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About Wolff-Parkinson-White's Electrocardiographic Model

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Abstract

Probably, Wolff-Parkinson-White syndrome (WPWS) is the most studied electrocardiographic model within the so-called cardiac arrhythmias. But there is still a long way to go to get to know it fully as well as to diagnose it efficiently and clearly.

There are still many WPW models that go underdiagnosed or misdiagnosed. The so-called "inapparent WPWS" is a paradigm of the previously written and deserves a detailed study of it.

Keywords: *Wolff-Parkinson-White syndrome (WPWS); Electrocardiographic Model; Cardiac Arrhythmias*

Introduction

Probably, Wolff-Parkinson-White syndrome (WPWS) is the most studied electrocardiographic model within the so-called cardiac arrhythmias. But there is still a long way to go to get to know it fully as well as to diagnose it efficiently and clearly.

There are still many WPW models that go underdiagnosed or misdiagnosed. The so-called "inapparent WPWS" is a paradigm of the previously written and deserves a detailed study of it.

Historically, the presence of atrioventricular (AV) conduction pathways considered as accessory was enunciated parallel to the discovery of the normal conduction system. At the end of the 19th century, the concept of conduction of the electrical impulse from the atria to the ventricles confronted the supporters of myogenous conduction and those of neurogenic conduction.

Before Kent, Paladin had declared himself a proponent of myogenic theory by studies conducted on several species [1]. Stanley Kent, in a series of manuscripts, described the presence of an atrioventricular connection in the heart of mammals [2]; thought that such pathways would form a nexus in the healthy heart. Cohn and Fraser [3] presented the first electrocardiographic tracing of ventricular pre-excitation.

In 1930, Wolff, Parkinson and White [4] published their classic manuscript about 11 cases of branch block with short PR interval in healthy young subjects predisposed to paroxysmal tachycardia. Anatomical confirmation of atrioventricular accessory pathways was the result of clinical descriptions of this condition.

Wood and Ohne [4,5] described atrioventricular muscle connections in patients with anterograde pre-excitation. Ohne introduces the term pre-excitation [4,6].

The advent of electrophysiological studies in humans ended up dissipating the dark spots of Wolff-Parkinson-White Syndrome (WPW) [5,7,8].

Durrer and Roos demonstrated the presence of ventricular pre-excitation in the human heart using epicardial mapping techniques. Cobb and colleagues precisely interrupted an atrioventricular accessory pathway during open heart surgery. In the laboratory, Durrer used programmed electrical stimulation to initiate and terminate tachycardia in patients with Wolff-Parkinson-White syndrome.

Wellens [13] used intracardiac electrograms to indicate that type A of Wolff-Parkinson-White syndrome depended on a left atrioventricular connection. Gallagher [11] has made countless contributions to the systematic study of pre-excitation syndromes [6,11].

It has been more than 75 years since Wolff, Parkinson and White described the fundamental characteristics of the syndrome that bears their names. The interest aroused by this entity has been enormous. It soon became clear that there were hidden accessory pathways, incapable of driving in the anterograde direction, but retaining retrograde conductibility, so they could cause tachycardias by re-entry of the atrioventricular junction.

More recently, atrioventricular accessory pathways with long and decremental driving times have been identified; this type of accessory pathway participates in the most common form of permanent supraventricular tachycardia. The information generated over the last 30 years concerning accessory pathways has been tremendous, and its diagnostic consequences, prognostic but fundamentally very important therapeutic.

Incidence

The incidence and prevalence of WPWS cannot be definitive as there are many clinically and electrocardiographically coincident processes with it and yet they are either underdiagnosed or not diagnosed as such.

Statistical data regarding the frequency of Wolff-Parkinson-White syndrome vary according to each author's material. On the whole, the actual incidence of the syndrome in the general population is considered to be around 1 per 1000; in hospitalized individuals it is higher (1.5:100).

Wolff-Parkinson-White syndrome has been seen at all ages, from birth to old age, although the majority of cases (90%) are younger than 50 years old. According to some authors, there is a predominance in the male sex, which varies from 60 to 75% of patients. According to our research, gender differences tend to decline.

Since Ohnell (1944) it has been admitted that Wolff-Parkinson-White syndrome is a hereditary anomaly, as demonstrated by the observation of ventricular pre-excitation in several members of an eight family and in univitelin twins [45,46]. In light of recent work, it seems clear that sometimes Wolff-Parkinson-White syndrome is a familial driving disorder, with a hereditary transmission of an autosomal recessive type [9].

Overall, it has been estimated that 70 - 85% of Wolff-Parkinson-White syndromes are observed in individuals with structurally healthy hearts.

The association of Wolff-Parkinson-White syndrome with congenital cardiac malformations has long been pointed out; its incidence varies, according to the authors, from 7.5 to 15%. In 1955, Sodi Pallares and colleagues were the first to draw attention to the association of Wolff-Parkinson-White syndrome type B with Ebstein's anomaly. Today this association is well proven, and pre-excitation is found in 6 to 26% of cases with such anomaly.

In a recent review of a total of 184 patients with Ebstein's anomaly, the authors found 45 cases (24%) of the pre-excitation syndrome [10].

It has also been observed in other congenital cardiopathies, such as the corrected transposition of great vessels, the tricuspid atresia, double exit chamber of the right ventricle, tetralogy of Fallot, ventricular septal defect, and other abnormalities, but it is difficult to establish that in these cases the association is not coincidental.

The association with acquired cardiopathy has also been known for many years, since the first case of Wolff-Parkinson-White syndrome associated with paroxysmal tachycardia, described by F.N. Wilson (1915), was accompanied by mitral stenosis. Its incidence varies between 15 and 30%. The cardiopathies most frequently observed are rheumatic, ischemic, cardiomyopathies and recently Gallagher [11] reported a rate of 6.75% in mitral valve prolapse and supraventricular paroxysmal tachycardias.

Histopathology

In the healthy adult heart, the muscular atrioventricular connection is limited to the single specific pathway of conduction through the atrioventricular junction, since, outside of it, the fibrous rings and the central fibrous body separate the myocardium from the atrium of the myocardium of the ventricle. However, in an unspecified percentage of macroscopically abnormal hearts, there are abnormal (or accessory) muscle connections. These can directly anastomose the atrial wall with the ventricular wall mounted on the fibrous rings, thus configuring a total "bypass" of the conduction system. These pathways are the so-called Kent bundles.

Concerning the histocytology of the accessory pathways, it should be noted first and foremost that the Kent bundles are, for the most part, made up of normal myocardium, with some sporadic transitional characteristic.

The topography of accessory pathways is varied; Kent bundles can be found at any point around the fibrous rings, although they are mostly located in the outer portion of the valvular insertion, in the subepicardial adipose tissue [12].

In the theoretical anatomy-functional plane, it seems evident that the common denominator of accessory pathways in the early activation of the ventricle by a "short circuit" of the auricular stimulus. It prevents, in full or in part, the physiological slowdown of the atrioventricular junction and conduction through the branches of the specific system. All this constitutes the physiopathological and anatomical archetype of the macro-reentry circuit and provides the key to the morphological interpretation of reentry and stimulus circle movement in the pathogenesis of the classic supraventricular paroxysmal tachyarrhythmias that complicate Wolff-Parkinson-White syndrome.

Electrocardiographic diagnosis

Under normal conditions, for an electrical impulse to be transmitted from the atria to the ventricles it has to pass through the standard conduction system; ventricular atrial node and His Purkinje system without producing premature activation of the surrounding myocardium and that almost simultaneous activation in the Purkinje network produces synchronized ventricular activation, this type of normal sequence produces in the surface electrocardiogram a habitual PR interval.

In Wolf Parkinson White syndrome the accessory pathway produces a ventricular pre-excitation is present if any part of the ventricles is activated earlier by the accessory pathway that can be done if the impulse reaches the ventricles exclusively over the regular conduction system.

It is called pre-excitation because it excites the ventricles before the time since the impulse when not entering the AV node does not suffer the delay in this one, for this reason, the PR interval can be short.

The typical electrocardiographic pattern of Wolff-Parkinson-White syndrome is characterized by:

- Short PR interval (less than 0.12 sec). This is not always the case, as we will explain later.
- Spread QRS (greater than 0.12 sec) with an initial paste that configures the typical delta wave. Delta wave is the fundamental hallmark for this type of syndrome.
- Secondary alterations of repolarization, with a T-axis opposing that of QRS.

Many patients with Wolff-Parkinson-White syndrome do not show, in sinus rhythm, all the typical electrocardiographic features of this entity. The duration of the PR interval and QRS complex, as well as the evidence of a delta wave, depends on four factors:

- Location of the accessory road.
- Driving times headphones,
- Driving times through the accessory road,
- Atrioventricular conduction times through the normal pathway [13].

An accessory pathway away from the sinus node, either anatomically (left lateral location) or functionally (intra-auricular or interauricular conduction disorder), makes pre-excitation little evident, or impossible to detect, in sinus rhythm.

The same is true in some patients with hyperconductive atrioventricular node and very short conduction times through the normal nodal pathway. Finally, accessory pathways with long driving times may show little or no signs of sinus rhythm. In the same subject, the degree of pre-excitation may vary spontaneously due to variations in driving times (mainly in nodal driving) induced by changes in vegetative tone or other factors. From Ohnell, these endless variations of PR and QRS in patients with pre-excitation have been referred to as the "concertin effect". According to the degree of contribution of the accessory pathway to ventricular activation and the constancy with which this phenomenon occurs, many varieties of Wolff-Parkinson-White syndrome can be distinguished:

Wolff-Parkinson-White manifest: a patient whose electrocardiogram in sinus rhythm shows a short (or normal) PR, delta wave and wide (or normal) QRS (Figure 1).

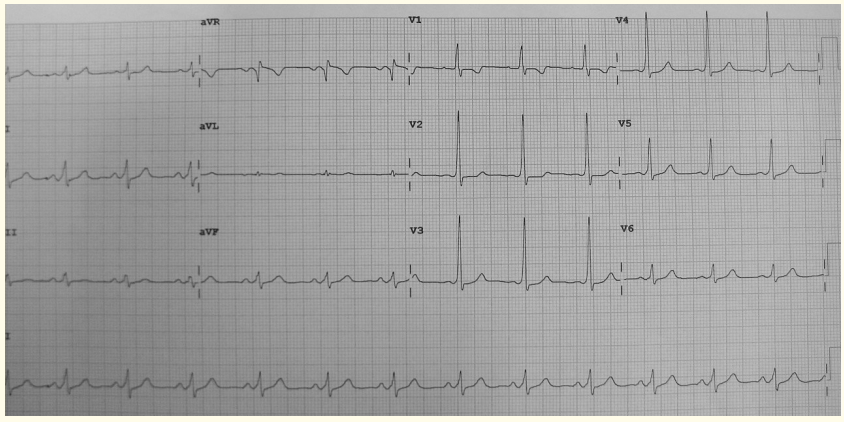

Figure 1: *A typical WPW electrocardiographic model with a shortened PR interval, an overt delta wave and discrete ventricular repolarization alterations.*

Intermittent Wolff-Parkinson-White: This term has been used generically to define the inconstancy of the pre-excitation image in sinus beats of the same or successive electrocardiographic traces. Intermittent pre-excitation can cause repolarization alterations in the beats usually driven by the so-called memory phenomenon [14].

Inapparent Wolff-Parkinson-White: Patients with Wolff-Parkinson-White syndrome who present with normal or near-normal PR and QRS without a limpid evidence of a delta wave as a result of similar atrioventricular conduction times through the normal pathway and accessory tract. It is important not to confuse these cases with those in which there is a hidden accessory pathway.

This so-called "Inapparent" WPW syndrome is much more frequent than one might think, in many more times than desired, it is frankly under-diagnosed.

Sometimes such underdiagnosis can lead to dire consequences (Figure 2).

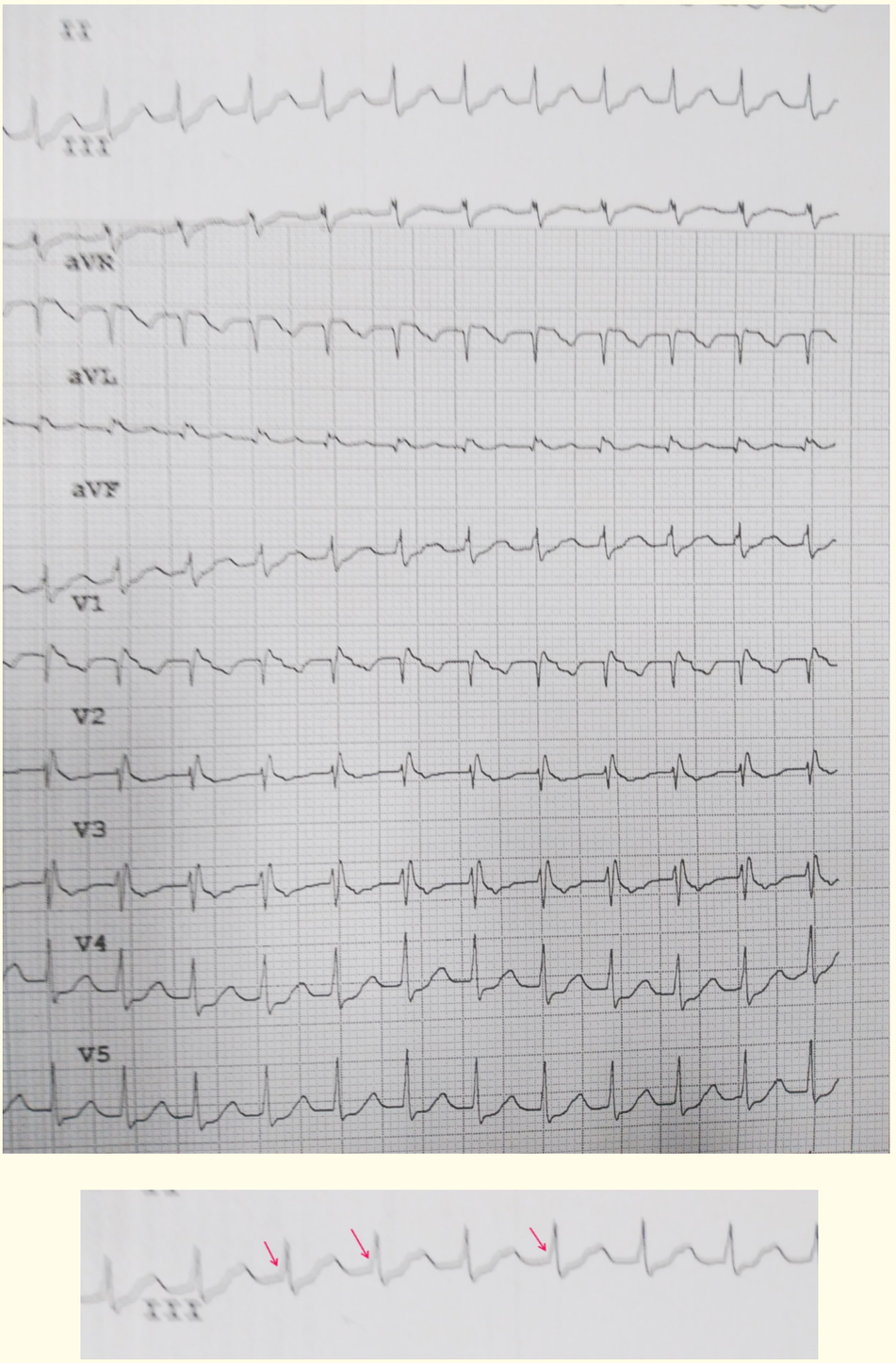

Figure 2: *A tachycardia 'crisis in a patient who'd already suffered three more episodes. Always nocturnal and suddenly. She was treated with 6 mg of adenosine triphosphate in rapid bolus that increased her heart rate. She was diagnosed as a paroxystic supraventricular tachycardia with a narrow QRS complex. However, the presence of delta waves can be seen (red arrows on D3). We recommend being very cautious before administering adenosine, since the result, depending on the causative entity, may worsening.*

The latter cannot lead in an anterograde direction, so not only is the electrocardiogram in sinus rhythm always healthy but in the case of developing atrial fibrillation, the ventricular response is determined by the electrophysiological properties of the atrioventricular node.

Topographical varieties

Since Rosenbaum [14,15], classically Wolff-Parkinson-White syndromes were divided into two groups, based on the form of QRS in the V1-V2 and esophageal derivations.

Two groups were distinguished: A, in which R waves predominate in these derivations, and B, in which the form of S or QS is the main one in at least one of the right precordial.

This classification is based on a global study of the QRS and Rosenbaum does not associate any particular topography to it.

The introduction of the programmed electrical stimulation made evident the inadequacies and limitations of this classification.

Electrophysiological studies and intraoperative mapping techniques allowed the definition of multiple electrocardiographic patterns, according to ten different locations of the accessory pathways along atrioventricular furrow [16].

Surgery on the Wolff-Parkinson-White simplified these initial classifications somewhat since four approaches of dissection were used according to the accessory pathways were anteroseptal, posteroseptal, lateral right and lateral left [17].

The electrocardiographic classifications are mainly based on the special orientation of the initial vectors, at 10, 20 and 40 msec of anomalous activation. Theoretically, the vectocardiogram should allow a more precise analysis of the spatial orientation of such vectors, due to the greater amplifications of the records obtained. Therefore, it is preferable to express the orientation of these vectors in the three planes to infer the site of the pre-excitation zone.

The left side type has a delta wave oriented in the frontal plane, down and to the right between +100 and +120 (negative delta in DI and aVL and positive in DII and DIII). In the horizontal plane, the delta wave is positive in V1, sometimes negative in V6 and the QRS is positive from V1 to V6.

The two posterior paraseptal types, right and left, have an up and left oriented delta wave between -75 and -30 (positive delta wave in DI, negative in DII, DIII and aVF). In the horizontal plane, the delta wave is positive in V1 in the left type, negative in V1 and positive in V2 in the right type. QRS is negative in V1 and positive from V2.

With the use of the thoracic circle it is possible to distinguish the right or left type with greater facility since in the right the delta wave is negative from V3R to V9R, while in the left the delta is positive from V3R to V9R [18] V3R to V7R. On the other hand, the possible variations induced by the dextrorotation of cardiac levorotation.

The right lateral type has a delta wave oriented to the left, between -30 and +30 (positive delta in DI and DII, negative in DIII) in the frontal plane and also to the left in the horizontal plane (negative delta in V1 and V2). The QRS is negative in V1 and V2.

The two previous paraseptals, right and left, have a delta wave oriented down and left in the frontal plane, between +60 and +80º (positive delta in DI, DII and DIII). In the horizontal plane, in V1 the delta wave is negative in the right type and positive in the left type [19-22].

In 1986, Gallagher [23] described what he called intermediate or parahisian septal accessory pathways, indicating that they are characterized by an electrocardiographic pattern of an anteroseptal path in sinus rhythm or the pre-excited beats during atrial fibrillation, while the retrograde atrial activation sequence was typical of a posteroseptal pathway. Epstein [24] has recently described two types of intermediate septal accessory pathways: the above, located at the vertex of the Koch triangle, in close relationship with the bundle of His, and the mideptals, situated at the midpoint of the Koch triangle, in the vicinity of the atrioventricular node.

Analyzing the QRS complex in derivations DIII, V1, and V2, the authors locate the pathways accessories [25] at five sites with 88% probability.

The new electrocardiographic algorithm which is presented, using a sequential analysis of the shape of the QRS complex in only three derivations, is simple and effective.

This facilitates electrocardiographic analysis in patients with Wolff-Parkinson-White syndrome and improves the results of radiofrequency ablation.

These findings are explained on the basis that early activation of the left free wall or left septum may result in earlier activation of the left ventricle and delayed activation of the right ventricle by the His-Purkinje normal conduction system. Accessory pathways at the left lateral level (free wall) produce a pattern of BRDHH by early activation of the left ventricle. The anterolateral accessory pathways were characterized by high R waves in the V1 shunt, reflecting earlier activation of part of the left ventricle away from the left side, as well as early transition in precordial shunts.

The right anteroseptal accessory pathways differed from other right septal channels because they had the positive QRS complex in the DIII shunt.

The top location of these pathways results from a vector of the lower QRS in the frontal plane, with positive QRS complexes in at least two of the three more inferior leads. The anteroseptal accessory pathways activate the right and left ventricles in an anteroposterior direction, resulting in a mean QRS axis.

Accessory pathways at the right lateral level (free wall) should be considered if a negative QRS is observed in leads DIII, V1, and V2 because the plane of the tricuspid ring extends in front of and below the interventricular septum. The spatial vector produced by the accessory pathways on the right free wall could be expected to result in a late horizontal transition vector with subsequent direction.

In the pre-excitation of the right free wall, the early forces in that location (directed forward) are predominantly the result of excitation on the accessory road, which causes an unopposed dominance of the strengths of the left free wall (directed backward), resulting in a deviation of the AQRS to the left.

Right posteroseptal or posterolateral accessory pathways may be present if a negative QRS complex is observed in DIII and V1 leads, but positive in V2. The pre-excitation of the right posterolateral accessory pathways, due to their inferior location in the tricuspid ring, should be expected to show a superior AQRS in the frontal plane. In the posteroseptal accessory pathways, the right and left ventricles are activated from the back to the front, which causes the QRS to deviate to the left.

In the left posteroseptal or posterolateral accessory pathways, a negative QRS complex is to be expected in DIII, but positive in V1. In the posteroseptal accessory pathways the ventricle is activated in the posterior-posterior direction, which produces a deviation of the QRS to the left, as well as an early transition in the horizontal plane.

Accessory pathways in Ebstein's anomaly

Electrocardiographic recognition of Wolff Parkinson White syndrome in patients with Ebstein anomaly [26] is difficult because its classic pattern of short PR with delta wave is not always evident.

Tachycardias in Wolff-Parkinson-White Syndrome

As Cárdenas mentions [27], pre-excitation syndromes would only be an electrocardiographic curiosity, were it not for the arrhythmias to which they give rise. Establishing the prevalence of arrhythmias in these patients is extremely difficult, as there is a large number of

patients who have pre-excitation and never suffer arrhythmias and, on the other hand, the natural history of pre-excitation is unpredictable. Patients with accessory pathways may develop different types of tachyarrhythmias. In some tachycardias, the accessory pathway is an indispensable component of the circuit (tachycardia by orthodromic re-entry, antidromic or multiple pathways).

In others, the accessory pathway is an "observer" of the arrhythmia and modifies its clinical consequences and its electrocardiographic expression (atrial fibrillation, atrial flutter, atrial tachycardia, intranodal re-entry). Finally, in a low percentage of patients, the existence of the accessory pathway is the cause of initiating and perhaps maintaining ventricular fibrillation, with a risk of sudden death [28].

Re-entry may occur when: a) there are at least two functionally different conduction pathways, b) a unidirectional blockage is induced in one pathway, and c) the driving time is slow enough in the unblocked pathway to allow recovery of excitability in the blocked pathway and retrograde conduction occurs in that pathway, thereby completing the circuit.

Circular motion tachycardia in Wolff-Parkinson-White syndrome is an example of re-entry. The orthodromic form is the most common: 85% re-entry tachycardia, in which the accessory pathway is used as part of the circuit. The term orthodromic denotes the propagation of an impulse in the normal direction, and in re-entry atrioventricular tachycardia arises when the antegrade conduction comes from the atrium, continues through the atrioventricular node and from there passes to the His-Purkinje system, ending in the ventricle. Retrograde conduction to the atrium is done by the accessory road closing the circuit. The shape of the resulting QRS is normal during tachycardia unless there is branch blocking of the His bundle (functional or fixed).

Accessory pathways that lead in both directions and those that only lead in the retrograde direction (hidden pathways) are the basis of this type of tachycardia in orthodromic circular motion. The duration of the tachycardia cycle depends on the total conduction time of all tissues in the re-entry circuit. The following electrocardiographic data suggest retrograde conduction via the accessory pathway during supraventricular tachycardia: a) a negative P wave during tachycardia, b) an RP interval greater than 70 msec, c) prolongation of the duration and length of the tachycardia cycle with functional branch blocking of the His bundle (see below). This sign is useful only when it appears. d) Electrical alternation of the QRS complex during tachycardia, a reasonably specific sign, but without sensitivity. e) Atrioventricular dissociation completely rules out this type of tachycardia. Accessory pathways leading in antegrade direction may result in antidromic circular moving tachycardias, in which atrioventricular conduction is via the Kent branch, while the normal conduction system is the retrograde arm of the re-entry circuit. During tachycardia the QRS is wide and shows maximum pre-excitation, as antegrade conduction is performed exclusively through the anomalous beam. This type of tachycardia occurs in only 5% of patients with Wolff-Parkinson-White syndrome and is commonly associated with the presence of multiple accessory pathways (60%) [29]. Some regular tachycardias, apparently antidromic, due to maximum pre-excitation, in addition to the fact that they may be due to two anomalous bundles (one that leads in antegrade form and the other in retrograde form), may also occur in cases of atrial flutter or atrial tachycardia, with antegrade conduction through the accessory pathway, or in intranodal tachycardia, which makes the front of activation reach the ventricles preferably through a Kent beam.

Incessant supraventricular tachycardia

A rare form of re-entry arrhythmia, which uses the accessory pathway for retrograde conduction, is incessant supraventricular tachycardia [30]. Often the accessory pathway is hidden, and its presence is identified by the electrophysiological study. Individuals with this arrhythmia may have tachycardia almost all day. The onset of the first crisis may require only a minimal increase in sinus frequency. More recently, atrioventricular accessory pathways with long and decremental driving times have been identified. This type of accessory pathway participates in the most common form of incessant supraventricular tachycardia [31]; the anterograde conduction of the circuit is through the normal conduction system, while the retrograde conduction is through the Kent branch. Thus, in the surface electrocardiogram is manifested as a tachycardia with narrow QRS, with interval RP > P'R. During tachycardia, the P wave is negative in DII, DIII and aVF and often the crisis starts without lengthening of the PR; tachycardias are more frequent in children. Most accessory pathways have a septal location with atrial insertion in the area of the coronary sinus opening, and their medical treatment is usually ineffective.

Atrial fibrillation

Of all the tachycardias that can be modulated via the accessory pathway, the most common is atrial fibrillation. Its actual incidence is difficult to establish; however, it has been around 12% [32].

The electrocardiographic diagnosis of atrial fibrillation related to Wolff-Parkinson-White syndrome should be suspected in the face of irregular frequency tachyarrhythmia with widened complexes presenting [33]:

a) High-speed ventricular frequency, above 185 beats per minute,

b) Presence of initial fillings (delta wave) in the extended complexes,

c) Location of an accessory pathway in the beats conducted with pre-excitation,

d) Different degrees of QRS widening.

e) Presence of fine complexes (without aberrant driving) which may be early or late. Sometimes they alternate series of beats with or without pre-excitation.

Regarding the production mechanism of atrial fibrillation, several factors are postulated, perhaps the most important being:

a) Spontaneous passage from reciprocating tachycardia to atrial fibrillation, considered the most frequent mechanism and probably responsible for spontaneous atrial fibrillation [34];

b) Exaggerated atrial vulnerability per se or due to the presence of the accessory pathway [34].

An important aspect is the behavior of the atrioventricular conduction during the atrial fibrillation crisis.

The production of very short RR cycles during atrial fibrillation appears to be the main determinant for the development of ventricular fibrillation. Klein [35] found in his series of patients with stimulation-induced atrial fibrillation a shorter RR interval between two pre-excited beats equal to or less than 250 msec.

Another type of arrhythmia seen in Wolff-Parkinson-White syndrome is atrial flutter. Typically, flutter impulses are transmitted to the ventricle through the anomalous bundle by the shortest refractory period of the accessory pathway compared to that of the atrioventricular node.

The result is tachycardia with very high ventricular frequency and entirely abnormal ventricular activation: complexes that are very crowded and of significant duration.

The same is true in cases of atrial tachycardia in the presence of Wolff-Parkinson-White syndrome. Another type of arrhythmia modulated by the accessory pathway is intranodal tachycardias associated with Wolff-Parkinson-White [36] syndrome. In intranodal reentrant tachycardias in patients with the accessory pathway, the QRS complex is normal, differential diagnosis with orthodromic atrioventricular supraventricular tachycardias is smooth, not the distinction between antidromic atrioventricular tachycardia and intranodal tachycardia with pre-excited QRS. The presence of two accessory routes Kent type can lead to tachycardias that use each of them in a sense [37]: Anterograde and retrograde.

Several cases have been published. In all of them, the frequency was the same or more than 180 beats per minute and the QRS was widened to look like maximum pre-excitation.

Ventricular fibrillation and sudden death

In patients with Wolff-Parkinson-White syndrome, sudden death is a very rare episode in asymptomatic patients; the risk is 0.1% per year per patient, increases to 1% in those who have reciprocating tachycardias and reaches 5.6% in those who have atrial fibrillation crisis with RR less than 250 msec [38].

Other predictors of risk for developing ventricular fibrillation are the presence of multiple accessory pathways and that the patient suffers not only from atrial fibrillation with rapid ventricular frequency but also tachycardias by circular movement through the accessory pathway.

However, it is essential to consider:

a) Ventricular fibrillation may be the most common form of ventricular fibrillation [35] first clinical manifestation of Wolff-Parkinson-White

b) Wolff-Parkinson-White syndrome it is not necessary that the WPW syndrome is evident for a patient to develop atrial fibrillation with a rapid ventricular response through an accessory pathway [39].

Torner [40], in a retrospective review of 23 patients who had ventricular fibrillation, found that only one patient was receiving antiarrhythmic drugs at the time of sudden death. It is possible that class I and DIII drugs, although in many cases they cannot prevent recurrence of atrial fibrillation crisis in subjects with accessory pathways, at least prevent them from developing ventricular fibrillation. As activation differences occur in atrial fibrillation from beat to beat, they cause a large dispersion of the ventricular refractory period, which favors the appearance of multiform ventricular tachycardias and ventricular fibrillation, the cause of sudden death in patients with Wolff-Parkinson-White syndrome [41].

Identification of multiple pathways

The incidence of multiple pathways [42,43] in patients with Wolff-Parkinson-White syndrome is approximately 5 - 15%. Gallagher and Sealy found that of 161 patients who underwent surgical ablation [17] (11%) had multiple pathways. Ebstein's anomaly [26] also has a higher incidence of various pathways. In the authors' center, the incidence of numerous pathways was 18/465 patients (3.9%), in six patients associated Ebstein anomaly was observed (33%). Wellens and Durrer [33] demonstrated that multiple pathways may have different electrophysiological properties and that antiarrhythmic drugs may have different effects on accessory pathways. Klein [35] observed that the existence of various pathways has adverse prognostic implications. The incidence of more than one accessory pathway is higher among patients with WPW syndrome who develop ventricular fibrillation.

History confirms the importance of definitive treatment in this subgroup of patients either by surgery or radiofrequency ablation [37,42,43].

Multiple pathways should be suspected when there are variations in the morphology of the polarity of the delta wave vector and QRS complex in sinus rhythm, or an antidromic tachycardia in a patient with WPW syndrome. In patients with WPW and atrial fibrillation, the presence of QRS complexes with changes in their morphology also suggests multiple accessory pathways. Such findings were found in 34% of patients.

The location of accessory pathways during the electrophysiological study is based on the following:

1. Two or more distinct patterns of ventricular pre-excitation are demonstrated in accessory pathways capable of anterograde driving:
   a) During stimulation at different frequencies from the same point;
   b) During stimulation at the same frequency from different points;
   c) After extra-stimulus points when reaching a critical coupling;
   d) After long diastolic pauses, as long as the phenomenon is reproducible and the AV intervals are constant; these diastolic pauses may be observed during vagal stimulation or after interruption of tachycardia;

e) Initiation of an antidromic tachycardia whose pre-excitation pattern is different from that detected in other pre-excited QRS complexes in the same patient.

2. In accessory pathways with retrograde conduction, two retrograde atrial activation sequences are observed, corresponding to ventricular conduction through an accessory pathway:
   a) Two sequences, both eccentric, either during ventricular stimulation or orthodromic tachycardia;
   b) Two retrograde atrial sequences, although one is concentric, if atrial pre-excitation is obtained when the ventricle is stimulated at the time the bundle of His is refractory.
3. Two accessory pathways should be suspected in patients with highly discordant anterograde and retrograde pre-excitation patterns, e.g., right-type anterograde pre-excitation with the left lateral sequence.
4. Presence of a new or different accessory pathway after radiofrequency ablation [48].

When both pathways are close to each other, their identification in the electrophysiology laboratory can be much more difficult.

However, radiofrequency ablation in one of them makes it possible to identify accessory pathways. Klein [35] reported a patient with three accessory pathways very close to each other in the right posterolateral atrioventricular region that he detected in a histopathological study. It should also not be forgotten that, on occasion, the fibers of Kent auriculoventricular.

May not be straight but oblique. In these cases, atrial and ventricular insertion may be different and simulate multiple pathways. Jackman used his orthogonal catheter in coronary sinus mapping and identified an oblique course of the accessory pathway in 39 of 43 recorded pathways. It must, therefore, be demonstrated in the electrophysiological study that there are indeed two accessory pathways. McClelland suggested that the same accessory pathway can often be composed of multiple fibers. That is why in some cases he performed radiofrequency ablation in a segmental form on the mitral or tricuspid atrioventricular rings. Tai and Lau found five 2 cm wide band left lateral accessory pathways that could only be removed by radiofrequency ablation in a long segment of the atrioventricular sulcus. Chen performed radiofrequency ablation on a patient presenting multiple pathway associations with intranodal re-entry tachycardia. Finally, cases are also reported of multiple accessory pathways that are associated with auriculofascicular fibers or Mahaim fibers.

Lethal potential of the accessory pathway

The electrophysiological study also makes it possible to evaluate the lethal potential of the accessory pathway, by analysis of the anterior refractory period of the Kent beam. In general, it can be said that refractory periods greater than 350 msec do not usually develop rapid ventricular responses in cases of atrial fibrillation. The ajmaline or procainamide [50] test has been used to identify patients with a short refractory period, and also helps predict what the oral effect will be. Accessory pathways with an anterior basal refractory period greater than 300 msec respond well and are frequently blocked with antiarrhythmics; when the figure is less than 300 msec they may or may not respond regardless of the value of the refractory period.

Establish the presence of the tachycardia mechanism not related to the accessory pathway (intranodal tachycardia, atrial tachycardias, ventricular tachycardia).

Finally, to perform definitive treatment by radiofrequency ablation of the accessory pathway and assess the outcome of treatment.

Natural history and prognosis

In neonates tachycardias have a low recurrence, which increases from the first month of life. If the tachycardia is rapid and coexists with congenital cardiopathies, it could become serious. In general, the behavior is not this and 50% have no episodes of tachycardia, and when they exist respond to drug treatment [45].

From the year of life onwards, the frequency and intensity of crises tends to decrease, to the extent that some patients do not require treatment. The electrocardiographic pattern of Wolff-Parkinson-White syndrome by 10 to 50% disappears [46].

This seems to be due to the complete development of the atrioventricular ring at this age, with separation of atria and ventricles and interruption of septal pathways. It has also been proven that cardiac parasympathetic hyperactivity can suppress the "occult" anterograde unidirectional blockade of the anomalous pathway and it has been postulated that the spontaneous remission, not uncommon, of Wolff-Parkinson-White syndrome in the newborn, when fetal vagal predominance may persist, may be due to a more mature type of vagosympathetic equilibrium being achieved [47].

The opposite mechanism would be the cause of the appearance of pre-excitation in acute infarction, attributed to vagal hyperactivity.

There are cases of spontaneous remission of Wolff-Parkinson-White syndrome in adults, in whom, from the clinical-anatomopathological point of view, the most convincing explanation is the described pictures of sclerosing alterations that affect the accessory pathway [48].

The overall prognosis for Wolff-Parkinson-White syndrome is usually good; the existence of a small number of cases of sudden death seems incontrovertible. This has motivated efforts to detect a subgroup of patients at high risk of sudden death [49].

Clinical approach

Asymptomatic patients

It is generally accepted that an asymptomatic patient with Wolff-Parkinson-White syndrome does not need any treatment. The universality of this assertion is questionable, since if the anterior refractory period of the accessory pathway is short and the patient presents with atrial fibrillation, the ventricular response could be very rapid and degenerate into ventricular fibrillation. In this sense, it is important to try to identify patients at high risk of sudden death. If the Wolff-Parkinson-White syndrome has intermittent behaviour, the refractory period is usually long and the patient should be considered low risk. In subjects with manifested Wolff-Parkinson-White ajmaline can be administered, 0.75 mg/kg in three minutes intravenously. If pre-excitation is maintained, the probability that the anterior refractory period of the accessory pathway is short (< 270 msec) is very high [50] and the patient should be sent to a specialized center for a more thorough assessment.

Many patients with overt pre-excitation remain asymptomatic throughout their lives. However, it is known that there is a very slight, but real, risk of cardiac death in patients with Wolf-Parkinson-White syndrome. This observation motivates interest in risk stratification in patients with pre-excitation to establish which may be considered "high risk" and the possibility that they may require further evaluation or treatment.

When patients with Wolf- Parkinson-White syndrome who were resuscitated from cardiac arrest are studied in the electrophysiology laboratory, a very brief anterograde refractory period of the accessory pathway is usually observed. It is presumed that in these patients atrial fibrillation is generated spontaneously, with rapid conduction to the ventricles due to the short refractory period. The arrhythmia then degenerates into ventricular fibrillation. In this situation it is possible hypothesize that asymptomatic patients with brief refractory periods of the antegrade accessory pathway may present a risk of sudden cardiac death. However, scientific data do not support this conclusion.

The risk of sudden cardiac death [39] in Wolff-Parkinson-White syndrome is approximately 1/1,000 patient-years, which is a very low number fortunately.

However, when invasive electrophysiological studies have been performed in groups of patients with pre-excitation but no history of arrhythmias, a considerable percentage of cases with short refractory periods of the accessory pathway has been observed. Evidently, if invasive tests are applied in a systematic manner, a percentage of patients with a possible high risk would be identified much higher than the percentage of patients actually at risk. In addition, most patients who are asymptomatic with pre-excitation will continue to be so in the future, regardless of the results of the electrophysiological study. Thus, there is no systematic indication for the application of electrophysiological studies in asymptomatic patients with pre-excitation.

Patients with palpitations without documented tachyarrhythmia

When the patient refers sudden onset and end palpitations without apparent precipitating factors and of a trimmed and recurrent nature, the possibility of having paroxysmal tachycardias is high, even if it has not been demonstrated electrocardiographically. When the duration and frequency of palpitations do not interfere with the patient's quality of life, acute pharmacological treatment is not necessary. An evaluation of the pre-excitation should be carried out, including the ajmaline test, and instructed in the practice of vagal maneuvers, recommending periodic revision.

Symptomatic patients with documented tachycardia

If the tachycardia is narrow QRS, it is most likely orthodromic tachycardia by circular motion. If the tachycardia has wide QRS, it may be a tachycardia similar to the previous one with aberrant conduction or with organic branch block; there is a possibility, however, that it is an antidromic tachycardia. This distinction is important, as the latter is much easier to control pharmacologically; however, they also require electrophysiological study, as they are often associated with multiple pathways. The treatment of the patient must be related to the clinical impact of the tachycardia. Only those cases in which the duration, incidence, age of the patient or clinical repercussions of tachycardia crises compromise the quality of life or life expectancy should receive antiarrhythmic treatment, although nowadays it is thought that radiofrequency ablation should be the therapy of choice. Symptomatic patients with documented atrial fibrillation are patients who must undergo electrophysiological study and radiofrequency ablation of the accessory pathway in the same session [51,52].

Conclusion

As a conclusion to the writing, we consider it very appropriate to comment that, on many occasions, patients with tachycardia greater than 160 bpm, with narrow QRS complexes, PR interval in ranges and the subtle presence of a delta wave are presented to us in the emergency department in too many occasions. When patients are assessed by physicians as a paroxystic tachycardia with narrow QRS complexes, the patient the patient is usually undergoing administration of Adenosine triphosphate in bolus rapid with the result of a major tachycardia. Although this effect is not pathognomonic of the WPWS, it can lead to a worsening of the picture. This was the case with the patient illustrated in figure 2.

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