Heart Rate and Rhythm Disorders

Philip Wackel, MD,* Bryan Cannon, MD*

**Pediatric Cardiology, Mayo Clinic, Rochester, MN*

Education Gaps

1. Proper evaluation of pediatric rhythm disorders can be challenging and requires knowledge of what tests are available and most appropriate to aid in this challenge. (1)
2. Correctly identifying common normal variants in the rhythm and distinguishing them from common abnormal findings is needed to properly triage and treat patients.

Objectives

After completing this article, readers should be able to:

1. Identify by history the symptoms concerning for rhythm disorders.
2. Select appropriate tests in the evaluation of suspected rhythm disorders.
3. Discuss common benign findings and appropriately limit testing/referrals.
4. Describe common rhythm disorders and potential associated underlying problems.

AUTHOR DISCLOSURE Dr Wackel has disclosed no financial relationships relevant to this article. Dr Cannon has disclosed that he serves on the Data Safety Monitoring Board of Medtronic and on the Board of Trustees of Mayo Support Services Texas. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

ABBREVIATIONS

AV	atrioventricular
AVR	accelerated ventricular rhythm
ECG	electrocardiogram
PAC	premature atrial contraction
PVC	premature ventricular contraction
QTc	corrected QT
SVT	supraventricular tachycardia
VT	ventricular tachycardia

INTRODUCTION

Rhythm disorders in pediatric patients encompass a vast and diverse group of problems that vary greatly not only in etiology but also in presentation. These disorders can range from bradycardia to tachycardia but also include inherited channelopathies as well as rhythm disorders associated with underlying congenital heart disease. With such a diverse group of potential problems and the high degree of variability in presenting symptoms, the evaluation and diagnosis of rhythm disorders can be difficult. As with most medical issues, starting with a thorough history and physical examination will help guide the need for further evaluation and testing to reach the appropriate diagnosis.

PRESENTATION

The presentation of arrhythmias varies depending on the type of arrhythmia, but even within the same arrhythmia there can be significant variation in presenting symptoms. Palpitations are a common complaint in the pediatric population, but most patients with palpitations do not have an underlying arrhythmia. Determining which patients need further evaluation and/or medical therapy can be challenging. Palpitations, or the feeling of abnormal heart beats, can be perceived

and described quite differently between patients. However, with a focused and detailed history it is often possible to significantly increase or decrease the suspicion for a true arrhythmia and, thus, direct the evaluation accordingly (Table 1). Typically, palpitations that present with sudden onset and sudden termination in a single beat are more consistent with an abnormal mechanism for tachycardia than when initiation and/or termination are gradual. In addition, palpitations that occur for only a single beat or last only a few seconds are less likely to correlate with a true tachyarrhythmia that is caused by an abnormal electrophysiologic mechanism. Small children may have difficulty describing symptoms and often note pain (or discomfort) over the chest because they lack the vocabulary to describe the sensation of palpitations.

Syncope is less commonly related to arrhythmias but should be taken seriously and investigated further because certain rhythm disorders may present with syncope. The most likely mechanism of syncope in an otherwise healthy child is benign autonomic reflex–mediated syncope and is not related to a rhythm disorder. (2) Benign syncope can usually be diagnosed largely based on history because there is often a prodrome before syncope and/or it is often associated with positional changes. Syncope during exercise, syncope with no prodrome, or syncope in a patient with a history of heart disease is more concerning for an underlying arrhythmia and should prompt further evaluation. Syncope that is preceded by palpitations may suggest a tachyarrhythmia, although this must be qualified by the circumstance as well as the description of the palpitations as oftentimes orthostatic changes can produce sinus tachycardia before reflex syncope or presyncope.

Rhythm disorders can be difficult to diagnose because patients may have completely normal examinations and evaluation findings when they are not actively having tachycardia. Patients in relatively slow tachycardia, with a heart rate that overlaps into the upper range of normal rates for age, may not perceive an abnormal rate and can be at risk for developing tachycardia-induced cardiomyopathy and symptoms of heart failure. (3) They can present with fatigue, shortness of breath, and edema. These findings may be subtle, and the presentation can overlap with many other common problems. Infants and nonverbal children with tachyarrhythmias may also present with signs of heart failure that can be subtle. Infants in tachycardia can present with poor feeding, diaphoresis, irritability, or somnolence before the abnormal mechanism of tachycardia is discovered.

TESTING

The evaluation for tachyarrhythmias can be divided into 2 aspects. The first is to make the appropriate diagnosis of the type of arrhythmia, if any, that is present. Depending on the type of arrhythmia, the second aspect of the evaluation may include trying to identify an underlying/precipitating cause or condition. The key to diagnosing or excluding arrhythmia is to record the rhythm during symptoms. If only sinus tachycardia is noted during symptoms, it may be reasonable to pursue an evaluation looking for the underlying cause of sinus tachycardia if this is not apparent from the history. Depending on the history and physical examination findings, this may include laboratory evaluation for hyperthyroidism and anemia. (4) If suspicion is raised clinically, testing for levels of urine catecholamines and/or urine vanillylmandelic acid and homovanillic acid to look for more rare causes of sinus tachycardia, such as pheochromocytoma and neuroblastoma, respectively, could be considered, although these conditions are exceptionally rare causes of tachycardia.

Recording the rhythm during symptoms can be accomplished in a variety of ways, and selection of the appropriate modality depends on the symptom characteristics. A resting electrocardiogram (ECG) should be acquired first, as this may provide information that could implicate certain arrhythmias even if the patient is not symptomatic at the time of the ECG. A 24- or 48-hour ambulatory cardiac rhythm monitor (Holter) may also be of value if symptoms are

TABLE 1. **Evaluation of Palpitations**

PALPITATIONS REQUIRING FURTHER EVALUATION	LESS CONCERNING PALPITATION CHARACTERISTICS
Abnormal electrocardiographic findings	Occurs with orthostatic changes
Associated with syncope	Preceded by lightheadedness/dizziness
Starts and stops in a single beat	Gradual onset and termination
Rate >220 beats/min or too fast to count	Rate <150 beats/min
Occurs during exercise	
Family history of sudden death	

occurring daily (or nearly daily) and it is likely that the patient will experience the symptoms while wearing the monitor. There are monitors that will continuously record the rhythm for up to 2 weeks if longer monitoring is needed to capture an event. For symptoms occurring less frequently, an event recorder may be more appropriate. There are different types of event recorders available but in general these are issued to patients to use in 30-day increments. There are 2 different types of event recorders. The first type continuously records the heart rhythm through electrodes connected to the chest (also known as a loop recorder). If the patient feels symptoms, he or she can push a button, and the monitor has the ability to retroactively store an ECG recording of the rhythm that occurred minutes before the patient activated the button. Loop recorders are particularly helpful for patients with infrequent and brief symptoms (<3 minutes). However, the downside is that the patient must be wearing the monitor to capture an event, and frequently the ECG electrodes and wires become bothersome with continual use for a month. Alternatively, other monitors (known as event recorders) can be used to capture ECG tracings during symptoms. Event recorders are handheld devices that are carried with the patient. These recorders contain ECG electrodes on the device that are capable of recording an ECG tracing when placed directly on the skin of the chest. When patients feel symptoms, they can pull out the monitor and place it on their chest and activate a button to record their rhythm during a symptom. This type of event recorder is useful for rare symptoms that are sustained. The patient must have sufficient time to recognize the palpitations, locate the event recorder, and properly place the monitor on the chest, which usually takes several minutes. This type of monitor may also be helpful in situations when the symptoms occur only in circumstances that would preclude constantly wearing a monitor, such as symptoms while swimming. There are also commercially available smartphone adapters and applications that are capable of electronically recording and storing a patient's cardiac rhythm. However, caution must be exercised when using certain programs because not all have been validated clinically for accuracy. (5) If unsuccessful in capturing a recording of the rhythm during symptoms despite attempts with ambulatory monitors, then implantable loop recorders and/or invasive testing by a pediatric electrophysiologist may be warranted.

RHYTHM DISORDERS

Normal Variations

On a baseline ECG there are common normal variants that are often encountered and must be interpreted correctly to avoid unnecessary further testing. One of the most common of these is sinus arrhythmia, also known as respiratory sinus arrhythmia. Sinus arrhythmia is a variation in the sinus rate during respiration that can sometime be quite abrupt and dramatic (Fig 1). The heart rate increases with inspiration and decreases with expiration in a reproducible pattern. Although labeled as an arrhythmia, this finding is a normal variant. The distinguishing characteristic in sinus arrhythmia is that the atrial contraction and resulting P wave on the ECG continues to originate from the sinus node and not from an ectopic atrial focus. The P wave morphology in sinus arrhythmia can vary slightly depending on the exit point from the sinus node, but the P wave morphology should not be drastically different than other sinus beats. In general, the P wave will remain upright in leads I and aVF, resulting in a normal P wave axis between 0 and 90 degrees.

Other normal variants can include early repolarization, which results in ST elevation in leads with upright T waves, a gradually upsloping ST segment, and normal-appearing T waves. This is almost universally considered a benign finding in pediatric patients and should be considered a normal variant. Also, an rSR′ pattern in lead V1 is a common normal variation seen primarily in adolescents. The features of a benign rSR′ pattern are a normal QRS duration for age and an R′ wave that is not taller than the initial R wave. Any of the aforementioned normal variants occurring in isolation should not automatically trigger further evaluation.

Extrasystoles

Premature atrial contractions (PACs) and premature ventricular contractions (PVCs) are referred to as extrasystoles and are generally noted incidentally as they only rarely cause symptoms. When symptoms are present, they are often described as single skipped beats or single hard beats. A PAC is an atrial beat that occurs before the next expected sinus beat and originates from atrial tissue that is not the sinus node, resulting in a P-wave morphology different than sinus. Most PACs will conduct normally to the ventricle, resulting in a QRS complex that appears identical to that

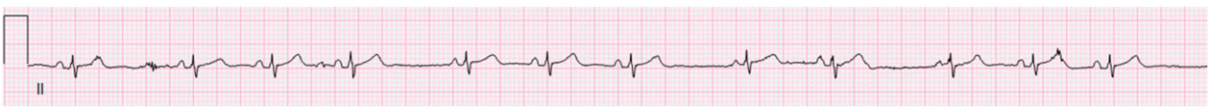


Figure 1. Sinus arrhythmia characterized by variation in the rate with no change in the P-wave morphology.

during normal sinus rhythm. However, some PACs may occur sufficiently early after the previous sinus beat that the atrioventricular (AV) node is still refractory, which results in no QRS complex and is referred to as a blocked PAC (Fig 2A). Also, if a PAC occurs while the His-Purkinje system is only partially refractory, this can result in aberrancy and a wide QRS complex following the PAC (Fig 2B). These PACs with aberrancy are often mistaken for PVCs due to the wide QRS complex, but careful inspection can reveal a P wave before these beats. A PAC in isolation does not by itself necessitate further evaluation, and most often, no treatment is required. Premature atrial contractions are particularly common in the newborn period and tend to resolve in the days or weeks after birth. A Holter monitor may be useful to try to correlate symptoms with PACs, quantify the PAC burden, and assess for occult atrial tachycardia but should be guided by clinical judgment.

A PVC is an early beat arising from the ventricular muscle resulting in a QRS complex that is different from sinus and is not preceded by a P wave. Generally, PVCs result in a wide QRS complex due to slower muscle-to-muscle conduction, but if they originate from or very near to the His-Purkinje fibers, the QRS complex can actually be fairly narrow, and in this case careful inspection of the QRS complex is required to identify a difference from a sinus QRS complex. Premature ventricular contractions are a common finding on an ECG or Holter monitor, occurring in up to 40% of pediatric patients. (6) Most patients with PVCs will have a structurally normal heart, but occasionally PVCs can be associated with myocarditis, cardiomyopathy, or channelopathies. Clinical features that may imply an underlying cardiac problem are PVCs of multiple morphologies, episodes of 3 or more PVCs in a row constituting ventricular tachycardia, or PVCs that increase in frequency with exercise. Patients with asymptomatic isolated PVCs that are of a single morphology, suppress with exercise, and occur in the presence of a structurally normal heart are almost always benign. These patients may be followed serially by a cardiologist if the ectopy burden is high (usually >10% PVCs on a 24-hour Holter monitor) to watch for development of arrhythmia-induced ventricular dysfunction, although this generally does not occur until the burden of ectopy is much higher. (7) Treatment of PVCs is reserved for those rare patients who are symptomatic from the PVCs or who develop ventricular dysfunction. First-line therapy can be antiarrhythmic agents, but some patients may elect to go straight for an ablation procedure to avoid medication use.

AV Block

Conduction abnormalities resulting in delayed or no conduction from the atrium to the ventricle are termed AV block. Categories of AV block include first-degree AV block, various types of second-degree AV block, and third-degree AV block, also known as complete AV block (Table 2). First-degree AV block is a misnomer because no AV block actually occurs but rather there is prolonged AV conduction resulting in a long PR interval for age (Fig 3). The normal PR interval varies according to age, with shorter PR intervals occurring in infants and longer PR intervals occurring in teenagers. First-degree AV block is most commonly seen as a normal variant in an otherwise healthy young person and is not always a sign of true conduction disease. (8) It may be associated with increased vagal tone and frequently will resolve when the sympathetic tone increases, such as during exercise. However, first-degree AV block can be associated with some types of congenital heart disease, infectious etiologies, or medication use, and further investigation may be warranted if clinical suspicion for an underlying

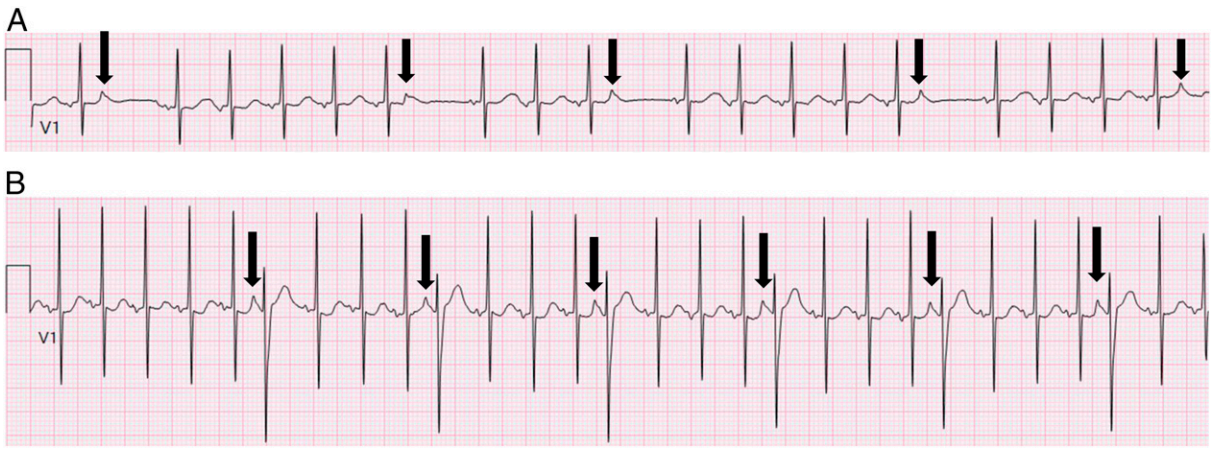


Figure 2. A. Blocked premature atrial contractions (PACs) (arrows). B. PACs (arrows) with aberrant conduction.

TABLE 2. **Types of AV Block**

TYPE OF AV BLOCK	DESCRIPTION
First-degree	PR interval prolongation only with no nonconducted P waves
Second-degree	
Mobitz type I	Progressive PR interval lengthening before a single nonconducted P wave
Mobitz type II	Sudden nonconducted P wave with no change in the PR on the previous beat
2:1	Every other P wave conducts to the ventricle
High-grade	More than a single nonconducted P wave in a row but some P waves do conduct
Third-degree	No P waves conduct to the ventricle

AV=atrioventricular.

problem is present. A Holter monitor may also be helpful to assess the AV conduction over 24 hours to screen for higher degrees of AV block that may be variably present throughout the day and to assess AV node conduction at higher heart rates. If first-degree AV block is present in isolation, it generally does not result in symptoms and generally does not require any treatment.

Mobitz type I second-degree AV block, or Wenckebach block, is a progressive lengthening in the PR interval before a single beat of AV block that is followed by a return of AV conduction on the following beat. This pattern may repeat itself and give the appearance of so-called grouped beating (Fig 4). This should be distinguished from sinus arrhythmia, which can also appear as grouped beating but without any block in AV conduction. Mobitz type I second-degree AV block is commonly seen in asymptomatic healthy young people at rest or during sleep secondary to high vagal tone affecting the AV node. (9) When it occurs during periods of high vagal tone, it almost always is an incidental finding without any symptoms and it does not require any further evaluation or treatment. Wenckebach is unusual to see in an individual who is awake unless the person is an elite athlete. A Holter monitor to assess for long pauses or higher degrees of AV block may be considered.

In contrast to Mobitz type I, which is common and usually benign, Mobitz type II second-degree AV block is always abnormal and always warrants further investigation. In Mobitz type II second-degree AV block there is no lengthening of the PR interval before AV block but rather abrupt AV block for a single beat. This generally correlates with AV conduction disease below the AV node in the His-Purkinje system and implies disease or damage to the AV conduction. (10) This finding is extremely rare in pediatrics and almost always occurs in the setting of an underlying bundle branch block. It is important when making the diagnosis to ensure that there is no prolongation of the PR interval before the dropped beat because the lengthening seen in Wenckebach block may be very subtle. In addition to an ECG, Holter monitor, and echocardiogram, further investigation of possible etiologies, including myocarditis, some forms of cardiomyopathy, or infiltrative disease, may be warranted. Presenting symptoms will vary depending on the underlying etiology but also may be absent. Pacemaker placement is indicated if the etiology is not reversible because Mobitz type II second-degree AV block is generally associated with unreliable AV conduction and can lead to sudden complete AV block.

There are other forms of second-degree AV block, including 2:1 AV block, characterized by every other atrial beat conducting to the ventricle, as well as high-grade AV block, in which 2 or more atrial beats in a row do not conduct to the ventricle. Depending on the frequency and the degree of AV

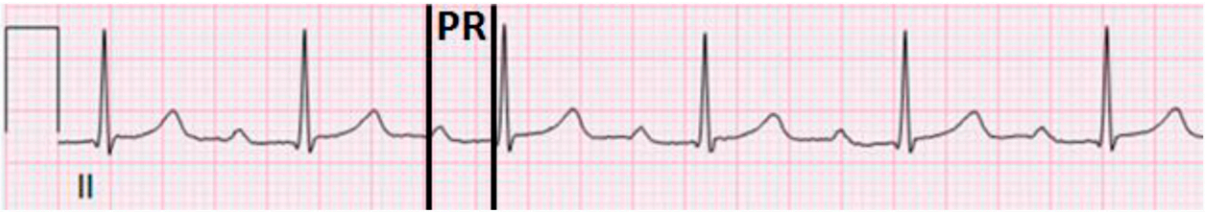


Figure 3. First-degree atrioventricular block with a PR interval of 252 milliseconds.

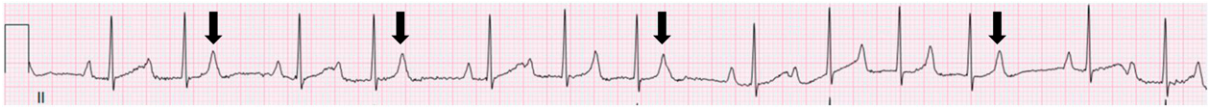


Figure 4. Type I second-degree atrioventricular (AV) (Wenckebach) block with progressive prolongation of the PR interval before a single beat of AV block (arrows).

block present, these forms of AV block may present incidentally, with palpitations, fatigue, lightheadedness, or syncope. Both of these other types of second-degree AV block also warrant evaluation by a cardiologist for the same indications as Mobitz type II second-degree AV block.

Third-degree AV block, also known as complete AV block, is seen when no atrial beats are conducted to the ventricle (Fig 5). Presenting symptoms will vary and will depend largely on the underlying escape rate and the presence or absence of associated cardiac abnormalities. Most often there will be a junctional, or possibly a ventricular, escape rhythm that provides the impulse to drive ventricular contraction. If the rate of the escape rhythm is fast enough, complete AV block may go unrecognized and lead to no symptoms. In others, the escape rhythm may be very slow, resulting in profound symptoms, including syncope, fatigue, signs of heart failure, or possibly sudden death if it occurs abruptly. Associated congenital heart disease or the presence of ventricular dysfunction will also influence the presenting symptoms.

In individuals presenting with third-degree AV block, the common etiologies include maternal autoimmune disorders, structural congenital heart disease, some types of cardiomyopathy, and acquired causes such as myocarditis, Lyme disease, or Chagas disease. An ECG, Holter monitor, and echocardiogram are all indicated in these patients. The acquired causes of complete AV block can occur at any age, and although congenital complete AV block is by definition present at the time of birth, it may not be identified in infancy if the underlying escape rate is fast enough. Therefore, in the presence of a structurally normal heart, further investigation into maternal autoimmune disorders is warranted, specifically to investigate for maternal systemic lupus erythematosus, maternal Sjögren syndrome, and other autoimmune diseases, including the presence of maternal anti-SSA/Ro and anti-SSB/La antibodies, which have all been linked to the development of congenital complete AV block. (11) Treatment is usually supportive, and if no reversible cause is found, permanent pacemaker placement may be warranted depending on several variables. If there is no congenital heart disease and there is normal ventricular function, a narrow QRS escape rhythm, an adequate rate (usually >55 beats/min), and the patient is hemodynamically stable, then a pacemaker may not be needed immediately. (12) However, patients with complete AV block will require ongoing reassessment from a pediatric cardiologist to determine when a pacemaker is indicated.

Prolonged QT Interval

The QT interval is measured from the onset of the QRS complex to the end of the T wave and is expressed in milliseconds (Fig 6). It is most reliably measured in leads II and/or V5. The QT interval changes with heart rate, which has led clinicians to "correct" it for heart rate, resulting in the corrected QT (QTc) interval. The Bazett formula ($QTc = \frac{QT}{\sqrt{R\text{-}R}}$) is the most commonly used formula to calculate the QTc interval and is done so by dividing the QT interval (in seconds) by the square root of the preceding R-R interval (in seconds). The QT interval automatically measured by the computer is often incorrect and should always be remeasured by hand before confirming that the QTc interval is prolonged. The exact cutoff value for what constitutes an abnormal QTc interval is debatable. In general, a QTc interval greater than 440 milliseconds in prepubertal boys and greater than 450 milliseconds in prepubertal girls is considered abnormal but does not necessarily indicate an underlying pathologic condition because these values significantly overlap with the upper percentiles of normal. (13) A QTc interval greater than 470 milliseconds should prompt an evaluation for long QT syndrome, and values greater than 500 milliseconds are virtually never seen in the absence of a pathologic condition.

Long QT syndrome is a channelopathy, or a disease caused by disturbed function of an ion channel involved in

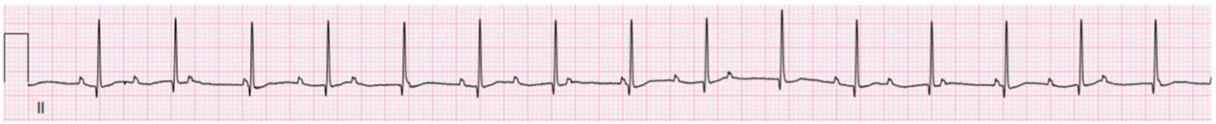


Figure 5. Complete atrioventricular block results in no relationship between the timing of the P wave and the QRS complex and no variation in the ventricular rate.

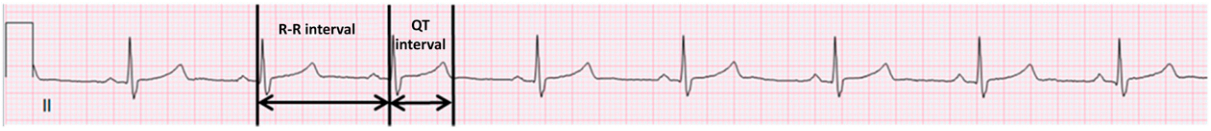


Figure 6. The R-R interval is measured from the beginning of the QRS complex to the beginning of the next QRS complex. The QT interval is measured from the beginning of the QRS complex to the end of the T wave. In this example, the R-R interval is 0.98 seconds and the QT interval is 0.47 seconds, resulting in a corrected QT interval of 475 milliseconds.

myocardial contraction. These are most commonly potassium channels or sodium channels. These disordered channels can result in electrical disturbances in the heart, including life-threatening ventricular arrhythmias. However, prolongation of the QTc interval can happen for a variety of reasons other than congenital long QT syndrome, including medications, electrolyte abnormalities, and neurologic abnormalities. It is, therefore, important to assess for these possibilities before labeling someone as having long QT syndrome because this diagnosis comes with the stigma of possible sudden death and can often be a difficult diagnosis to reverse if incorrect. However, QTc intervals greater than 500 milliseconds predispose the patient to dangerous ventricular arrhythmias regardless of the underlying cause.

Most patients who have long QT syndrome are asymptomatic, and the presenting symptoms of long QT syndrome are typically related to the occurrence of torsades de pointes, which is a specific type of polymorphic ventricular tachycardia where the QRS axis gradually changes in amplitude (Fig 7). Torsades de pointes will have a sudden onset and can present as palpitations, syncope, cardiac arrest, or sudden death. If long QT syndrome is suspected, a referral to a cardiologist is indicated before genetic testing is pursued. However, if sudden death occurs it is crucial to order genetic testing for channelopathies on the decedent's blood saved from the autopsy if no other cause of death is identified. This so-called molecular autopsy could be vital to the evaluation and treatment of family members. The treatment of long QT syndrome varies depending on multiple factors, including history, type of mutation, and response to previous treatment. However, β-blockers are almost always first-line therapy in all patients with diagnosed long QT syndrome. An implantable cardioverter-defibrillator may be indicated in high-risk long QT syndrome after careful consideration of the risks and options.

Supraventricular Tachycardia

Supraventricular tachycardia (SVT) is the most common type of abnormal tachycardia present in pediatric patients. (14) Most often it presents as sudden-onset narrow complex tachycardia, but it may also have a wide QRS complex in certain scenarios. Symptoms are classically described as having an abrupt onset and abrupt termination of rapid heart rates. The abrupt nature of the palpitations stems from the fact that more than 90% of pediatric SVT has a reentrant mechanism using an accessory pathway or 2 inputs into the AV node. (15) Most commonly, reentrant SVT consists of conduction down the AV node, resulting in ventricular contraction followed by conduction "reentering" the atrium using one of the aforementioned abnormal structures, which then facilitates conduction back down the AV node and subsequent "reentry" back to the atrium, thereby perpetuating this cycle of activation and reactivation and leading

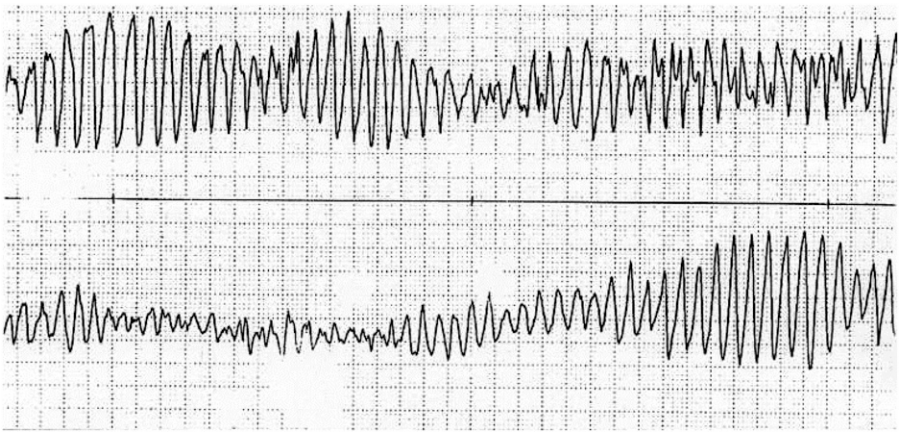


Figure 7. Torsades de pointes.

to abrupt tachycardia. Generally, pediatric patients who are otherwise healthy and have structurally normal hearts hemodynamically tolerate SVT for short periods, even at rates well above 200 beats/min. In addition to the rapid heart rates they may report diaphoresis, nausea, a pale appearance, and/or presyncopal feelings, but true syncope is rare. Supraventricular tachycardia can be a difficult diagnosis to make because palpitations are common in the pediatric population. Table 1 describes some features to help distinguish benign palpitations from an abnormal mechanism of tachycardia. Recording a rhythm strip or ECG while in tachycardia is vital before initiating acute treatment in an otherwise stable patient so that the tachycardia is documented and can be reviewed later if needed. Acute treatment with vagal maneuvers such as ice to the face or performing a Valsalva maneuver may abruptly terminate SVT. Vagal maneuvers such as gagging and ocular massage should be avoided in young patients because these can cause physical harm when not done carefully/properly. If needed, intravenous adenosine can usually abruptly terminate SVT due to its transient effect on the AV node. If giving intravenous adenosine, it is again important to record the rhythm while administering the adenosine because this may lend information as to the exact mechanism of SVT when it terminates. Also, adenosine must be delivered rapidly via a large-bore intravenous push with an immediate saline flush behind it due to the extremely rapid metabolism of adenosine, resulting in a half-life of 1 to 6 seconds. The possible adverse effects of adenosine therapy include flushing, brief anxiety or a sense of impending doom, nausea, chest pain, bronchospasm, initiation of atrial fibrillation, transient AV block, and transient hypotension, so before administering adenosine, one needs to be prepared to manage the possible adverse effects.

In 90% of pediatric patients, SVT is from either AV reentry tachycardia using an accessory pathway or AV nodal reentry tachycardia using 2 inputs into the AV node. (15) Once tachycardia has terminated, a baseline ECG should be obtained to look for a Wolff-Parkinson-White pattern, which consists of a short PR interval, a delta wave, and a wide QRS complex (Fig 8). Electrolyte disturbances and hyperthyroidism rarely play a role in the development of SVT and are not routinely evaluated at presentation unless other clinical signs or predisposing factors are present. The goal of long-term management of SVT is to prevent recurrence, and this can be accomplished with either medications or an ablation procedure. First-line medications include β-blockers or calcium channel blockers. Once a patient weighs more than 15 kg, it is reasonable to pursue an ablation to obtain a permanent cure. Below 15 kg, medical therapy options are usually exhausted before considering an ablation unless there is poor ventricular function. In general, ablation procedures have a very high likelihood of permanently eliminating the abnormal substrate needed for tachycardia, and the risks of an ablation are fortunately very low in the modern era. (16)(17) This makes an ablation procedure an attractive option for many patients who want to avoid the lifelong cost and inconvenience of taking medications indefinitely.

Ventricular Tachycardia

Accelerated ventricular rhythm (AVR) or ventricular tachycardia (VT) occurs when 3 or more beats in a row originate from the ventricular myocardium at a rate faster than sinus. Depending on the rate it will be classified as either AVR or VT. It is important to differentiate AVR from VT in pediatric patients because there are different prognostic implications of each. The exact rate at which this distinction is made varies with age and is based on the rate of the ventricular arrhythmia compared with the sinus rate. Generally, AVR is defined as rates up to 15% to 20% faster than the expected sinus rate or a ventricular rate less than 120 beats/min in the older teenager. Typically, AVR is benign, rarely requires treatment, and carries the same excellent prognosis as PVCs. (18) An AVR can occur at any age and may present as early as the first hours after birth. In the absence of structural heart disease and/or metabolic/electrolyte abnormalities, AVR is thought to be benign but does require longitudinal follow-up to assess for resolution and to monitor for the development of ventricular dysfunction. In a neonate with AVR, most will self-resolve within the first year after birth, and treatment is needed only if ventricular dysfunction occurs. In older children, AVR can be followed much like that of isolated PVCs, as discussed previously herein, with longitudinal follow-up to assess burden of ectopy and for the potential development of ventricular dysfunction in the setting of frequent ectopy.

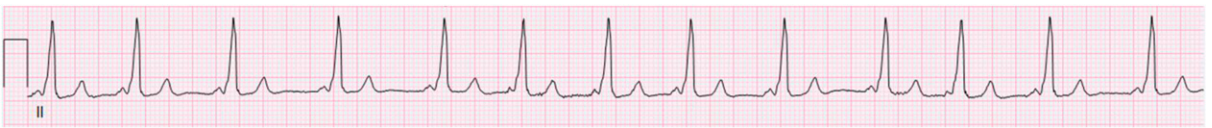


Figure 8. Wolff-Parkinson-White pattern characterized by a short PR interval, sloping of the initial QRS complex known as a delta wave, and a wide QRS complex.

Ventricular tachycardia can be divided or categorized in different ways. It can be described as monomorphic versus polymorphic and also divided into VT in the normal heart versus the abnormal heart. Monomorphic VT in the presence of a normal heart is most likely benign, whereas polymorphic VT or VT in the presence of heart disease is more concerning for a pathologic cause. Although ventricular tachycardia requires a thorough cardiovascular evaluation by a cardiologist, not all causes are life-threatening. Therefore, establishing that the heart is structurally and electrically normal is vital before entertaining one of the common causes of benign VT in a child. The initial evaluation should, thus, include an echocardiogram, ECG, and Holter monitor. If there is a clinical reason to suspect an electrolyte abnormality, then laboratory values should also be obtained, but most pediatric VT will occur in the face of normal electrolytes. The appearance of some types of benign VT can overlap with that of arrhythmogenic right ventricular cardiomyopathy, myocarditis, cardiomyopathy, or various channelopathies. Therefore, ensuring that there is no underlying heart disease in the face of VT may also require additional testing with an exercise stress test and/or cardiac magnetic resonance imaging, depending on the clinical scenario. Once there is establishment of a completely normal heart, the causes of VT are generally benign and include outflow tract VT and posterior fascicular VT. These 2 types of VT are monomorphic and may be sustained or nonsustained. Oftentimes they present with palpitations but can also present as an incidental finding. Treatments of these types of VT are dictated by their frequency, the presence of symptoms, and any effect on ventricular function. Treatment includes medical therapy with antiarrhythmic agents and/or an ablation procedure, but no treatment may be necessary in the face of a normal heart, slow ventricular tachycardia, and the absence of symptoms.

Polymorphic VT, defined as beat-to-beat variations in the QRS morphology and/or QRS axis during VT, is rare in childhood and generally carries a worse prognosis than monomorphic VT. It is extremely rare to have polymorphic VT in children with no underlying heart disease, and it is almost always associated with an underlying cardiac abnormality. Therefore, the presence of multiform ventricular ectopy or polymorphic VT in a child warrants a thorough investigation into underlying structural heart disease, cardiomyopathy, myocarditis, and channelopathies. In addition, metabolic/electrolyte abnormalities and toxicity from medications or recreational drugs should be investigated as well. Even if no etiology is identified, these patients warrant longitudinal follow-up and, depending on their degree of ectopy and symptoms, may require antiarrhythmic agents or even implantation of an internal cardioverter-defibrillator depending on the severity of the presentation.

Summary

On the basis of:

- Good evidence and expert opinion, the history will help determine the level of suspicion for a rhythm disorder and help guide appropriate testing. (2)
- Expert consensus, an electrocardiogram (ECG) is indicated in all patients suspected of having a rhythm disorder. (4)
- Expert opinion, palpitations that are associated with abnormal ECG findings associated with syncope, occur at a rate greater than 220 beats/min, have an abrupt onset and termination, or occur during exercise require further investigation. (1)
- Strong evidence, it is important to properly identify normal variants seen on an ECG or rhythm tracing that do not require further evaluation or referral. (6)(8)(9)
- Good evidence and expert opinion, proper interpretation of a rhythm disorder will determine the need for referral or further testing. (12)(13)(14)

References and Suggested Readings for this article are at http://pedsinreview.aappublications.org/content/38/6/243.

PIR Quiz

There are two ways to access the journal CME quizzes:
1. Individual CME quizzes are available via a handy blue CME link under the article title in the Table of Contents of any issue.
2. To access all CME articles, click "Journal CME" from Gateway's orange main menu or go directly to: http://www.aappublications.org/content/journal-cme.

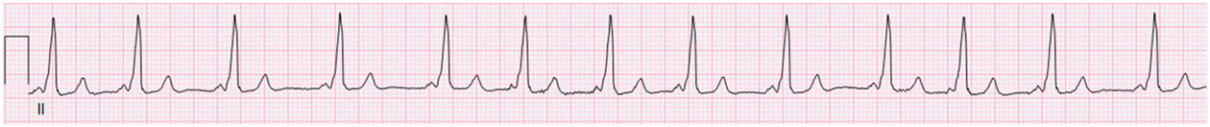
1. A previously healthy 15-year-old boy returns to school a day after a 4-day episode of vomiting and diarrhea. While he was getting up from his chair to go to his next class he remarked that he felt dizzy and then fainted. After 10 to 15 seconds, he woke up and was oriented to person, place, and time. Which of the following is the most likely diagnosis?
   A. Benign autonomic reflex–mediated syncope.
   B. Idiopathic hypertrophic cardiomyopathy.
   C. Mobitz type II second-degree atrioventricular (AV) block.
   D. Prolonged QT interval.
   E. Supraventricular tachycardia.
2. A previously healthy 14-year-old girl is brought to the office by her mother because her "heart feels like it is beating fast." She recently was selected to be on her high school debate team and during 2 recent competitions during the past 8 days she stated that she felt her heart beating rapidly. She did not faint and she did not feel dizzy. She did have facial blushing. She thinks the episodes lasted less than 5 minutes. On examination, her heart rate is 82 beats/min, respiratory rate is 20 breaths/min, and blood pressure is 106/64 mm Hg. The remainder of her examination findings are normal. An electrocardiogram (ECG) shows a heart rate of 86 beats/min that increases with inspiration and decreases with expiration. P waves are upright in leads I and aVF. The PR interval is 0.15 seconds, QRS duration is 0.10 seconds, and QRS axis is +60. T waves and ST segments are normal. The ECG results are most consistent with which of the following?
   A. Accelerated ventricular rhythm.
   B. First-degree heart block.
   C. Polymorphic ventricular tachycardia.
   D. Sinus arrhythmia.
   E. Supraventricular tachycardia with aberrant conduction.
3. A healthy 14-year-old boy (75th percentile for weight and height) is referred to a pediatric cardiologist following the sudden death of his 16-year-old brother while playing in a pickup basketball game with friends. An autopsy was performed that included a "molecular autopsy" that identified a mutation in the *KCNH2* gene consistent with a diagnosis of long QT type 2 syndrome. An ECG on the 14-year-old boy reveals a corrected QT interval of 510 milliseconds. Which of the following is the most appropriate next step in management?
   A. β-Blocker therapy.
   B. Dietary potassium restriction.
   C. Dietary sodium supplementation.
   D. Digoxin.
   E. Left cardiac sympathetic denervation.

REQUIREMENTS: Learners can take *Pediatrics in Review* quizzes and claim credit online only at: http://pedsinreview.org.

To successfully complete 2017 *Pediatrics in Review* articles for *AMA PRA Category 1 Credit™*, learners must demonstrate a minimum performance level of 60% or higher on this assessment, which measures achievement of the educational purpose and/or objectives of this activity. If you score less than 60% on the assessment, you will be given additional opportunities to answer questions until an overall 60% or greater score is achieved.

This journal-based CME activity is available through Dec. 31, 2019, however, credit will be recorded in the year in which the learner completes the quiz.

2017 *Pediatrics in Review* now is approved for a total of 30 Maintenance of Certification (MOC) Part 2 credits by the American Board of Pediatrics through the AAP MOC Portfolio Program. Complete the first 10 issues or a total of 30 quizzes of journal CME credits, achieve a 60% passing score on each, and start claiming MOC credits as early as October 2017.



4. A previously healthy 12-year-old girl is brought to the emergency department by her parents after fainting. Before fainting she stated that she could feel her heart beating and was light headed. After awakening she was oriented to person, place, and time and could answer questions. On physical examination, she weighs 40 kg (50th percentile), her heart rate is 80 beats/min, and her blood pressure is 101/62 mm Hg. Lead II of an ECG is shown above. Which of the following is the diagnosis?
   A. First-degree AV block.
   B. Mobitz type II second-degree AV block.
   C. Monomorphic ventricular tachycardia.
   D. Polymorphic ventricular tachycardia.
   E. Wolf-Parkinson-White syndrome.
5. For the same 12-year-old girl as in the previous question, which of the following is the most appropriate treatment to prevent tachycardia and achieve cure?
   A. Adenosine.
   B. Amitriptyline.
   C. Digoxin.
   D. Procainamide.
   E. Radiofrequency ablation.

Additional Resources for Pediatricians

AAP Textbook of Pediatric Care, 2nd Edition

- Chapter 132: Cardiac Arrhythmias - https://pediatriccare.solutions.aap.org/chapter.aspx?sectionId=107998264&bookId=1626

Point-of-Care Quick Reference

- Cardiac Arrhythmias - https://pediatriccare.solutions.aap.org/Content.aspx?gbosid=165412

Parent Resources from the AAP at HealthyChildren.org

- Irregular Heartbeat (Arrhythmia): https://www.healthychildren.org/English/health-issues/conditions/heart/Pages/Irregular-Heartbeat-Arrhythmia.aspx

For a comprehensive library of AAP parent handouts, please go to the *Pediatric Patient Education* site at http://patiented.aap.org.

Heart Rate and Rhythm Disorders

Philip Wackel and Bryan Cannon

Pediatrics in Review 2017;38;243
DOI: 10.1542/pir.2016-0119

Heart Rate and Rhythm Disorders

Philip Wackel and Bryan Cannon

Pediatrics in Review 2017;38;243
DOI: 10.1542/pir.2016-0119