Review

Paroxysmal supraventricular tachycardia: physiopathology and management

Paola Neroni, Giovanni Ottonello, Danila Manus, Alessandra Atzei, Elisabetta Trudu, Susanna Floris, Vassilios Fanos

Neonatal Intensive Care Unit, Neonatal Pathology, Puericulture Institute and Neonatal Section, AOU and University of Cagliari, Italy

Proceedings

Abstract

Paroxysmal supraventricular tachycardia (PSVT) is the most frequent arrhythmia in newborns and infants. Most supraventricular tachycardias affect structurally healthy hearts. Apart from occasional detection by parents, most tachycardias in this age group are revealed by heart failure signs, such as poor feeding, sweating and shortness of breath. The main symptom reported by school-age children is palpitations. The chronic tachycardia causes a secondary form of dilative cardiomyopathy. Treatment of acute episode usually has an excellent outcome. Vagal manoeuvres are effective in patients with atrioventricular reentrant tachycardia. Adenosine is the drug of choice at all ages for tachycardias involving the atrioventricular node. Its key advantage is its short half life and minimum or no negative inotropic effects. Verapamil is not indicated in newborns and children as it poses a high risk of electromechanical dissociation. Antiarrhythmic prophylaxis of PSVT recurrence is usually recommended in the first year of life, because the diagnosis of tachycardia may be delayed up to the appearance of symptoms. Digoxin can be administered in all forms of PSVT involving the atrioventricular node, except for patients with Wolff-Parkinson-White syndrome below one year of age. Patients with atrioventricular reentrant PSVT can be treated effectively by class Ic drugs, such as propaphenone and flecainide. Amiodarone has the greatest antiarrhythmic effect, but should be used with caution owing to the high incidence of side effects.

Keywords

Corresponding author

Dr. Paola Neroni, Neonatal Intensive Care Unit, Neonatal Pathology, Puericulture Institute and Neonatal Section, Department of Surgery, University of Cagliari, SS 554 bivio Sestu, Monserrato (CA), 09042, Italy; email: paolaneroni123@gmail.com.

How to cite

Introduction

Paroxysmal supraventricular tachycardia (PSVT) is the second most common arrhythmia in children after sinus tachycardia. A heart rate (HR) higher than 220 bpm in children below one year of age and higher than 180 bpm in children above one year is suspicious for PSVT. In 40% of cases, PSVT arises in the first year of life [1]. Its incidence in the neonatal period has not been estimated adequately (1 out of 15,000-25,000 live births) [2]. A predisposing condition (congenital heart disease, medications, concomitant infection) is found in 15% of cases [1-3].

Pathophysiological mechanism

According to their underlying electro-physiological mechanism, supraventricular tachycardias are divided into 1) reentrant tachycardia and 2) automatic tachycardia [4] (**Fig. 1**). Reentrant tachycardia occurs when the electrical stimulus is conducted more than once in a closed circuit. This occurs in the presence of a functional-anatomic substrate able to be activated by appropriate stimuli, e.g. ectopic heartbeats. This substrate must consist of two pathways having the following features: different duration of the refractory periods, appearance of a unidirectional block on one pathway, concurrent slowdown of conduction speed on the other pathway. Reentrant circuits can differ in extent and involve several heart structures. They may be limited to a few myocardial fibres in
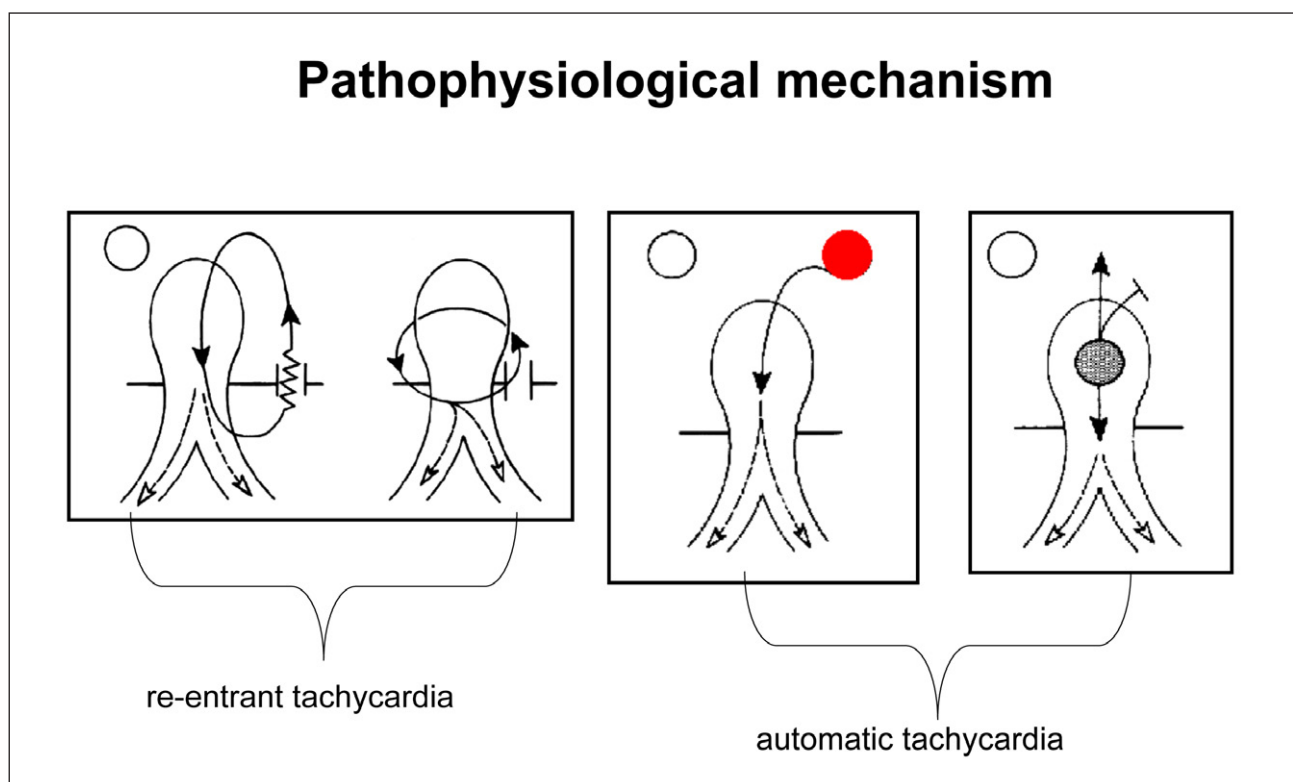

Figure 1. According to their underlying electrophysiological mechanism, supraventricular tachycardias are divided into reentrant tachycardia and automatic tachycardia.

the context of the atria or of the atrioventricular junction (micro reentry), or may be more extensive and involve on the one hand the normal His bundle conduction axis and, on the other, the atrioventricular accessory pathways (macro reentry). The accessory atrioventricular pathways are the most frequent cause of supraventricular tachycardia in paediatric age, and give rise to atrioventricular reciprocating tachycardia (AVRT); on the other hand, children have a much lower incidence than adults of atrioventricular nodal reentrant tachycardia (AVNRT) whose only reentry circuit is the atrioventricular node [4]. Most PSVTs affect structurally healthy hearts, even though various structural heart diseases are potentially exposed to the presence of accessory circuits; they include Ebstein's anomaly, the congenitally corrected transposition of the great arteries and tricuspid atresia. A particular subgroup are cardiac tumours (rhabdomyoma) and glycogen storage heart disease (Pompe or Danon disease).

Automaticity is a property which in physiological conditions is only manifested by the sinus node and by conduction tissue. It consists in slow, spontaneous diastolic depolarisation until the threshold potential is reached, which conditions the onset of the action potential. Activation frequency decreases from the sinus node, which is the heart's pacemaker, to the Purkinje fibres in the ventricle. In certain situations, for example after damage to the myocardium, or following changes in extracellular fluids or, more often, without a known reason, some heart cells may themselves acquire automaticity (abnormal automaticity) and, if their rate is higher than that of the sinus node, they may take over as the heart's main pacemaker. One such tachycardia is ectopic atrial tachycardia, which may originate at any point in the atria without involving in its mechanism the sinoatrial node, the normal atrioventricular junction or the accessory atrioventricular pathways. This is a well-organised tachycardia, whose normal sinus rhythm is replaced by high-frequency impulses (in the newborn up to 250-300/min) originating from a small area of the atrium. The P wave's morphology and axis allow us to locate the site of origin of the ectopic focus. Characteristic phenomena respectively at the beginning and at the end of the tachycardia are a warming-up and a cooling-down phase. Atrioventricular conduction may be 2:1, 3:1; alternatively, an atrioventricular block (AVB) may occur (periodic AVB) [5].

Atrial ectopic tachycardia is a relatively infrequent arrhythmia in newborns; it accounts for not more than 5-10% of all supraventricular tachycardias [5]. It may occur in isolation or in association with organic heart disease. The types of heart disease most frequently associated with this arrhythmia are: congenital heart disease, myocarditis and cardiomyopathy [6]. The electrophysiological mechanism underlying individual supraventricular tachycardias also affects their response to attempts to induce and interrupt tachycardia in the course of electrophysiology studies. Thus, reentrant and triggered forms can be both induced and interrupted, whereas those due to increased automaticity can be neither induced nor interrupted, but can be recorded only if the atrium is stimulated at a rate faster than the tachycardia. In paediatric age, the most frequent form (about 80%) is reentrant through an accessory pathway.

Symptoms and clinical signs

In newborns and small children with a structurally normal heart, recognising a single, short PSVT episode is often difficult, and diagnosis may be delayed, owing to the non-specific signs and symptoms. Newborns tolerate very well HR values above 250 bpm, provided the episodes are not protracted; indeed, after 12-24 hours the left ventricular diastole is damaged and cardiac output decreases, leading to progressive haemodynamic impairment which in turn causes the onset of the symptoms and clinical signs of heart failure (up to 30% in PSVT in infants below 12 months of age). Signs of congestive heart failure such as vomiting, sweating, tachypnea and pale-greyish, moist and cold skin can develop rapidly. A careful, objective heart examination should be performed, assessing: intensity of the first and second heart sound, presence of pathological murmurs, radial and femoral pulses and arterial pressure.

Electrocardiography diagnosis of paroxysmal supraventricular tachycardia

Electrocardiography (ECG) evaluation must cover: HR (in PSVT HR > 220/min), QRS complex morphology (whether wide or narrow, in order to differentiate between ventricular and supraventricular tachycardia), the regularity of QRS complexes, and possible identification of the P wave, its morphology and its relationship with the QRS complexes. Nodal reentrant tachycardia, similarly to that mediated by an accessory pathway tends to be highly regular, with a stable rate, while

sinus tachycardia exhibits a fluctuating rate. At the ECG the common finding of the reentrant form is narrow QRS; in most cases the P wave is distinct from QRS and is identified between the end of the QRS and the ascending branch of the T wave; however, the high frequency makes it difficult to identify [7].

Drug treatment

Early management

If the patient is clinically stable, one should firstly attempt vagus nerve stimulation manoeuvres, such as the "diving reflex", obtained by applying ice for 15-20 seconds to the newborn's face (region of the mouth and nose) or massage of the carotid sinus or Valsalva manoeuvres in older cooperating children (e.g. making them blow energetically into a straw, causing them to vomit); eyeball compression is always contraindicated in paediatric age as it carries the risk of eyeball-retina damage. If vagal manoeuvres prove ineffective, adenosine should be administered: its effectiveness in reentrant arrhythmias is in excess of 98%. Adenosine is a drug with a short half-life, which causes a block at the level of the sinoatrial node and of the atrioventricular node. Given its short half-life, it must be administered in a fast bolus (intravenous or intraosseous) followed by saline wash-out (2-2.5 ml). The initial recommended dose is 0.1 mg/kg; if ineffective it can subsequently be increased to 0.2 mg/kg and up to a maximum of 0.5 mg/kg [8]. Should adenosine prove ineffective, propaphenone or flecainide (class Ic drugs) are useful at a dose of 1 mg/kg intravenously. They are effective where the reentry circuit is supported by an accessory pathway. They should not be administered to patients with organic heart disease or heart failure as they can depress ventricle function further. In neonates with the signs and symptoms of tachycardiomyopathy who do not respond to adenosine, amiodarone may be useful (a bolus of 5 mg/kg in 20 minutes followed by an infusion of 10 mg/kg over the next 24 h). In patients with severe cardiorespiratory impairment or sudden clinical worsening, adenosine should be administered only if intravenous access and the drug are immediately available; when this is not the case (or if adenosine is ineffective) synchronised cardioversion (0.5-1 J/kg) after inducing deep sedation is indicated; cardioversion should never be delayed in unstable patients who already show the signs of systemic hypoperfusion.

Maintenance therapy

The natural history of PSVT presenting in the neonate suggests that spontaneous resolution usually occurs in the first year of life [9]. For infants born with a history of fetal PSVT, doctors choose to give maintenance antiarrhythmic therapy postnatally to 52-63% of non-hydropic fetuses and to about 80% of hydropic fetuses [10-12]. Digoxin has long been the drug of choice in antiarrhythmic prophylaxis for PSVT. Success rates range between 42% and 75% [13]. Close monitoring of serum concentration is recommended in order to prevent negative side effects, given that digoxin is no longer used in children below one years of age with Wolff-Parkinson-White syndrome owing to the potential risk of ventricular fibrillation. Beta-blockers, in particular propranolole, are commonly used to prevent the recurrence of neonatal PSVT in cases where digoxin is contraindicated. Their success rates range from 50% to 90%. Flecainide is a class Ic drug with a success rate in preventing recurrences of about 100%; however, it also carries a risk of pro-arrhythmic effects of up to 7.5%. Another effective drug is propaphenone, another class Ic drug. In recent years, oral sotalol, a class III drug with beta-blocking properties, has been used increasingly often in the long-term treatment of paediatric supraventricular arrhythmias. Amiodarone is another class III antiarrhythmic drug. The main electrophysiological effect of amiodarone is to increase duration of the action potential and the refractoriness of all cardiac cells [14]. Amiodarone does not significantly affect myocardial contractility. Ablation should be reserved to cases of complete lack of response to all antiarrhythmic treatments coupled with heart failure.

Follow-up in the first year of life

Most (60-90%) infants with Wolff-Parkinson-White syndrome undergo spontaneous resolution by 1 year of age [15, 16]. Most infants with ectopic atrial tachycardia at < 6 months of age will be free from atrial tachycardia after 12 months of antiarrhythmic therapy [17-18]. Patients under antiarrhythmic treatment should be monitored monthly to adapt the drug dosage to the natural increase in body weight. In patients with abnormal pathway reentrant PSVT, antiarrhythmic treatment can be suspended after the first 8-12 months of life; in these cases it may be useful to perform transesophageal atrial stimulation during the therapeutic wash-out to assess the

inducibility of tachycardia. If tachycardia is still inducible, the physician should consider whether to resume treatment [16-19]. Parents should be trained to measure HR (at the wrist, using the stethoscope).

Conclusions

Neonatal-onset PSVT is frequently found in the clinical practice of paediatric cardiologists. The acute treatment of a single episode usually has an excellent prognosis. In any case, antiarrhythmic prophylaxis is recommended throughout the first year of life. The antiarrhythmic drug should be chosen carefully by considering the pathophysiological mechanism underlying the tachycardia, the presence of ventricular pre-excitation signs in the basal ECG and the risk/benefit ratio of each case.

Declaration of interest

The Authors declare that there is no conflict of interest.

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