Preexcitation syndromes: diagnostic consideration in the ED

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Abstract Preexcitation syndromes are a common cause of paroxysmal tachycardias presenting to the ED. Emergency physicians should be familiar with the common electrocardiographic manifestations of preexcitation, particularly the Wolff-Parkinson-White abnormality, as these conditions require specific therapeutic management. This article reviews the pathophysiology of preexcitation, along with the electrocardiographic findings of Wolff-Parkinson-White and its associated tachyarrhythmias. © 2009 Elsevier Inc. All rights reserved.

1. Introduction

Preexcitation is defined as a “premature activation of the ventricular myocardium by an impulse that travels by an anomalous path and...avoids physiologic delay in the atrioventricular junction” [1]. Preexcitation as a finding on electrocardiography (ECG) has come to be known as the “Wolff-Parkinson-White abnormality,” after the authors who first systematically described the appearance of preexcitation and its association with paroxysmal tachycardia. Several variations of preexcitation syndrome have been described, being differentiated from one another by the anatomy of the anomalous path, or accessory pathway. Examples of accessory pathways include atrioventricular (AV) tracks (extensions of atrial myocardium that connect to ventricular myocardium by bridging the electrically insulating fibrous AV septum), nodofascicular tracks (connection between the AV node and a distal portion of the ventricular fascicles), and other variations such as nodoventricular, fasciculoventricular, and atriofascicular tracks. Most of these bypass tracts can result in ventricular preexcitation and can predispose to reentry arrhythmias manifested as the clinical syndrome of preexcitation, paroxysmal tachycardia, and its associated symptoms of palpitations, dizziness, and/or syncope.

Accessory AV pathways are responsible for most cases of Wolff-Parkinson-White (WPW) syndrome. Wolff-Parkinson-White syndrome is commonly used in reference to the finding of preexcitation on the ECG, and syndrome is attached if paroxysms of tachycardia occur. Findings suggestive of WPW syndrome on 12-lead ECG analysis are found in 1.5 to 3.1 per 1000 persons in Western populations [2-4,8]. The actual incidence of WPW syndrome may be higher because not all cases of preexcitation are evident on standard 12-lead ECG in the absence of paroxysmal tachycardia. Some cases require high-resolution ECG or invasive electrophysiologic studies for diagnosis (referred to as “concealed” WPW syndrome) [5].

An accessory pathway creates ventricular preexcitation by conducting atrial impulses to the ventricle faster than...
the native atrioventricular node, potentially providing a substrate for reentry arrhythmias and paroxysmal tachycardia. Paroxysmal tachycardia can present clinically as palpitations, dizziness, or frank syncope. Sudden death is also possible with WPW syndrome in the setting of extremely rapid and sustained tachycardia. However, this is rare with an estimated incidence of 1 per 1000 patient-years in persons with known ventricular preexcitation [6-11]. This article will primarily focus on a description of the pathophysiology of and 12-lead ECG manifestations of WPW syndrome.

2. A brief history of WPW syndrome

Drs Wolff, Parkinson, and White first described the syndrome now known as WPW syndrome in 1930 in a report of 11 patients with a short P-R interval, “bundle-branch block,” and paroxysmal supraventricular tachycardia and/or atrial fibrillation [12]. Descriptions of similar ECG and clinical findings had been published as early as 1915 but were not ascribed to a single syndrome [13]. It was not until 1959 that accessory AV pathways were determined to be responsible for the findings in WPW syndrome, although such pathways had been described in 1893 by Stanley Kent [14]. This discovery has allowed electrophysiologists to map out and ablate accessory pathways, albeit initially with varying rates of success and complications depending on the location of the pathway. The introduction of radiofrequency energy for use in ablative procedures in the late 1980s allowed for tremendous improvements in the success rates of this procedure. In clinical practice today, nearly all accessory pathway locations are amenable to attempts at ablative therapy, with reported recurrence rates between 2% and 9% [15]. More recently, advances in the understanding of WPW syndrome have been made on the molecular level with the discovery of a familial form of WPW syndrome linked to a single gene [16].

3. Demographics

Although in most cases there is no strong pattern of genetic inheritance, some cases of WPW syndrome occur either as part of a single inherited trait of preexcitation or as part of a broader genetic syndrome with more extensive cardiac disease [17]. Emergency physicians should have a heightened awareness for the possibility of preexcitation when evaluating these patients. Table 1 lists those syndromes associated with WPW syndrome.

4. Pathophysiology

Before discussing abnormal conduction, it is illustrative to review normal heart conduction and ECG findings (Fig. 1). Preexcitation occurs when an atrial impulse is conducted in an anterograde (atria to ventricle) fashion down an AV accessory pathway before the AV node-His-Purkinje axis begins depolarization. How this manifests clinically and on the ECG depends on how much faster the atrial impulse is conducted down the accessory pathway into the ventricular myocardium as opposed to the AV node-His-Purkinje axis. The classic “short P-R” interval description of WPW syndrome is a result of premature ventricular depolarization mediated by the AV accessory pathway. This early ventricular depolarization manifests on the ECG as both a short P-R interval and a slurred upstroke in the R

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Fig. 1 The impulse formation and conduction process in a patient with normal sinus rhythm. “A” denotes transmission of the impulse through intraatrial pathways to the AV node. “B” denotes transmission of the impulse through the AV node and into the His-Purkinje system. “C” represents the propagation of the impulse through the ventricular myocardium via the specialized conduction system. The ECG complex is the surface electrical manifestation of this process with a normal PR interval and QRS complex, exhibiting the normal conduction through the atria to the AV node (“A” and “B”) and ventricles (“C”), respectively.
wave, referred to as the delta wave. The delta wave represents relatively disorganized and slow ventricular depolarization via the accessory AV pathway and ends at the point at which AV nodal conduction begins to successfully propagate, thus creating a terminal narrow QRS complex (Fig. 2). Other ECG manifestations may include a prolonged (>120 milliseconds) QRS complex and secondary ST-T waves changes that are discordant to the QRS vector.

The presence of a delta wave indicates that AV accessory pathway conduction is faster than AV node conduction, creating a substrate that can predispose to preexcitation arrhythmias in the setting of either AV nodal blockade or rapid rates of atrial depolarization, such as atrial fibrillation. Rates of pre-excited tachycardias depend on the length of the refractory period of the accessory pathway tissue, which is usually longer than that of the AV nodal tissue. This difference in refractory period length provides the substrate by which a premature atrial depolarization can incite AV reentry tachycardia (AVRT).

5. Electrocardiographic features of WPW syndrome

There are 3 ECG criteria for diagnosing WPW syndrome; a short (<120 milliseconds) PR interval, a
delta wave, and an anomalous QRS configuration. The last of these is the most variable factor and depends on the location of the bypass tract. The most common locations for accessory AV pathways are, in decreasing order of frequency, left lateral (free wall), posteroseptal, right free wall, and anteroseptal.

Fig. 4  Right posteroseptal accessory AV pathway. Occasional narrow QRS complexes without evidence of preexcitation are seen (arrows). The QRS and delta waves in the inferior leads are negative during preexcitation, simulating prior inferior myocardial infarction. The right ventricular origin is indicated by the negative delta wave in V1, which promptly becomes upright in V2. Delta waves are best seen in the lateral precordial leads.

Fig. 5  Left posteroseptal accessory AV pathway. As in Fig. 4, negative delta and QRS deflections are seen in leads III and aVF, mimicking prior inferior myocardial infarction. The left ventricular origin of this pathway is indicated by the positive delta wave in V1.
5.1. Left lateral accessory pathway

The most commonly found accessory AV pathways insert into the lateral free wall of the left ventricle (Fig. 3). On the ECG, preexcitation via this pathway results in a positive delta wave and QRS in V1, upright delta and QRS in the inferior leads (as opposed to posteroseptal locations, see below), and Q waves with negative to isoelectric deflections.

Fig. 6 The same patient in Fig. 4, now with suppressed accessory pathway conduction. Procainamide was administered to this patient after an episode of atrial fibrillation with rapid ventricular conduction and a wide QRS complex. Note the inverted T waves in the inferior leads, which are the result of ventricular memory of preexcitation in these leads.

Fig. 7 Wide QRS complex tachycardia (WCT) with retrograde AV node conduction in the patient with WPW syndrome, an ART. This form of WCT uses the accessory pathway as the anterograde limb of the reentry loop (“A”) with the AV node functioning as the retrograde limb (“C”). The reentry loop involves the ventricular (“B”) and atrial (“D”) myocardium. The impulse arrives at the ventricle via the accessory pathway and is propagated distally via the ventricular myocardium; the QRS complex is widened in that the His-Purkinje system is not used to rapidly and efficiently propagate the impulse, thereby producing a wide QRS complex.

Fig. 8 Narrow QRS complex tachycardia (NCT) with antegrade AV node conduction in the patient with WPW syndrome, an orthodromic reciprocating tachycardia (ORT). This form of NCT uses the AV node as the anterograde limb of the reentry loop (“A”) with the accessory pathway functioning as the retrograde limb (“C”). The reentry loop involves the ventricular (“B”) and atrial (“D”) myocardium. The impulse arrives at the ventricle via the AV node and is propagated distally using the His-Purkinje system, thereby producing a narrow QRS complex.
in any of the lateral leads (I, aVL, V5, or V6). These Q waves can give the mistaken impression of a former high lateral myocardial infarction and can give the appearance of right axis deviation.

5.2. Posteroseptal accessory pathway

The second most common location for an AV bypass tract is in the posteroseptal region of the AV groove. Ventricular preexcitation from this pathway manifests on the ECG with negative deflecting delta waves and QRS complexes in the inferior leads (II, III, aVF). Preexcitation via this pathway is thus often mistaken for prior inferior myocardial infarction and can give the appearance of left axis deviation. Those that are situated in the right ventricle show negative delta and QRS deflections in V1 (Fig. 4), whereas those in the left ventricle show positive deflections (Fig. 5). Acute suppression of accessory pathway conduction can also result in ST-T wave abnormalities suggestive of ischemia (Fig. 6).

6. Electrocardiographic features of WPW syndrome

Accessory AV pathway activity is determined by the electrophysiological properties of the pathway tissue, including the rate of tissue depolarization, length of the refractory
period, and differentials in these properties when confronted with anterograde (atria to ventricle) or retrograde (ventricle to atria) directions of conduction. An accessory AV pathway can be seen as 2 separate tracks, one running from the atria to the ventricle and the other heading in the opposite direction. The clinical syndrome of WPW can likewise be categorized into 2 separate disorders: (1) those arrhythmias that result from anterograde accessory AV pathway conduction (resulting in a wide QRS complex) and (2) those that use retrograde accessory AV pathway conduction (resulting in a narrow QRS complex). Examples of the first include atrial tachycardias (ie, atrial fibrillation) that result in premature and disorganized ventricular depolarization via the accessory AV pathway or the uncommon antidromic reciprocating tachycardia (ART) in which an impulse becomes entrained in a circuit involving the accessory pathway and the AV node, traveling in a retrograde (ie, antidromic) fashion up the AV node from ventricle to atria, and in turn down the accessory pathway from atria to ventricle (Fig. 7). The same circuit may run in the opposite direction, in which case it is called orthodromic reciprocating tachycardia (ORT), in which the retrograde conduction from ventricle to atria occurs through the accessory AV pathway instead of through the AV node (Fig. 8). Orthodromic reciprocating tachycardia is much more common than ART and largely encompasses the narrow QRS complex variety of WPW syndromes. These 2 entities are clinically distinct, as the acute management of wide and narrow QRS arrhythmias mediated by accessory AV pathways is different in the acute setting. However, the therapeutic implications of ablative therapy are the same for both entities and both warrant referral for electrophysiologic evaluation.

6.1. Orthodromic reciprocating tachycardia or AVRT

ORT, or AVRT, is a common presentation of WPW syndrome, as most accessory AV pathways are in fact only capable of retrograde conduction. Atrioventricular reentry tachycardia can be initiated in the setting of a premature atrial complex when the refractory period of the accessory AV

Fig. 11 Preexcited atrial fibrillation in the patient with WPW syndrome. The ECG waveform is an irregular, wide QRS complex tachycardia with delta wave and beat-to-beat variation in QRS complex morphology. The delta wave, not present in all beats, signifies the depolarization of the ventricular myocardium due to the impulse arriving via the accessory pathway. The beat-to-beat variation in the QRS complex results from the varying contribution to ventricular depolarization arriving via the accessory pathway and the AV node.

Fig. 12 Atrial fibrillation with WPW syndrome. Note the variety of QRS morphologies, but the relatively constant amplitude of voltages, in contrast to the wide variation in QRS voltages seen with PVT. A probable fusion beat is seen (arrow).
Fig. 13  Continued.
pathway is longer than that of the AV node. For this to occur, the premature atrial complex must occur early enough in the cardiac cycle to be blocked from anterograde conduction down the accessory pathway but late enough to allow for successful conduction through the AV node. As a result, a normally conducted impulse from the atria is spread throughout the ventricles by the AV node-His-Purkinje axis (referred to as orthodromic conduction) and passes back into the atria in a retrograde fashion though the now excitable accessory AV pathway (Fig. 8). As long as the atrial fibers are not refractory to depolarization at the time this impulse arrives, the atria will depolarize prematurely (ie, before the sinus node depolarizes), setting up a reentrant circuit. Rates of ventricular depolarization typically range between 150 and 250 beats per minute. A representative ECG of AVRT is shown (Fig. 9). Note that the p wave is distinctly seen following the QRS, giving a “short r-p interval” tachycardia. This is in contrast to the typical form of AV nodal reentry tachycardia (AVNRT), in which the p wave is often buried in the QRS complex due to simultaneous activation of both atria and ventricles. However, a discrete p wave can be appreciated following the QRS complex in up to 30% of AVNRT, making this an imperfect discriminator between AVRT and AVNRT, and thus a nonspecific indicator for the presence of an accessory AV pathway [18].

Another confounding factor in detecting the presence or absence of an accessory AV pathway is that accessory AV pathways do not always conduct in an anterograde direction and thus do not demonstrate preexcitation on standard 12-lead ECGs [5]. These are commonly referred to as “concealed” accessory pathways. Concealed accessory AV pathways may still facilitate arrhythmias or simply lay inactive and/or dormant. Atioventricular reentry tachycardia in the absence of findings of WPW syndrome on a sinus rhythm ECG is thus also referred to as “concealed” WPW syndrome.

6.2. Wolff-Parkinson-White syndrome with atrial fibrillation

Another concerning feature of AVRT is that it can disorganize into atrial fibrillation, which can have disastrous consequences in patients with accessory AV pathways capable of anterograde conduction (Fig. 10). Atrial impulses can reach an accessory pathway at a rate of 300 to 400 times per minute and may result in hemodynamic instability due to rapid rates of ventricular response, far in excess of that allowed by the AV node-His-Purkinje axis. Although accessory pathways typically have longer refractory periods than the AV node, and thus should not be capable of pacing the ventricles faster than the AV node, this refractory period can shorten markedly at high rates and allow for a rapid rate of conduction than the AV node. This manifests as a wide QRS complex tachyarrhythmia with an irregular rate, the differential of which includes WPW syndrome with atrial fibrillation, polymorphic ventricular tachycardia (PVT), and atrial fibrillation with interventricular conduction delay. Distinguishing between these rhythms can be extremely difficult. Having access to a previously recorded ECG in these cases is critical.

Polymorphic ventricular tachycardia tends to exhibit chaotic and haphazard QRS complex voltages as opposed to the other 2 entities and thus is the most likely diagnosis if wide voltage variation in QRS complexes is present. These patients may have a history of alcoholism, poor nutritional status, or excessive diuretic use, all of which predispose to hypomagnesemia and the torsades de pointes variety of PVT. An ECG obtained before the tachycardia may reveal a prolonged QT interval in these individuals. Alternately, PVT can be sequelae of preexisting ischemic heart disease, congenital long QT syndrome, or acute coronary syndromes. In most cases, the ECG will suggest the correct diagnosis based on the chaotic electrical nature of this rhythm.

In contrast, AF with interventricular conduction delay usually manifests QRS complexes that are consistent in appearance across the ECG. Rates of ventricular depolarization in these cases tend not to exceed 190 to 200 beats per minute, which is usually the upper limit of AV nodal conduction cycles. Such ECGs in rare instances may also show an occasional narrow complex QRS without a delta wave if the conduction delay is rate induced, which would alternatively suggest the correct diagnosis. Prior ECGs showing sinus rhythms with similar morphologies of ventricular conduction delays strongly suggest this diagnosis.

Wolff-Parkinson-White syndrome with AF should be suspected based on clinical history and ECG features. Patients who present with a history of multiple episodes of sudden syncope in the absence of known cardiovascular disease are particularly suspicious. The ECG can demonstrate a variety of findings, with QRS morphologies varying widely depending on the location of the accessory pathway and the relative rate of AV nodal conduction (Fig. 11). The presence of any simultaneous conduction via the AV nodal
pathway may manifest as a fusion beat QRS complex, which can be an additional clue to this rhythm (Fig. 12). Patients in this rhythm can deteriorate quickly and require emergent management, either with antiarrhythmic therapy (Figs. 13A-D) or electrical cardioversion.

7. Pediatric considerations

Wolff-Parkinson-White syndrome is a relatively common cause of supraventricular tachycardia (SVT) in young children. Approximately 60% of children with SVT will manifest their arrhythmia within the first year of life, most often by 3 to 4 months of life [19]. Up to 20% of newly diagnosed SVT in children will reveal WPW syndrome after conversion to sinus rhythm [20]. Many of these children will lose their predisposition to tachycardia, as the electrical properties of the accessory pathways can change with age. These patients may present with new onset tachyarrhythmias to the ED, and emergency physicians should be familiar with the treatment in this population. Major deviations from adult management include a higher reliance on vagal maneuvers (ie, ice bags applied to the face to stimulate a diving reflex), more reliance on medical management, and the use of digoxin as a means of arrhythmia suppression. Syndromic associations are of particular importance in this group (Table 1).

8. Summary

Wolff-Parkinson-White syndrome is a rare but potentially lethal conduction disturbance that requires specific emergency management. Emergency physicians should thus be intimately familiar with common ECG manifestations. Asymptomatic cases warrant prompt cardiology referral on an outpatient basis, whereas symptomatic cases should be hospitalized in conjunction with electrophysiologic evaluation.

References