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REVIEW
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Common Supraventricular and Ventricular Arrhythmias in Children

Hasan Candaş Kafalı[1], Yakup Ergül[2]

[1]Department of Pediatric Cardiology, Pediatric Cardiologist, University of Health Sciences, İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey
[2]Department of Pediatric Cardiology, Pediatric Cardiac Electrophysiologist, University of Health Sciences, İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

ABSTRACT

The most common pediatric arrhythmias are tachycardias, and the most common type is supraventricular tachycardia, originating from or above the atrioventricular node and HIS bundle. Ventricular tachycardias are less common but more dangerous. Supraventricular tachycardias usually cause a narrow complex tachycardia unless there is a basal bundle branch block or rate-dependent aberration. A wide QRS tachycardia should be treated as ventricular tachycardias unless proven to be an supraventricular tachycardia with aberration. Diagnosis of both tachyarrhythmia types depends mainly on 12-lead electrocardiography. The most common supraventricular tachycardia type in newborns and infants is atrioventricular reentry tachycardia, related to manifest or concealed accessory pathways and in adolescent atrioventricular nodal reentry tachycardia, whereas focal atrial tachycardias consist of 10%-15% of supraventricular tachycardias during all ages. Supraventricular tachycardias have a low risk of morbidity, and ablation therapy is successful in most types with success rates over 90%. Ventricular tachycardias can be monomorphic or polymorphic, nonsustained or sustained, and can cause more hemodynamic instability than supraventricular tachycardias, requiring more close monitoring and urgent therapies. If hemodynamically unstable, synchronized cardioversion must be performed. Polymorphic ventricular tachycardias are very dangerous and often associated with primary ion channel defects (channelopathies), which can cause sudden cardiac death.

Keywords: Supraventricular tachycardia, ventricular tachycardia, pediatric arrhythmia

INTRODUCTION

Pediatric arrhythmias are essential in pediatric cardiology practice and are of interest to pediatric electrophysiologists after the new diagnosis and treatment methods in recent years. However, with basic diagnostic methods, especially electrocardiography (ECG), pediatricians should be able to guide the diagnosis and treatment of pediatric arrhythmias, which they frequently encounter in their clinics.

Pediatric arrhythmias are generally classified under 3 headings: tachyarrhythmias, bradyarrhythmias (sinus node dysfunction, and atrioventricular (AV) blocks), and primary ion channel defects (channelopathies) with sudden cardiac arrest and death (SCD).[1,2] In this review, tachyarrhythmias, which are frequently seen in the pediatric clinic, will be discussed.

It should be noted that the normal heart rate values in children decrease with increasing age and approach adult values in adolescents (Table 1). Tachyarrhythmia means that the heart rate is above the average values determined according to the patient's age.

Sinus arrhythmia, nodal rhythm, ectopic atrial rhythm, and wandering atrial pacing activity ("wandering atrial pacemaker") are normal rhythm variants that can be seen in 15%-25% of healthy children and do not require further examination and treatment (Figure 1).[1,2]

Corresponding author:
Yakup Ergul
✉yakupergul77@gmail.com
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Table 1. Normal Heart Rates for Age in Children (3)	
Age	**Lower and Upper Limits (bpm)**
0-3 months	110-180
3-12 months	100-170
1-2 years	100-150
2-5 years	95-140
5-12 years	80-120
12-18 years	60-100
bpm, beat per minute.	

SUPRAVENTRICULAR ARRHYTHMIAS

Premature Atrial Contractions

Premature atrial contractions (PACs) are premature beats originating from an ectopic atrial focus above the AV node/HIS bundle. Mostly seen as a narrow QRS complex following an abnormal p wave, sometimes a wide QRS complex may develop due to aberration because of refractoriness of one of the main branches under the HIS bundle. Sometimes even the hole HIS bundle may be refractory, so the ectopic beat will not be conducted to ventricles, called a "non-conducted" PAC (Figure 2A and B). Although it is more common in newborns and infants, it is seen at rates up to 50% throughout childhood.[1,2]

Premature atrial contractions are usually asymptomatic and are discovered incidentally during a routine examination. However, they may also occur in accordance with predisposing factors such as myocarditis, sympathomimetics, digitalis, tricyclic antidepressants, caffeine intake, central venous catheters, hypokalemia, hypercalcemia, hypoxia, and hypoglycemia. Especially in the neonatal period, 2 : 1 non-conducted PACs may cause bradycardia and can be confused with AV blocks since they are often associated with sinus node reset. Premature atrial contractions generally do not require further examination and treatment.[1,2]

Sinus Tachycardia

Sinus tachycardia is a narrow QRS tachycardia with p waves identical to sinus p waves, originating from the sinus node (Figure 3A). Sinus tachycardia can be considered a physiological response to events such as exercise, pain, anxiety, and fever. However, it can also be a response to nonphysiological conditions like anemia, hyperthyroidism, hypovolemia, and various drugs or substance abuse (beta-agonists such as adrenaline, isoproterenol, dobutamine, and salbutamol, sympathomimetics such as amphetamine, methylphenidate, also atropine, antihistamines, caffeine, theophylline, and marijuana). Unlike paroxysmal supraventricular tachycardias (SVTs), there is usually a gradual onset and termination, and the tachycardia rate is not constant. In cases other than the physiological sinus tachycardia response, treatment should be directed toward the underlying cause.[1,2,4]

Supraventricular Tachycardias

Supraventricular tachycardias originate from the atrium or AV node/HIS bundle. Heart rate during SVT is 200-300 bpm in infants and 180-250 bpm in older children[1,4] (Figure 3B). The term "paroxysmal" is used to describe abrupt onset and end.

Supraventricular tachycardia is the most common type of arrhythmia seen in children. Its incidence has been reported as 13/100 000 and its prevalence as 2.25/1000. It is usually sporadic, and SVT frequency in first-degree relatives has been reported as 5.5%-7%.[1,2,4,5]

The first episode of SVT in children is diagnosed in the first year of life in 50%-70% of cases. There is no recurrence in 30%-50% of cases after 18 months. On the contrary, if the first symptomatic attack occurs later, the probability of spontaneous disappearance is low.[1,4]

The patient's age is an essential factor in the clinical findings. Congestive heart failure findings develop in half of the patients due to newborns' and infants' high heart rates (220-280 bpm). In 20% of cases, it can be detected incidentally during a routine examination without any complaints. Palpitation, chest pain, abdominal pain, pallor, and sweating are among the most common symptoms in older children, while syncope and heart failure are rare.[4,5] Other factors determining the clinic are the duration of the tachyarrhythmia (insidious SVTs that continue at relatively slow rates for a long time may cause

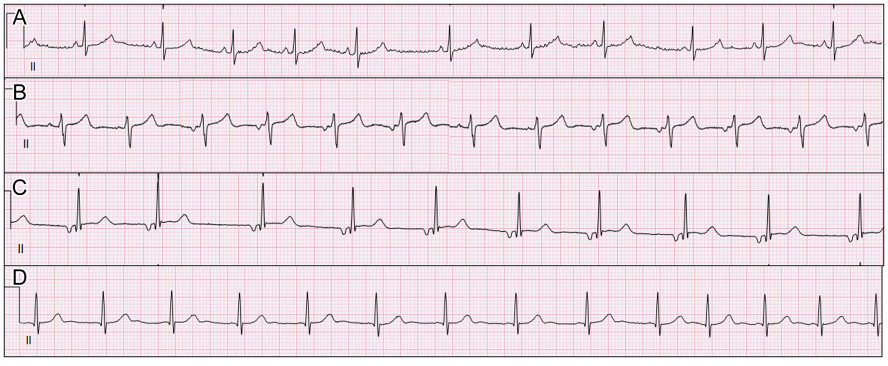

Figure 1. Normal rhythm variants that can be seen in healthy children. (A) Sinus arrhythmia ("respiratory arrhythmia") in a 14-year-old boy athlete (normal sinus p waves with regular fixed P-R interval and cyclic and gradually lengthening and shortening of P-P intervals causing the heart rate to increase with inspiration, and decrease with expiration, but always within normal limits) with unchanged morphology are seen. (B) Wandering atrial pacemaker activity seen intermittently during daytime in a 4-year-old female patient (R-R distances are generally equal, as in normal sinus rhythm, with gradually changing p-wave morphology). (C) Ectopic atrial rhythm in a 9-year-old female patient at a rate similar to the sinus rate, mainly occurring at night (originates from a different atrial focus than sinus node, with a rate close to the average sinus rate). (D) Intermittent nodal rhythm seen in a 12- year-old male patient at night (the same narrow QRS as sinus rhythm, without any P waves in front of it).

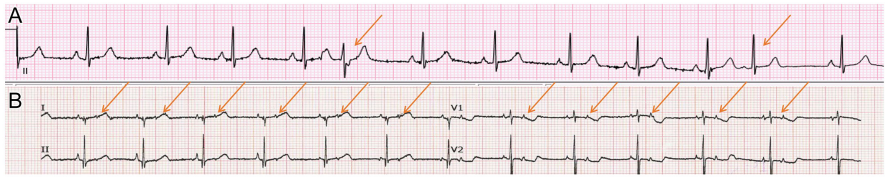

Figure 2. (A) Premature atrial contractions (PACs) with aberrant and normal conduction to ventricle shown with red arrows. (B) Electrocardiography at the border of bradycardia (100 bpm) in a newborn with non-conducted bigeminy PACs.

"tachycardia-induced cardiomyopathy" (TIC)) and the underlying heart disease.[4,6] The mortality rate in SVTs is low (1% in those with concomitant heart disease, 0.25% if the heart is structurally normal).[2]

Twelve-channel ECG recording is the primary tool for the diagnosis of SVT. Although, in most cases, a narrow QRS complex tachycardia is encountered on ECG (Figure 3B), sometimes a wide QRS complex tachycardia can also be seen due to a bundle branch block found at baseline or as a result of high-rate related aberration. The ECG during an attack contains many essential clues for differential diagnosis of SVT (Figure 4). Basal ECG is generally normal, except in manifest ventricular preexcitation.[5,7]

Twenty-four-hour ambulatory Holter ECG monitoring and event recorder devices, which provide rhythm monitoring for up to 15-30 days, are used to detect more rare SVT attacks. Blood count, electrolytes, and thyroid tests are recommended for laboratory investigations. Telecardiography and echocardiography are recommended, especially in patients with congestive heart failure (CHF) findings and to exclude structural heart disease.

As a rule, medical treatment is not recommended for palpitations without a documented SVT. If an SVT has been documented before, especially if it recurs, antiarrhythmics are recommended as medical treatment.[1,4] If SVT is detected during an acute palpitation attack, the patient's general condition and hemodynamic status are evaluated first. Today, electrophysiologic study (EPS) and ablation therapy are leading treatment options that provide permanent treatment in almost all types of SVT.[5-7]

Acute Medical treatment in SVT: The first thing to do in a patient presenting with SVT should be to monitor the patient quickly and to determine the hemodynamic status (perfusion defect, hypotension, heart failure or shock findings, and acute-altered consciousness) while recording a 12-channel ECG. If the hemodynamic status is stable, the first treatment should be vagal stimulus (ice bag/cold application to the face and other valsalva maneuvers; 20%-40% effective). If vagal stimulation is ineffective, the first-line drug is IVadenosine[1,4,7] (Table 2).

Adenosine is the first choice in the acute medical treatment of SVT. It provides acute treatment by terminating reentrant SVTs containing AV node through the temporary AV block. Although it cannot terminate arrhythmia in other SVT types, such as automatic atrial tachycardias and atrial flutter/fibrillation, the p waves appear obviously during the temporary AV block, helping diagnose these SVT types. Since the half-life is very short (5-7 seconds), in order to be effective, it should be administered by a central venous route as close as possible to the heart and administered as a rapid push and undiluted, followed by sufficient saline washing. As a first dose, 100 μg/kg is recommended, up to 200-300 μg/kg doses in case of no response (maximum 6 mg for the first dose and maximum 12 mg for the

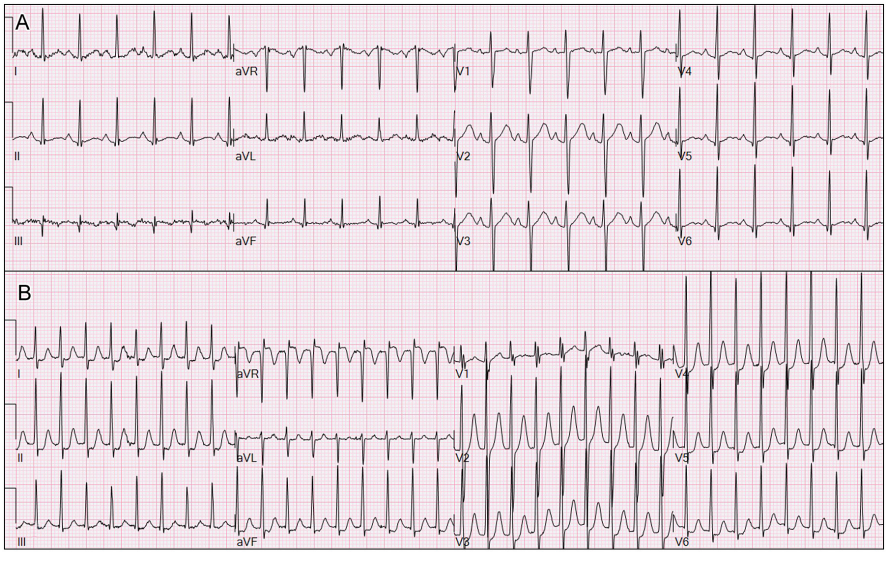

Figure 3. (A) Sinus tachycardia at a rate of 150/min in a 14-year-old male patient; p waves are the same as sinus p waves. (B) A 12-year-old female patient with narrow QRS tachycardia, where p waves can not be discriminated, and at a rate of 210 bpm.

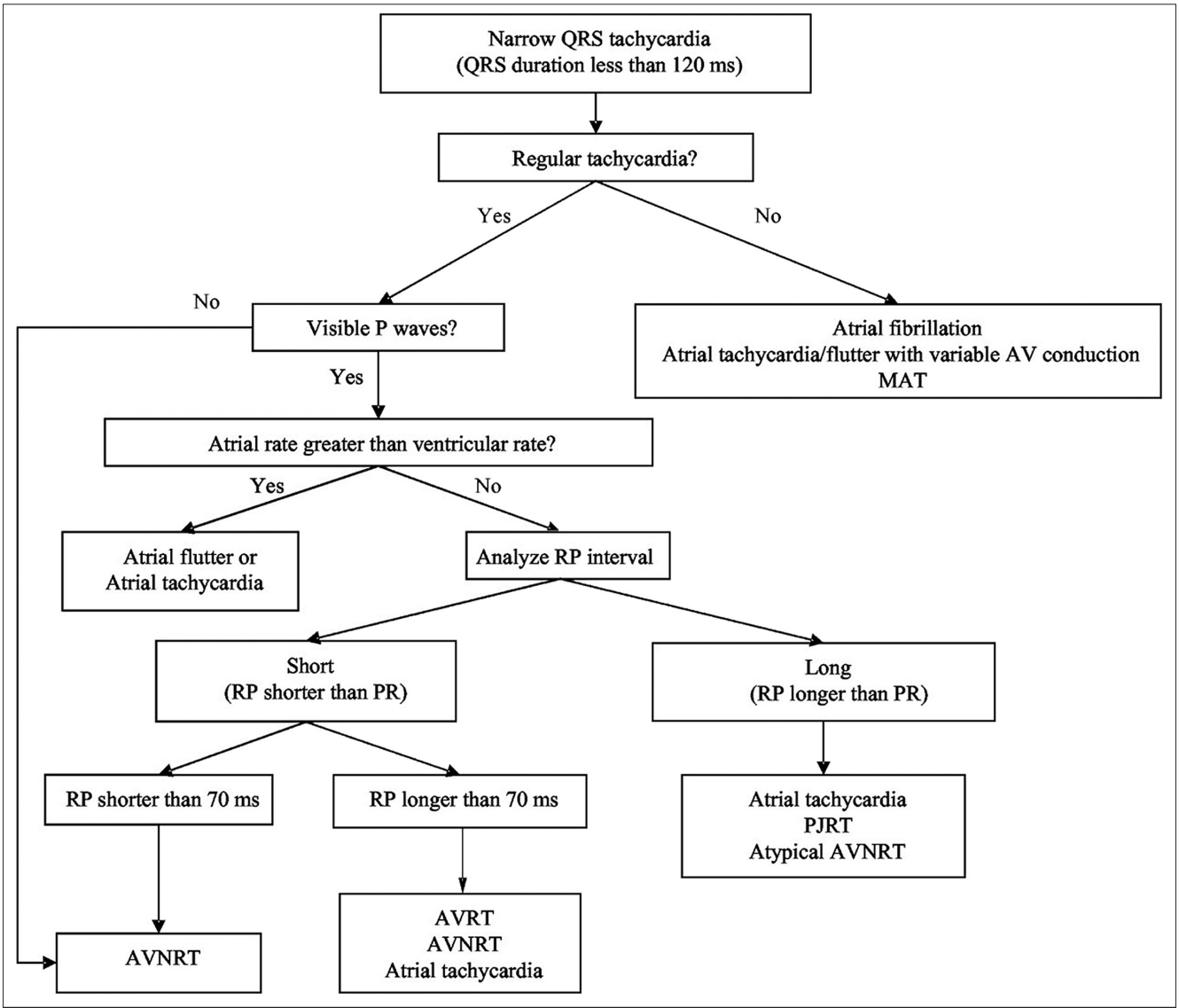

Figure 4. Differential diagnosis of narrow QRS-complex tachycardias (6).

Table 2. Recommendations for Acute Treatment of Hemodynamically Stable Regular Narrow QRS Tachycardia in Infants and Children (1)

Drug/Intervention	Dosage (iv)	Class	Level
Vagal manuevres	Ice immersion, gastric tube insertion in infants, Valsalva, and headstand in older children	I	B
Transesophageal atrial overdrive pacing[a]		I	B
Adenosine	Rapid bolus starting dosages: For infants: 0.15 mg/kg. For .1 year of age: 0.1 mg/kg Increasing dosage up to 0.3 mg/kg	I	B
Verapamil[b,c]	0.1 mg/kg slowly over 2 minutes	I	B
Flecainide[b]	1.5-2 mg/kg over 5 minutes	IIa	B
Propafenone[b]	Loading: 2 mg/kg over 2 hours. Maintenance: 4-7 μg/kg/min	IIa	B
Amiodarone	Loading: 5-10 mg/kg over 60 minutes. Maintenance infusion: 5-15 μg/kg/min	IIb	B

[a]Most effective if AV reentrant tachycardias or atrial flutter.
[b]Myocardial depressant effect.
[c]Contraindicated in infants<1 year of age. iv, intravenous; class, recommendation class; level, level of evidence.

second and subsequent doses). The effect of adenosine should be documented by recording continuous ECG while giving it.[1,4]

The most common side effects of adenosine are shortness of breath, chest discomfort, and flushing. Care should be taken in patients with a history of allergy (acute bronchospasm), patients with borderline cardiac reserve (SCD due to coronary steal syndrome), and patients with Wolff-Parkinson-White (WPW) syndrome (induction of atrial fibrillation and causing rapid conduction to the ventricle, which in turn degenerates to ventricular fibrillation).[5,7,8]

If it is not possible to stop SVT with adenosine, esmolol (50-200 µg/kg bolus in 10 minutes, and 50-100 µg/kg/min infusion for maintenance dose; normal cardiac functions as a prerequisite), amiodarone (5 mg/kg loading dose in 1 hour, 10-15 mg/kg/day infusion for maintenance dose; requiring close blood pressure monitoring), or verapamil (0.1 mg/kg IV bolus, can be repeated up to max. 5 mg dose; not used under 1 year old because of cardiac depressant effects) can be used.[4,7]

Synchronized direct current (DC) cardioversion (1-2 J/kg) should be the first-choice treatment in hemodynamically unstable patients.[7,8]

Chronic Treatment in SVTs: The most crucial disadvantage of chronic treatment is the frequent side effects of the medications. Treatment should be stopped after a few years, except for risky patients, and whether arrhythmia persists, or its characters are altered should be evaluated because arrhythmia symptoms may change or even disappear as children get older.[5-7]

EPS and Ablation in the Treatment of SVTs: It is the treatment of choice nowadays, even in arrhythmias controlled by chronic drug therapy, and can be used in a significant portion of SVTs. Although success rates vary depending on the arrhythmia substrate, it is around 90% in patients with SVT. When the focus causing SVT is detected during EPS, this area is reached with special catheters, and ablation can be performed.[8,9,10]

Atrioventricular Reentrant Tachycardia (Concealed or Manifest Accessory Pathway-Dependent SVTs): In the presence of the manifest accessory pathway (AP), the ventricles are prematurely preexcited through the AP, which conducts the impulse more rapidly than the AV node. Thus, the first part of the QRS is a sloping delta wave and, consequently, a short PR interval and wide QRS complex, called WPW preexcitation, is the most common type of manifest preexcitation (Figure 5A).

The prevalence of the disease is found as 1-3/1000 children in large series of pediatric and adult studies. Atrioventricular reentrant tachycardia (AVRT) is the most common SVT type seen in newborn and infants. The frequency of AVRT decreases in 90% of patients after the first year of life, but tachycardias recur in approximately 30% of patients at the age of 7-8 years. Antegrade AP disappears spontaneously in 40% of WPW patients in the first year of life. In WPW patients with fast conducting APs, there is a risk of SCD after rapid antegrade conduction during atrial fibrillation, which can degenerate to ventricular fibrillation (Figure 5B). SCD rate has been reported at around 0.6% in various studies. The annual risk of sudden death in symptomatic WPW patients is 0.25%, and the lifetime risk is 3%-4%.[11,12] The most definitive way of risk assessment is the EPS.

Mostly an orthodromic tachycardia (antegrade conduction through the AV node and retrograde through AP; resulting in narrow QRS tachycardia) and rarely an antidromic tachycardia (antegrade conduction through the AP and retrograde through the AV node; resulting in wide QRS tachycardia) can be seen. Digoxin and calcium channel blockers, which increase accessory conduction by shortening the effective refractory period of the AP, are contraindicated in pre-excited patients because they can cause rapid ventricular conduction during atrial fibrillation. In the acute phase, adenosine bolus should be performed only in places where defibrillator and emergency equipment are available, as it increases the passage through the AP by AV blocking. Adenosine should not be given in a case of irregular wide QRS tachycardia (considering it could be an antidromic tachycardia accompanied by atrial fibrillation). Catheter ablation should be performed to permanently treat the AP in patients with a history of syncope and severe SVT who meet the intermediate and high-risk criteria in EPS. According to the latest guidelines, since there may be a risk of sudden death in asymptomatic pre-excitation, ablation is recommended in children older than 5 years and over 15 kg, considering the risk.[12]

Some APs only work retrogradely (concealed AP). They can only cause orthodromic tachycardia, and there is no increased risk of sudden death as they do not allow antegrade conduction.

Permanent junctional reciprocal tachycardia (PJRT) is a rare long RP tachycardia caused by a concealed AP with decremental conduction features and is usually localized in the right posteroseptal region (Figure 5C). It tends to be incessant and may cause TIC. Ablation is recommended in symptomatic cases.[13,14]

Atrioventricular Nodal Reentrant Tachycardia

Atrioventricular nodal reentrant tachycardia (AVNRT) is the most common type of SVT in older children and adolescents and is more common in girls.[15] Atrioventricular nodal reentrant tachycardia results from a re-entering loop using extensions within or immediately adjacent to the atrioventricular node. The circuit usually includes 2o paths, called "fast" and "slow," with different conduction rates and refractory periods. In the more commonly seen typical AVNRT, where antegrade conduction is through the slow pathway and retrograde is through the fast pathway, atrial and ventricular contractions will occur near-simultaneously, resulting in tachycardia with a very short RP (<70 ms) interval (Figure 5D). In atypical AVNRT, the descending pathway is the fast pathway, and the ascending pathway is the slow pathway. Therefore, the RP interval is long, and a differential diagnosis with other long RP tachycardias such as PJRT and focal atrial tachycardia (FAT) is needed (Figure 5E). In typical AVNRT, P waves may not be seen when the P waves coincide with the QRS complex due to almost simultaneous contraction of the atria and ventricles. If it appears, it can be seen as a slight deflection at the end of the QRS, causing "pseudo-S" in the inferior leads

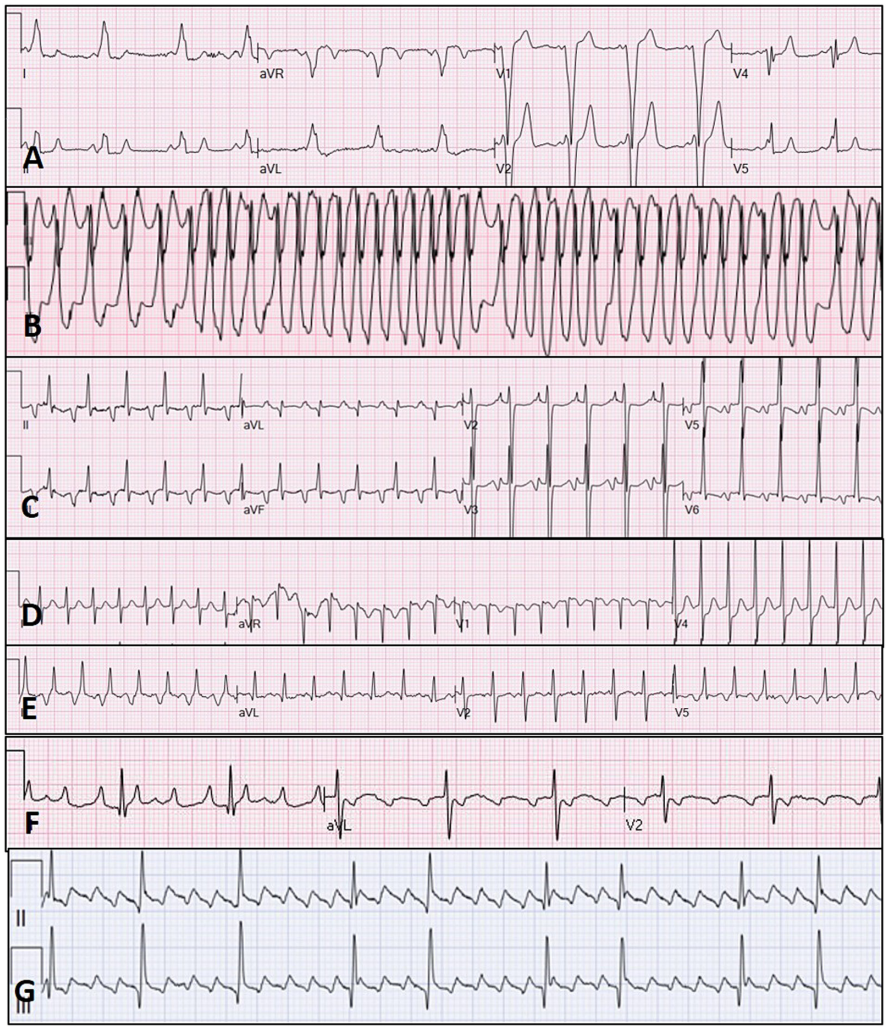

Figure 5. (A) Manifest pre-excitation pattern consisting of short PR interval, delta wave, and wide QRS tirade on 12-channel electrocardiography (ECG); Wolff-Parkinson-White (WPW) pre-excitation. (B) Fast conduction to ventricles of atrial fibrillation in WPW, which can degenerate to ventricular fibrillation. (C) Permanent junctional reciprocal tachycardia (PJRT) recording with long RP interval and negative P waves in the inferior leads (DII-III and aVF) are seen. (D) Very short RP interval in ECG (retrograde p waves seen immediately after the QRS as "pseudo-r" in lead V-1 and a "pseudo-s" in lead D-III) seen in typical AVNRT. (E) Atypical AVNRT ECG with long R-P interval is seen (differential diagnosis includes other long R-P interval SVTs such as focal atrial tachycardia and PJRT). (F) Focal atrial tachycardia ECG; p waves seen on the ground after adenosine administration, returning to the isoelectric line. (G) Atrial flutter ECG with typical saw-tooth-like p waves.

and "pseudo-R" in V1. Vagal maneuvers, adenosine bolus, and IV beta-blockers can be used in acute treatment. Catheter ablation provides a permanent solution in recurrent patients, especially those over 5 years. Cryoablation is a good option in children because it is safe and does not have the risk of permanent AV block.[16]

Focal Atrial Tachycardia

It constitutes 11%-16% of SVTs in children and is the most common cause of incessant tachycardias and TIC. Starting from a focus different than the sinus node, the tachycardia propagates centrifugally into the atria. Since it is not an AV node-dependent SVT, in the ECG, a long RP tachycardia with a p-wave axis and morphology different from the sinus p-wave is seen. Sometimes AV dissociation or irregular R-Rs can be seen. It is mostly unresponsive to adenosine (Figure 5F). After it starts, FAT gradually accelerates (warm-up) and slows down to stop (cool-down).[1,4,8]

Focal atrial tachycardias are usually refractory to medical therapy. In stable patients, intravenous beta-blockers can be used, and intravenous group III antiarrhythmic agents can be used in unresponsive cases. It often requires combination therapy. If resistant, ventricular rate control with beta-blockers should also be considered. Catheter ablation is the first choice in resistant cases.

Atrial Flutter

It occurs with a macro-reentering mechanism independent of the AV node, where the atrial rate can be between 250 bpm and 400 bpm. Due to the high atrial rates, no isoelectric line can be seen between the P waves, a typical sawtooth appearance peculiar to flutter (Figure 5G).[1,17]

Incisional macroreentrant tachycardias arising around the scar tissue formed from previous cardiac surgery are called intraatrial reentry tachycardia (IART). The rate in IART is

slightly lower than in atrial flutter. An isoelectric line may be found between the P waves on the EKG.

Synchronized cardioversion (CV) is used in the acute treatment of atrial flutter. If the duration of flutter or IART is greater than 48 hours or the duration is unknown, and the hemodynamics is stable, DC CV or medical CV such as amiodarone is not performed. Before this, the presence of any thrombus should be checked with transthoracic and transesophageal echocardiography. Cardioversion can be tried after 3 weeks of anticoagulation therapy in cases with longstanding IART or in whom thrombus is detected by echocardiography. Beta-blockers or calcium channel blockers can be used to reduce rapid ventricular conduction for rate control during these 3 weeks. In chronic treatment, catheter ablation is the first option.[7,8,17]

VENTRICULAR ARRHYTHMIAS

Premature Ventricular Contractions

Premature ventricular contraction (PVC) is characterized by a premature wide QRS complex (frequently with a compensatory pause) with an abnormal T wave (usually opposite the QRS direction) and without a preceding P wave on the ECG. Its incidence in healthy children has been reported to be between 0.3% and 2.2% in routine ECG.[1,2,18]

Premature ventricular contractions are called uniform if they have the same morphology (emerging from the same ectopic focus) and multiform (from different foci) if they are of different morphology.

Although isolated PVCs are usually asymptomatic, they can sometimes lead to palpitations, chest pain, and if frequent, impaired cardiac function. The presence of concomitant structural heart disease or concomitant VTs and symptoms such as exercise-related syncope in the history should be investigated carefully. Twelve-channel ECG, Holter, and if age-appropriate, exercise testing should be included in the initial evaluation. In addition, the presence of predisposing heart diseases such as congenital heart disease, mitral valve prolapse, arrhythmogenic right ventricular dysplasia (ARVD), cardiomyopathies, cardiac tumors, coronary artery anomalies, and myocarditis (including COVID-19) should be investigated with echocardiography.[1,18,19]

Premature ventricular contractions are considered "benign" if morphologically uniform, disappear with exercise, and have no underlying structural heart disease or channelopathy (Figure 6A). Treatment is usually unnecessary. However, if PVCs are frequent or in couplet- or triplet-form, follow-up is recommended (Figure 6B).

Premature ventricular contractions with concomitant structural heart disease, syncope in the history, or sudden death in the family, PVCs increasing with exercise, PVCs as frequent 3-5 "Salvo" beats (Figure 6C), and PVC morphologies with poor prognostic features (R on T phenomenon, multifocal origin, short coupling interval, interpolated PVC) are considered "malignant" (Figure 6D-F), and follow-up and treatment in a Pediatric Cardiology/Arrhythmia center are recommended.[18,19]

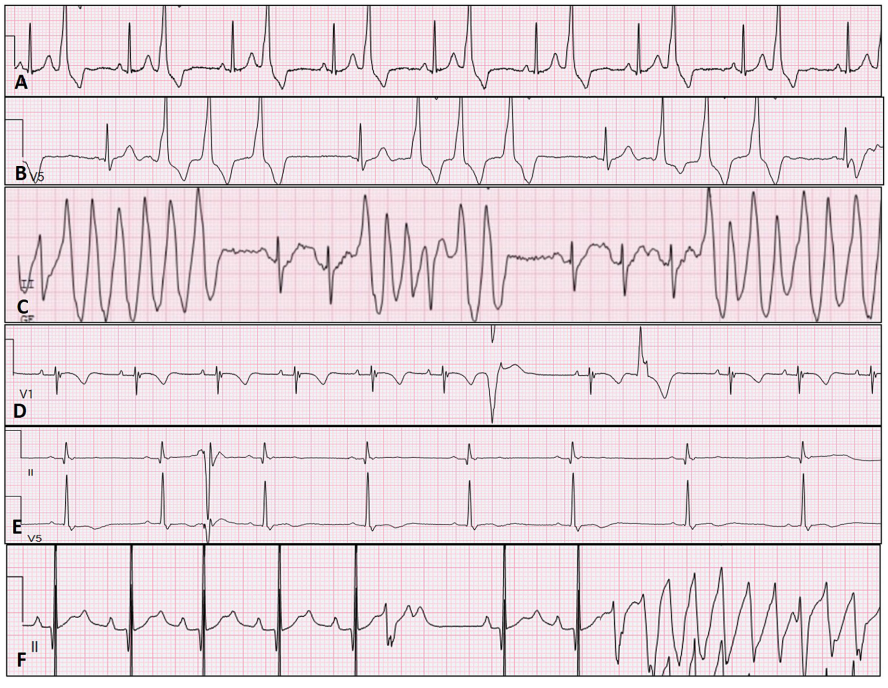

Figure 6. (A) Premature ventricular contractions (PVCs) uniform in morphology and "bigeminy" in frequency, without any other poor prognostic features in the examination, considered "benign." (B) Triplet monomorphic PVCs (also called ventricular tachycardia (VT)) at a slow rate. (C) Four to six PVCs in polymorphic "Salvo" form, requring careful evaluation. (D) Multiform (polymorphic) PVCs in 2 different morphologies. They became more frequent and finally evolved into polymorphic VT in the exercise test and are considered "malignant." (E) An interpolated PVC requires more attention, as the probability of coinciding with the T wave in front of it increases ("R on T" phenomenon). (F) Another PVC with a malignant feature; a very short coupling interval causes the "R on T" phenomenon, initiating polymorphic VT.

Ventricular Tachycardias

Ventricular tachycardias (VTs) are less common than SVTs, with an incidence of 1/100 000 in the general pediatric population and a prevalence of 2-8/100 000 in school age. On Holter ECG monitoring, PVCs or short VT attacks can be detected in 1%-5% of healthy adolescents. As with PVCs, there are VTs that have a "benign" character and do not require treatment and those with severe hemodynamic consequences and require an aggressive approach.[18,20,21]

Ventricular tachycardia can be defined as tachycardia originating from the ventricular myocardium and conduction system under the atrioventricular node/His bundle. Electrocardiographically, ≥3 PVCs with a rate 20%-25% faster than the basal sinus rate or >120 bpm can be defined as VT. If the rate is equal to the basal sinus rate, it is called an "idioventricular rhythm," and if the rate is 10%-20% higher than the sinus rate is called an "accelerated idioventricular rhythm."[20,22]

Ventricular tachycardia can be morphologically monomorphic (each QRS complex is identical and the rhythm is regular) or polymorphic (multiple different QRS morphologies are present, and the rhythm is irregular). It can be "non-sustained" (<30 seconds) or "sustained" (≥30 seconds)

Most VTs occur against the background of an identifiable cause or structural heart disease. Ventricular tachycardia's acute and chronic causes are listed in Table 3.[2,18,23,24]

Clinical manifestations vary widely according to the rate/duration of VT and the presence of underlying structural heart disease. While the patient may be asymptomatic at diagnosis, palpitations, dizziness, pre-syncope, chest pain, and shortness of breath may be observed. Incessant VTs can cause TIC. The most severe and rare clinical finding is SCD (mainly seen in primary ion channel diseases).[24-26]

The presence of any cause listed in the Table 2 in the patient's history, arrhythmia, cardiomyopathy, relatives with an implantable cardiac defibrillator (ICD) or sudden/early death in family history, and signs of accompanying congenital heart disease or cardiomyopathy in the physical examination should be investigated. Serum calcium, potassium, magnesium levels, thyroid function tests, troponin-T for myocarditis, and toxicological tests should be done if necessary.

In the case of wide QRS tachycardia, ECG findings supporting VT are AV dissociation with V>A, intermittent fusion and sinus capture beats (Figure 7A and B), VT morphology similar to the background PVC, positive or negative concordance in the chest leads and "northwest" QRS axis.[19,25]

Twenty-four-hour Holter ECG monitoring can determine the frequency of VT attacks and evaluate the response to treatment, while exercise test can be used to diagnose exercise-triggered VTs (CPVT, "triggered" right ventricular outflow tract VTs). Echocardiography should be performed in every patient to evaluate cardiac functions and investigate concomitant structural cardiac defects, cardiomyopathy, and tumors. Electrophysiological study can be performed mainly in patients with congenital heart disease (CHD) (±surgery), some CMP and ion channel diseases, and unexplained life-threatening syncopes. Depending on the arrhythmia induced during EPS, ablation or ICD implantation can be decided in the same session.[20-26]

Any wide QRS tachycardia should be considered VT until proven otherwise, and synchronized cardioversion (1-2 j/kg,

Table 3. Acute and Chronic Causes of Ventricular Tachycardias

Acute Causes (Mostly Temporary/Treatable)		**Chronic Causes**	
Metabolic*	Hypoxia	***Congenital****	*Tetralogy of fallot*
	Acidosis		*Coronary anomalies*
	Hypoglycemia		*Mitral valve prolapse*
	Hypokalemia	***Cardiomyopathy (CMP)****	*Dilated CMP*
	Hypocalcemia		*Hypertrophic KMP*
	Hypomagnesemia		*Aritmogenic KMP*
Iscemic	Coronary anomalies		*Left ventricular noncompaction*
	Kawasaki disease	***Channelopathies****	*Long QT syndrome*
Traumatic	Cardiac surgery		*Catecholaminergic polymorphic VT*
	Trauma		*Brugada syndrome*
Infective	*Myocarditis**	**Acquired**	*Cardiac tumors**
	Rheumatic fever	**Idiopathic**	Idiopathic outflow VTs • Right/left ventricule • Aortic sinus valsalva • His-bundle
Toxic*	Drugs (digoxin, tricyclic antidepresssants, sotalol, flecainid, phenothiazines)		Idiopathic left ventricular VT • Left posterior/anterior fascicular
	Substance abuse ("bonzai," cocain and heroin)		Others • Mitral–tricuspid annulus

*Most frequently seen.

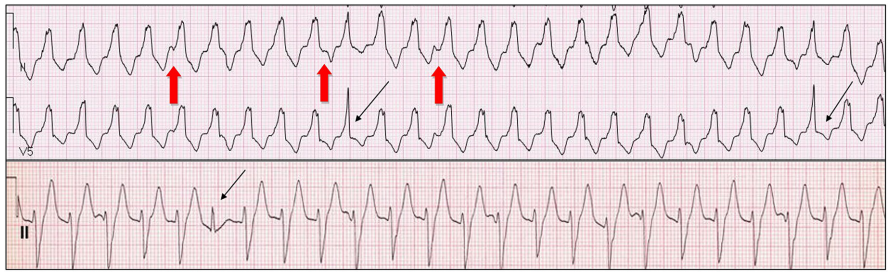

Figure 7. (A) Atrioventricular dissociation in the form of p waves (thick red arrows) that can be distinguished from time to time between the QRS and T waves, and 2 fusion beats (thin black arrows) in the middle and near the end of the ECG, characterized by a narrower QRS than the VT but wider than the normal QRS. (B) Atrioventricular dissociation and a "capture" beat at the sixth beat from the beginning (in normal sinus QRS morphology).

under sedation) should be performed if it causes hemodynamic compromise. Underlying treatable causes (such as electrolyte disorders, hypothermia, hypovolemia) can be corrected, and drug therapy can be initiated. Amiodarone (5 mg/kg IV with a max. 300 mg given in 20-60 min) or, if available, procainamide IV can be given. In addition, lidocaine can be given as a 1 mg/kg IV bolus followed by a maintenance infusion dose of 20-50 µg/kg/min.[1,18,19]

In a wide QRS tachycardia that does not cause hemodynamic impairment, at first 12-channel ECG should be taken to differentiate between SVT and VT (Figure 8). If ECG cannot differentiate it, vagal maneuvers or adenosine IV can be used. Procainamide IV is the first choice in treatment. If not available, esmolol, lidocaine, and amiodarone IV are recommended. If drug therapy fails, synchronized cardioversion should be performed[1,18] (Table 4).

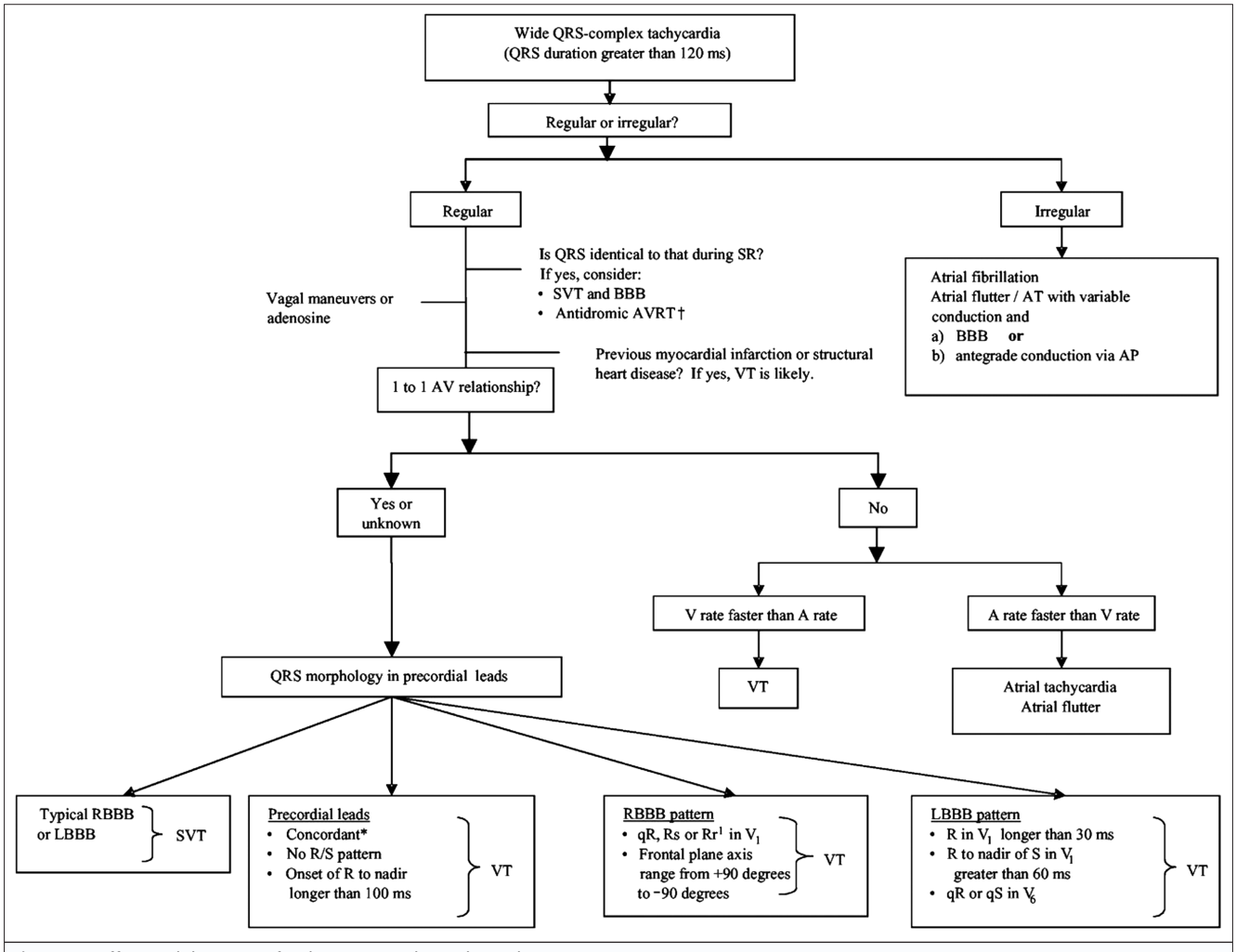

Figure 8. Differential diagnosis of wide QRS-complex tachycardia (6).

Table 4. Recommendations for Acute Treatment of Wide QRS Tachycardia in Infants and Children (1)

Wide QRS Tachycardia	Drug/Intervention (Dosages See Table 1).	Class	Level
Wide QRS tachycardia of unknown mechanism	Electrical cardioversion	I	C
	Lidocaine iv bolus starting at 1 mg/kg (up to 3 doses in 10 minute interval); followed by infusion of 20-50 μg/kg/min	IIa	C
	Amiodarone iv loading: 5-10 mg/kg over 60 minutes, followed by maintenance infusion of 10 mg/kg/day (5-15 μg/kg/min).	IIb	
	Procainaimide iv	IIb	
	Esmolol iv bolus 500 μg/kg	IIb	
	Magnesium sulphate iv	IIb	
Antidromic tachycardia, pre-excited AF	Electrical cardioversion	I	B
	Flecainide iv	IIa	C
SVT with bundle branch block	See **Table 2** for acute treatment of SVT		
Monomorphic ventricular tachycardia	Electrical cardioversion	I	C
	Propranolol iv	IIb	C
	Lidocaine iv		
	Sotalol iv		
Polymorphic ventricular tachycardia	Electrical cardioversion	I	C
	Propranolol iv	IIb	C
	Deepsedationor general anesthesia	IIb	C
	Potassium and magnesium iv.	IIb	C

iv, intravenous; Class, recommendationclass; Level, level of evidence; AF, atrialfibrillation; SVT, supraventricular tachycardia.

Ablation indications are being symptomatic (chest pain, shortness of breath, pre-syncope, or syncope), frequent PVC/VT, drug-resistant VT, refusal of medication, and VT that causes deterioration of heart functions.[4,7]

Three frequently seen and well-known "benign" types of monomorphic VTs in childhood (in the presence of a structurally normal heart) are "incessant" idiopathic infantile VT, idiopathic right ventricle (RV) outflow tract VT, and idiopathic left ventricle (LV) posterior fascicular VT.[27,28] Monomorphic rapid VTs, on the other hand, are more malignant as they rapidly disrupt hemodynamics and require more advanced and invasive treatments. Polymorphic VTs are considered malignant because they originate from different foci and generally progress rapidly. Two known typical examples are "bidirectional" VT and "Torsades de Pointes (TdP)." They are primarily associated with primary ion channel defects.

Idiopathic RIght Ventricle Outflow Tract Ventricular Tachycardia

It is the most common type of VT in children and adolescents. Delayed after-depolarization due to triggered activity is the mechanism (increase in the amount of cAMP with beta-adrenergic stimuli such as exercise and anxiety).[18,19,27] It can be terminated by vlsalva maneuver, adenosine IV, beta-blockers, or calcium channel blockers. In 12-channel ECG, VT morphology is typically in the left bundle branch block pattern with an inferior axis (Figure 9A). Nowadays, the ablation method replaces medical treatment in suitable patients.

Idiopathic Left Ventricle Posterior Fasicular Ventricular Tachycardia

It is also known as the "Verapamil-responsive idiopathic LV." It is mainly seen in men aged between 15 and 40 years. The reentry mechanism in the left branch posterior fascicle is the responsible mechanism.[18,29] In the 12-channel ECG, the VT morphology typically has a right bundle branch block pattern and superior axis (Figure 9B). Since VT partially uses the HIS-Purkinje system, the QRS is relatively narrow compared to other VTs. Mostly, there is no structural heart disease, except accessory bands in the left ventricle. It is sensitive to IV verapamil and may also be terminated with IV adenosine. While verapamil constitutes the primary option in medical treatment, ablation by transcatheter route gives very successful results.

"Bidirectional" Ventricular Tachycardia

It is a particular type of polymorphic VT with alternating QRS morphology. It shows right-left axis deviation due to QRS complex alternans. They are primarily seen with exercise and fear-related syncope. Three typical causes are CPVT (including Andersen Tawil syndrome), digoxin toxicity, and some types of ARVD.[1,2,30]

Catecholaminergic polymorphic ventricular tachycardia: It is a rare primary ion channel disease that causes syncope and SCD, characterized by bidirectional VES/VT (polymorphic) on exertion or sudden excitement (Figure 9C) and without any accompanying structural heart disease. The frequency is 1/10 000. Family history is of sudden death, syncope, and treatment-resistant seizures-epilepsy under the age of 40 should be investigated. Mortality up to 30 years of age in symptomatic patients is 30%-50%.[31,32,33]

The disease is frequently inherited in autosomal dominant pattern (Ryanodine Receptor-2; RyR2 mutation) and less frequently autosomal recessive pattern (Calsequestrin-2; CASQ2) is observed (both mutations in 65% of all cases).

The most common symptom is syncope triggered by exercise or emotional stimulation (53%-80%). It usually starts between the ages of 7 and 12 years. They are often misdiagnosed as "epilepsy." In approximately 30% of patients, the first symptom is

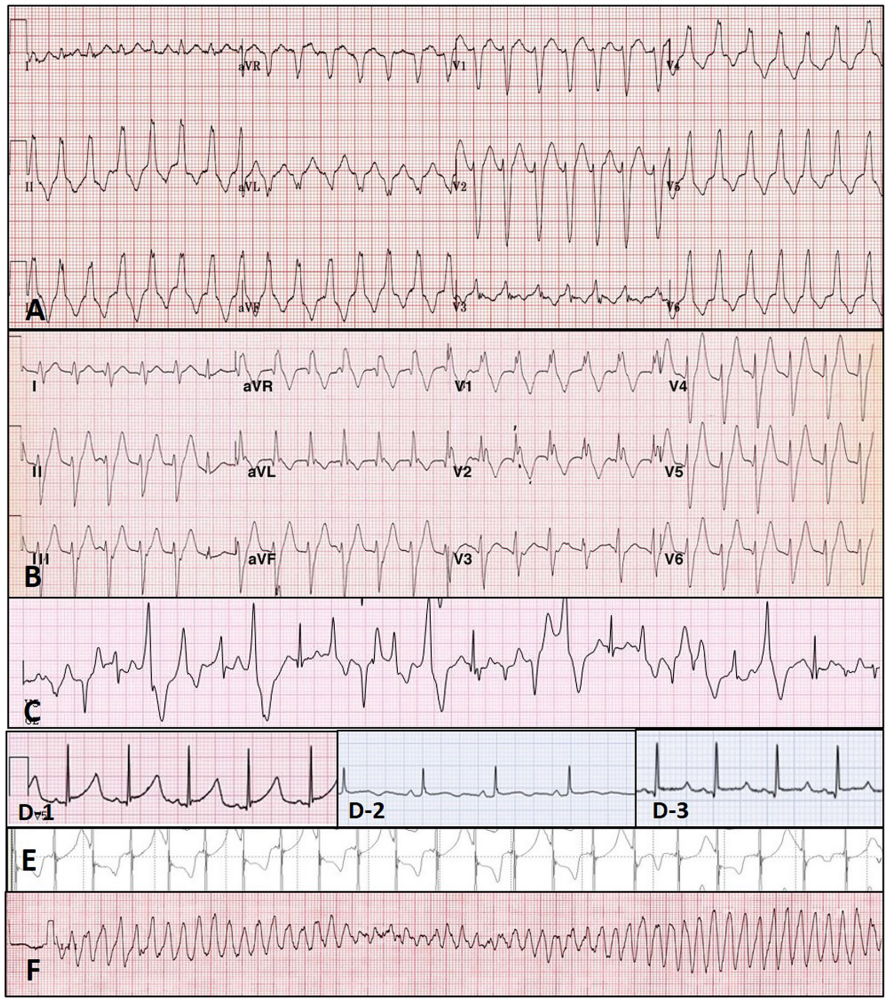

Figure 9. (A) Idiopathic right ventricle outflow tract ventricular tachycardia (VT) electrocardiography (ECG) shows typical left bundle branch block pattern and inferior axis (positive QRS in leads D-II, D-III, and aVF). (B) In idiopathic LV posterior fascicular VT ECG, typically right bundle branch block pattern and superior axis (negative QRS in leads D-II, D-III, and avF) are seen, and the QRS is relatively narrow. (C) Superior-inferior alternating QRS is seen in "bidirectional" (polymorphic) VT (typically seen in CPVT). (D1) Long QT type-1 ECG sample with broad-based T waves are typical. (D2) Example of a long QT type-2 ECG with typical biphasic and low amplitude T waves. (D3) Example of long QT type-3 with normal-width T wave but prolonged "ST interval." (E) T wave alternans, which is considered as a high-risk factor in Holter recording (major changes from beat to beat). (F) "Torsades de Pointes" (polymorphic VT), mostly initiated by a premature ventricular contraction that creates an "R on T" phenomenon by overlapping the T wave of the previous QRS due to prolonged QT.

cardiac arrest.[32,33] Diagnosis is made by detecting bidirectional VT or polymorphic PVCs during exercise or catecholamine infusion in patients under 40 years of age who do not have structural heart disease and have a normal resting ECG.

Treatment includes lifestyle changes (avoiding all kinds of effort, including competitive sports and drugs that cause sympathetic stimulation), pharmacological treatment (beta-blockers -especially nadolol and ropranolol-, and flecainide), left cardiac sympathectomy (LCSD) (in patients with syncope despite adequate medical treatment and to prevent unnecessary ICD shocks; nowadays LCSD is becoming the primary therapy even before ICD), and ICD implantation (in those with a history of cardiac arrest).[30-34]

Torsades de Pointes

Torsades de Pointes literally means "twisting/dancing dots" in French. It is a malignant polymorphic type of VT with increasing and decreasing QRS amplitude on ECG. Among the important causes are drug effects (antiarrhythmics such as amiodarone and procainamide, antidepressants), and electrolyte imbalances such as hypokalemia, hypocalcemia, and hypomagnesemia also constitute the typical ventricular arrhythmia seen in long QT syndrome. Prolonged TdP should be treated with cardioversion as it will cause hemodynamic impairment. If TdP does not disrupt hemodynamics, magnesium sulfate 25-50 g/kg should be given as an IV infusion in 10-15 minutes.[30,31]

Long QT Syndrome (LQTS): It is the most common inherited genetic arrhythmia syndrome characterized by prolonged QTc interval and typical TdP that can result in sudden death. The prevalence is 1/2000-2500, and the 1-year risk of death is 20% in untreated cases, while the 15-year risk of death is approximately 53%.[31,34]

Long QT syndrome is a single gene disease that is often inherited in an autosomal dominant pattern. Genetic mutations can be shown in 80%-90% of patients. There are 17 known

subtypes, but type-1, type-2, and type-3 cases constitute 90%-95% of all patients (Figure 9D1-D3). The most common mutations are in the KCNQ1 (type-1), KCNH2 (type-2), and SCN5A (type-3) genes. Torsades de Pointes is triggered by physical (especially swimming) or emotional stress in type-1, stress, sudden loud noise (alarm clock), and fear (dog barking, horror movie) in type-2, and during sleep in type-3.[31,34]

The most common symptoms are syncope, convulsions (misdiagnosed as epilepsy!), aborted cardiac arrest, and SCD. Bradycardia, T wave alternans (Figure 9E), TdP (Figure 9F), PVCs (monomorphic-polymorphic), and AV block (usually in the form of 2 : 1 and especially in newborns-infants) can be seen in ECG, Holter, and exercise tests.

Diagnosis is made by evaluating ECG, family history, and genetic data (Schwartz scoring).

Apart from the 3 most common types, a rare but high-risk type of LQTS, accompanied by bilateral sensorineural hearing loss and inherited in an autosomal recessive pattern, is called "Jervell Lange Nielsen" (JLN) syndrome.[30,31]

Treatment includes lifestyle changes (prohibition of competitive sports and swimming and avoidance of all drugs that prolong QT), pharmacological treatment (β-adrenergic receptor blockers propranolol or nadolol and mexiletine in type-2 and 3), ICD implantation (in previous cardiac arrests and high-risk types such as JLN, type -3, female gender, and type-2, QTc consistently >500-550 ms), and LCSD (in addition to ICD in patients in whom beta-blockers are ineffective/intolerable).[34]

CONCLUSION

A better understanding of the pathophysiology and recent developments in the treatment of pediatric arrhythmias in the last 2 decades has yielded better management of these tachyarrhythmias by pediatric cardiologists and electrophysiologists. Most of these patients can be correctly diagnosed, successfully treated, and followed up by pediatricians. Therefore, every pediatrician facing pediatric arrhythmia patients should know about the diseases, ECG findings, and the therapeutic options available to avoid disease-related mortality and morbidity.

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