Arrhythmia Identification: Stabilization and Treatment

8

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Introduction

Of the over 25 million yearly pediatric emergency room (ER) visits in the United States, primary cardiac-related issues in children <18 years of age are much less common than seen in adults who frequently present with acute coronary issues [1]. However, heart rate changes and arrhythmias occur not infrequently, and monitoring the rate and rhythm and obtaining an electrocardiogram (ECG) are often useful. Since children are seen in many ER settings from those in a dedicated children's hospital to primarily adult-oriented hospitals, to urgent care facilities, and to rural areas where pediatric specialty services may not be readily available, interpretation of pediatric ECGs often becomes the responsibility of physicians who may have limited experience with the age-related variances of ECG patterns among children with and without congenital heart defects (CHD [2]. In addition, with the increasing numbers of patients with repaired congenital heart defects surviving into adulthood who present to emergency rooms with various complaints, variations of their ECGs associated with their underlying anatomical/structural CHD may mimic ECG findings associated with ischemic coronary disease. In this regard, close examination of the full 15-lead ECG, rather than a single rhythm strip, is often required for proper interpretation. In addition the computer interpretation of ECGs may be erroneous

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with a reported error rate up to 75% which can result in unnecessary additional testing and costs, as well as patient distress and inappropriate referrals to cardiac specialists [3, 4].

The purpose of this chapter is to enable the reader to identify commonly seen normal and abnormal ECGs and the diverse rhythm disturbances seen in children and adults with various congenital heart defects. In addition, specific pathophysiologies of arrhythmias and appropriate therapies will be presented.

1-Normal Variants

Case 1

An 8-year-old patient presented to the emergency department with a 1-week history of chest pain localized to the left lower chest. The pain is stabbing in nature and occurs at rest with no radiation. No associated symptoms are present except for occasional palpitations. Physical examination is unremarkable except for chest wall tenderness on palpation. He was seen 2 days ago at another facility where an EKG was performed (Fig. 8.1). Repeat EKG at the time of presentation is shown in Fig. 8.3. What is the next appropriate approach?

(A) Place a 24 h Holter monitor.
(B) Order an echocardiogram.
(C) Reassurance of non-cardiac chest pain.
(D) Check electrolytes.
(E) Obtain troponin levels.

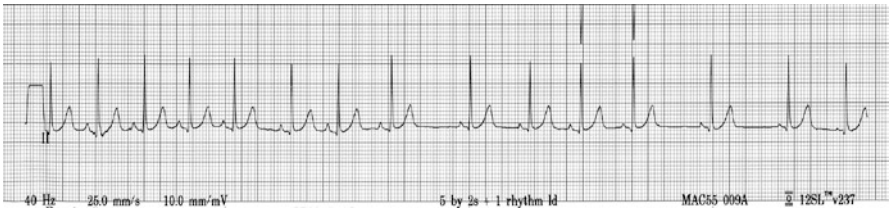


Fig. 8.1 Sinus arrhythmia. Lead II: normal P waves and PR intervals with rate changes

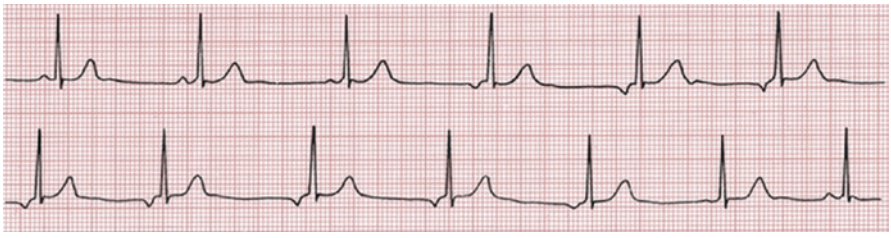


Fig. 8.2 Wandering atrial pacemaker. Lead II: P waves change from upright to inverted and back to upright. The PR interval shortens with the inverted P waves

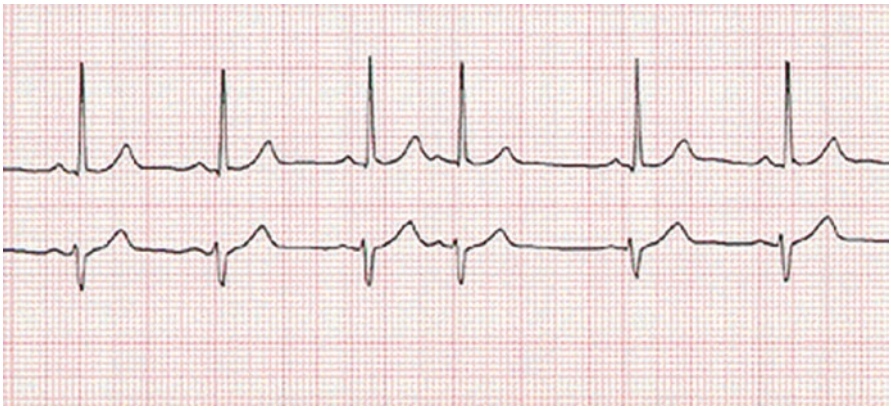


Fig. 8.3 Three sinus complexes (left) are followed by a PAC, which then conducts to the ventricle. This is followed by a compensatory pause before sinus rhythm resumes

Discussion

When interpreting ECGs, it is important to determine if any particular pattern is normal or not. Heart rate variability is normal, and often a child or teenager's complaint of sensed tachycardia or palpitations does not have a primary cardiac etiology. Sinus arrhythmia is normal and is a benign condition. The heart rate varies with respiration, and this is more pronounced in children. It is a normal physiologic finding caused by changes in parasympathetic input of the vagal tone to the heart during respiration. The ECG pattern shows normal P waves and PR intervals with rate variability (Fig. 8.1). Another common finding among children is a wandering atrial pacemaker. This benign variant is characterized by changes in P wave morphology and PR intervals and reflects atrial activation along the preferential conduction pathways (Fig. 8.2).

Premature atrial complexes (PAC) are typically benign and common especially in infants and young children. They can occur throughout childhood [5–7]. No treatment is required. Typically, the P wave morphology of the premature complex is different from sinus rhythm. It may be visible immediately after the T wave or be hidden in and distort the T wave of the preceding QRST complex. Careful examination of preceding T waves will often disclose "humps-n-bumps" distortions of atrial P waves, which may or may not conduct to the ventricles (Figs. 8.3 and 8.4). At times, the single premature atrial impulse (PAC) is conducted through the AV node before the ventricle has recovered electrically from the previous impulse and results in an abnormal QRS morphology (PAC with aberration) mimicking a premature ventricular complex (PVC) (Fig. 8.5).

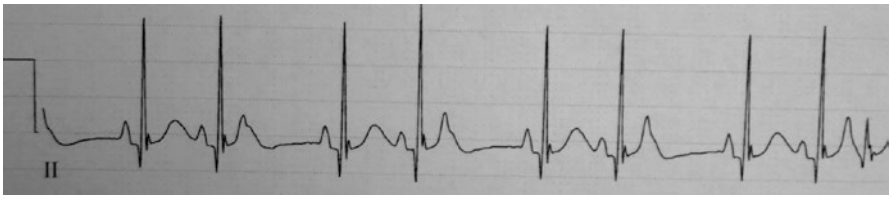


Fig. 8.4 Sinus rhythm with PACs falling on top of the T wave causing distortion. The P wave is blocked from conducting to the ventricle due to refractoriness of the AV node. The last complex conducts with aberrancy

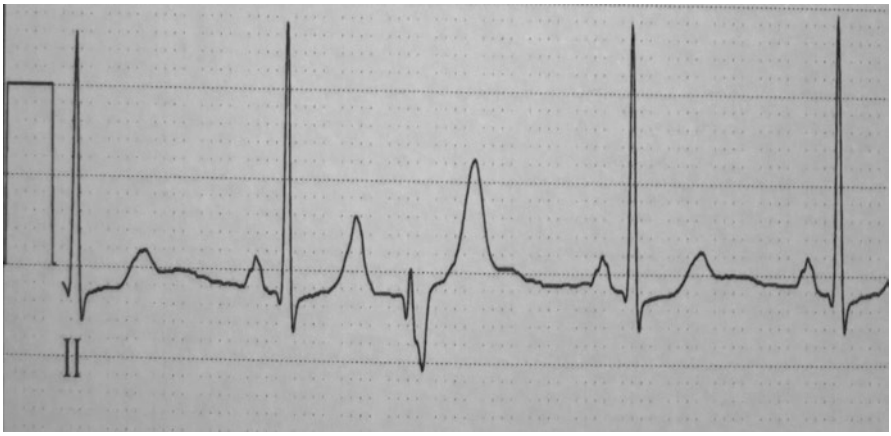


Fig. 8.5 PAC with aberration. The third QRS complex is abnormal and wide as a result of the PAC being conducted before the ventricle has completely recovered from the previous impulse. Note that the T wave of the preceding QRS is peaked and taller than the other sinus complex T waves, indicating distortion by a hidden P wave

Answer: C. The ECG findings of sinus arrhythmia and PACs are benign variants in children, and the localized chest pain that is reproducible is typical of chest wall discomfort and not cardiac in origin.

Case 2

A 10-year-old patient with a past medical history of diabetes mellitus presented to the emergency department and is found to be in diabetic ketoacidosis. His cardiovascular examination is remarkable for tachycardia. His initial lab work-up shows potassium at 6 meq/L.

What is the first EKG finding of hyperkalemia?

(A) Sinus tachycardia
(B) Prolonged QT interval
(C) U waves
(D) Peaked T waves
(E) Inverted T waves

Discussion

1. **Electrolyte Imbalance**

   It must be remembered that the cardiac cellular action potential that initiates electrical activity is electrolyte dependent. Alterations in potassium and calcium currents can have profound effects on the ECG. Thus, non-cardiac etiologies may be responsible for significant ECG changes. A tall, peaked, and symmetrical T wave is the first change seen on the EKG on a patient with hyperkalemia (Fig. 8.6). As the hyperkalemia increases, there is further slowing of conduction through the myocardium, manifested by a prolonged PR interval with eventual absence of P waves, and an increase in QRS duration. Ultimately the QRS complex widens to a severe conduction delay and becomes a "sine wave" mimicking ventricular tachycardia (Fig. 8.7). If untreated, true ventricular tachycardia and fibrillation can result.

   Hypokalemia causes ST segment depression, decrease in the amplitude of the T wave, and increase in the amplitude of the U wave (Fig. 8.8). These changes are best seen in the precordial leads V4–V6. The T and following U waves can merge resulting in the misdiagnosis of prolonged QT interval.

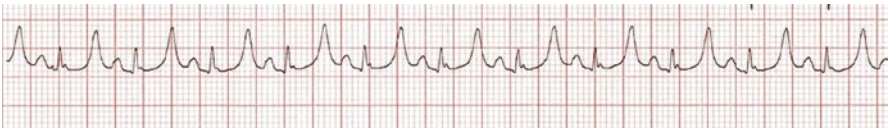


Fig. 8.6 Peaked T waves are very evident

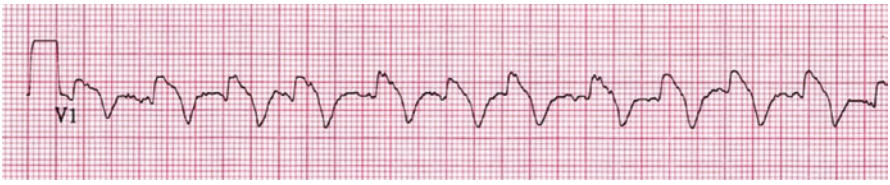


Fig. 8.7 QRS "sine wave" appearance associated with a potassium >8 meq/L

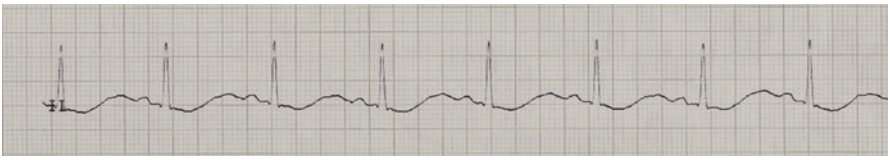


Fig. 8.8 A 5-year-old patient on oral furosemide with a 4-day history of vomiting and diarrhea. His potassium level was 2.5 meq/L. Diffuse ST-T slurring is evident

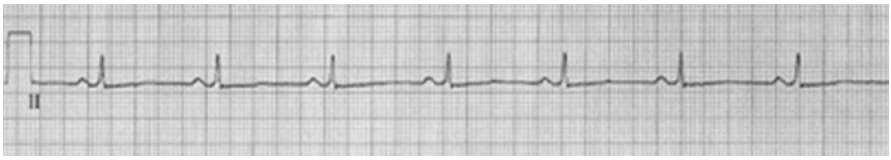


Fig. 8.9 ECG from a child with a calcium of 7.5 mg/dL. The QT interval is prolonged and the T waves are flat

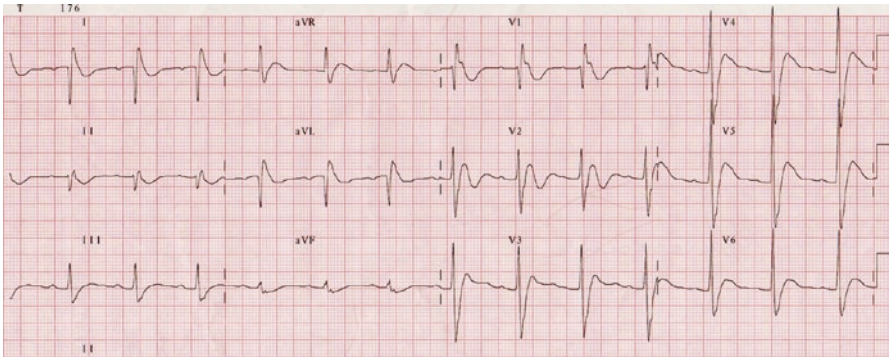


Fig. 8.10 Hypercalcemia results in shortening of the ST and the QT segments. There is rapid upstroke of the initial portion of the T wave. Another finding is the ST segment elevation in V1–V2

Calcium affects the cardiac action potential during phase 2 ("plateau phase"). This is reflected on the surface ECG by changes in the QT interval and the ST segments. Hypocalcemia can cause a lengthening of the QT interval (Fig. 8.9), while hypercalcemia will be associated with the opposite effect and a shortened QT interval (Fig. 8.10).

2. **Drug Related**

The ECG is sensitive to pharmacologic agents that interfere or alter ion transport in cardiac tissue. These agents include antimicrobials, antifungals, antidepressants, antipsychotics, and antiarrhythmics. Listing of these agents can be found elsewhere [8, 9]. Typically, the QT interval can be prolonged (Fig. 8.11) putting patients at risk for ventricular arrhythmias. The measured QT interval is age- and gender-dependent [10] and is thus corrected for heart rate using Bazett's formula: $QTc = QT/\sqrt{RR}$. A corrected QT interval of more than 0.47 s is considered abnormal in males and more than 0.48 s in females. Genetically inherited long QT syndrome is discussed below.

3. **Early Repolarization**

A current controversy exists as to the implications of early repolarization seen on the terminal part of the QRS and any predisposition to potentially serious arrhythmias (Fig. 8.12). This QRST wave variant is characterized by at least a 0.1 mV J elevation at the terminal part of QRS in two contiguous inferior and/or

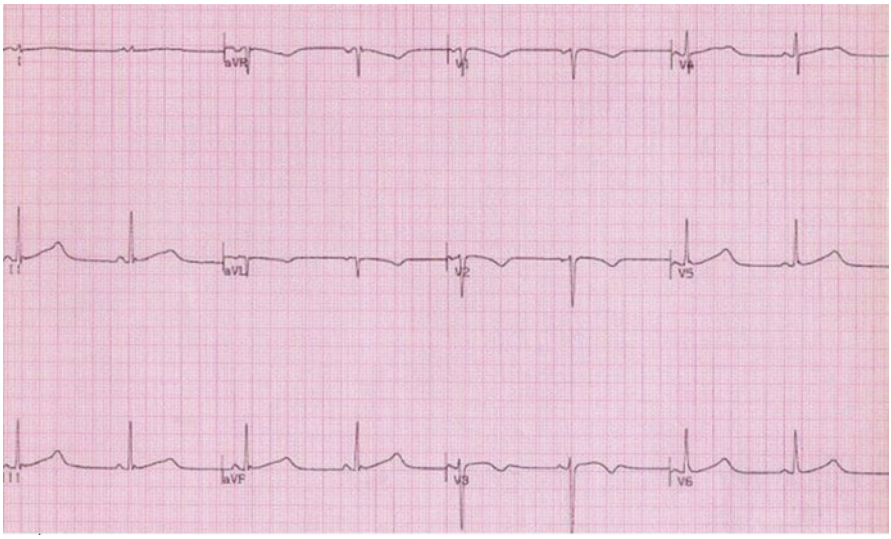


Fig. 8.11 ECG from a 9-year-old patient with pneumonia, treated with a macrolide antibiotic. The QTc interval measures >0.5 s

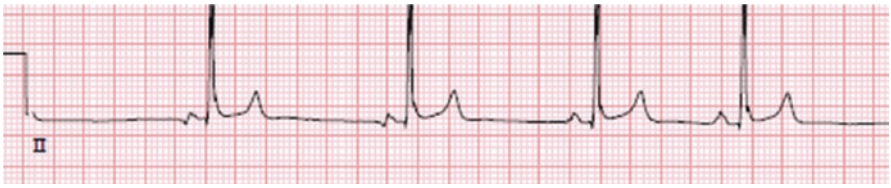


Fig. 8.12 Early repolarization pattern. Note the terminal slurring upstroke of the QRS as it reaches the ST segment

lateral ECG leads. This is in contrast to ST-T wave changes seen with myocardial injury (see below). The prevalence is highly variable between 1 and 13% of children. It is more common in males, young athletes, and those of African-American descent, although it can be seen in any ethnic group. Although there may be an association with arrhythmias among adults with early repolarization, to date, there have been no definitive adverse findings reported in children, and it is thus considered a benign condition [11].

4. **Dextrocardia**

As a result of normal embryologic development, the cardiac silhouette and apex are to the left side of the chest. However, at times, normal heart development occurs but with the cardiac silhouette to the right (mirror image dextrocardia). If this occurs, the position of the sinus node and right atrium is on the left side of the heart, and the resultant P wave axis will be inverted in lead I. In this condition, the

right-sided precordial leads (V1–V3) will show increased QRS voltages, and left precordial leads will have diminished voltages (V5–V7) (Fig. 8.13). This can be confirmed by chest radiography. It is important to confirm that the negative p waves in lead I are not due to lead misplacement (see section on "Artifact").

Answer: D. A tall, peaked, and symmetrical T wave is the first change seen on the EKG on a patient with hyperkalemia.

Ventricular Premature Contractions

Ventricular premature contractions (PVCs) are early depolarizations of the ventricles leading to early systolic ventricular contractions. They are usually followed by a pause resulting in an irregular heart rate (Fig. 8.14). They are usually benign in an asymptomatic healthy child and disappear with exercise. In symptomatic patients who present with low cardiac output syndrome, frequent ventricular premature contractions may be associated with depressed ventricular function predisposing to malignant arrhythmias. They will need cardiology consultation with an echocardiogram to assess the function.

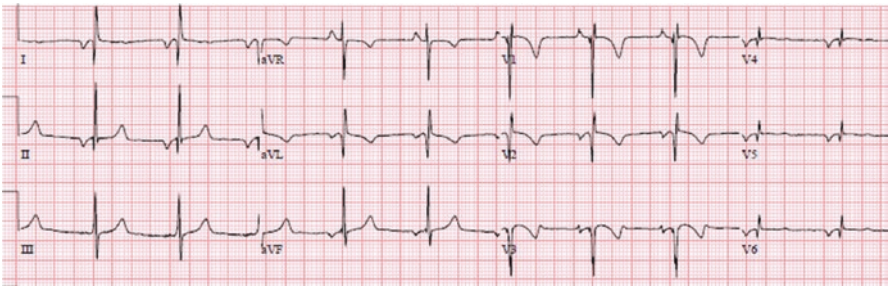


Fig. 8.13 ECG in a child with dextrocardia showing a left sinus node rhythm with negative P waves in leads I, II, and AVL and positive in AVR with low-voltage QRS complexes in the left precordial leads (V5–V6)

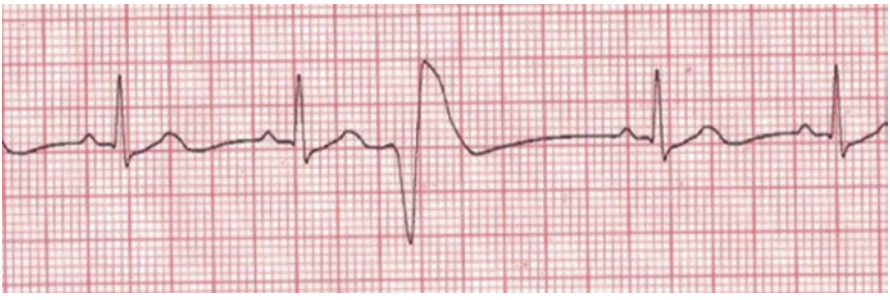


Fig. 8.14 The third QRS complex is wide and occurs earlier than the sinus rate. Depending on location in the respective ventricle, the QRS can exhibit a right, left, or nonspecific bundle branch block pattern

Artifact/Lead Misplacement

A common cause of concern is apparent ECG changes that are artefactual and may lead to unnecessary cardiology consultation and patient and family distress. Limb lead reversal, especially the arm leads, can present a QRS pattern that mimics dextrocardia or a lateral infarct (Fig. 8.15). This may compound anxiety if the patient presents with unrelated nonspecific chest wall pain or costochondritis. This is a common error despite current ECG limb leads that are both color-coded and embossed with letters indicating on which limb to place the electrode (RA, LA, RL, LL). The negative P wave deflection in leads I and AVL indicates that the P wave axis is reversed and should immediately prompt a repeat ECG with proper lead placement.

Everything that appears as an ECG abnormality is not always a real rhythm abnormality. In Fig. 8.16, even the monitor erroneously recorded a wide complex

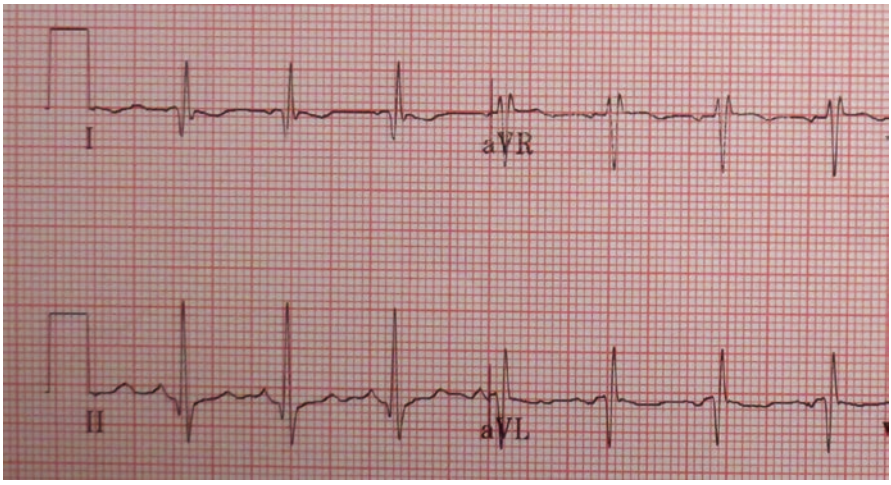


Fig. 8.15 Typical appearance of an arm limb lead reversal. In leads I and AVL, there are negative P waves as well as abnormal Q waves. The appearance of Q waves in these leads is consistent with a lateral infarct. However, the inverted P waves suggest a simple lead placement error

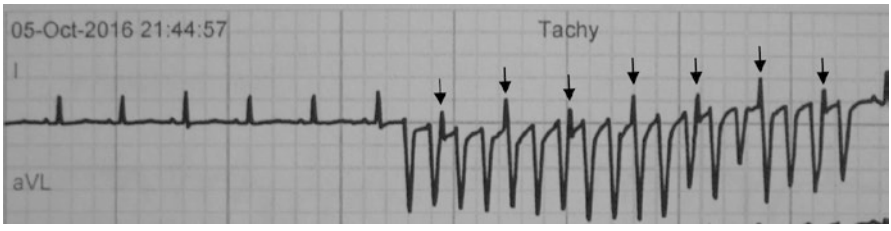


Fig. 8.16 Baseline artifact being detected as an abnormal tachycardia. Close examination shows that the ECG rate is constant as evidenced by the regular upright QRS complexes (arrows) as seen in lead I

tachycardia, when the deep negative deflections are artifactual. This is evidenced by the regular upright QRS complexes which march through at the same rate as the sinus rhythm and are distorted by the artifact.

Case 3

A 1-month-old baby presented to the emergency department with 1-week history of poor feedings and irritability. There are no recent illness, fever, or other associated symptoms. Physical examination: vital signs – afebrile; HR, 290/min; BP, 59/40 mmHg; and RR 40/min. Lungs: clear to auscultation. No murmurs heard. Abdomen: soft and the liver 3 cm below the right costal margin. Peripheral perfusion is decreased with a capillary refill time of 4 s. EKG is performed, and it shows narrow complex tachycardia at a rate of 290/min.

What would be the next appropriate step?

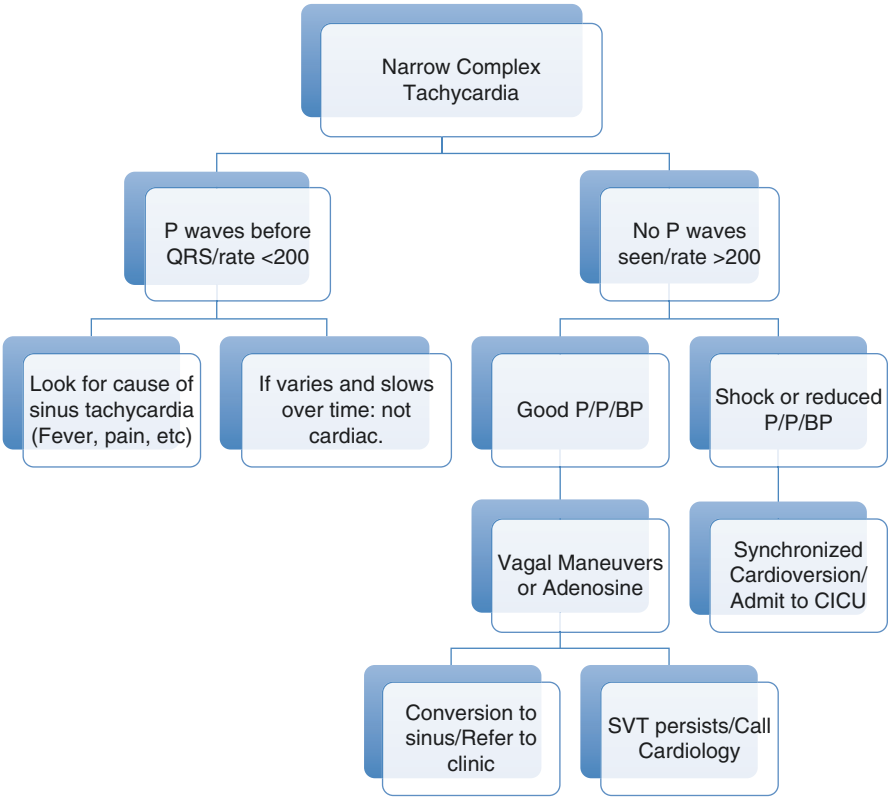
(A) Give adenosine.
(B) B- Synchronized cardioversion.
(C) C- IV antibiotics.
(D) D- Order an echocardiogram.
(E) CXR.

Supraventricular/Atrial Tachycardias

Among children presenting to emergency rooms (ER) with possible rhythm issues, supraventricular tachycardia (SVT) remains the most common etiology presenting at any age. However, the term "SVT" is not in itself a distinct diagnosis but rather a general term simply implying an atrial origin to the arrhythmia.

The atria do not possess a distinct electrical system comparable to the ventricular His-Purkinje system. Therefore, electrical impulse conduction is transmitted via muscle cell- to-cell interaction. Preferential muscle alignments permit rapid conduction to extend from the sinus node, located close to the superior vena caval-atrial junction to the AV node located near the septal leaflet of the tricuspid valve. This electrical impulse propagation can be compared to an ocean wave and is typically uniform. However, when the wave meets an obstacle, it bounces back. As a result of individual anatomy and muscle alignments in otherwise normal heart or due to scar tissue in patients with repaired CHD, atrial tissue conduction at times may not be uniform. This can result in pathways of both "slow" and "fast" conduction. The most common reason for any arrhythmia is "reentry" in which an electrical impulse propagates down one pathway, then backs up another, and circles around to reenter the first. This reentry phenomenon may be confined to the atrial tissue (atrial reentry), has an atrioventricular node (AVN) reentry pathway, or occurs through a direct atrioventricular tissue component as found with pre-excitation conditions such as Wolff-Parkinson-White.

In the current era, correct identification of the reentry site is important as therapeutic options differ based on location. For example, adenosine is commonly administered to attempt termination of SVT but is ineffective in terminating a primary atrial arrhythmia such as atrial flutter. Adenosine may still be useful as it may block the AVN conduction enough to expose the flutter waves to facilitate the proper diagnosis. The QRS morphology during the various forms of SVT is typically narrow or normal unless the patient has preexisting damage to the conduction system, such as following congenital heart surgery, or has any of the pre-excitation syndromes. Immediate therapeutic options will depend on clinical presentations. If there is evidence of impeding shock/heart failure, synchronized D/C cardioversion should be done immediately. However, if the child is hemodynamically stable, adenosine may be an effective first choice of therapy. This is given as a rapid bolus intravenous injection followed by a rapid saline flush as the half-life of adenosine is very short. The dose is 0.05–0.1 mg/kg which can be increased by 0.05–0.1 mg/kg per dose every 1–2 min up to 0.3 mg/kg/dose. For older children >50 kg, start with 6 mg IV, and then try 12 mg rapid IV push.



Algorithm for management of narrow complex tachycardia
P/P/BP = perfusion/peripheral pulses/blood pressure

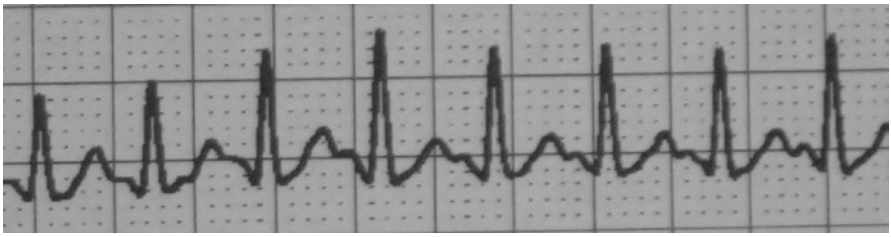


Fig. 8.17 Lead I. Sinus tachycardia. The heart rate is 220 bpm, there are P waves preceding every QRS, and the P axis and morphology are normal

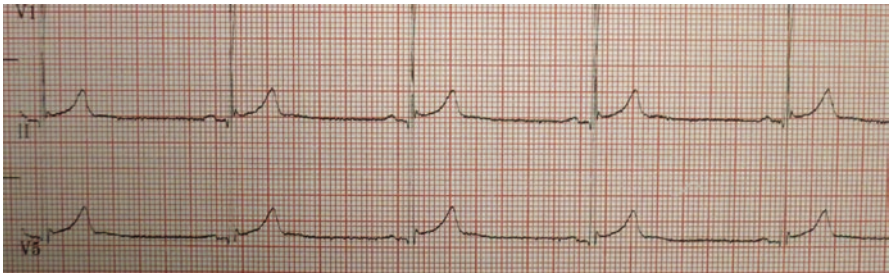


Fig. 8.18 Sinus bradycardia at a rate of about 40 bpm in an asymptomatic 16-year-old who runs varsity cross country and track. Improved physical conditioning results in efficient cardiac performance and slower heart rates. No therapy is required

Primary Atrial Rhythms

Since the sinus node impulse normally travels right to left across the atrial tissue, the P wave of the ECG will be positive in lead I during normal sinus rhythm. This will be irrespective of any changes in rate often up to 230 bpm in an infant and 220 bpm in the young child. The rate is dependent on contributing physical factors and will increase with fever, anxiety, or a high metabolic state [7]. In this regard, sinus tachycardia is associated with a normal P wave axis and morphology and accounts for the majority of narrow QRS tachycardias (Fig. 8.17).

Another frequent area of concern is a slow heart rate or bradycardia (Fig. 8.18). Knowledge of normal heart rate ranges for age is mandatory to be able to recognize heart rates out of ordinary for the child's age. The most common cause of a slow heart rate is sinus bradycardia due to high vagal tone such as seen in adolescents who undergo extensive athletic training which will result in slow resting heart rates. Sinus bradycardia is rarely a cause for concern or need for a referral to a cardiologist unless the child has symptoms of syncope or near syncope.

If the patient has an intrinsic bundle branch block, as is frequently seen following repair of congenital heart defects such as tetralogy of Fallot, the wide complex QRS pattern of right bundle branch block may mimic ventricular tachycardia (Fig. 8.19). Prompt history taking and clinical examination can help to distinguish between SVT and VT.

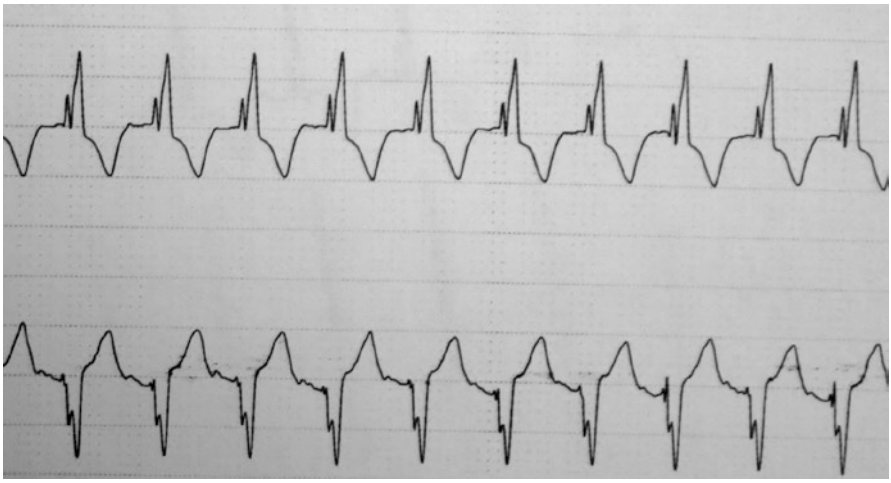


Fig. 8.19 Top, lead V1; bottom, lead II. Many patients with repaired CHD will have a complete right bundle branch QRS pattern, and rapid atrial rates may mimic ventricular tachycardia. Although the rhythm in lead V1 (top) appears to be a wide QRS ventricular tachycardia, a careful search shows normal P waves in lead II (bottom) confirming sinus tachycardia

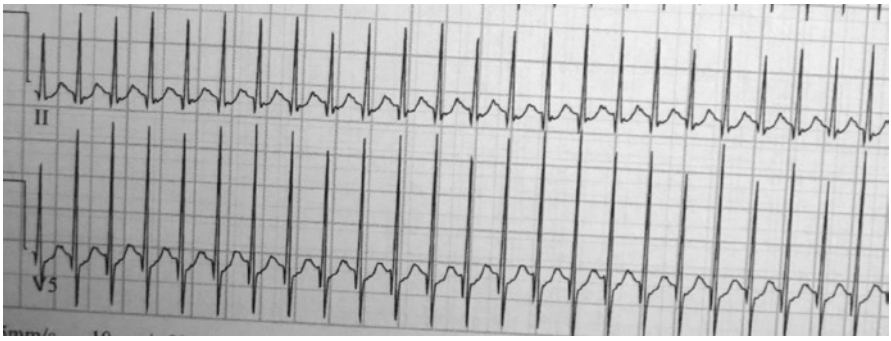


Fig. 8.20 Atrial ectopic tachycardia at 290 bpm. This can easily be confused with sinus tachycardia, but the incessant fixed rate that is much too fast for sinus indicates its origin from an abnormal focus elsewhere in the atria

Atrial ectopic tachycardia or ectopic atrial tachycardia (AET/EAT) is caused by instability of tissue membrane potentials in the atrium and can have heart rates reaching 250–270 bpm. This rhythm is typically faster than can be reached with sinus tachycardia. AET/EAT tachycardia rates are fixed without the usual rate variability seen with sinus rhythms (Fig. 8.20). It results from an automatic focus within the atria. They tend to accelerate (warm up) and get faster after the first few beats or decelerate (cool down) before reverting to sinus rhythm. They tend to have insidious presentation, with a gradual onset of heart failure or even dilated cardiomyopathy. Adenosine and direct current cardioversion are not effective in converting this rhythm. Cardiology consult should be immediately performed for pharmacological control.

Answer: B. In this infant with signs of shock, immediate synchronized cardioversion should be performed. If there was no sign of shock, then a rapid adenosine bolus can be given.

Atrial ectopic tachycardias can have a sudden onset, but it is typically faster than sinus tachycardia, and there is usually an abnormal P wave present (Fig. 8.21).

Primary Atrial Tachycardia (Flutter, IART, Fibrillation)

Case 4

A 45-year-old patient with single ventricle physiology presents with a 24 -h history of sudden-onset palpitations with no other associated symptoms. Physical exam: afebrile, HR100/min, BP 110/70 mmHg, and RR 20/min. Normal S1 and single S2. Lungs: clear to auscultation, no crackles, and no wheezing. Abdomen: soft, no tenderness or hepatomegaly, and well perfused. EKG performed showed the findings in Fig. 8.22.

What would be the most appropriate next step?

(A) Give adenosine.
(B) Admit to cardiology and start IV heparin.
(C) Synchronized cardioversion.
(D) Order an echocardiogram.
(E) External pacing.

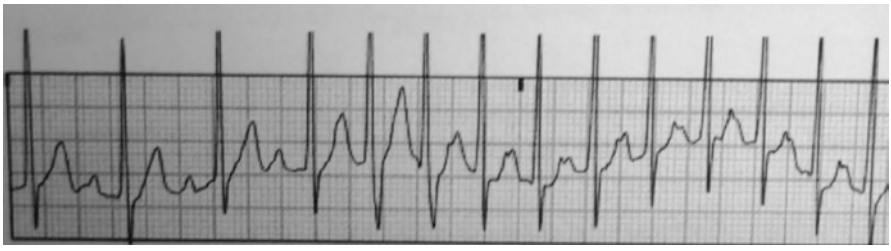


Fig. 8.21 Lead II. In this figure, sinus rhythm at a rate of 100 bpm is evident on the left. There is then a single premature atrial complex with the P wave on top of the T wave causing distortion of the usual rounded T wave appearance. This initiates a rapid atrial tachycardia at 250 bpm

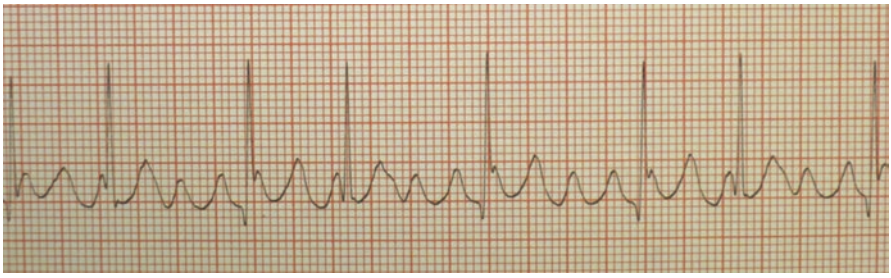


Fig. 8.22 Lead II. Typical "saw tooth" appearance of atrial flutter waves (typically about 300 bpm) with variable AV conduction, in a 3 or 4 to 1 ratio

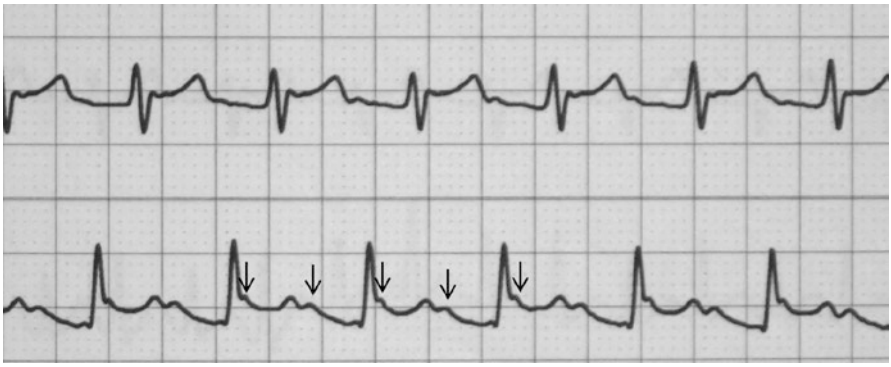


Fig. 8.23 Among patients with repaired CHD with stretched and thickened atria, a slower form of atrial flutter, IART, can occur. As seen, this rhythm can be misinterpreted as sinus tachycardia with 1° AV block. Careful examination of QRS complexes in the bottom lead shows a second atrial wave (black arrow) buried at the end of the QRS complex confirming 2:1 block of this slow atrial flutter

As previously discussed, the atria do not possess special conduction pathways for smooth electrical propagation. Anatomical landmarks such as the tricuspid valve ring, venae cavae, muscle ridges (such as the crista terminalis), and pulmonary vein openings all contribute to interrupt the electrical impulse to create potential re-entrant circuits [12]. In addition, children with repaired CHD have surgical incisions which leave behind scars which can alter electrical conduction. A common result is atrial flutter which can be diagnosed by examining leads II and III for the "saw tooth" pattern of the flutter waves (Fig. 8.22). Any adult with a repaired congenital heart defect presenting with a fast heart rate for no apparent reason should be evaluated for atrial flutter.

Atrial flutter rates range from 240 to 340 bpm and conduct to the ventricles at variable ratios (1:1–4:1) depending on AVN conduction, yielding ventricular rates of 150–200 bpm [9]. Although flutter waves with AVN conduction ratios of 4:1 can readily be appreciated, conduction ratios of 1:1 or 2:1 may be misinterpreted as sinus tachycardia. In such instances, the "T wave" of the QRST may be erroneously identified as a "P wave." Remembering that every QRS has a T wave following it and P waves can be independent will help to make the correct diagnosis.

Among patients with certain repaired CHD, such as single ventricles (Fontan-type surgical repair) a variant of atrial flutter, intra-atrial re-entrant tachycardia (IART), is commonly seen. The atrial rate is typically slower than flutter (150–200 bpm) and, therefore, so is the concomitant ventricular response (75–100 bpm). A misdiagnosis of sinus tachycardia with 1° AV block is often made (Fig. 8.23).

Although less common in children than adults, atrial fibrillation does occur, especially among patients with repaired CHD. The ECG typically shows an irregularly irregular ventricular rate with nonspecific undulating atrial activity (Fig. 8.24).

In instances of primary atrial tachycardias, the administration of adenosine may not terminate the arrhythmia; however, the drug can transiently block AVN conduction and expose the underlying atrial rhythm (Fig. 8.25). This patient can be referred to an electrophysiologist for medical therapy or, if resistant, for an EP study.

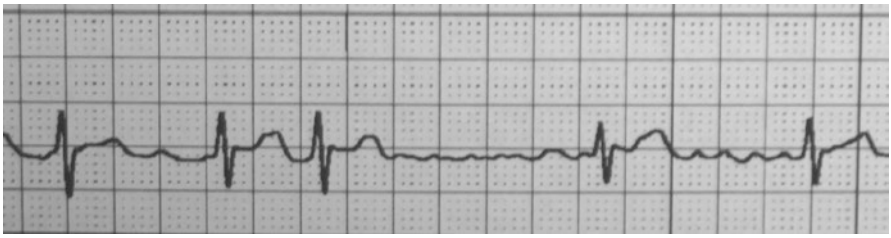


Fig. 8.24 Atrial fibrillation. Coarse wavy baseline is evident with a highly variable rate of QRS complexes

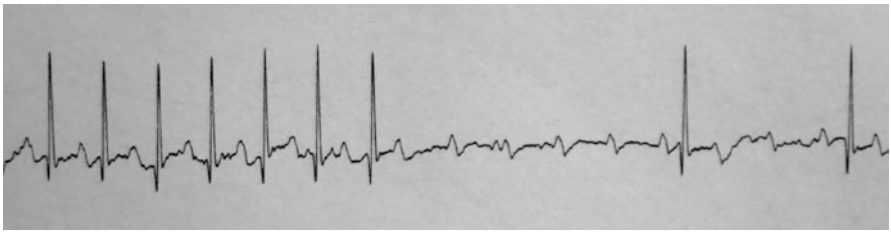


Fig. 8.25 Administration of adenosine during a primary atrial tachycardia typically does not terminate the arrhythmia since the AV node is not part of the circuit. However, it can aid in making the proper diagnosis by transiently delaying AV node conduction where the underlying atrial tachycardia is exposed

AV Node Re-entrant Tachycardia (AVNRT)

AVNRT can be best understood with the concept of "slow" and "fast" conduction as described above. Some children have more than one pathway for electrical impulses to cross the AV node. Atrial tissue connections to the AV node conduct at variable speeds and create a reentry circuit. Typically, the sinus impulse reaches the AV node via the "slow" atrial tissue pathway. It then exits via the "fast" pathway back into the atrium only to reenter the "slow" circuit again. Often, this circuit can be interrupted by vagal maneuvers or medication. The typical ECG shows a rapid narrow QRS (typically >220 bpm) with no visible retrograde P waves (Fig. 8.26). Tachycardia exhibits the qualities of both starting and stopping abruptly. Since the AV node is involved in the arrhythmia circuit, medications such as adenosine can be effective in acutely breaking the re-entrant circuit. The incidence of this type of SVT increases with age and becomes the most common form of SVT by adolescence [13].

Answer: B. Admit to cardiology and start IV heparin. The most likely cause for the sudden onset of a regular tachycardia in an older patient who had a Fontan operation for a "single ventricle" is a slow atrial flutter. This carries a risk for atrial thrombosis, and thus the patient should be admitted for anticoagulation and elective cardioversion once a transesophageal echocardiogram is done to look for a thrombus.

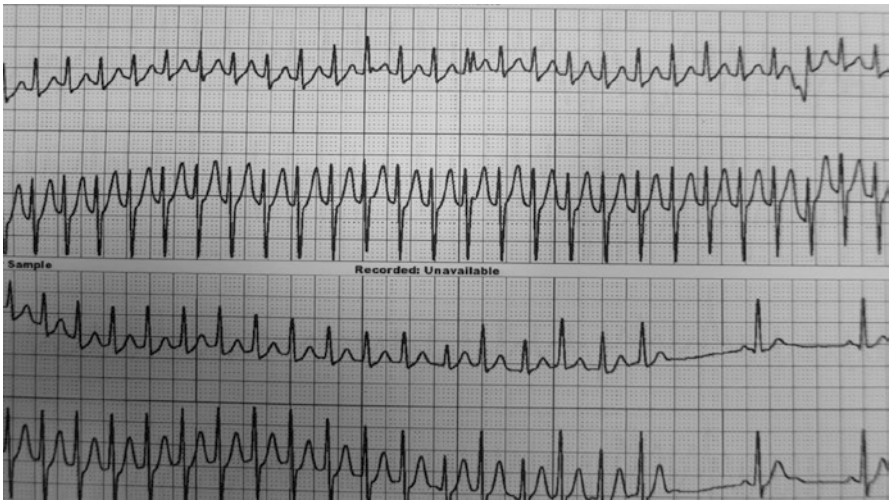


Fig. 8.26 Typical appearance of AVNRT illustrating a narrow QRS tachycardia without evidence of P waves. As shown at the end of the strip, tachycardia terminates abruptly

Atrioventricular Reciprocating Tachycardia (AVRT): Pre-excitation Syndromes

AVRT is the most common form of SVT among infants and represents persistence of embryologic atrioventricular muscle connections. Since any direct tissue-tissue connection may not possess the property of slowing of conduction found in the AV node, the PR interval is typically shorter than normal. Since these direct muscle connections will activate the ventricle earlier than electrical impulses arising via the AV node, the resultant QRS will exhibit varying degrees of pre-excitation or a slur at the onset of the QRS complex referred to as a "delta" (Δ) wave. The QRS may exhibit only a mild slur or a complete bundle branch block pattern, depending on how much of the ventricle is activated via the accessory connection. In addition, the delta wave may be negative, depending on the particular lead. Careful examination of all leads is mandatory to make the correct diagnosis (Fig. 8.27).

In addition, there may be competitive AV conduction between the normal AV node and accessory pathway so that both a normal and a preexcited QRS pattern can occur (Fig. 8.28).

The most common clinical pre-excitation condition is the Wolff-Parkinson-White syndrome (WPW) although there are other types of pre-excitation (Mahaim variants, atrio-nodal and atrio-junctional), which create accessory electrical conduction pathways and predispose to reentry arrhythmias. Among anatomical congenital heart defects, the most common association with WPW is Ebstein anomaly of the tricuspid valve. Other associated congenital cardiac conditions with WPW include Noonan's syndrome and various cardiomyopathies.

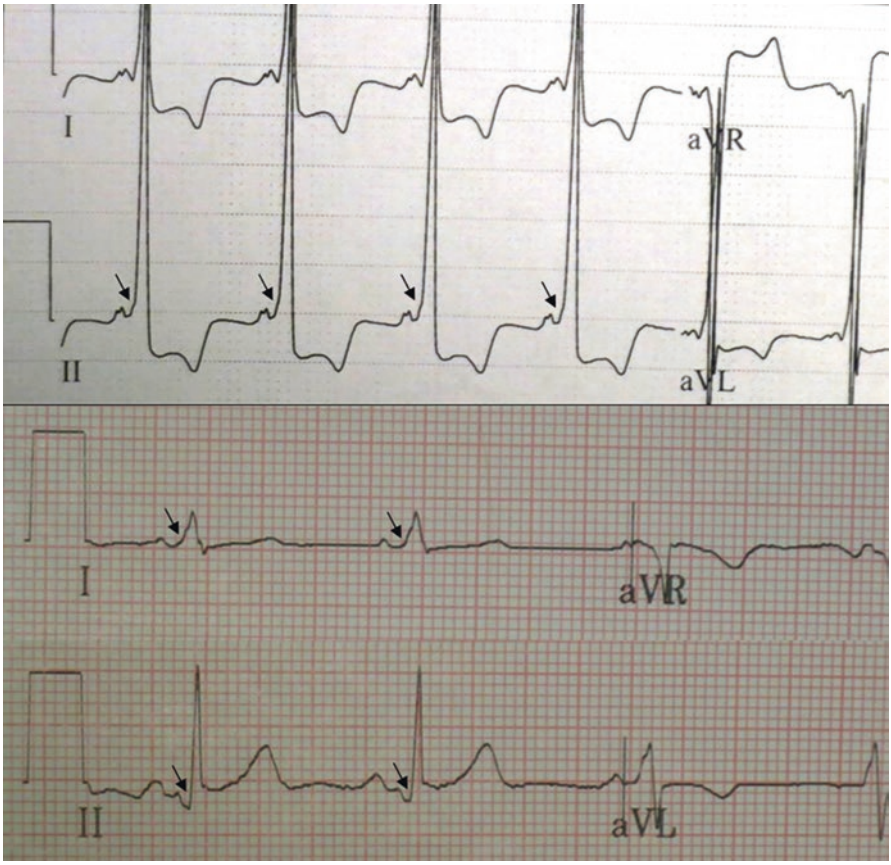


Fig. 8.27 The "delta" wave at the onset of the QRS characteristic of ventricular pre-excitation can be quite obvious or very subtle depending on the particular lead and location of the accessory connection in the heart. The delta wave can be either a positive (top) giving an upward slant to the QRS or a negative deflection as seen in lead II on the bottom tracing

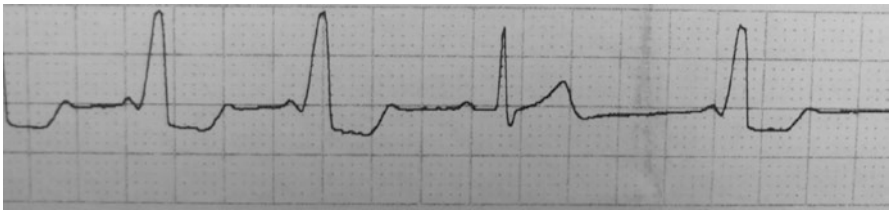


Fig. 8.28 The pre-excitation QRS pattern is not necessarily an "all-or-none" phenomenon. Depending on conduction and refractoriness of tissue, both normal and pre-excitation QRS patterns can occur in the same patient

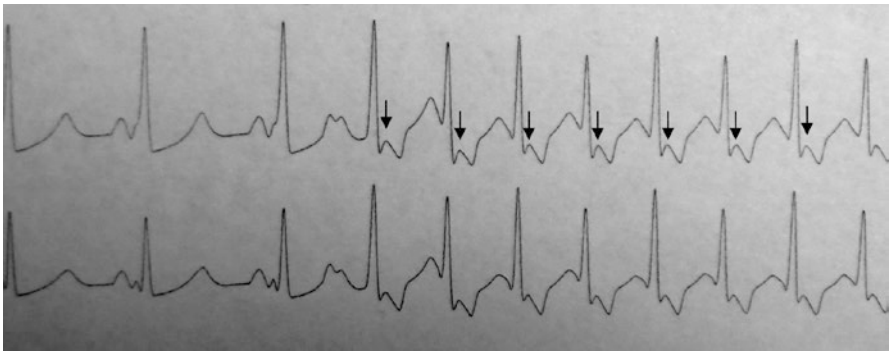


Fig. 8.29 Typical "orthodromic" AVRT (WPW) showing sinus rhythm with preexcited QRS complexes on the left and then a single PAC which is conducted to the ventricle via the AV node with a normal QRS duration. This initiates a narrow QRS tachycardia with retrograde P waves (arrows) evident just following the QRS and distorting the ST segment

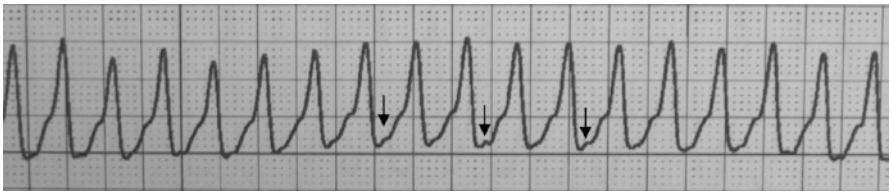


Fig. 8.30 "Antidromic" AVRT can mimic monomorphic ventricular tachycardia. Retrograde P waves are visible following the wide QRS complexes as indicated by the arrows

In the typical form of AVRT, antegrade AV conduction is via the AV node (orthodromic conduction). The accessory pathway then acts to permit retrograde VA conduction, and the circuit is created. The QRS is typically narrow with a heart rate of ≥250 bpm. Due to retrograde atrial activation from the circuit, retrograde P waves are commonly visible immediately after the QRS (Fig. 8.29).

Rarely in children, the antegrade AV conduction utilizes the accessory pathway (antidromic conduction) with the AV node serving to create the retrograde circuit. In these instances, ventricular activation begins where the accessory pathway connects and causes an altered QRS pattern. This may mimic ventricular tachycardia (Fig. 8.30). Since the AV node plays a role in the re-entrant circuit, adenosine can be an effective therapy. Of note, since the accessory tissue connections do not slow the atrial rate, sudden death (about 0.8% of cases) can result if atrial flutter occurs with a very rapid ventricular response (Fig. 8.31) [14].

The flow chart gives a suggested evaluation and therapeutic pathway for patients who present with a wide complex QRS tachycardia. In general, if they are in any distress or show signs of shock, immediate synchronized electrical cardioversion should be performed, with simultaneous emergent cardiology

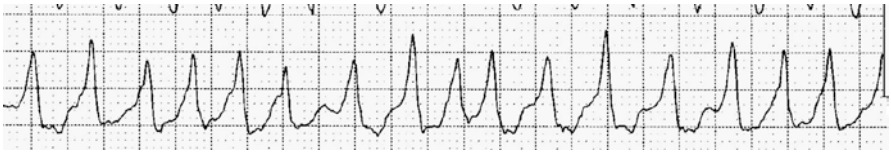
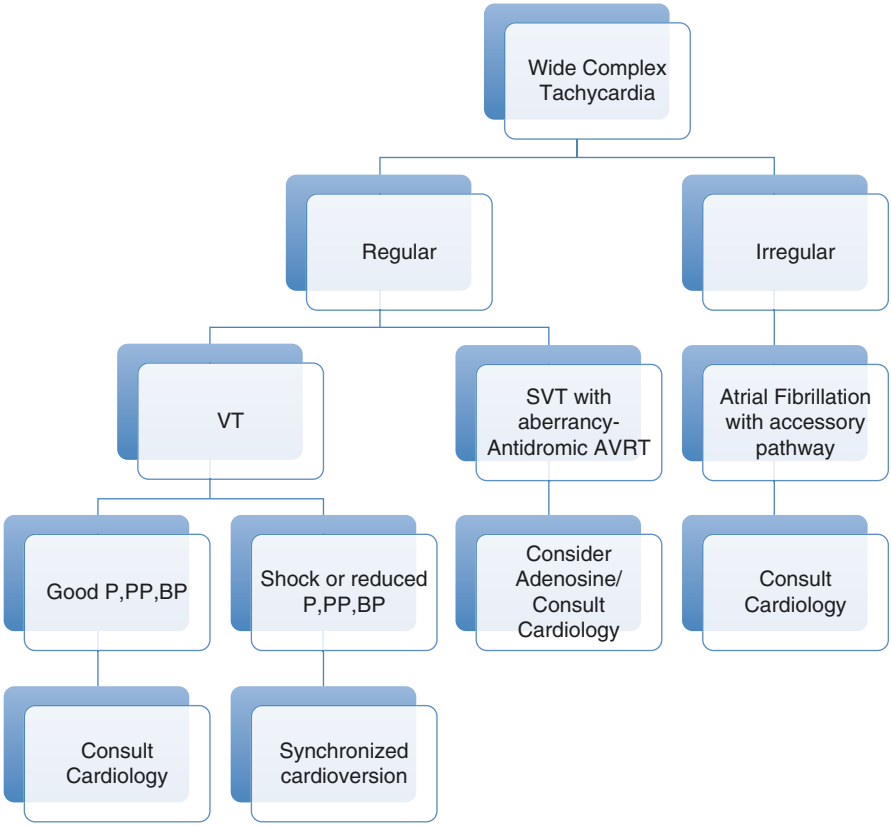


Fig. 8.31 Atrial fibrillation in a patient with WPW and antidromic conduction. Compared with Fig. 8.29, the R-R intervals are variable. The shortest R-R measures about 200 ms (heart rate 300 bpm) which can contribute to sudden death if that conduction rate is persistent

consultation. If they are clinically well, then a more detailed evaluation of the ECG recordings can be done and reviewed with a cardiology consultant (by phone, scanning, or email) to determine if medical therapy may be beneficial in converting the abnormal rhythm.



Algorithm for Management of Wide Complex Tachycardia
P/PP/BP = Perfusion/peripheral pulses/blood pressure

AV Node Conduction

First-Degree AV Block

The AV node protects the ventricle from rapid atrial rhythms due to its inherent anatomy and tissue conduction properties which slows the electrical propagation from the atria to the ventricles. This is reflected by the PR interval on the resting ECG. Age-related changes in the PR occur. In addition, changes in the PR interval can be the result of normal vagal tone, medications, myocardial damage, and infections. AVN conduction problems can be associated with certain congenital heart lesions such as septal defects and congenitally corrected transposition of the great arteries as well as with repaired CHD.

First-degree AV block is defined as a PR interval that is prolonged for age (usually >180 ms in young children and >200 ms in older children and adults), and every P wave is followed by a QRS (Fig. 8.32) Typically, no treatment is required although clinical correlation is important if the patient is on any medications that might cause AVN delay (beta- or calcium blockade), has a fever (possible myocarditis or acute rheumatic fever), or has evidence of Lyme disease.

Case 5
A 14-year-old girl sinks to the bottom of the pool while swimming a competitive meet. She is rescued, given CPR, and brought to the ED. She revives but has no memory of what occurred. On examination, her heart rate is 35 and regular, BP is 86/54, and her perfusion is reduced. Her ECG is shown in Fig. 8.35. The first step in her management is:

(A) Emergent temporary pacemaker insertion
(B) IV epinephrine
(C) Oral steroids
(D) Synchronized cardioversion
(E) Chest CT scan

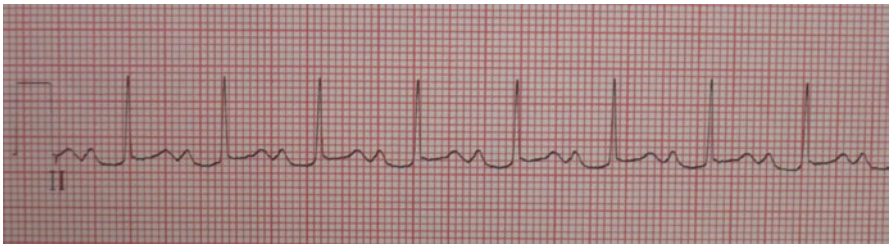


Fig. 8.32 First-degree AV block. P wave axis and morphology are normal. PR interval is prolonged (240 ms); however, every P wave is followed by a QRS complex

Second-Degree AV Block

Progression of the AV node delay which results in intermittently non-conducted P waves is referred to as second-degree AV block (2° AV block). Two distinct patterns are observed: Mobitz type 1 or Wenckebach has progressive PR interval prolongation preceding the non-conducted P wave (Fig. 8.33), while Mobitz type 2 has a fixed PR interval with dropped beats (Fig. 8.34). Most commonly, Wenckebach second-degree block is a benign finding, while Mobitz type 2 is not as this type may progress to third-degree or complete heart block.

Complete or Third-Degree AV Block

Complete independence of the P wave from the QRS is never normal (Fig. 8.35). Complete heart block (3° AV block) may be congenital or the result of inflammation, disease, surgery, CHD, or drug therapy. Complete congenital heart block

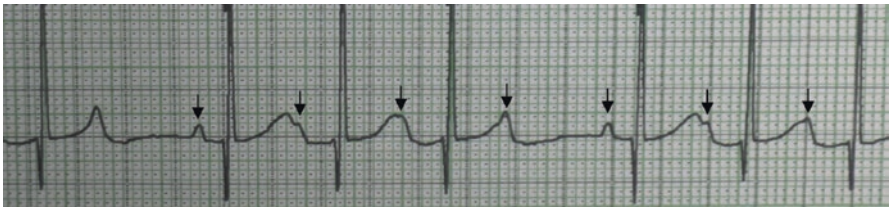


Fig. 8.33 2° AV block Wenckebach (Mobitz type I) is characterized by progressive PR prolongation that precedes the non-conducted P wave (arrows indicate the p waves). It is secondary to increased vagal tone and can occur in normal subjects and athletes without cardiac disease

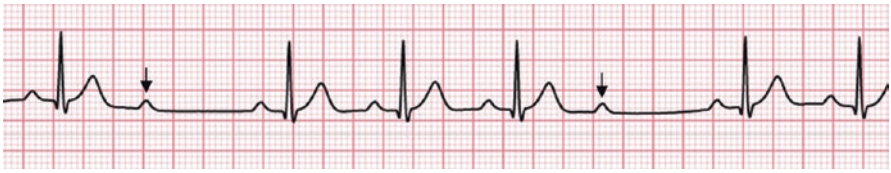


Fig. 8.34 Mobitz type 2 is characterized by a constant PR interval prior to the non-conducted P wave (arrows). It results from conduction system disease below the level of the AV node. This is typically abnormal and rarely seen in children without underlying heart disease

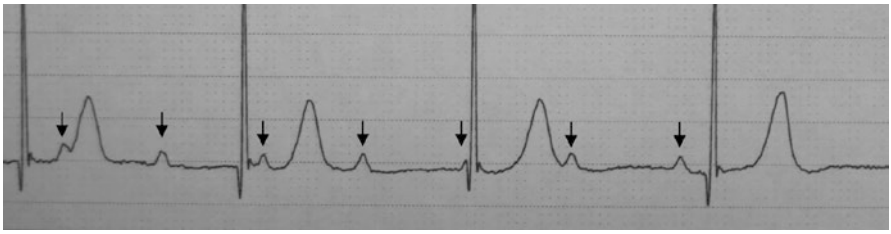
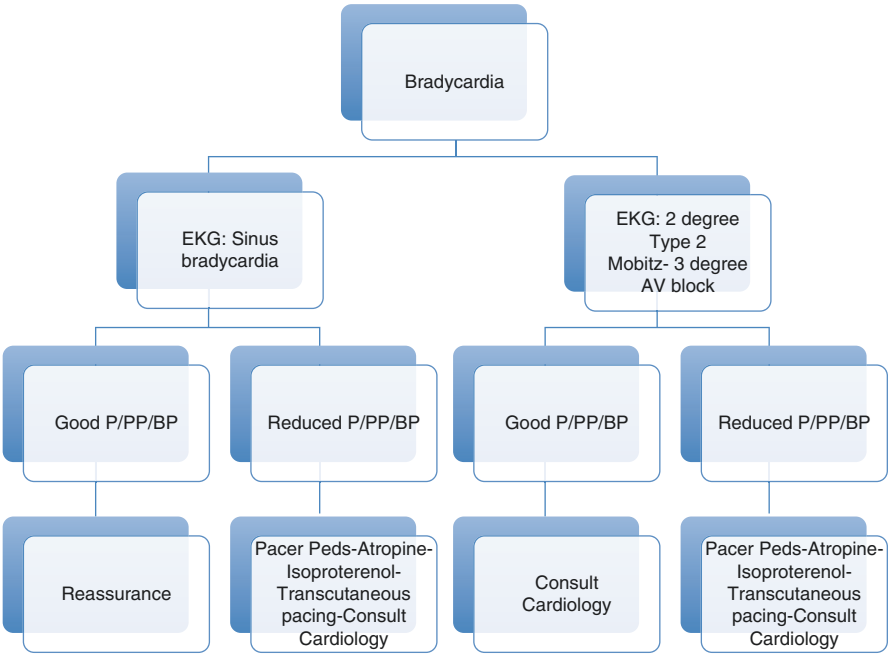


Fig. 8.35 Complete or 3° AV block with independent atrial P waves (arrows) and narrow QRS ventricular complexes

occurs sometimes in babies born to mothers with systemic lupus who have antibodies that affect the developing conduction system in the fetus. If the ventricular rate is too slow, the patient may develop heart failure and require pacemaker therapy.

The flow chart presents a method of quickly evaluating patients with bradycardia. In general, there is urgency in cardiology evaluation and intervention only when shock or near shock accompanies the slow heart rate.



Algorithm for Management of Bradycardia
P/PP/BP = Perfusion/peripheral pulses/blood pressure

Cardiac Devices (Pacemaker/Defibrillator)

Children, even infants, do receive pacemakers and defibrillators (ICD) as a result of AV node issues or ventricular arrhythmias typically resulting from CHD surgery or inherited abnormalities of the conduction system. A full description of device therapy among children and patients with CHD is beyond the scope of this chapter and can be found elsewhere [15]. Basically, a pacemaker emits an electrical pulse which then elicits a cardiac muscle response. Patients who receive a pacemaker are given ID cards to help identify the manufacturer and the device model type. On the ECG, when the device emits an electrical discharge, a spike or sharp signal is seen. Normally, this is followed by either an atrial P or ventricular QRS complex, depending if the device is single (pacing only the ventricle) or dual chamber pacing both the atria and ventricles (Fig. 8.36). ICDs are used for internal defibrillation of potentially life-threatening arrhythmias for those at greatest risk. However, they have risks associated with them. The faster heart rates of children compared with adults may be interpreted as an arrhythmia by the ICD,

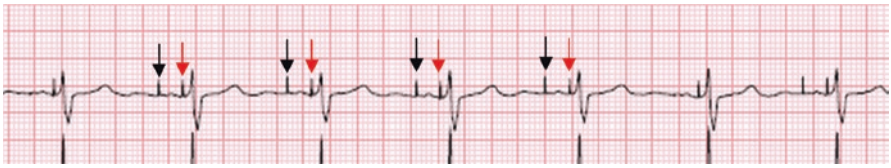


Fig. 8.36 Lead II: normal dual-chamber pacemaker function. Every electrical spike is followed by either an atrial P wave or ventricular QRS complex. Black arrow-atrial spike; Red arrow-ventricular spike

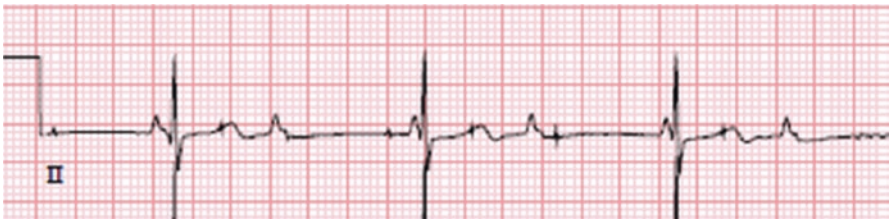


Fig. 8.37 ECG from a 5-year-old with congenital AV block and a fractured pacemaker lead. Note the complete AV block, slow ventricular rate, independent P waves, and intermittent pacemaker spikes that are not followed by any cardiac muscle response

causing an inappropriate ICD discharge. The important concept for the ER physician is to determine if the device is functioning properly (Fig. 8.37). Often a chest radiograph will show a displaced or fractured pacemaker/ICD lead. In instances of suspected device malfunction, or inappropriate ICD discharges, a thorough evaluation of the device is mandatory. Suspected pacemaker malfunction should prompt an urgent cardiology consultation to one familiar with implanted cardiac device technology and has access to the pacemaker interrogating device.

Answer: A. Newly diagnosed complete heart block is a true cardiac emergency requiring cardiac pacing. In the teenager described, this may be due to myocarditis which has affected the conduction system. The cardiology consultant will be able to insert a temporary pacemaker through a femoral vein or jugular approach. If cardiologist is not available, then external pacing may be required until a transvenous pacemaker insertion is available and if the low rate does not maintain adequate cardiac output. Newborns with congenital complete heart block may tolerate ventricular escape rates in the 50s but may require pacemaker insertion for lower rates or if associated decreased ventricular function is present.

Ventricular Arrhythmias

While abnormal and worrisome, ventricular arrhythmias are fortunately less common in children than in adults; it is imperative that the primary care provider is able to identify these findings and provide appropriate initial management [16]. Patients with a history of known and/or repaired CHD represent a high-risk population for serious arrhythmias. However, patients with no prior cardiac history may also sometimes present with serious ventricular rhythm abnormalities.

Repaired Congenital Heart Disease and Bundle Branch Blocks

Following intracardiac repair of CHD or in certain unrepaired defects, the baseline ECG is often abnormal due to surgical incisions and/or altered anatomy. This can cause a bundle branch block or nonspecific intraventricular conduction delay and serve as a diagnostic clue even before a full history is elucidated. In the presence of concerning symptoms (e.g., pre-syncope/syncope and sensed tachycardia), the ER physician should have a high index of suspicion for the presence of serious arrhythmias in such patients even when no arrhythmia has been documented in the ER setting.

Complete right bundle branch block (RBBB) is common in patients with repaired ventricular septal defects and tetralogy of Fallot due to surgical incisions on the right ventricular wall. Complete RBBB (Fig. 8.38) produces a typical "rsR" prime pattern ("rabbit ears") in lead V1 with a wide QRS (>120 ms) with a terminal delay in the left precordial leads representing delayed activation of the right ventricle. A cause of diagnostic error often arises among patients with repaired atrioventricular canal defects in whom the QRS axis is "leftward" or "superior." This finding, combined with a RBBB pattern, often leads to the erroneous diagnosis of bifascicular coronary disease.

A true left bundle branch block (LBBB) is rare and always pathological in the pediatric population and merits full investigation in conjunction with a cardiologist. This is often associated with intrinsic myocardial issues or Kawasaki disease and coronary involvement (Fig. 8.39). The LBBB is characterized by a wide QRS (> 120 ms) with a negative deflection in V1 (QS pattern or sometimes a tiny r wave with rS pattern), late precordial transition (positive R waves beyond lead V3 or V4), and often a notched "M" pattern in the middle of the R wave in lateral precordial leads.

Nonspecific intraventricular conduction delay (IVCD) not meeting the criteria for either a RBBB or a LBBB pattern may be seen in patients with some repaired congenital heart defects, especially patients with a single ventricle (Fontan-type palliation). The QRS duration is typically <120 ms (Fig. 8.40). This finding may not be of concern unless clinical findings suggest an arrhythmia.

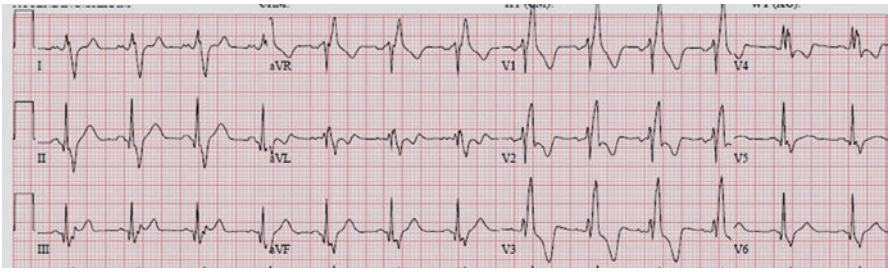


Fig. 8.38 Typical appearance of a RBBB pattern seen among patients with repaired CHD, most commonly tetralogy of Fallot

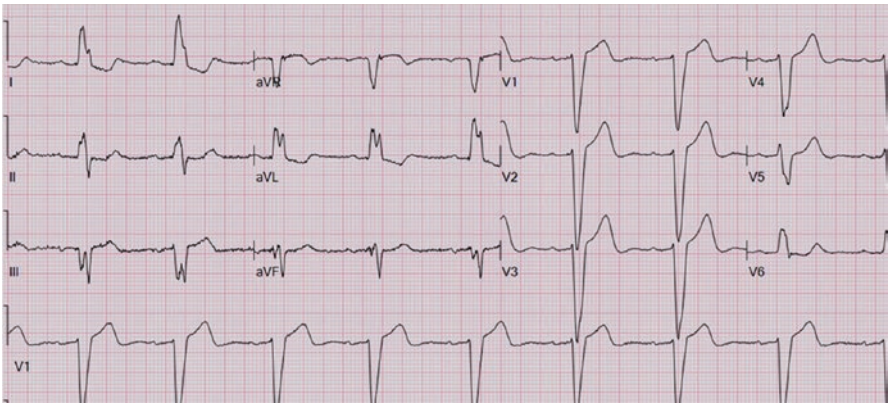


Fig. 8.39 LBBB pattern. In comparison to the more common RBBB pattern, the "rabbit ears" are absent in the right precordial leads

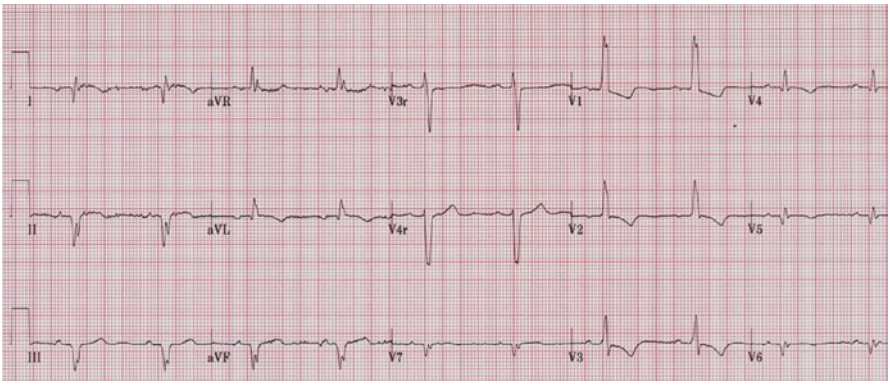


Fig. 8.40 Nonspecific QRS pattern in IVCD showing neither a right nor left bundle branch block pattern. This is common among patients with single ventricle anatomy

Myocardial Injury

Myocardial injury creates a distinct pattern of ST changes, seen at the terminal part of the QRS. Typically, there is diffuse ST segment elevation seen in multiple leads (Fig. 8.41). This is different from the early repolarization pattern described above. Clinical suspicions of inflammatory muscle conditions such as myocarditis or coronary artery problems should be explored. This pattern is always abnormal and requires urgent cardiology consultation and ICU admission. Confirmatory blood tests including cardiac troponins are helpful in this situation.

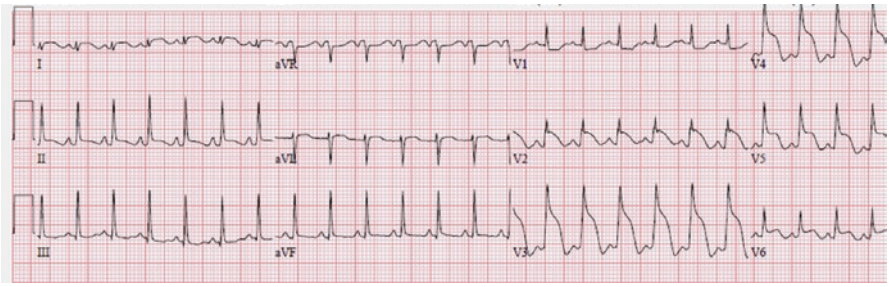


Fig. 8.41 Diffuse ST elevation is seen across the precordial leads in a child presenting with myocarditis

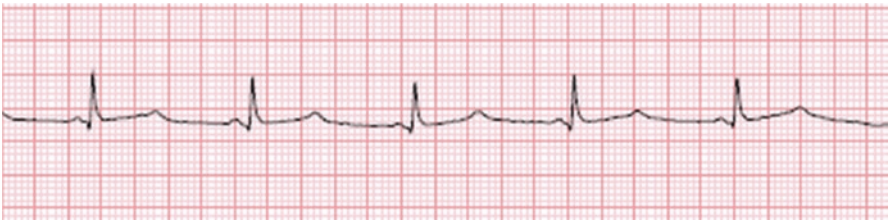


Fig. 8.42 Lead II: QTc >500 ms in a child with familial LQTc syndrome

Genetic Cardiac Channelopathies

A detailed description of the cardiac channelopathies that predispose the patient to serious ventricular arrhythmias and sudden death is beyond the scope of this chapter and can be found elsewhere [17]. A high index of suspicion is needed for the diagnosis in the presence of concerning symptoms and no documented arrhythmia.

A QT interval corrected for heart rate (QTc) ≥ 470 ms in adolescent males or ≥480 ms in adolescent females is considered to be prolonged [18]. A measured prolonged QT interval in the setting of seizures, syncope, or pre-syncope associated with exertional or a sudden startle merits referral to the cardiologist for further evaluation. Discussions should include a family history of long QT syndrome (LQTS) or sudden unexpected death in first-degree relatives. Familial LQTS should be suspected when the QTc interval is ≥500 ms in the absence of any drugs or electrolyte abnormalities which can prolong the QT interval (see above). Other non-cardiac conditions (e.g., trauma, acute intracranial pathology) can also cause QT prolongation (Fig. 8.42).

Another cause of syncope is the Brugada syndrome (BrS) which is rare among children. It is one of the inherited sodium channelopathies and is associated with ventricular arrhythmias. The typical Brugada type I ECG pattern (Fig. 8.43) is denoted by a coved ST segment elevation ≥2 mm in leads V1 and/or V2. Placement of the precordial ECG leads at a higher intercostal space than normal (second intercostal space) improves detection.

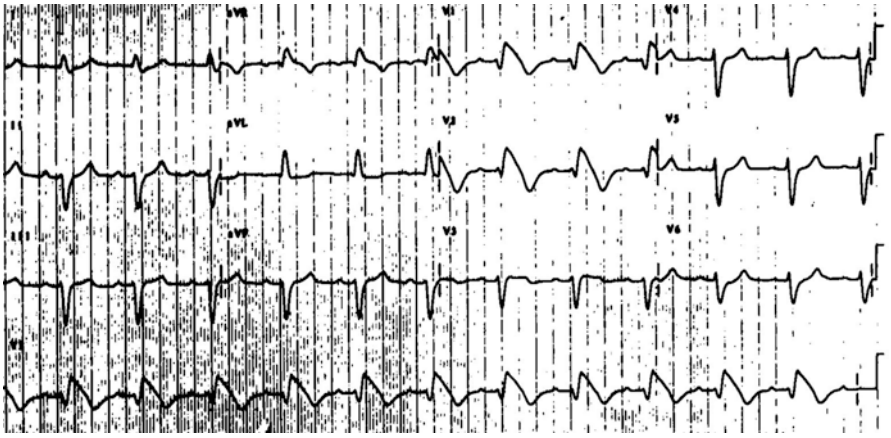


Fig. 8.43 Typical "coved" QRS pattern in lead V1 of Brugada syndrome. Compare this to a RBBB pattern above

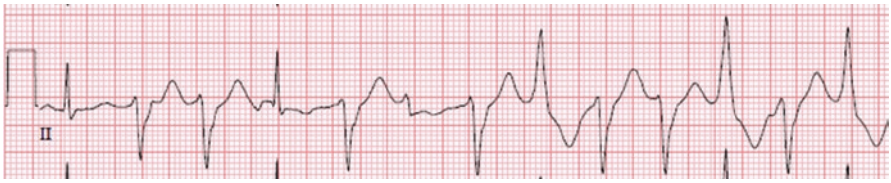


Fig. 8.44 Bidirectional ventricular tachycardia in a patient with CPVT. Note the initial normal QRS pattern (first beat on the left) prior to the arrhythmia onset

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is another inherited myocardial channelopathy associated with sudden death (Fig. 8.44). However, the baseline ECG is normal. Ventricular ectopy and arrhythmias may occur spontaneously but happen more commonly with exercise.

Ventricular Tachycardia

As indicated above, premature ventricular contractions (PVC) are common in children even with structurally normal hearts. However, ventricular tachycardia (VT) is rare. VT can be non-sustained (<30 s and no significant symptoms) or sustained (≥30 s and/or significant symptoms such pre-syncope or syncope). VT can be classified as monomorphic (all QRS complexes with the same morphology), bidirectional (positive and negative QRS), or polymorphic (variable morphologies). The diagnostic criteria for VT include a wide QRS tachycardia without a typical right or left bundle branch block pattern and the presence of AV dissociation. The right ventricular outflow tract (RVOT) is a common site for arrhythmia (Fig. 8.45). Management depends on type of arrhythmia [19, 20].

While polymorphic VT/VF can be the result of channelopathies such as BrS and CPVT, it can also be seen in the setting of ischemia as illustrated in Fig. 8.46 from

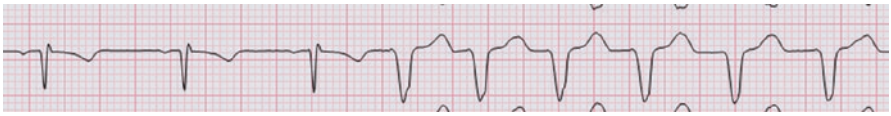


Fig. 8.45 Lead V1: Onset of a wide QRS with LBBB pattern in a teenager arising from the RVOT

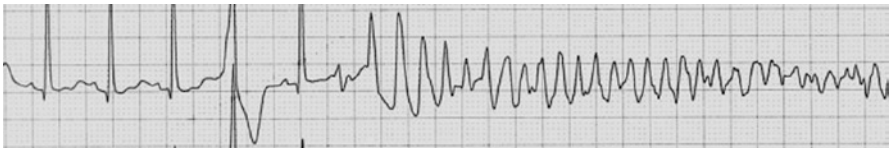


Fig. 8.46 Sinus rhythm followed by a PVC and subsequently the Torsades de pointes variety of VT

a child with coronary artery occlusion following a surgical "unroofing" for anomalous origin of left coronary artery from right aortic sinus. Torsades de pointes is a type of polymorphic VT with gradual variation in the amplitude and "twisting" of QRS axis around the isoelectric line and is the type of VT associated with the long QT syndrome and certain medications. Sustained VT with decreased perfusion always requires immediate action, including electrical cardioversion and emergent cardiology consultation. This most often would be followed by medications in an intensive care setting as recurrence of the arrhythmia is common.

Conclusion

Children at any age can present with arrhythmias. It must be remembered that a repaired congenital heart defect is never normal and even with excellent surgical repair, residual hemodynamic changes and scar tissue can predispose to arrhythmias. In addition, genetically inherited arrhythmogenic conditions can be expressed at any age, and the developing cardiac conduction system predisposes infants and young children to rhythm disturbances. Computer-generated ECG interpretations are frequently erroneous and can cause unnecessary patient anxiety and inappropriate hospital admissions and consultations. Knowledge to interpret real or perceived abnormalities of the ECG is necessary to make appropriate clinical decisions as to management or appropriate cardiology referrals.

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