

Supraventricular Tachycardia in the Pediatric Primary Care Setting: Age-related Presentation, Diagnosis, and Management

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ABSTRACT

As many as 1 in 250 children experience supraventricular tachycardia (SVT), but its presentation is often vague and its symptoms mistakenly attributed to other common pediatric conditions. If SVT is correctly identified in a timely manner, most children will go on to live normal healthy lives. SVT is not covered in depth in most pediatric advanced practice nursing programs, but because of its prevalence, it should be familiar to all pediatric primary care providers. This article reviews common mechanisms of SVT and their age-related presentation, diagnosis, and

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management. A case study of an 8-year-old boy with SVT is presented. *J Pediatr Health Care.* (2008) 22, 289-299.

Key words: Supraventricular tachycardia, atrioventricular nodal re-entry tachycardia, wolff-parkinson-white, atrial ectopic tachycardia, atrioventricular reciprocating tachycardia, accessory pathway-mediated re-entry tachycardia, automatic tachycardia, permanent junctional reciprocating tachycardia, electrophysiology, SVT, AVNRT, WPW, AET, AVRT, PJRT

Nick, an 8-year-old star hockey player, presented to his pediatric primary care provider with a 1-month history of decreased energy and shortness of breath. His father had noted his decreasing ability to keep up with his teammates during the past 4 months. In the office, Nick's temperature was 97.1°F, his heart rate was 160 beats per minute (BPM), his respiratory rate was 30 breaths per minute, and his blood pressure was 100/70 mm Hg. Although his primary care provider had only a limited knowledge of supraventricular tachycardia (SVT), she suspected that this could be the cause of his problems and sent him directly to the emergency department for evaluation.

INTRODUCTION

SVT is the most common symptomatic pediatric arrhythmia (Vos, Pulles-Heintzberger, & Delhaas, 2003). It is defined as a sustained tachyarrhythmia originating above the bundle of His (Hanisch, 2001). SVT is caused by different physiologic conditions that result in similar electrocardiogram (ECG) features (Grossman, 1997). Although its exact incidence is unknown, it has been estimated to affect between 1 in 25,000 to 1 in 250 children (Chun & Van Hare, 2004). The purpose of this article is to increase awareness of SVT among pediatric primary care providers by

reviewing common types of SVT and presenting an age-based framework of common presentations, diagnosis, and management.

As many as 16 different mechanisms of pediatric SVT exist (Grossman, 1997). This article will focus on the three most common groups of SVT mechanisms: accessory pathway-mediated re-entry tachycardia, atrioventricular nodal re-entry tachycardia (AVNRT), and atrial ectopic tachycardia (AET) (Doniger & Sharieff, 2006). These distinct mechanisms have different age-related

experiencing more frequent and uncomfortable symptoms may benefit from pharmacologic management or curative invasive therapy (Chun & Van Hare, 2004).

SVT may be difficult for the pediatric primary care clinician to assess because infrequent episodes of short duration are difficult to document (Kantoch, 2005). SVT is usually paroxysmal and characterized by an abrupt and unpredictable onset and termination of palpitations, making it unlikely that an episode will happen to oc-

cur during a routine office visit (Perry, 1997). Because of its precipitous cessation, the arrhythmia may already be absent upon arrival if a patient goes to the clinic after the onset of symptoms. At that point, an ECG during normal sinus rhythm may be unremarkable, especially if the mechanism is AVNRT (Wiest & Uber, 2005). Pediatric primary care clinicians often need to rely on symptom report to recognize SVT. Reports of symptoms may be difficult to elicit and can be unreliable depending on the age and developmental status of a patient. Further complicating the evaluation is the similarity of some SVT symptoms to stress, anxiety, panic disorders, and other more common conditions with which the primary care provider may be familiar (Lessmeier et al., 1997). Although rare in the school-aged child or adolescent, prolonged and/or untreated episodes of SVT may lead to cardiomyopathy with or without congestive heart failure (CHF) (Boramanand & Perry, 2005). Be-

cause of its prevalence, potentially difficult diagnosis, and rare but serious complications, SVT is an important condition for pediatric primary care clinicians to recognize and manage, and patients with SVT should be referred to a cardiologist as early as possible.

PATHOPHYSIOLOGY

Types of SVT

Accessory pathway-mediated re-entry tachycardia. Also known as atrioventricular reciprocating tachycardia (AVRT), accessory pathway-mediated re-entry tachycardia is the cause of approximately 75% of cases of pediatric SVT (Hanisch, 2001). AVRT is characterized by the presence of one or more accessory conduction pathways that are anatomically separate from the normal cardiac conduction system (Doniger & Sharieff, 2006). The accessory pathway (AP) is a myocardial connection capable of conduction between the atria and ventricles at a point other than the AV node (Perry, 1997). Conduction travels down either the AV node or the AP and up the other, forming a re-entrant circuit. In other words, an unconventional conduction loop is formed (Figure 1, D). Most often, AVRT is orthodromic, meaning the electrical conduction travels antegrade down the AV node and retrograde up the AP (Hanisch). These pathways may be overt or concealed (Perry). Evidence of overt pathways can be seen on 12-lead ECGs, whereas concealed pathways cannot be identified on the ECG (Hanisch).

Wolff-Parkinson-White (WPW) syndrome is a common type of orthodromic AVRT that is usually overt and, therefore, identifiable on an ECG (Hanisch, 2001). In WPW syndrome, the AP is known as either the Kent bundle or a bypass tract and it may activate the ventricle, leading to partial or total ventricular depolarization (Goldberger, 2006). During SVT, a conduction loop is formed between the Kent

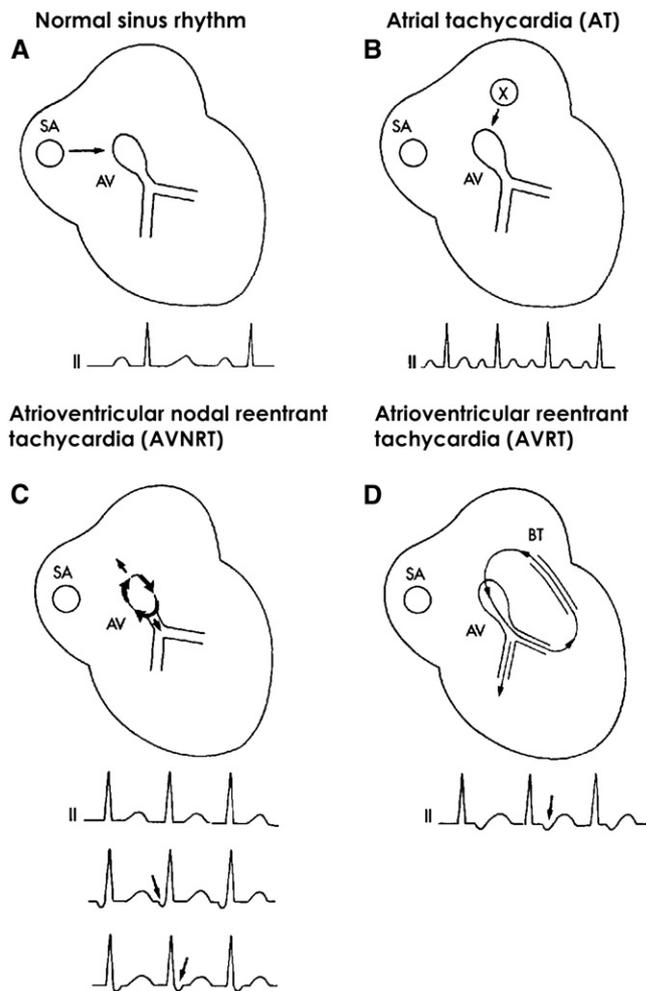
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prevalence (Kantoch, 2005). Pediatric patients also will vary in clinical presentation depending on age, mechanism of SVT, and duration and rate of the tachyarrhythmia (Perry, 1997). Infants and neurologically impaired children may sustain SVT for several days before it is symptomatically apparent, whereas children and adolescents experiencing SVT are better able to alert their caregivers to their discomfort and seek prompt medical attention (Grossman).

Although some mechanisms of SVT are associated with congenital heart disease, most children with SVT have structurally normal hearts (Green, Kitchen, & Ray, 2005). Mortality resulting from SVT is relatively low, with estimates at about 1% in patients with associated structural congenital heart defects and 0.25% in those without other cardiac abnormalities (Vos et al., 2003). Pediatric patients who are minimally symptomatic are generally easy to manage and often do not require any medical treatment (Kantoch, 2005). However, patients

cur during a routine office visit (Perry, 1997). Because of its precipitous cessation, the arrhythmia may already be absent upon arrival if a patient goes to the clinic after the onset of symptoms. At that point, an ECG during normal sinus rhythm may be unremarkable, especially if the mechanism is AVNRT (Wiest & Uber, 2005). Pediatric primary care clinicians often need to rely on symptom report to recognize SVT. Reports of symptoms may be difficult to elicit and can be unreliable depending on the age and developmental status of a patient. Further complicating the evaluation is the similarity of some SVT symptoms to stress, anxiety, panic disorders, and other more common conditions with which the primary care provider may be familiar (Lessmeier et al., 1997). Although rare in the school-aged child or adolescent, prolonged and/or untreated episodes of SVT may lead to cardiomyopathy with or without congestive heart failure (CHF) (Boramanand & Perry, 2005). Be-

FIGURE 1. Major causes of supraventricular tachycardia and their conduction pathways. (Adapted with permission from Goldberger, 2006.)



bundle and the AV node (Figure 1, D). In a small percentage of patients with WPW syndrome, atrial fibrillation may occur. Given the antegrade conduction properties of the AP, rapid conduction of atrial fibrillation over the AP to the ventricles can result in ventricular fibrillation, which can lead to syncope and sudden death if the arrhythmia does not spontaneously terminate. Although the incidence is low, this well-known association with sudden cardiac death makes prompt diagnosis of WPW syndrome essential (Chun & Van Hare, 2004). The incidence of WPW syndrome is increased in patients with Ebstein's anomaly, tricuspid atresia, double-outlet right ventricle, and hypertrophic cardiomyopathy (Hanisch).

A small portion of patients experience SVT caused by familial WPW syndrome. This type of WPW syndrome has a pattern of autosomal dominant inheritance and has been isolated in several

AV block occur in addition to pre-excitation. The AV block, which usually occurs in the fourth decade of life, often means the patient will require permanent pacemaker implantation (Sidhu & Roberts, 2003).

AVRT also may be due to concealed APs that are only orthodromic (Perry, 1997). Permanent junctional reciprocating tachycardia (PJRT) is a specific type of concealed AP AVRT. PJRT is a rare but important mechanism of SVT that is generally without the spontaneous resolution characteristics of most other types of AVRT and, therefore, is almost always incessant (Vaksmann et al., 2005). In fact, PJRT is the most common form of incessant SVT in children, and the tachyarrhythmia is present for more than 75% of the day in most affected patients. In contrast to WPW, the AP found in PJRT has slow conduction properties (Perry). The slow conduction properties and incessant nature of PJRT can result in a dilated cardiomyopathy in patients who remain undiagnosed. Therefore, a child who consistently presents with tachycardia at rates that are even slightly higher than normal for age warrants a more in-depth cardiac evaluation.

Atrioventricular nodal re-entry tachycardia. Atrioventricular nodal re-entry tachycardia (AVNRT) is the second most common mechanism of SVT in children, accounting for about 15% of cases of pediatric SVT (Hanisch, 2001).

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families. Its clinical course varies from typical WPW syndrome because cardiac hypertrophy secondary to abnormal cell growth and conduction abnormalities such as

The AV node is comprised of "slow" pathway and "fast" pathway inputs. AVNRT occurs when there is antegrade conduction block over one pathway (usually the fast

pathway), resulting in conduction over the other (usually the slow pathway). By the time antegrade conduction over the slow pathway (as seen in *typical* AVNRT) has occurred, the fast pathway has recovered and conduction is able to proceed *retrograde* over the fast pathway. This situation results in a re-entrant SVT circuit (Figure 1, C). AVNRT often is triggered when a premature impulse, such as a premature atrial or ventricular contraction, occurs during the refractory phase of the fast pathway, where it thus encounters unidirectional block and initiates a reentrant circuit (Van Der Merwe & Van Der Merwe, 2004). Atypical AVNRT is characterized by other variations of AV node conduction in which, for example, the electrical impulse travels antegrade down the fast pathway and retrograde up the slow pathway (Scheinman & Yang, 2005). On rare occasions, atypical AVNRT may be incessant, as with a *slow-slow* variant (Perry, 1997).

Automatic tachycardia. Automatic tachycardias are caused by an abnormal electrical impulse that originates outside of the sinus node but still within the cardiac area (Figure 1, B) (Hanisch, 2001; Perry, 1997). Atrial ectopic tachycardia (AET), an automatic tachycardia, is rare overall but is one of the most common mechanisms of pediatric incessant SVTs and can result in a dilated cardiomyopathy if it remains undetected. The exact mechanism of AET is unknown, but it appears to be due to abnormal automaticity, perhaps caused by remnant embryonic cells with automatic qualities (Perry).

With the exception of familial WPW syndrome, the mechanisms of SVT discussed in this article are not generally considered to be genetic conditions. Some anecdotal evidence exists regarding heritability of other causes of SVT, but clear evidence to confirm or refute this claim does not exist. The substrate for the causes of SVT discussed in

this article is believed to be present at birth, although symptoms may arise at any point in the life span.

Age-related Incidence

Infants and toddlers. It is estimated that 50% to 60% of cases of pediatric SVT present in the first year of life (Perry, 1997). Accessory pathway-mediated re-entry tachycardias are the most common mechanism of SVT in infants and children (Sanatani, Hamilton, & Gross, 2002). This mechanism accounts for as many as 90% of cases of SVT in infants (Kugler & Danford, 1996). AET is another cause of SVT that is more common in young children (Chun & Van Hare, 2004). However, the incidence of AET in the pediatric population remains very small (Vos et al., 2003). AVNRT is almost completely absent in infants (Hanisch, 2001). There is a gradual increase in the frequency of AVNRT in patients older than 1 year of age (Perry). It is important to note that approximately 15% of cases of SVT in infants are associated with other conditions such as heart disease, drug administration, or fever (Kantoch, 2005).

SVT will resolve spontaneously in many infants by the age of 6 to 12 months (Hanisch, 2001). However, up to 33% of those patients will experience a recurrence at a mean of 8 years of age (Perry, 1997). SVT presentation when the child is older than 1 year of age is a predictor of recurrence (Sanatani et al., 2002).

School-aged children and adolescents. Accessory pathway-

mediated re-entry tachycardia remains the most common mechanism of SVT in the younger adolescent population (Brembilla-Perrot, Houriez et al., 2001). However, the incidence of AVNRT as the primary mechanism of SVT increases with age (Doniger & Sharieff, 2006). As many as one third of cases of SVT in adolescents are caused by AVNRT (Kugler & Danford, 1996). The incidence and frequency of different mechanisms of SVT is similar to those of adults by the late teenage years (Perry, 1997). Therefore, by late adolescence, AVNRT is the most frequent cause of SVT (Brembilla-Perrot, Houriez et al.).

Presentation

The presentation of SVT is related to the age of the patient and the rate, duration, and mechanism of the tachyarrhythmia (Box 1). The presence of underlying heart disease also may be a factor (Grossman, 1997). There are little data specific to the presentation of SVT in children aged 1 to 6. This could be related, in part, to the natural history of "remission" in this age group (Perry, 1997).

Infants and toddlers. The heart rate in infants with SVT frequently ranges from 220 to 280 BPM (Doniger & Sharieff, 2006). The heart rate in children older than 1 year generally is slower, ranging from 180 to 240 BPM (Perry, 1997). Signs and symptoms of SVT in infants include poor feeding, vomiting, diaphoresis, increased sleeping, and irritability. These indicators of SVT in infants and toddlers often are difficult to recognize, are nonspecific, and may mimic the symptoms of other common childhood illnesses (Kantoch,

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2005). In a study by Vos, Pulles-Heintzberger, and Delhaas (2003), 74% of parents or nurses of infants with SVT did not notice or report any signs at all.

BOX 1. Signs and symptoms of pediatric supraventricular tachycardia

Infants

- Poor feeding, vomiting, irritability, increased sleepiness, syncope, and diaphoresis
- Pallor, cough, respiratory distress, and cyanosis if congestive heart failure is present

Toddlers and school-aged children

- Palpitations, chest pain, dizziness, shortness of breath, and syncope

Adolescents

- Palpitations, chest pain, dizziness, shortness of breath, pallor, diaphoresis, decline in exercise tolerance, fatigue, anxiety, and syncope

Data from Boramanand & Perry, 2005; Doniger & Sharieff, 2006; Grossman, 1997; Hanisch, 2001; Kantoch, 2005; Perry, 1997.

Because infants cannot report their symptoms, their tachycardia may not become evident until they have experienced prolonged periods of SVT. This hemodynamic decompensation results from decreased ventricular diastolic filling time, stroke volume, coronary artery perfusion, and cardiac output (Grossman, 1997). CHF can occur after 24 to 48 hours of untreated SVT. CHF may occur sooner in the presence of underlying congenital heart disease (Hanisch, 2001). Prolonged heart rates greater than 280 BPM also are associated with the occurrence of CHF (Perry, 1997). Pallor, cough, respiratory distress, and cyanosis may be noted if the infant is experiencing CHF (Doniger & Sharieff, 2006). These signs are directly related to the duration of SVT in addition to the extent of CHF present (Perry). Once CHF develops, the infant's condition may deteriorate rapidly (Green et al., 2005).

School-aged children. The presentation of SVT in school-aged children is significantly different than that of infants and toddlers. This difference in presentation is perhaps the result of an increased ability to report symptoms and is evidenced by the near absence of CHF as a complication of SVT in any age group other than infants (Perry, 1997). Symptoms include palpitations, chest pain, dizziness, or shortness of breath (Doniger & Sharieff, 2006). Incessant forms of slow SVT, such as PJRT and AET, may present as a gradual decrease in exercise tolerance, easy fatigability, and shortness of breath. These symptoms in an otherwise previously healthy child with a persistently elevated heart rate may represent a tachycardia-induced cardiomyopathy and should prompt immediate cardiac evaluation.

Adolescents. Symptom report is an important diagnostic tool in determining the presence of true SVT, and the majority of adolescents are capable of providing very accurate descriptions of their symptoms (Kantoch, 2005). Adolescents may experience the same symptoms as school-aged children, such as palpitations, chest pain, dizziness, and shortness of breath. They also may exhibit pallor, presyncope, diaphoresis, and clammy skin, which are signs of SVT in adults. Be-

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cause the incidence of AVNRT increases in this population, it is perhaps helpful to note that anec-

BOX 2. Differential diagnosis of supraventricular tachycardia

Cardiac

- Congenital heart disease and myocarditis

Diet or medication

- Caffeine consumption, over-the-counter medications, prescription medications, and illicit drugs

Psychological

- Stress, anxiety attack or disorder, and panic attack or disorder

Other

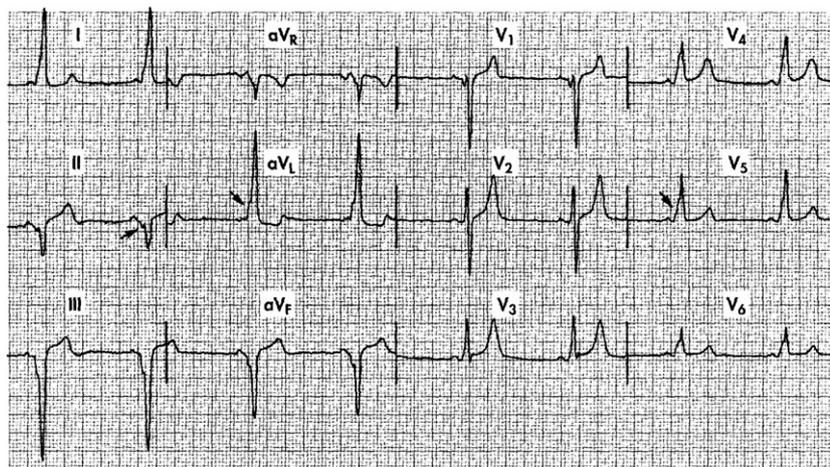
- Hyperthyroidism, electrolyte abnormalities, febrile illness, and dehydration

Data from Brembilla-Perrot & Marcon et al., 2001; Delacrétaz, 2006; Doniger & Sharieff, 2006; Grossman, 1997.

dotal evidence suggests that a feeling of palpitations in the neck region is associated with this mechanism of SVT (Perry, 1997). Heart rates tend to be the same or slightly slower than those in school-aged children experiencing SVT, with a range that can go as low as 150 BPM (Hanisch, 2001). Syncope is a rare but potential symptom of SVT and may represent a life-threatening arrhythmia

in patients with WPW or underlying congenital heart disease (Kantoch).

FIGURE 2. Electrocardiogram features of Wolf-Parkinson-White (WPW) syndrome. The classic WPW triad of a short PR interval (0.08 seconds), wide QRS complex (0.12 seconds), and delta wave (indicated by arrows) are visible. (Adapted with permission from Goldberger, 2006.)



DIAGNOSIS

Differential Diagnosis

Many conditions should be considered in the differential diagnosis of the cause of SVT (Box 2). These conditions include underlying congenital heart disease; stress, anxiety, and panic disorders; hyperthyroidism; electrolyte abnormalities; febrile illness; dehydration; use of over-the-counter, prescription, and illicit drugs; caffeine consumption; and myocarditis (Brembilla-Perrot, Marcon et al., 2001; Delacrétaz, 2006; Doniger & Sharieff, 2006; Grossman, 1997). This wide range of potential causes emphasizes the importance of a thorough history when evaluating a patient experiencing SVT.

ECG Features

A 12-lead ECG should be performed, when possible, on any patient who presents with symptoms or complaints of previous symptomatic episodes consistent with SVT. It is a relatively inexpensive, noninvasive test that may help to identify or rule out potential causes of SVT.

Accessory pathway-mediated re-entry tachycardia. The ECG is diagnostic in patients with WPW syndrome and is, in fact, the

preferred diagnostic method. However, this diagnosis may be difficult to make in young patients with very high heart rates and/or APs on the left side of the heart (Perry, 1997). The baseline ECG in normal sinus rhythm is characterized by a shortened PR interval and a delta wave, which is a slurred initial portion of the QRS complex indicating pre-excitation (Figure 2) (Hanisch, 2001). During orthodromic SVT, this slurring disappears, and there is a narrow complex QRS tachycardia, usually without visible P waves. The RP interval is shorter than the PR interval. In *antidromic* SVT, where conduction occurs antegrade over the AP and retrograde over the AV node, pre-excitation will be manifest and a *wide* complex tachycardia will appear. This presentation often can be mistaken for ventricular tachycardia. In a primary care office setting, a wide complex tachycardia should always be considered a medical emergency. The ECG in SVT for patients with a concealed AP is similar to that of orthodromic SVT in a WPW patient: there is a narrow QRS tachycardia with difficult to discern P waves and an RP interval that is shorter than the PR interval.

PJRT and other concealed accessory pathway-mediated mechanisms of SVT do not have evidence of pre-excitation on the baseline ECG in normal sinus rhythm. In tachycardia, the ventricular rate of PJRT is 140 to 250 BPM but is usually toward the slower end of this spectrum. The RP interval is usually equal to or longer than the PR interval, with deeply negative P waves in leads II, III, and aVF present (Perry, 1997), indicating the posterior septal location of this concealed AP.

AVNRT. Heart rates during SVT tend to be slower than in accessory pathway-mediated SVT, with the exception of PJRT (Perry, 1997). During tachycardia at faster heart rates, P waves are buried in the QRS complex but are usually identifiable (Figure 3) (Hanisch, 2001). An initiating event, such as a premature atrial contraction, may be seen on the ECG recording (Perry). The baseline 12-lead ECG during normal sinus rhythm is not always useful in identifying patients with AVNRT because it is usually unremarkable (Wiest & Uber, 2005). However, patients with a borderline first-degree AV block (prolonged PR interval) during normal sinus rhythm may raise the index of suspicion for dual AV node physiology and may be at higher risk for clinical AVNRT.

AET. The hallmarks of AET on the ECG during SVT are abnormal rate and P wave shape that is different than normal sinus P waves (Figure 4) (Perry, 1997). The heart rate often is age-dependent and ranges from 90 to 330 BPM (Hanisch, 2001). AET is usually incessant, or occurs in frequent salvos. A baseline ECG and rhythm strip may be helpful in identifying this arrhythmia in the infant or young child with symptoms such as fussiness, irritability, lethargy, and poor feeding. Early diagnosis of AET is critical, because it is not uncommon for a tachycardia-induced cardiomyopathy to develop in these patients if they are not treated.

FIGURE 3. Electrocardiogram features of supraventricular tachycardia caused by atrioventricular nodal re-entry tachycardia. Note that the P waves are buried in the QRS complex. (Adapted with permission from Goldberger, 2006.)

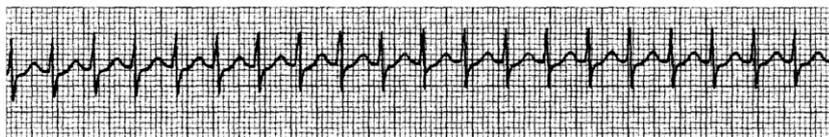
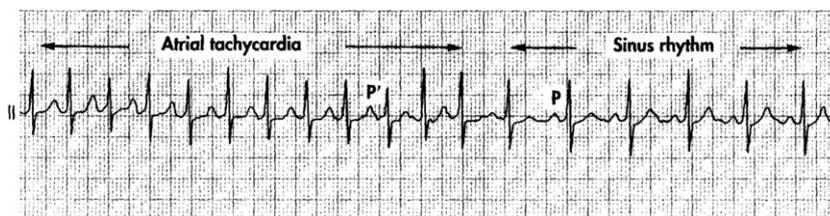


FIGURE 4. Electrocardiogram features of supraventricular tachycardia caused by atrial tachycardia and spontaneous conversion to normal sinus rhythm. Note the P wave shape (P') that differs from normal sinus P waves. (Adapted with permission from Goldberger, 2006.)



Ambulatory ECG Monitoring Devices

Ambulatory ECG monitors are portable devices that are worn by the patient during routine activities to capture ECG recordings of symptoms that may or may not represent true arrhythmia. These devices are either continuous recorders, also known as Holter monitors, or intermittent event recorders (Funk, Chrostowski, Richards, & Serling, 2005).

Holter monitor. A Holter monitor is a portable device that records a continuous ECG tracing. It is attached to the patient with ECG wires and electrodes and typically is worn by the patient for 24 to 48 hours to provide information about symptoms that are likely to occur during that short period (Funk et al., 2005). The most commonly used Holter monitor uses only two or three leads. Therefore, it is fairly efficient in arrhythmia identification but does not provide as much information on the location of arrhythmogenic focus as does a 12-lead ECG. However, 12-lead Holter monitors are now available and are beginning to be used in

some settings (Emmel, Sreeram, Schickendantz, & Brockmeier, 2005). Obviously, a Holter monitor is not a good diagnostic or monitoring option for a patient who experiences infrequent episodes of SVT (Kantoch, 2005). However, it is a useful tool for frequently occurring SVT and incessant mechanisms like PJRT because they are present the majority of the day (Perry, 1997).

Intermittent recorders. Transtelephonic electrocardiographic event monitoring is another helpful diagnostic tool for the evaluation of patients with suspected arrhythmias. Intermittent recorders, also known as ambulatory ECG monitoring devices, may be used for longer periods (weeks or months) than Holter monitors. They are activated by the patient, so are only practical if the child is old enough and developmentally able to understand the directions, recognize symptoms, and activate the recorder himself or herself. The recorded rhythms are transmitted over a telephone to a receiving center (Funk et al., 2005).

Two types of intermittent recorders exist: presymptom memory loop recorders and postsymptom

recorders. Presymptom memory loop recorders are always attached to the patient with ECG wires and electrodes. The ECG is constantly being recorded and erased in a continuous loop. When a patient experiences symptoms, he or she presses a button that will cause the device to capture a recording of the rhythm. The device can be set to record the rhythm from a pre-specified amount of time before the patient presses the button. This feature is important in identifying some SVTs where the onset of the SVT can be diagnostic, such as premature atrial contractions at the onset of AVNRT. Some models have an autotrigger feature that automatically records for a period when the device senses a heart rate outside of the programmed normal limits (Funk et al., 2005).

Another type of intermittent recorder is the postsymptom recorder. This device is carried by the patient but not attached via electrodes or continuously recording. Instead, the patient places the device on his or her chest when he/she experiences symptoms, presses the "record" button, and an ECG is recorded. Although this device does not have the advantage of capturing a presymptom period, it is often selected because it is smaller and does not have to be worn continuously (Funk et al., 2005).

Based on their study of 495 children and adolescents with suspected arrhythmias, Saarel et al. (2004) recommended continuous loop recording for patients who experienced episodes lasting less than 20 seconds, had one to two episodes per week, and/or experienced syncope. Otherwise, the authors suggested that the most useful and effective strategy for detecting SVT in pediatric patients is the use of 4-week, postevent transtelephonic monitoring. In general, these monitoring devices will be prescribed and interpreted by a pediatric cardiologist, so prompt referral of the patient with

suspected arrhythmia is crucial to appropriate care and management.

Electrophysiology Study

Electrophysiology study (EPS) is the most definitive diagnostic procedure for the classification of different mechanisms of SVT (Gross, Epstein, Walsh, & Saul, 1998). EPS is usually performed under conscious sedation or general anesthesia, depending on the age of the child. Under fluoroscopic guidance, electrocatheters are inserted through peripheral vessels, most often the femoral veins, and advanced into the heart. Intracardiac signals are amplified, recorded, and analyzed to map the electrophysiologic characteristics of the heart and determine the site and mechanism of the arrhythmia (Bottoni et al., 2003). Ablation, a curative measure for many mechanisms of SVT, generally is performed in conjunction with EPS (Kantoch, 2005) and will be discussed in the section on management. Another means of EPS access is transesophageal, but this method generally is limited to diagnostic procedures in infants with suspected or known SVT (Etheridge & Judd, 1999) and for risk-stratification in patients with WPW.

Other Testing

When symptoms suggestive of tachycardia occur, additional tests may be performed to rule out other conditions included in the differential diagnosis. Laboratory tests include a complete blood cell count (CBC) with differential to rule out infectious processes or hematologic abnormalities such as anemia, electrolyte values to identify imbalances that may lead to cardiac manifestations, toxicology screen, arterial blood gas, thyroid function tests, and a chest x-ray with lateral and anteroposterior views to rule out cardiomyopathy and CHF (Doniger & Sharieff, 2006). Screening for anxiety and panic disorders also should be performed in appropriate age groups,

with care not to misdiagnose cardiac symptoms as psychological or vice versa.

MANAGEMENT

Immediate

Nonpharmacologic Therapies

Immediate therapies are episodic and are designed for symptom relief, rather than addressing the actual substrate of SVT. Physical maneuvers that enhance vagal activity may be successful in terminating the tachyarrhythmia during episodes of SVT by increasing the refractoriness of the AV node (Box 3) (Wen et al., 1999). In infants and younger children who cannot be taught specific techniques, the vagal-dive reflex maneuver consisting of an ice-cold wet cloth or ice bag placed on the patient's face may be attempted (Kugler & Danford, 1996). This technique decreases the risk of aspiration associated with complete facial submersion in ice water, a technique that is used with decreasing frequency. The most effective of these maneuvers in older children and adolescents is the Valsalva maneuver, which involves bearing down as if having a bowel movement (Wen et al.). Other techniques include the Muller's maneuver, which is deep inspiration against a closed glottis in which the nose is pinched and inspiration is attempted with the mouth closed after a forced expiration; gagging; forceful coughing; pulling of the tongue; or stimulation of the gag reflex (Bosen, 2002). Retching, coughing, vomiting, physically induced gagging, a handstand or headstand held for 30 seconds, breath holding, deep breathing, and bending over to touch the floor also may be effective (Grossman, 1997). Historically, carotid sinus massage was performed as an alternative means of stimulating vagal activity. However, it has been proven ineffective in several studies, is less effective than other vagal maneuvers overall, and is unsafe if not performed properly by a trained

BOX 3. Maneuvers to enhance vagal activity

- Ice-cold wet cloth or ice bag on the face
- Bearing down as if having a bowel movement
- Deep inspiration with nose pinched and mouth closed after expiration
- Forceful coughing
- Pulling of the tongue
- Gagging, retching, or vomiting via physical stimulation of the gag reflex
- Breath holding
- Deep breathing
- Handstand or headstand held for 30 seconds
- Bending over to touch the floor

Data from Bosen, 2002; Grossman, 1997; Kugler & Danford, 1996; Wen et al., 1999.

and highly skilled care provider. It should never be performed in a primary care setting (Wen et al.). Maneuvers that enhance vagal activity are effective at terminating episodes of SVT as much as 53% of the time (Wen et al.).

Electrical cardioversion should be performed when life-threatening conditions, such as loss of consciousness, shock, or CHF, arise as a result of prolonged and/or especially high rates of SVT (Kugler & Danford, 1996). In rare cases of extreme cardiovascular compromise and decompensation, extracorporeal life support may be used to stabilize the patient and sustain life and has been used successfully during EPS and ablation (Walker et al., 2003).

Medical Therapy

Intravenous adenosine is the first-line pharmacologic measure for termination of SVT in infants and children in the emergency setting (Dixon, Foster, Wyllie, & Wren, 2005). Calcium channel blockers also may be used in the acute setting to terminate SVT

(Chun & Van Hare, 2004). Importantly, the use of calcium channel blockers is *contraindicated* in children younger than 1 year because of their hemodynamic dependence on heart rate and inability to augment stroke volume. Administration of a calcium channel blocker in this age group may lead to rapid hemodynamic compromise and death (American Heart Association, 2005).

Traditional first-line medications for chronic management of SVT include digoxin and/or β -blockers such as atenolol. In the event of failure of these agents, the second line of pharmacologic therapy usually consists of propranolol, amiodarone, flecainide, or sotalol (Wong, Potts, Etheridge, & Sanatani, 2006). It is important to note that digoxin is *contraindicated* in patients with WPW because it enhances conduction properties of the AP and predisposes the patient to rapid conduction of atrial fibrillation and sudden death (Kugler & Danford, 1996). Flecainide has also been shown to increase sudden death in patients with underlying congenital heart disease and, therefore, should be used with caution in this population (Price, Kertesz, Snyder, Friedman, & Fenrich, 2002).

In cases of refractory SVT, a combination of flecainide and sotalol has been shown to be successful in managing the tachyarrhythmia (Price et al., 2002). Other combinations of medications may also be attempted (Wong et al., 2006) for refractory arrhythmias in young infants in whom EPS and ablation are high risk.

Most infants with SVT will not require medication after 9 months of age (Etheridge & Judd, 1999). Patients in this age group are often allowed to "outgrow" their medication dose and are monitored closely with Holter monitors. As previously stated, some of these patients will have recurrence of arrhythmia in later childhood, so parents and providers should have a heightened awareness should the child present

with complaints of palpitations. Pharmacologic therapy achieves successful control in approximately 68% of children (Chun & Van Hare, 2004). However, if pharmacologic therapy is ineffective or is not acceptable to the family, ablation can be performed (Price et al., 2002).

EPS and Ablation

EPS generally is performed under the assumption that ablation will immediately follow if appropriate. This means that the mechanism of SVT is isolated, its location and electrophysiologic properties are deemed acceptable for ablation, and the patient is physiologically capable of undergoing the procedure (Kantoch, 2005). Because of the increasing understanding of ablation and experience level of providers, ablation is often the first line of therapy for many children with SVT, has relatively low morbidity and mortality, and it results in a low rate of recurrence of SVT (Chun & Van Hare, 2004).

The traditional method of ablation uses radiofrequency energy to heat and obliterate the site of origin of the SVT mechanism (Kantoch, 2005). However, a newer procedure is cryoablation, in which liquid nitrous oxide is used to cool the catheter to sub-freezing temperatures. This catheter is targeted at the area of interest and destroys the tissue by freezing. The main advantage of cryoablation is that it allows reversible cooling so that the electrophysiologist can test the area first for accuracy of location and risk of leading to heart block before freezing the tissue to an even cooler level, at which point a permanent lesion will form (Chun & Van Hare, 2004).

Ablation is successful during the acute phase approximately 95% of the time (Kugler, Danford, Houston, & Felix, 2002). However, in a retrospective study of children undergoing radiofrequency ablation between 1989 and 1991, Collins, Chiesa, Dubin, and Van Hare (2002) reported the recurrence

rate of SVT in children with structurally normal hearts having undergone ablation 10 or more years earlier to be 38%. The Pediatric Radiofrequency Ablation Registry, which contains data on 4135 patients who underwent 4651 procedures in 46 centers between 1991 and 1996, provided additional insight on complications and mortality. Using data from this registry, Van Hare et al. (2004) reported that permanent second-degree or third-degree heart block occurred in approximately 1% to 3% of pediatric patients undergoing ablation. Schaffer, Gow, Moak, and Saul (2000) reported that mortality caused by radiofrequency ablation occurred approximately 0.12% of the time in patients with structurally normal hearts and 0.89% of the time in patients with structural heart defects. Mortality can be due to ventricular arrhythmia, coronary or cerebellar thromboembolism, traumatic injury, myocardial perforation, or hemopericardium. The risk of mortality from radiofrequency ablation increases with underlying heart disease, lower patient weight, left-sided procedures, and increased number of radiofrequency energy applications (Schaffer et al.).

Much of the available data on complications and mortality represent the early experience of ablation. Some of these centers performed only a few ablations annually. Ablations in low-volume centers are likely to result in higher rates of complications and recurrence of SVT. Overall knowledge of and experience with pediatric ablation has increased dramatically since 1996. The actual rates of recurrence and nonlethal complications, especially in high-volume centers, seem to be considerably lower now, although confirmation from more recent registry data has not been published (Chun & Van Hare, 2004).

In infants, pharmacologic therapy usually is preferred to ablation because the risk of morbidity and

mortality related to the ablation procedure is increased in this population (Price et al., 2002). Pharmacologic therapy also is preferred because of the lack of data regarding long-term effects of ablation at a young age when cardiac structures may be undeveloped and the fact that many infants will experience either complete absence of or an extended remission of SVT by 1 year of age, thus eliminating the need to risk an invasive procedure (Perry, 1997).

Implications for the Primary Care Setting

It is important that all pediatric primary care providers understand thoroughly which signs and symptoms warrant a baseline primary care cardiac work-up and which findings should lead to a referral to a cardiology specialist.

There is much debate as to what warrants a cardiology referral. An adolescent's report of associated symptoms alone may be enough for a practitioner to suspect SVT. In younger children, the primary care provider may not see the child until hemodynamic instability occurs, and a referral would more likely be to the emergency department for acute management and subsequent cardiology referral. If SVT is found incidentally on physical examination or after a child experiencing related symptoms is brought to the primary care office, the patient requires further evaluation. If the patient is not believed to be hemodynamically compromised and the hospital is very close, a parent may bring the child straight to the emergency department. Otherwise, an ambulance should be called. The child should be observed to ensure maintenance of airway, breathing, and circulation. While waiting for the ambulance to arrive, the primary care provider can obtain a 12-lead ECG if the equipment is available and instruct the patient to perform various vagal maneuvers in an attempt to terminate the arrhythmia.

Parental distress regarding a potential cardiac diagnosis is likely and must be taken into account by the primary care provider when providing that information (Green et al., 2005). When SVT related to one of the common mechanisms discussed in this article is suspected, the low rate of mortality and high rate of effective treatment or cure should be stressed. Prompt referral is important, however, because it is not possible for a primary care provider to determine the mechanism responsible for the arrhythmia and whether it is potentially fatal.

Once the patient has been diagnosed with a cardiac condition that leads to SVT, it is important for the primary care provider to become informed of the pathophysiology of the condition and how it and its treatment affect routine health care management. Although no primary care provider can be expected to be an expert on every medical condition affecting the population in which he or she practices, a basic working knowledge will be helpful not only for medical management but also in the event that patients and their families wish to involve the primary care provider in their discussions regarding treatment choices.

Because the primary care provider is expected to maintain all records and have an understanding of the patient's overall health status, an increased awareness and understanding of conditions typically managed by specialists is valuable. The role of pediatric primary care providers in the management of cardiac conditions often is underestimated. However, a small amount of additional knowledge transforms them into valuable contributors to the outcomes of pediatric patients who experience SVT, as was true for 8-year-old Nick.

CASE CONCLUSION

Upon arrival at the emergency department, Nick had blood

drawn for a CBC with differential and electrolytes and had an ECG and a chest x-ray done. Nick's resting ECG showed a narrow QRS-complex tachycardia at a rate of 176 BPM. His chest x-ray demonstrated cardiomegaly and mild pulmonary edema. Given these findings, an echocardiogram was performed that showed severe global right and left ventricular systolic dysfunction with a left ventricular ejection fraction of 14% and severe mitral regurgitation. The arrhythmia was diagnosed as AET, and he was admitted to the pediatric intensive care unit for management of tachycardia-induced cardiomyopathy.

In the intensive care unit, dopamine, milrinone, digoxin, and Lasix were administered. For arrhythmia control, Nick received intravenous procainamide and amiodarone. Once adequate rhythm control was achieved, he was weaned from intravenous medication and was discharged home with procainamide and amiodarone for antiarrhythmic therapy and with an anti-congestive regimen consisting of captopril, furosemide, carvedilol, and aldactone. After a normal rhythm was achieved and his ventricular function was recovered, Nick was brought to the cardiac catheterization laboratory to undergo EPS and radiofrequency ablation. The electrophysiologist found an abnormal atrial ectopic focus in the right atrial appendage, confirming the diagnosis of AET, and successfully ablated it. Following resolution of the arrhythmia, Nick's echocardiogram demonstrated a normal ejection fraction with trace physiologic mitral regurgitation. He returned to his hockey team and one year later was voted the league's Most Valuable Player.

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