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An Interesting Case of Wolf – Parkinson White Syndrome in a Young Patient with Personal and Family History of Sensorineural Deafness.

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Summary

We report a case of pre-excitation WPW syndrome in a 17-year old man who was brought to the hospital by ambulance following the collapse. Wolff-Parkinson-White Syndrome type A was diagnosed from precordial leads. His electrocardiogram revealed short PR interval, delta waves and positive waves with dominant R in all pericardial leads. His blood results showed an isolated raised ALT level. Subsequent echocardiography was unremarkable with an ejection fraction of 55% apart from septal and inferior wall dyssynchrony. With regards to the past medical history, he had sensorineural deafness (SND) since childhood, and he also had a family history of SND. Consequently, he was transferred to the cardiac electrophysiology department at another hospital after consultation and underwent ablation. A successful post-ablation electrocardiogram revealed resolution of WPW syndrome signs and post-ablation features such as peak T waves.

Background

In Wolff–Parkinson–White (WPW) syndrome, accessory atrioventricular pathways (AP) are fibers that connect the atrium to the ventricle outside the normal atrioventricular nodal-His-Purkinje conduction system with an incidence of 0.1–0.3% in the general population and an associated sudden cardiac death risk of less than 0.6%. The atria and ventricles are electrically isolated from each other in a normal heart by nonconductive fibrous atrioventricular (AV) ring except at the AV node and bundle of His. Impulses are typically initiated from within the sinoatrial node, and conduction propagates to the ventricles via the His-Purkinje system. Patients with WPW have at least one additional accessory electrical pathway between the atria and ventricles bypassing the AV node resulting in premature electrical impulses and ventricular pre-excitation that also allows retrograde propagation of impulses. The accessory pathway can be associated with reentrant SVT and sudden death depending on the conduction characteristics.

The syndrome is further categorized into type A or B based on conventional 12-lead electrocardiography (ECG), with pre-excitation from the posterolateral base of the left ventricle (LV) in Type A and the right ventricular free wall or interventricular septum in Type B **[**1**]**. Patients may experience palpitations, dizziness, syncope, congestive heart failure, or sudden cardiac death (SCD). The ECG characteristics of WPW show a short PR interval of <0.12 seconds, slurring and the slow rise of the initial QRS complex (delta wave), a widened QRS complex with a total duration >0.12 seconds and abnormal ventricular repolarization. Atrioventricular pathways result in abnormal pre-excitation around the atrioventricular annuli and produce a dyssynchronous contraction of cardiac chambers **[**2**]**. Radiofrequency catheter ablation (RFCA) is a well-established treatment for patients with WPW syndrome associated with a tachyarrhythmias recurrence risk of less than 5%. The effectiveness of ablation is dependent on the precise localization of the AP **[**3**]**.

Case Presentation

A 17-year-old male patient was brought to the emergency department following collapse at home by ambulance service. He regained consciousness after two minutes and on further questioning, he alluded to the feeling of palpitation for the last three weeks. He was studying currently and was not involved in any strenuous physical activities. In terms of past medical history, he had sensorineural deafness since childhood. He also had two first-degree relatives with sensorineural deafness, however, there was no family history of sudden cardiac death. He denied any use of medications, alcohol, or illicit drugs.

His initial electrocardiogram illustrated features of WPW syndrome such as short PR interval, normal QRS complexes and delta wave. (ECG in figure 1). Cardiovascular examination was otherwise

unremarkable; chest X-ray and routine blood tests were normal apart from the raised level of ALT. The patient was discussed with the Tertiary Cardiac electrophysiology team and the patient was transferred for RFCA. He underwent electrophysiological studies and successful accessory pathway ablation (figure 2).

Investigations

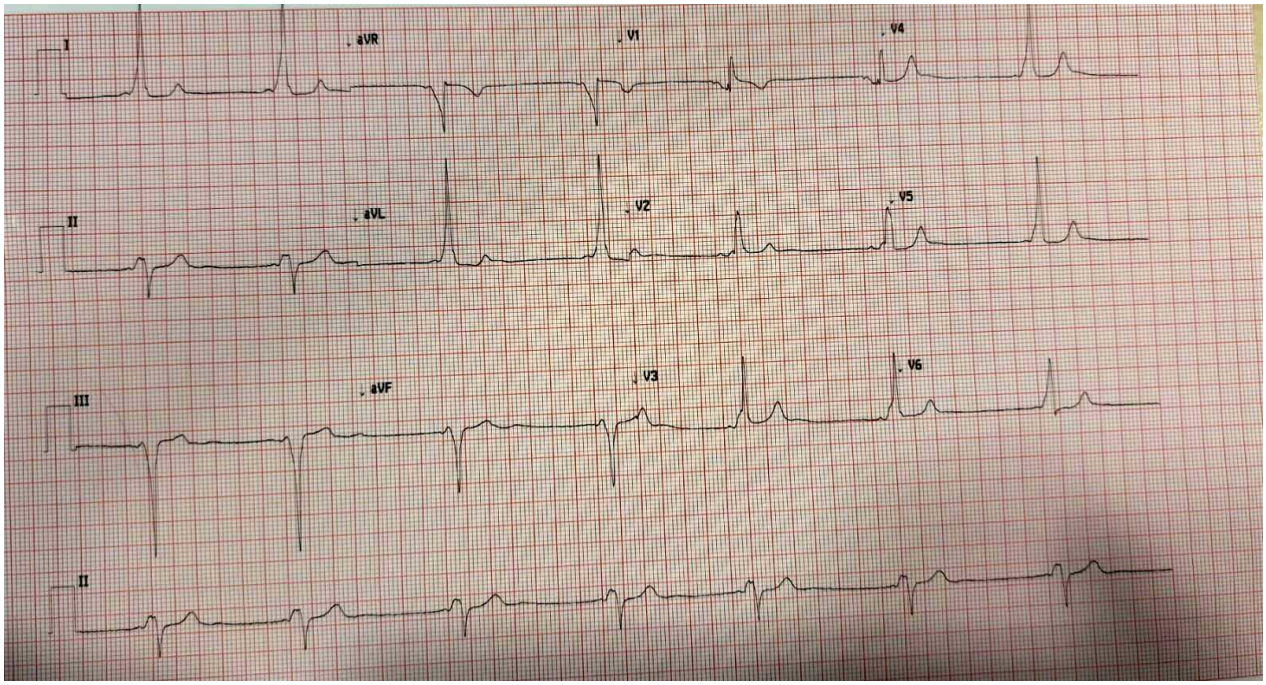

Figure1: The initial electrocardiogram which was taken in emergency department showed short PR interval, delta waves and positive waves with dominant R on all pericardial leads.

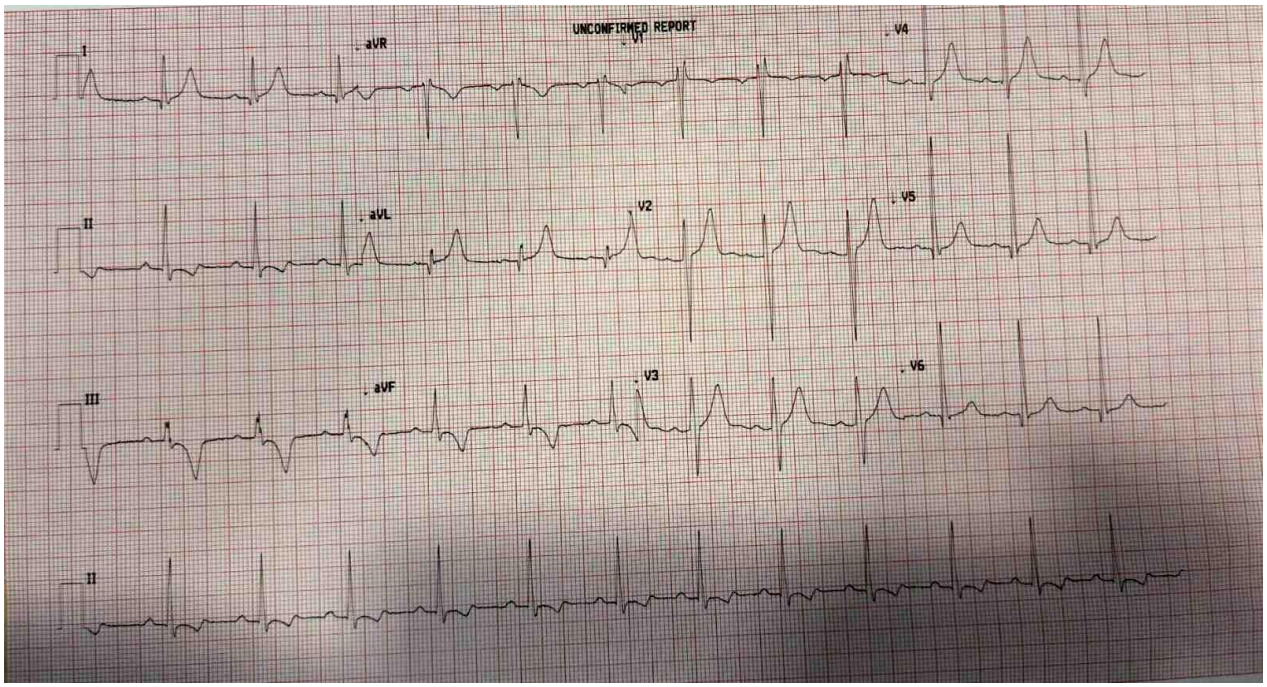

Figure 2: After successful ablation of postero-septal accessary pathway, the electrocardiogram showed sinus rhythm with no delta wave; peaked and deep T waves can be appreciated in the anterior and inferior leads respectively (classic post-ablation "pseudo inferior wall myocardial infarction" pattern), deemed as the evidence of a successful ablation.

Differential Diagnosis

The differential diagnosis of WPW includes structural heart diseases and arrhythmias. An important differential to rule out is hypertrophic obstructive cardiomyopathy (HOCM). Patients with HOCM can also present collapse and loss of consciousness. Patients with HOCM usually have hypertrophied left ventricular septum in the basal left ventricular (LV) segment and they may also have left ventricular outflow tract obstruction (LVOT), diastolic dysfunction, mitral regurgitation and left atrial enlargement if they have diastolic dysfunction. This patient had a normal echocardiogram ruling out HOCM.

The other differential diagnosis can be broadly divided into narrow complex and broad complex regular and irregular tachycardias. Regular narrow complex tachycardia may include sinus tachycardia, atrial tachycardia, atrial flutter (with regular AV block), AVNRT, AVRT, junctional tachycardia. The ECG and electrophysiology study supported the diagnosis of WPW in this patient.

Irregular narrow complex tachycardia can include atrial fibrillation, atrial flutter (with variable AV block), multifocal atrial tachycardia, sinus tachycardia with ectopic beats. Regular wide complex tachycardia includes ventricular tachycardia, accelerated idioventricular rhythm, paced rhythm,

artifact, and any SVT associated with aberrant ventricular conduction or accessory pathway that can be triggered by drugs, metabolic and electrolyte abnormalities. Finally, the Irregular wide complex tachycardia includes torsade de pointes, non-sustained ventricular tachycardia, any cause of an irregular narrow complex tachycardia associated with abnormally aberrant conduction.

Brugada syndrome is another possible differential diagnosis in young patients presenting with collapse. It has three types of repolarization patterns previously described. Type 1 is characterized by a prominent coved ST-segment elevation displaying J wave amplitude or ST-segment elevation ≥2 mm or 0.2 mV at its peak followed by a negative T-wave, with little or no isoelectric separation. Type 2 also has a high take-off ST-segment elevation, but the, J wave amplitude (≥2 mm) gives rise to a gradually descending ST-segment elevation in this case that remains ≥1 mm above the baseline, followed by a positive or biphasic T-wave that results in a saddleback configuration. Type 3 is a right precordial ST-segment elevation of <1 mm of saddleback type, coved type, or both **[**4**]**.

Treatment

After confirmation of the diagnosis and discussion with the electrophysiology department, the patient underwent a successful radiofrequency ablation of WPW syndrome.

Outcome and Follow-Up

With regards to the treatment, the patient had successfully RFCA and converted to sinus rhythm. He was followed up in the outpatient department in 3 months and was symptom-free now.

Discussion

The prevalence of WPW syndrome is 2 per 1000 general population. This case serves as an important message that tachyarrhythmias in the presence of an accessory pathway may present with distinct electrocardiographic features, potentially leading to incorrect diagnoses and treatments that may be life-threatening.

To review the classical manifestations of WPW syndrome, it is significant to recall the presence of the bundle of Kent, also known as an accessory pathway through which fast anterograde conduction can outpace slower atrioventricular (AV) node conduction. This pathway results in a relatively quick depolarization of the ventricles, resulting in distinct ECG changes like a short PR interval, wide QRS complex, and the virtually pathognomonic delta wave. Moreover, in “concealed” WPW syndrome, it is difficult to discern any electrocardiographic anomalies at baseline, as the accessory pathway may not

conduct in an anterograde fashion. In the majority of WPW patients, paroxysmal AV reentrant tachycardia (AVRT) occurs via anterograde conduction through the AV node, followed by retrograde conduction through the bundle of Kent (orthodromic AVRT), producing a tachycardia with narrow QRS morphology. These patients typically do not demonstrate rapid preexcitation responses during AF, likely due to either anterograde conduction delay of the accessory pathway relative to the AV node or block (figure 3 & 4). Wide QRS tachycardia, in contrast, can occur in patients with antidromic AVRT, whereby anterograde conduction through the accessory pathway is followed by retrograde conduction through the AV node. This circuit may also transpire in patients with preexisting bundle branch blocks **[**5,6**]**.

During pre-excited AF, the atria can discharge at a rate higher than 300 impulses per minute, obscuring delta waves—the key electrocardiographic feature of WPW syndrome. The AV node normally blocks most of these impulses due to decremental conduction, an intrinsic repolarization property that allows the node to conduct more slowly when it receives faster signals. However, an accessory pathway without such a built-in delay makes 1:1 conduction possible, with ventricular rates reaching 300 bpm. Pre-excited AF is thus characterized as a malignant arrhythmia, as sudden cardiac death may result from this rhythm degenerating into ventricular fibrillation **[**6,7**]**. It is also important to remember that following ablation large, peaked T waves may appear in leads along with the delta wave, which was also notable, with the concordant polarity. Namely, in leads where the delta wave was positive (leads I, aVL, and V2-6 in our patient), T waves are positive as well. A classic post-ablation memory T wave pattern often perceived evidence of successful ablation.

Certain medications such as Beta-blockers, calcium channel blockers such as Diltiazem, Verapamil, Digoxin, and adenosine should be avoided in patients with WPW as they can accelerate the accessory pathway. Additionally, young patients with WPW may not show typical short PR interval and delta waves initially and young patients presenting with collapse should be properly investigated. Patients with WPW and tachycardia may have an underlying infection, pulmonary embolism, or dehydration. It is possible to have multiple accessory pathways in about 10% of cases. Two population studies put the rate of sudden cardiac death between 0.0002 to 0.0015 per patient-years for patients with WPW pattern and some risk factors place a patient at higher risk for sudden cardiac death, including male gender, age less than 35 years, history of atrial fibrillation or AVRT, multiple accessory pathways, septal location of the accessory pathway. Although WPW is a dangerous condition in certain cases, however prognosis for patients with WPW patterns has improved significantly as antiarrhythmic medications, and ablation techniques recently. RFCA can be curative in these patients and has high success rates with low rates of complications.

Another interesting fact about our patient was his strong family history of sensorineural deafness and he also suffered from sensorineural deafness. However, we could not establish any link between the two.

Figures 3 & 4 show the WPW accessory pathway.

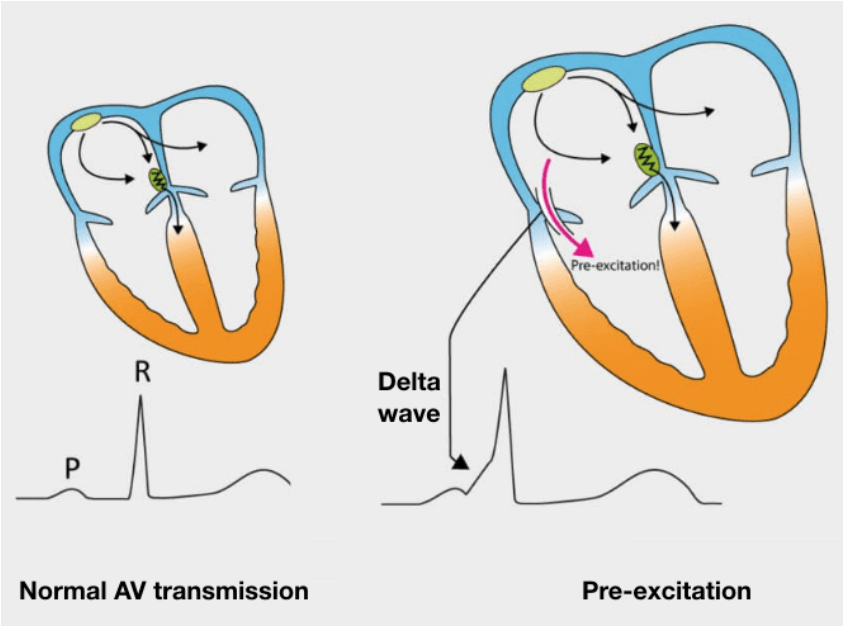

Figure 3

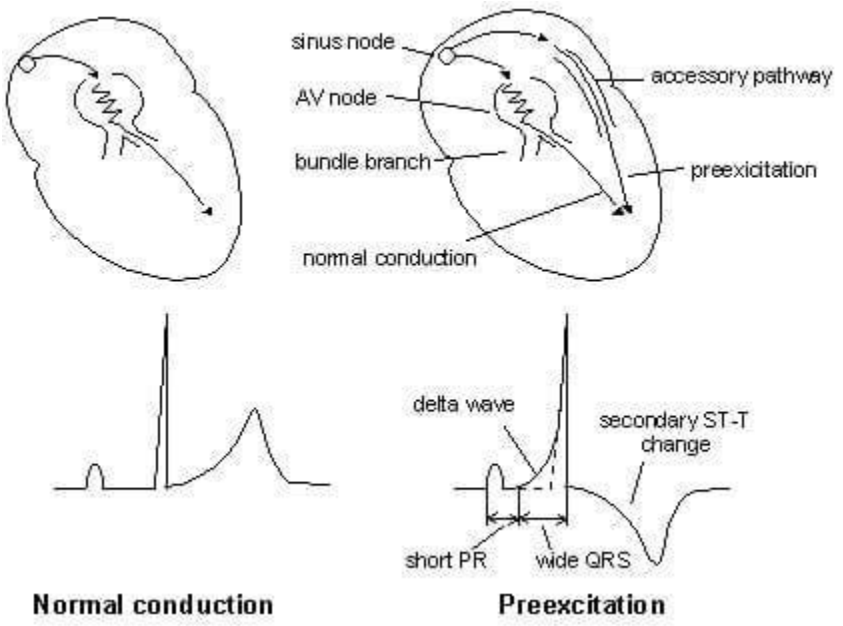

Figure 4

Learning Points/Take Home Messages

- Wolff-Parkinson-White (WPW) syndrome should always be suspected in a young patient presenting with symptomatic palpitations or collapses.
- It is always recommended to seek specialist advice in complex cases of tachyarrhythmias who need further evaluation and treatment accordingly.
- In Wolff–Parkinson–White (WPW) syndrome, the effectiveness of radiofrequency ablation treatment is dependent on the exact localization of the accessory atrioventricular pathway (AP) from where impulses generate.
- Young patients with WPW syndrome can die due to tachyarrhythmias and ventricular fibrillation (VF) if misdiagnosed particularly in athletes.

Patient's Perspective

It was a frightening experience for me to suddenly pass out like this. I started to notice racing of heart a few times for three weeks before my sudden passing out. I was completely shocked when I was told that my heart was having a rhythm problem and that I required surgery. I was glad to have had successful surgery and my symptoms completely resolved with that. I am thankful to the cardiology team at the hospital who looked after me and kept me updated all this time. They were very reassuring and did not make me more nervous.

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