Focus on Diagnosis : Cardiac Arrhythmias in Children

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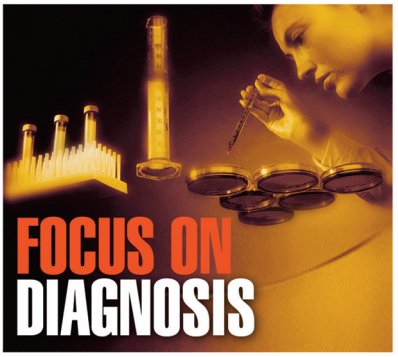

Cardiac Arrhythmias in Children

Eric A. Biondi, MD*

Author Disclosure

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Introduction

Although most childhood arrhythmias are benign, prompt and correct diagnosis of a serious rhythm disturbance in a child can be lifesaving. Such rhythm disturbances may arise at any age and have a wide variety of presentations. This article discusses various pediatric arrhythmias that may be encountered by the community pediatrician, highlighting their presentation, findings on electrocardiography (ECG), and when to refer for additional evaluation.

The Sinoatrial Node

Sinus Rhythm and Sinus Arrhythmia

Some rhythm disturbances originate within the sinoatrial (SA) node. This cardiac pacemaker is located in the upper wall of the right atrium and initiates electrical conduction through the cardiac muscle. The term sinus rhythm designates normal heart rhythm controlled by this node. ECG shows a P wave with a leftward and inferior axis before each QRS complex and a normal PR interval (120 to 200 msec).

Sinus arrhythmia occurs in healthy children and is described as a decrease in SA node firing subsequent to activation of the vagus nerve by exhalation. The heart rate, thus, varies with respiration, and ECG shows sinus rhythm with a prolongation of the R-R interval during exhalation. Such prolongation may be suppressed with exercise or other causes of sinus tachycardia. This finding is normal and is not a reason for referral.

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Sick Sinus Syndrome

Although most significant arrhythmias occur below the SA node, one emanating from the SA node, sick sinus syndrome (SSS), is worth mentioning briefly. This rhythm is a result of SA nodal dysfunction and is seen most often in patients who had prior cardiac (especially extensive atrial) surgery or cardiomyopathy. Although many forms of SSS are asymptomatic, common clinical manifestations include shortness of breath, chest pain, and syncope. The rhythm is characterized by brady- and tachyarrhythmias. ECG may show SA block, atrial fibrillation (AF), or supraventricular tachycardia (SVT). Patients suspected of having SSS should be referred to a cardiologist for additional evaluation.

The Atria

Several common rhythm disturbances can arise within the atria. ECG findings associated with atrial rhythms generally involve changes to the P wave or P-R interval.

Premature Atrial Contractions

Premature atrial contractions (PACs) are very common in asymptomatic pediatric patients and are benign. They arise when an ectopic focus stimulates the atria without input from the SA node. Although they may be caused by drug use, caffeine, or electrolyte imbalances, the inciting factor usually is unknown. Patients infrequently report feeling a "skipped beat" or "pause," often followed by a strong beat, which is the result of prolonged filling time before resumption of sinus rhythm. If history, physical findings, and ECG are diagnostic, the patient can be reassured, and no additional evaluation is necessary. If the patient is bothered

Abbreviations

AET:	atrial ectopic tachycardia
AF:	atrial fibrillation
AP:	accessory pathway
AV:	atrioventricular
AVNRT:	atrioventricular nodal tachycardia
ECG:	electrocardiography
LQTS:	long QT syndrome
PAC:	premature atrial contraction
PVC:	premature ventricular contraction
QTc:	corrected QT interval
SA:	sinoatrial
SSS:	sick sinus syndrome
SVT:	suptraventricular tachycardia
VF:	ventricular fibrillation
VT:	ventricular tachycardia

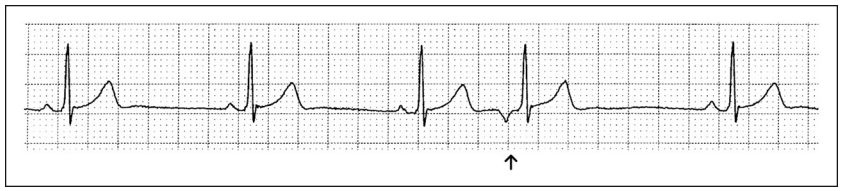

Figure 1. There is a premature atrial complex (PAC) after the third sinus QRS. Notice the premature, inverted P wave. The prolonged pause before the next beat suggests that the ectopic beat has reset the sinoatrial node. The QRS complex is normal, indicating that both bundle branches were polarized before the PAC.

by PACs, known inciting events should be avoided.

Common ECG findings of PACs include premature, inverted, or oddly shaped P waves, indicative of an ectopic atrial focus, and sharp inflections, often within the T waves (Fig. 1). If the premature beat occurs while both bundle branches are polarized, it is conducted to both ventricles simultaneously, resulting in a normal QRS complex. If one of the branches is refractory, the beat conducts along the opposite bundle branch, resulting in a wide QRS complex. Finally, if both ventricles are refractory, the beat is not conducted and no QRS complex is formed. This is known as a blocked PAC. In older patients, such episodes are commonly but not invariably suppressed during exercise as a result of the sinus tachycardia. Referral to a pediatric cardiologist is unnecessary.

It is important to remember that although most PACs are benign, they may occur rarely in infants in a bigeminal, blocked fashion, causing feeding intolerance and decreased cardiac output because of the slow heart rate. These unusual patients should be referred to a pediatric cardiologist.

Atrial Flutter and Fibrillation

Atrial flutter is another relatively common arrhythmia that is characterized by atrial rates of 250 to 400 beats/min. Arising in newborns who have normal hearts and older children born with structural heart disease, atrial flutter is caused by a reentrant circuit confined to the right atrium. Infants may present with congestive heart failure, and older children may have dizziness, syncope, chest pain, and shortness of breath. The major clinical clue is the heart rate. In children, atrial flutter can be conducted to the ventricles in a 1:1 fashion, resulting in ventricular contractions of more than 300 beats/min, or in a ratio of 1:2, causing rates of 150 to 200 beats/min. In infants, the ECG often shows classic inverted "saw-tooth" deflections, best seen in leads II, III, and aVF, that usually are inverted if the patient has the typical, counterclockwise reentrant pathway. The patient should be referred for urgent cardiac evaluation and treatment. Neonatal atrial flutter rarely reoccurs after sinus rhythm is restored. It is important to note that atypical atrial flutter, characterized by slower, more rounded P waves of lower voltage separated by an isoelectric line, is a potentially lethal arrhythmia, usually occurring in the setting of complex heart disease in older children.

AF is uncommon in young children, although there is evidence to suggest that it is underreported in adolescents. The rhythm derives its name from rapid fibrillation of the atrial muscle without coordinated contraction and most often is the result of structural heart disease causing stretching of the atria. AF generally is not life-threatening but can cause palpitations, chest pain, or syncope. Careful examination of a patient's pulse shows an irregularly irregular rhythm. ECG showing absent or very low-voltage P waves and an irregular R-R interval confirms the diagnosis. If AF is suspected but the ECG tracing is normal in the office, 24-hour outpatient Holter monitoring or the use of event recorders may be of assistance. Any patient who has the new diagnosis of AF should be referred to a pediatric cardiologist. It is very important to have the patient seen urgently because prolonged (usually >24 hour) fibrillation or flutter can result in clot development within the left atrium. With resumption of sinus rhythm, the clots can embolize, resulting in stroke, myocardial damage, or other end-organ infarctions.

The Atrioventricular Node and Supraventricular Tachycardias

Supraventricular Tachycardia

SVT is defined as a rapid tachycardia originating above the bundle of His. It occurs in as many as 1 in 250 children but often is misdiagnosed due to the variety of presentations it may cause. There are many different mechanisms for SVT, but they can be divided into three major categories: reentrant tachycardia using an accessory pathway (AP); reentrant atrioventricular nodal tachycardia (AVNRT), typically seen in adolescents; and atrial ectopic tachycardia (AET).

In infants, SVT may present with heart rates of 220 to 270 beats/min. Infants who experience prolonged SVT may have a history of poor feeding, pallor, irritability, and lethargy. The arrhythmia often is diagnosed after 24 or 48 hours of sustained SVT, when hemodynamic decom-

pensation arises and congestive heart failure develops.

School-age children can verbalize symptoms and, therefore, usually are seen before developing heart failure. They may complain of "beeping in my chest," heart pounding, chest pain or fullness, shortness of breath, sweating, or exercise intolerance. They almost never experience syncope. The tachycardia rate is slower, usually 180 to 240 beats/min rather than the approximately 220 to 270 beats/min seen in infants.

Adolescents experience similar signs and symptoms as seen with school-age children, but they are more capable of precise descriptions. It is useful to ask the patient to describe the heart rate during the episodes, which typically last from a few seconds to a few hours. History of a heart rate that is "too fast to count," a pounding sensation in the neck, or an abrupt resolution of palpitations, often after vagal maneuvers, is helpful. Another clue is the description of a "switch on-switch off" tachycardia rather than a pattern of progressive acceleration or deceleration. Occasionally, a school nurse or coach will have counted the pulse rate. Such findings help to distinguish SVT from other common causes of similar symptoms in adolescents such as anxiety, stress, caffeine consumption, or dehydration, all of which cause sinus tachycardia.

SVT in infants may be difficult to differentiate from sinus tachycardia by ECG. SVT usually manifests as a narrow complex (<80 msec) tachycardia with a nonvariable heart rate greater than 220 beats/min. P waves often are difficult to see but may be seen as sharp deflections within the T waves.

In older children, ECG findings vary, based on the mechanism of the SVT. In patients who have AP-mediated reentry tachycardia, ECG characteristics differ based on whether the antegrade current is carried by the atrioventricular (AV) node or the accessory pathway. If the AP carries such a current, as in Wolff-Parkinson-White syndrome (a condition in which an aberrant AP causes pre-excitation of the ventricles), the classic findings are a shortened PR interval and "delta waves," a gradual sloping of the R wave caused by the pre-excitation (Fig. 2). If the AV node carries the antegrade current, ECG shows a narrow complex tachycardia and typically lacks obvious P waves. In this case, sharp upward deflections in the T waves, representing retrograde conduction through the AP, may be seen.

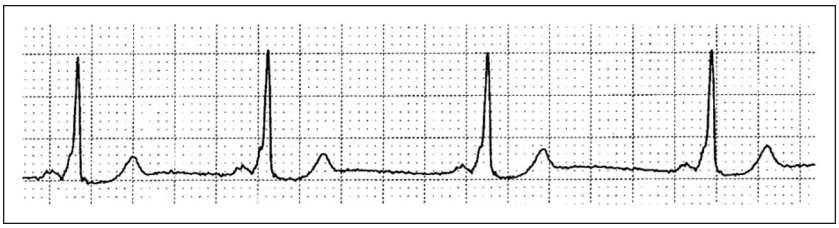

Figure 2. Wolff-Parkinson-White (WPW) syndrome. Delta waves, a wide QRS complex, and a short P-R interval (here it is 40 msec) are classic ECG findings in WPW syndrome.

In AVNRT, which occurs more commonly in older children, heart rates often are slower without visible P waves, which are buried within the QRS complexes, and an initiating event such as a PAC may be identified. Lastly, the ECG in AET may show a variable heart rate of up to 330 beats/min with abnormal P waves. This form of SVT is important to identify because rates this fast are poorly tolerated and affected patients can develop a cardiomyopathy.

When a patient is suspected of having any form of SVT, cardiac referral is indicated. Ambulatory ECG monitoring devices (Holter monitors or event recorders) are useful for diagnosing SVT in patients who have sporadic episodes. Compared with Holter monitors that capture events in only a 24- or 48-hour period, event recorders can operate for up to 1 month and are activated by the patient when symptoms occur. The recorded ECG is sent via telephone to a cardiologist for analysis. Electrophysiologic study is the definitive method of diagnosing the mechanism underlying the SVT and is used for identification of the AP, which can be treated with radiofrequency ablation.

The Ventricles

The ventricles comprise the final cardiac area in which arrhythmias can develop, and although several dangerous arrhythmias can develop here, this discussion focuses on the ventricular disturbances most likely to present to an outpatient office. Two uncommon but potentially lethal arrhythmias also are mentioned.

Premature Ventricular Contractions

Premature ventricular contractions (PVCs) are caused by ectopic firings within the ventricle and, although less common than PACs, may occur in as many as 25% of healthy children. Patients usually are asymptomatic but may report chest fullness, dizziness, or a feeling that the "heart skips" and then resumes with a strong beat.

Twelve-lead ECG always should be obtained in a patient suspected of having PVCs to allow the clinician to assess PVC morphology. Holter

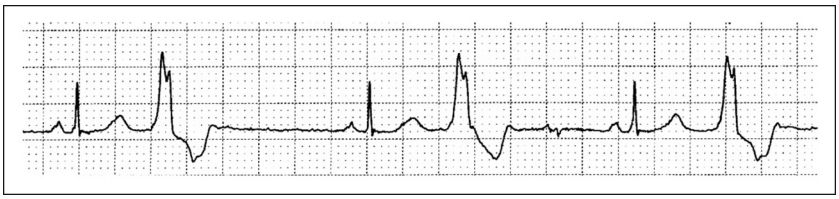

Figure 3. Ventricular bigeminy with premature ventricular beats (PVCs) occurring after each sinus beat. The PVCs are uniform, are bizarre, demonstrate wide QRS complexes without a preceding P wave, and show T-wave inversion. There is also a prolonged pause after each PVC.

or event monitoring may be useful in documenting infrequent episodes. The PVC itself appears as a premature, bizarre, wide QRS complex not preceded by a P wave and often followed by a compensatory pause (Fig. 3). The pause is associated with increased ventricular filling and increased stroke volume of the next beat that may be noticed by the patient as a pause followed by a strong beat. If the PVC occurs close enough to the next sinus beat, a fusion beat may occur that has characteristics of both a PVC and a normal QRS complex. If they are frequent, PVCs may occur with every other beat (bigeminy) or every third beat (trigeminy).

PVCs are benign if they are single, uniform in appearance, and suppressed or at least not aggravated by exercise and there is no evidence of underlying heart disease or family history of sudden, early death. For patients who have abnormal family histories, the clinician should be more suspicious of the potential for dangerous ventricular arrhythmias, and those patients should be referred to a pediatric cardiologist for additional evaluation.

Long QTc Syndrome

The long QT syndrome (LQTS) is associated with a potentially dangerous ventricular arrhythmia, torsades de pointes. Although not every patient who has a prolonged corrected QT interval (QTc) has LQTS, an interval of more than 450 msec is suggestive of LQTS and more than 470 msec is considered abnormal (Fig. 4). Using the QTc, calculated as the QT interval divided by the square root of the previous R-R interval, is important because that value "corrects" the QT interval for the patient's heart rate. There is often a family history of unexplained sudden death (50% in symptomatic patients). Patients can present with syncope, seizures, palpitations, and cardiac arrest. As many as 10% have episodes of sudden death. Frequently, a previously healthy patient reports fainting spells while swimming, playing sports, or exercising. Several genetic cases of LQTS have been identified. Specifically asking about congenital deafness in the family can provide a clue to the diagnosis because deafness often is associated with a particularly malignant form of hereditary LQTS. Any patient who has symptoms and even a borderline prolonged QTc should be referred to a pediatric cardiologist.

Ventricular Tachycardia

Ventricular tachycardia (VT) in children is defined as a tachycardia of at least three successive ventricular beats. It is referred to as nonsustained if the rhythm lasts less than 30 seconds and terminates spontaneously. If it lasts longer than 30 seconds, it is considered sustained and usually requires therapeutic intervention.

VT in the pediatric population occurs most commonly in children who have abnormal hearts. Although many patients are asymptomatic, symptoms such as pallor, fatigue, and chest palpitations may occur. In infants, VT often manifests as feeding intolerance. Among children who have healthy hearts, VT carries a good prognosis, in contrast to VT in children who have abnormal hearts or a history of cardiac dysrhythmias. Causative factors include use of drugs, caffeine, and decongestants as well as electrolyte imbalances and underlying cardiac disease.

On physical examination, there may be evidence of unsuspected congenital or acquired cardiac disease. ECG shows a bizarre, wide QRS complex (>120 msec) tachycardia, which usually has a regular rhythm (Fig. 5). P waves may or may not be recognizable, depending on the ventricular rate, and T waves typically are opposite in polarization to the QRS. The QRS complexes may vary in appearance if the ectopic input is multifocal. Any patient identified as

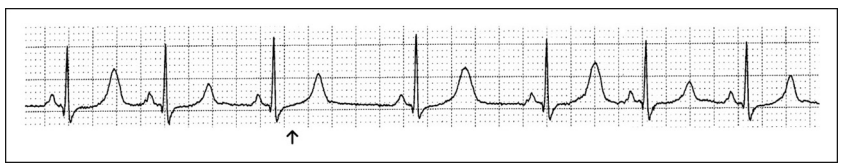

Figure 4. Long QT syndrome. The interval from the Q wave to the time at which the T wave returns to the isoelectric point is prolonged. This ECG also demonstrates sinus arrhythmia, a normal finding, with prolongation of the R-R interval during exhalation. To calculate the corrected QT (QTc) interval, the formula QT/$\sqrt{}$ previous R-R is used. The corrected QT interval (QTc) here is 505 msec. (48/$\sqrt{}$90)

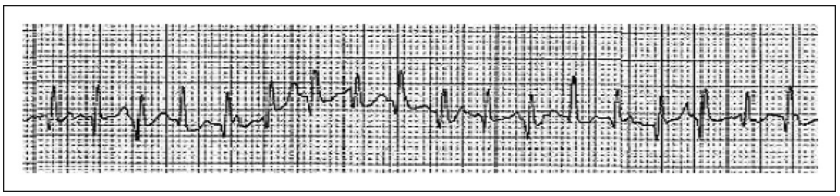

Figure 5. Ventricular tachycardia. The heart rate is approximately 200 beats/min, and the QRS complexes are wide (>120 msec). The QRS complexes vary in appearance, suggesting multifocal ectopic input.

having VT should be assessed immediately for hemodynamic instability. Once clinically stable, such patients require a cardiac evaluation, including radiography, echocardiography, exercise stress testing, and 24-hour Holter monitoring.

Ventricular Fibrillation

Ventricular fibrillation (VF) is a rare cardiac emergency caused by uncoordinated activity of the cardiac muscle fibers, often resulting in cardiac arrest. The heart tremors rather than contracts and, therefore, pulses are nonpalpable. The confirmatory diagnostic test is ECG, which shows a bizarre, random waveform without clearly identifiable P waves or QRS complexes and a roaming baseline. Any patient suspected of having VF requires advanced cardiac life support intervention because circulation may cease within seconds of onset. In the acute setting, such treatment involves the use of an electric defibrillator.

Although many clinicians may go an entire career without seeing an episode of VF, its rarity makes it all the more dangerous. Because this arrhythmia occurs most commonly in children after heart surgery, and the number of children surviving operative congenital heart disease is increasing, it is important to have at least a cursory knowledge of the acute diagnosis and management of VF.

Summary

- As in many aspects of medicine, a thorough history is vital for identification and diagnosis of cardiac arrhythmias in children and can help differentiate a benign arrhythmia from a pathologic one.
- In most cases, ECG is satisfactory for diagnosis. However, if the pediatrician feels that ECG is insufficient, it is best to refer the patient to a cardiologist for further evaluation.

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