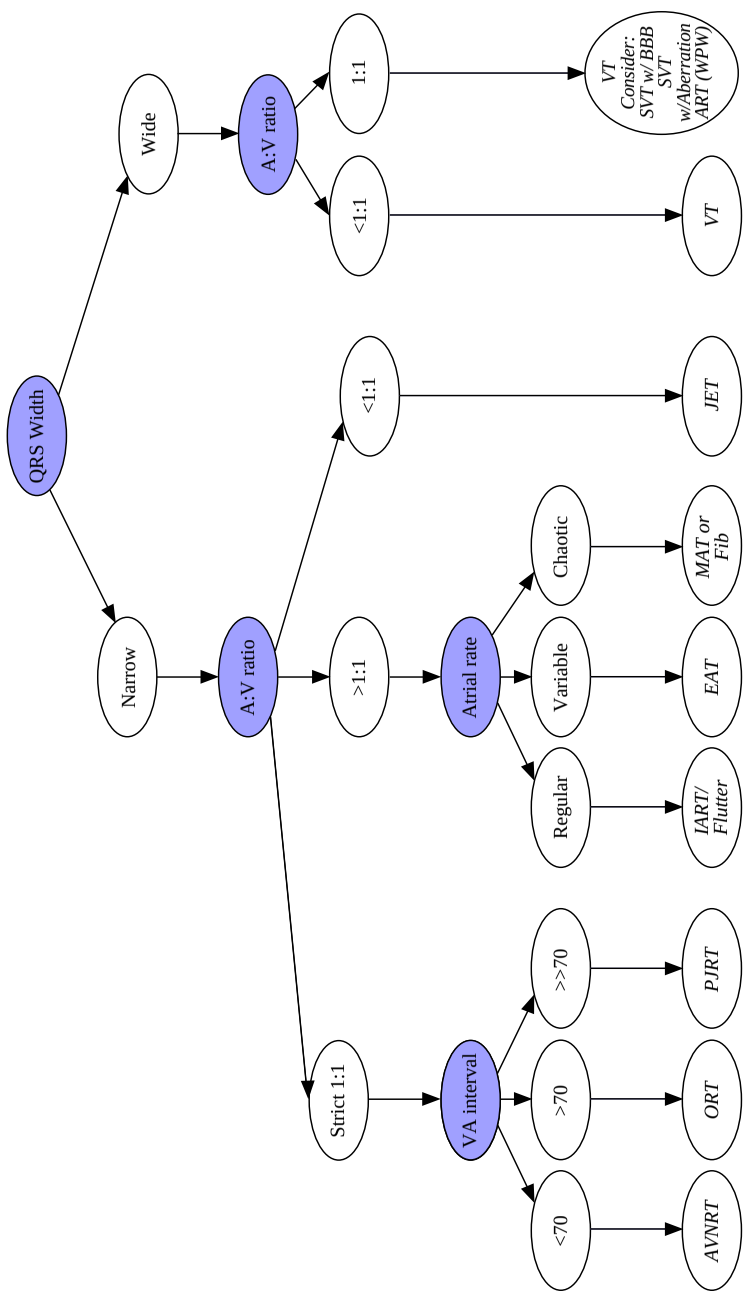
If these modalities are inconclusive because the symptoms are erratic and uncommon, the newly developed implantable loop recorder may be useful. These monitors are generally implanted in the pectoral region in a subcutaneous or subpectoral pocket and are active for 14 months. The monitor records a single-lead ECG after activation by the patient or parent or automatically if a preset heart rate algorithm is triggered. Recently, a retrospective, multicenter study showed that the recorders are able to facilitate correlation of rhythm with symptoms in all patients who had symptoms following implantation [1].

Diagnostic approach

Once the arrhythmia has been recorded, a diagnostic algorithm can be used to determine the exact diagnosis [2].

Tachycardia

A general approach to diagnosing tachycardias is shown in Fig. 2. Tachycardias can be divided into two general groups: narrow complex and wide complex. Narrow-complex tachycardias imply normal conduction through the atrioventricular (AV) node and His-Purkinje system. These supraventricular tachycardias (SVTs) are the most common arrhythmias in children. Wide-complex tachycardias typically originate in the ventricle, with certain exceptions. These exceptions include supraventricular tachycardias with pre-existing bundle branch block or rate-dependent aberrancy and antidromic reciprocating tachy-


QRS Width
Wide
A:V ratio
1:1
VT
Consider:
SVT w/ BBB
SVT
w/Aberration
ART (WPW)
<1:1
VT
Narrow
A:V ratio
<1:1
JET
>1:1
Atrial rate
Chaotic
MAT or
Fib
Variable
EAT
Regular
IART/
Flutter
Strict 1:1
VA interval
>>70
PJRT
>70
ORT
<70
AVNRT

cardia. In reentrant tachycardias involving an accessory pathway, conduction around the circuit can occur in two different ways. The first involves conduction down the AV node (antegrade) and up the accessory pathway (retrograde). This narrow-complex SVT is termed ''orthodromic reciprocating tachycardia'' (ORT). The second type involves conduction down the accessory pathway and up the AV node. This wide-complex SVT is termed ''antidromic reciprocating tachycardia.'' The next diagnostic step is to determine the relationship of A/V activation [3]. If the ratio of A/V activation is exactly 1 (number of P waves/number of QRS complexes = 1), then ORT or AV nodal reentrant tachycardia (AVNRT) is the likely cause.

Sufficiently rapid sinus tachycardia may be confused with SVT because they are both characterized by a narrow-complex tachycardia with an A/V ratio of 1. The P wave axis in sinus tachycardia will be 0° to $+90^\circ$, whereas the P wave axis in AVNRT or ORT will be 0° to -90°. An A/V conduction ratio greater than 1 suggests primary atrial tachycardia. If the atrial rate is very regular, atrial flutter is likely. An irregular atrial rate with a single P-wave morphology that differs from the normal sinus P-wave morphology suggests ectopic atrial tachycardia. Finally, an irregular atrial rate with three or more P-wave morphologies indicates multifocal atrial tachycardia. An A/V ratio less than 1 distinguishes junctional ectopic tachycardia.

Determining the relationship of atrial to ventricular activation can sometimes be difficult during tachycardia because P waves are not readily apparent on the ECG tracing. To enhance the sensitivity of this test, several maneuvers can be performed. Transesophageal recording involves placing a bipolar electrode catheter into the esophagus of the patient, typically through a nasopharyngeal approach. The catheter leads are then connected to a recording device, such as the right and left arm leads of an ECG machine. When properly positioned, the electrodes lie just behind the left atrium and will record a high-frequency, local atrial electrogram. The lead with the atrial electrogram can then be compared with the surface ECG leads to determine the relationship of atrial and ventricular activation. In addition, with an esophageal electrode in place, therapeutic pacing maneuvers can be used to terminate tachycardia [4]. Another method of ''bringing out the P wave'' is administration of adenosine. Adenosine is a nucleotide that transiently inhibits conduction through the AV node. It is administered intravenously, and, because it is rapidly metabolized by red blood cells, it must be given as a rapid bolus and as close to the central circulation as possible. If AV conduction is blocked without interrupting the rapid atrial rate, then a primary

Fig. 2. Diagnostic approach for determining tachycardia mechanism. ART, antidromic reciprocating tachycardia; AV, atrioventricular; AVNRT, atrioventricular nodal reentrant tachycardia; BBB, bundle branch block; EAT, ectopic atrial tachycardia; IART, intra-atrial reentrant tachycardia; JET, junctional ectopic tachycardia; MAT, multifocal atrial tachycardia; ORT, orthodromic reciprocating tachycardia; PJRT, permanent junctional reciprocating tachycardia; SVT, supraventricular tachycardia; VA, ventriculo-atrial; VT, ventricular tachycardia; WPW, Wolff-Parkinson-White syndrome. (*From* Walsh EP, Saul JP, Triedman JK, editors. Cardiac arrhythmias in children and young adults with congenital heart disease. Philadelphia: Lippincott Williams & Wilkins; 2001. p. 103; with permission.)

atrial tachycardia is suggested. If tachycardia breaks with adenosine, then the AV node is a required limb of the reentrant loop, signifying an A/V ratio of 1. Interruption or delay of conduction through the AV node can also be accomplished with vagal maneuvers, such as eliciting the diving reflex or the Valsalva maneuver.

Wide-complex tachycardias should be assumed to be ventricular, especially in the emergent setting, until proven otherwise. Ventricular tachycardia is characterized by a wide-complex tachycardia with AV dissociation (ie, no relationship between atrial and ventricular activation) and an A/V ratio less than 1. Ventricular tachycardia can also be confirmed by the presence of fusion beats, which represents a fusing of the ventricular beat with a sinus beat. The differential diagnosis of a wide-complex tachycardia with an A/V ratio of 1 is ventricular tachycardia with retrograde atrial conduction, SVT with aberrancy, SVT with bundle branch block, or antidromic reciprocating tachycardia.

Bradycardia

The diagnostic possibilities for abnormally slow heart rates in children are more limited. Sinus node dysfunction consists of sinus bradycardia, chronotropic incompetence during exercise, and sinus pauses. In the pediatric setting, sinus node dysfunction is typically seen following surgical repair of congenital heart disease. Complete heart block, defined as the inability of an atrial impulse to be conducted to the ventricle through the AV node, is another possible cause of bradycardia. It is represented on the ECG as AV dissociation with the atrial rate faster than the ventricular rate. Complete heart block may be congenital (maternal systemic lupus erythematosus, maternal Sjögren's syndrome) or acquired (typically a complication of congenital heart disease surgery or infectious causes such as viral myocarditis, Lyme disease, and others).

An important cause of bradycardia in the pediatric population is ingestion. Medications commonly found in the home, such as digoxin, β-blockers, clonidine, opioids, and sedative-hypnotics, can cause bradycardia. Organophosphate pesticides can also cause sinus bradycardia. Digoxin, calcium-channel blockers, tricyclic antidepressants, and lithium can cause AV block. A toxicology screen should be considered in pediatric patients presenting to the emergency room with a new-onset bradycardia.

Referral to a pediatric cardiologist

There are a number of benign causes of irregular heart rhythms observed in childhood. Sinus arrhythmia refers to the normal reflex-derived changes in heart rate in response to the respiratory cycle. During inspiration, the heart rate accelerates, and during expiration the heart rate decelerates. Sinus arrhythmia is typically more pronounced in school-aged children and adolescents. Sinus arrhythmia is a normal rhythm and does not require further work-up or therapy.

Premature atrial contractions (PACs) are often seen in children and especially in infants. In the setting of a normal cardiovascular examination and normal electrolytes, PACs are generally benign. Closely coupled PACs may block in the

AV node. Frequently blocked PACs is one of the most common causes of bradycardia in the newborn. If the infant is otherwise well and without hemodynamic compromise, therapy is often not needed. PACs rarely require further cardiologic evaluation.

Premature ventricular contractions (PVCs) are also frequently observed in normal infants and adolescents. The reported incidence of isolated PVCs in infants is 10% to 15% and in adolescents is 20% to 35% [5]. PVCs generally require a more thorough cardiovascular work-up, which can include measurement of serum electrolytes, 24-hour Holter monitoring, and an echocardiogram, along with referral to a pediatric cardiologist. These tests are meant primarily to rule out structural heart disease or subclinical ventricular dysfunction. If a thorough work-up reveals no identifiable cause, the diagnosis of idiopathic PVCs is given. Idiopathic PVCs are generally benign and self-limited. Characteristically, benign PVCs are of a single-QRS morphology (monomorphic) and are easily suppressed with exercise. The prognosis is quite favorable, and treatment is rarely warranted.

Any sustained or frequently recurrent arrhythmia should be referred to a pediatric cardiologist or pediatric electrophysiologist. The mechanism and cause of the arrhythmia will be evaluated, any associated structural heart disease will be ruled out, and therapeutic alternatives will be determined. Any child presenting with syncope, as a potential presenting sign of an arrhythmia, also requires an evaluation by a pediatric cardiologist.

Treatment

Tachycardia

Acute therapy

Medical treatment of tachycardia can be divided into acute and chronic therapy [2]. Acute therapy seeks to interrupt the arrhythmia or slow the ventricular response rate and, ultimately, restore sinus rhythm. Acute therapy depends on the clinical situation. A patient who presents in extremis as a consequence of tachycardia should be approached differently from one who is in stable condition. In the emergent setting, the ABCs of resuscitation must be followed. Once the airway is secured and ventilation is assured, attention can be paid to circulation. If the patient has stable vital signs during the tachycardia, time can be spent evaluating the rhythm using the diagnostic approach outlined previously.

In the authors' practice, adenosine is the medication most commonly used for rapid treatment of reentrant tachycardias in which the AV node is part of the circuit. Adenosine works by interrupting conduction through the AV node. Its rapid half-life (in seconds), however, requires that a sufficient bolus reach the heart quickly for it to work. Digoxin is also effective and is especially useful in the patient with decreased myocardial function. Other pharmacologic therapies used in the acute setting include intravenous β-blockers, such as esmolol, intravenous procainamide, and intravenous amiodarone. Other than digoxin, these

medications should be used with caution because they all have negative inotropic effects. Other therapeutic modalities include transesophageal pacing and vagal maneuvers. Vagal maneuvers for adolescents and older children include the Valsalva maneuver and the headstand. In infants, the diving reflex, elicited by administering a bag of ice to the center of the face, is often successful in terminating the SVT. Although intravenous calcium-channel blockers are an important therapy for SVT in adults, they are contraindicated in children, especially those under 1 year of age. There have been reports of hemodynamic decompensation and sudden death in infants who were given verapamil [6]. If these maneuvers fail, or the patient is hemodynamically unstable, direct current (DC) cardioversion is the next option. Synchronized cardioversion should be performed with an energy output of 0.5 to 1 J/kg. The output can be doubled to a maximum of 5 to 6 J/kg until the treatment is effective.

The acute management of ventricular tachycardia requires a full evaluation of the ABCs of resuscitation. Any acute and reversible causes of the ventricular tachycardia, such as electrolyte abnormalities or acidosis, should be sought and treated. Intravenous lidocaine, administered at a dose of 1 mg/kg, is often first-line therapy. If this measure is successful, lidocaine infusion should be started. If the patient is severely compromised, or the lidocaine was not successful or available, synchronized cardioversion at 1 to 2 J/kg should be performed. Other medications that have proved effective in the emergent setting include intravenous procainamide and amiodarone.

Chronic therapy

Chronic therapy attempts to prevent recurrence of the tachycardia and is targeted to the type and mechanism of the tachycardia. Therapies for reentrant supraventricular tachycardias that use the AV node as part of the reentrant circuit attempt to modify conduction through the AV node. Digoxin and β-blockers are first-line oral therapy for these types of SVT. Digoxin and calcium-channel blockers are contraindicated in patients with Wolff-Parkinson-White syndrome. These medicines enhance antegrade conduction down the accessory pathway. This enhancement allows a more rapid ventricular response during atrial flutter or fibrillation, which can then precipitate ventricular fibrillation. For SVT refractory to first-line therapy, other antiarrhythmics such as flecainide (a class IC agent), procainamide (a class IA agent), sotalol, amiodarone (a class III agent), or verapamil (a class IV agent) can be employed. For primary atrial tachycardias, two strategies are used to address the arrhythmia. The first involves decreasing the ventricular response rate by slowing AV conduction with digoxin or calcium-channel blockers. The second strategy attempts to alter the electrophysiology of the atrial substrate. Commonly used medications include β-blockers, class IA agents such as procainamide, class IC agents such as flecainide and propafenone, and class III agents such as amiodarone and sotalol.

Chronic therapy for ventricular tachycardia often requires that the underlying cause, such as poor ventricular function, inflammation, ischemia, electrolyte abnormalities, and others, be addressed in addition to treating the arrhythmia. If

the arrhythmia persists, and the patient initially responded to lidocaine, mexiletine may prove useful as a chronic agent. Catecholamine-sensitive arrhythmias, such as the ventricular tachycardia of long QT syndrome, often respond well to β-blockers. Class I agents and amiodarone are second-line medications.

Table 1 lists doses and side effects of commonly used antiarrhythmic agents in the pediatric population that will likely be encountered in a general pediatric practice.

Table 1
Commonly used antiarrhythmic agents in the pediatric population

Antiarrhythmic agent	Dose	Side effects
Class I		
Procainamide	Oral: 30–100 mg/kg/d ÷ every 6 hours (or every 12 hours for Procanbid (two times a day)	Nausea, vomiting, lupuslike syndrome, pancytopenia, agranulocytosis, confusion
Lidocaine	Intravenous: 1 mg/kg	Seizure, central nervous system symptoms, arrhythmias, respiratory distress, hypotension
Mexiletine	Oral: 5–15mg/kg/d ÷ every 8 hours	Nausea, vomiting, headache, tremor, dizziness, paresthesia, rash
Phenytoin	Oral: 2–5mg/kg/d ÷ every 12 hours following load of 10–15mg/kg ÷ every 6 hours	Rash, Stevens-Johnson syndrome, neuropathy, gingival hypertrophy
Flecainide	Oral: 2–6 mg/kg/d ÷ every 8 hours	Arrhythmia, conduction disturbances, dizziness, blurred vision, headache
Class II		
Propranolol	Oral: 2–4 mg/kg/d ÷ every 6 hours	Hypotension, bronchospasm, hypoglycemia (in neonates), lethargy, depression
Nadolol	Oral: 1–2 mg/kg/d ÷ every day or every 12 hours	Bradycardia, lethargy, depression
Atenolol	Oral: 1–2 mg/kg/d ÷ every 12 hours	Lethargy, depression, bradycardia, postural hypotension, agranulocytosis
Class III		
Amiodarone	Oral: load of 10 mg/kg every 12 hours × 5–14 days followed by maintenance of 5–7 mg/kg/day every day	Arrhythmias, prolonged QTc interval, hepatotoxicity, hypo- and hyperthyroidism, corneal microdeposits, photosensitivity, pulmonary fibrosis
Sotalol	Oral: 80–160 mg/m^2/d ÷ every 8 hours for infants or every 12 hours for older children	Arrhythmia, prolonged QTc interval, bradycardia, fatigue, dyspnea
Class IV		
Verapamil	Oral: 4–8 mg/kg/d ÷ every 8 hours or every day SR form	Hypotension, bradycardia, cardiac decompensation (esp. in infants)
Other agents		
Adenosine	Intravenous: 0.1 mg/kg rapid bolus	Transient bradycardia and tachycardia, transient AV block
Digoxin	Oral: Load of 30–40 mcg/kg (depending upon age) over 1st day followed by maintenance of 5–10 μg/kg/day every day or ÷ every 12 hours	AV block, arrhythmias, nausea, vomiting

Catheter ablation

Catheter ablation has become an increasingly popular therapeutic modality for all types of tachycardia. The singular advantage of this therapeutic approach is that it is curative. Catheter ablation using radiofrequency energy was first performed in a child in the early 1990s. Catheter ablation involves application of radiofrequency energy through a steerable electrode catheter to the arrhythmia substrate (ie, accessory pathway, AV nodal slow pathway, ectopic focus). Radiofrequency energy causes tissue heating and necrosis. To locate the site of the arrhythmia substrate, electrophysiologic mapping is performed. This mapping is conventionally done using two to four multielectrode catheters placed at locations of normal conduction tissue. Electrograms recorded from the electrode catheters represent local myocardial activation. Therefore, activation of the heart can be observed in both time and two-dimensional (2-D) space for normal sinus rhythm. Then, using certain pacing maneuvers, the arrhythmia can be elicited, and the abnormal activation sequence can also be observed. From this information the mechanism of the arrhythmia and the location of the substrate can determined and subsequently eliminated.

Radiofrequency catheter ablation has been used successfully to cure various types of arrhythmias in pediatric patients. Success rates between 81% and 97% have been reported for accessory pathway–mediated SVT, depending on the location of the accessory pathway. Success rates higher than 95% have been reported for AVNRT and higher than 87% for ectopic atrial tachycardia [7]. Complication rates are low—3% to 4% [7]. The major complications are AV block, cardiac perforation/pericardial effusion, and thromboembolic phenomenon. Common indications for radiofrequency ablation include life-threatening arrhythmias, medically recalcitrant tachycardias, adverse drug reactions, tachycardia-induced cardiomyopathy, impending cardiac surgery, and patient choice. With the advent of newer technology, indications for the procedure have become broader, and complex arrhythmias, such as atrial flutter caused by postoperative atrial scar and ventricular tachycardia, can be successfully treated.

Several advanced mapping systems have been introduced in the past several years. Electroanatomic mapping systems (CARTO, Biosense Webster, Inc., Diamond Bar, California) use the principles of triangulation in a magnetic field to increase the navigational precision of the ablation catheter. The mapping catheter is placed at known anatomic locations within the heart, and local activation, referenced to a stationary electrode, is recorded. These data are then represented as a three-dimensional (3-D) model of the heart on which the activation sequence of the arrhythmia is superimposed. In addition, voltage maps can be created that represent areas of varying voltage amplitudes. Voltage maps are useful for showing areas of diseased or scarred tissue. Noncontact mapping systems (Ensite 3000, Endocardial Solutions, St. Paul, Minnesota) use a balloon catheter with a multielectrode array that records electrical activity from the endocardial surface of a heart chamber. A virtual 3-D model of the heart chamber is established using a steerable contact-mapping catheter. Algorithms then allow electrical activity at nearly 3000 points on the heart chamber wall to be superimposed on the 3-D

model. Single-beat mapping can be performed so that the patient does not have to remain in a sustained arrhythmia to localize its activation. The theoretic advantage of these systems is that they provide a 3-D representation of electrical activation of the heart, allow the mapping of complex arrhythmias in the setting of complex anatomy such as after the Fontan or Mustard/Senning operations, decrease fluoroscopy time, and allow the mapping of unstable or nonpersistent arrhythmias (noncontact systems). These systems have already been shown to be useful for mapping complex atrial arrhythmias in patients who have undergone surgery for congenital heart disease [8,9] and for mapping ventricular tachycardia [10,11].

Advances in ablative technology have also been made. Cooled ablation catheters (Chilli Cooled Ablation Catheters, Boston Scientific, Natick, Massachusetts) have an internal flow mechanism that decreases the temperature of the catheter tip. The cooler tip allows an increased delivery of total energy that increases lesion size and depth. This technique has proved useful in thickened endocardium, such as atrial muscle incorporated into the Fontan pathway [9] or ventricular muscle [12]. In addition, some centers are using cryoablation (CryoCath Technology, Montreal, Quebec, Canada) to destroy tissue [13]. Cryotherapy removes heat from the tissue. At certain temperatures reversible cellular changes occur, and at lower temperatures permanent damage occurs. The theoretic advantages of cryoablation are that its effects can be reversible, so that potential ablation sites can be tested, and that it allows more precise site selection.

Implantable cardioverter defibrillators

Implantable cardioverter defibrillators (ICDs) are implantable devices connected to epicardial or intravascular leads that can sense ventricular arrhythmias and deliver a shock within the heart to restore normal sinus rhythm. Several large, controlled trials in adults have shown a clear survival advantage of ICDs compared with conventional therapy for the prevention of sudden cardiac death. ICDs have also been shown to be effective in treating malignant ventricular arrhythmias in pediatric patients [14,15]. A recent, retrospective study showed that 28% of patients received appropriate shocks for ventricular tachycardia over a 2-year follow-up period [14]. As of 2004, however, there have been no controlled trials in pediatric patients in which a survival benefit could be tested. The indications for ICD implantation have been poorly defined in pediatric patients because the incidence of sudden cardiac death is low, and appropriate risk-stratification methods have been elusive. Nevertheless, the commonly accepted practice is to place ICDs in patients who have had aborted sudden death and for whom no reliable treatment is available, in patients who have recurrent, symptomatic ventricular arrhythmias poorly controlled by medications, and in asymptomatic patients who have strong risk factors for sudden cardiac death, such as a patient with long QT syndrome and a family history of sudden death or a patient with hypertrophic cardiomyopathy and ventricular tachycardia.

Special consideration is required for implantation of these devices in children. The smaller size of the patient requires the use of smaller devices as well as novel

lead configurations. Somatic growth of the child is strongly correlated to lead failure [14]. Inappropriate shocks, caused by inappropriate sensing, lead failure, or high sinus rates, can have significant psychologic impact on the child or adolescent.

Bradycardia

Pacemakers

Pacemakers are the primary treatment for symptomatic bradycardia. The goal of pacemaker therapy is to restore as close to a physiologic rhythm as possible within the confines of the available technology. In the most recent report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines [16], there is general agreement for recommending a pacemaker in the following conditions:

1. Advanced second- or third-degree AV block associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output
2. Symptomatic sinus node dysfunction during age-appropriate bradycardia
3. Postoperative second- or third-degree AV block that persists for at least 7 days
4. Congenital complete AV block with a wide QRS escape rhythm, complex ventricular ectopy, ventricular dysfunction, or ventricular rate less than 50 to 55 beats/minute (bpm) in an infant or less than 70 bpm in a patient with congenital heart disease
5. Sustained pause-dependent ventricular tachycardia, such as in long QT syndrome

In older children and adolescents, the pacemaker generator is typically implanted in the subpectoral region with the leads attached to the endocardium using a transvenous route. Pacemaker generators are typically implanted in the abdomen in infants and small children with the leads attached to the epicardial surface of the heart. This configuration is primarily dictated by the smaller size of the major veins to the heart, which may narrow following intravascular placement of the pacemaker leads. In addition, in patients of any size with a surgically altered systemic venous return, pacemakers are typically implanted in an epicardial fashion. Pacemaker leads may be attached either to a single chamber (atrium or ventricle) or to both chambers, depending on the rhythm causing the bradycardia and the size of the patient.

The generator is then programmed to provide sufficient rate and rhythm support for the patient. Newer pacemaker models have more advanced programmable capabilities. These capabilities include rate responsiveness [17], which senses patient movement and allows activity-related rate increases. Other features include arrhythmia monitoring and documentation and self-testing capabilities for battery conservation [18].

With advances in technology, new indications for pacemakers in pediatrics are constantly being developed. Cardiac resynchronization therapy (CRT) involves simultaneous pacing of the left and right ventricle (biventricular pacing) in the setting of heart failure with intraventricular conduction delay. CRT has been shown to have a positive impact on cardiac output and quality-of-life indicators in adult patients. Biventricular pacing and a modification of this principle, multisite pacing, has been preliminarily shown to benefit patients following repair of congenital heart disease [19–21]. In addition, pacemakers that are programmed to detect and pace terminate atrial tachycardias have recently been shown to have modest success in treating atrial arrhythmias in patients with postoperative congenital heart disease [22].

Pacemakers and implantable cardioverter defibrillators in the general pediatric clinic

As pacemakers and ICDs become more widely used in the pediatric population, patients with these devices will be seen more commonly in the general practitioner's practice. Some practical aspects of these patients' care are therefore important for the general pediatrician. These patients are restricted from activities that could cause direct damage to the pacemaker. These restrictions include contact sports, such as football, wrestling, and rugby. Other restrictions on participation in sports are based on the cardiac disease and not on the presence of the implanted device. The authors follow many patients in their pacemaker clinic who are active participants in soccer, basketball, softball, and other sports. There are some commercially available protective garments that can be worn to help shield the device.

Although it is somewhat controversial, the authors' practice is to recommend prophylaxis for subacute bacterial endocarditis (SBE) for patients who have intravascular pacemakers or ICDs. SBE prophylaxis is not required for epicardial devices.

MR imaging scans are contraindicated in all patients with pacemakers and ICDs. Patients with these devices can use cellular telephones, but it is recommended that the patient use the ear contralateral to the position of the generator. Although several studies have shown that it is safe for pacemaker patients to pass through metal detectors at airports, the authors recommend that the patients notify security and undergo a manual search. Hand-held metal detectors can also cause pacemaker and ICD malfunction. Another source of electromagnetic interference are newer roller coasters, which patients with implanted devices should avoid.

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