# Aborted Sudden Cardiac Death Resulting in Permanent Hypoxic Encephalopathy in a Patient with Wolff-Parkinson-White Syndrome

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Sudden cardiac death (SCD) is a rare complication of Wolff-Parkinson-White (WPW) syndrome. We report a 32-year-old man with WPW syndrome and an eight-year history of recurrent, sudden onset of palpitation who presented to our emergency department with SCD. Despite successful resuscitation, permanent hypoxic encephalopathy and chronic respiratory failure developed. **( Mid Taiwan J Med 2008;13:216-20 )**

**Key words**

Wolff-Parkinson-White syndrome, sudden cardiac death, supraventricular tachycardia

## INTRODUCTION

Wolff-Parkinson-White (WPW) syndrome is characterized by the presence of the bundle of Kent, which comprises one or more accessory pathways (AP) between the atria and ventricle. The syndrome was first described by Louis Wolff, Sir John Parkinson, and Paul Dudley White in 1930.

The incidence of newly diagnosed cases of WPW syndrome in the general population is around 4.4 per 100,000 people per year [1]. The incidence of WPW syndrome is twice as high among men as among women (6.8/100,000/year versus 2.2/100,000/year) and typically affects young subjects. The most common arrhythmia of WPW syndrome is paroxysmal supraventricular tachycardia (SVT), which can lead to chest tightness, shortness of breath, dizziness and anxiety.

The incidence of sudden cardiac death (SCD) in patients with WPW syndrome is estimated to range from 0.15 to 0.39% [2-4]. Possible factors for SCD in patients with WPW syndrome include a very short AP anterograde refractory period (less than 250 ms) during atrial fibrillation (AF), a history of symptomatic tachycardia, multiple APs, and Ebstein's anomaly [4].

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## CASE REPORT

A 32 year-old man had an eight-year history of recurrent palpitation, which could be self-terminated by performing the Valsalva maneuver or carotid sinus massage. He had been advised by general practitioners to undergo an electrophysiologic study (EPS) and radiofrequency catheter ablation (RFCA) but the patient refused because he thought he could easily control the arrhythmia. One year prior to this hospitalization, the frequency and duration of palpitation episodes began to increase gradually (at least 1 episode per week). The patient continued to control the episodes with the Valsalva maneuver and at times took medication. Six hours prior to arriving at our emergency department (ED), he suffered from recurrent

palpitation before going to bed that night; the episodes could not be terminated by medication or by performing the Valsalva maneuver. His wife witnessed that the patient had shortness of breath the entire night. Early the next morning, the patient was cyanotic and in a comatose state. Neither spontaneous respirations nor heart beats were noticed upon arriving at our ED and the electrocardiography (ECG) revealed a bizarre wide complex rhythm (Fig. 1A). Cardiopulmonary resuscitation (CPR) was performed immediately. The rhythm soon changed to ventricular fibrillation (VF) (Fig. 1B) followed by pre-excited AF with rapid ventricular response (Fig. 1C), which spontaneously converted to normal sinus rhythm (Fig. 1D) shortly after repeated defibrillation (200 Joules × 1, 300 Joules x 1 and 360 Joules × 4). The complete ECG revealed a short PR interval and a wide QRS complex with initial delta waves compatible with WPW syndrome (Fig. 2). Echocardiography revealed a structurally normal heart. The patient was placed on ventilator support and transferred to the intensive care unit in a comatose state. One episode of paroxysmal SVT recurred but did not respond to intravenous adenosine injection (Fig. 3); it was finally terminated by cardioversion of 100 Joules. The patient had irreversible brain damage and needed long-term mechanical ventilator support. He did

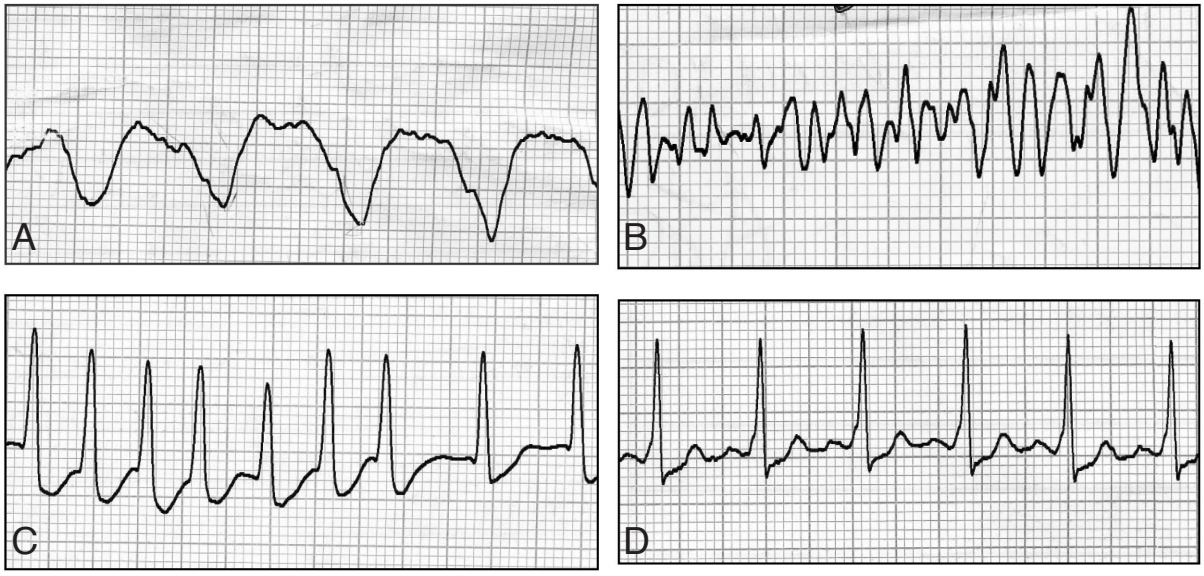


Fig. 1. A: Initial EKG shows a bizarre wide QRS complex rhythm. B: VF appeared after cardiac massage and intravenous injection of epinephrine. C: The rhythm converted to atrial fibrillation from ventricular fibrillation after defibrillation. D: The rhythm spontaneously converted to sinus tachycardia. (25 mm/s, 10 mm/mV)

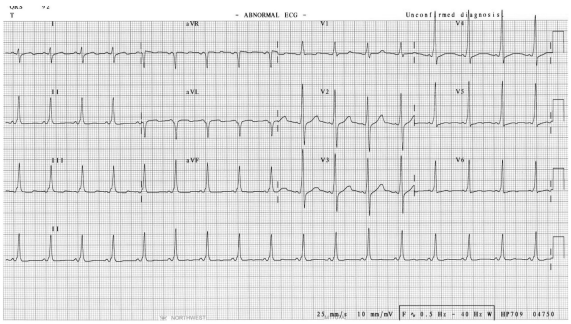


Fig. 2. The complete electrocardiogram reveals a typical WPW pattern.

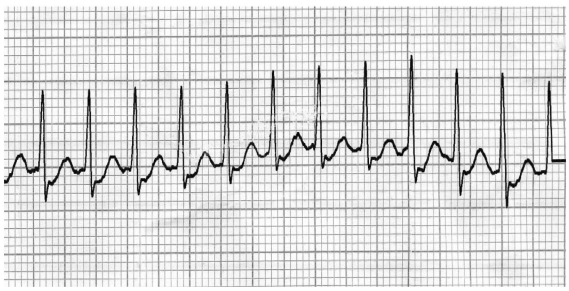

Fig. 3. One episode of orthodromic atrioventricular reentrant tachycardia occurred after admission. The average heart rate was 200/min. (25 mm/s, 10 mm/mV).

not receive further EPS and was transferred to another respiratory care center.

## DISCUSSION

Treatment of WPW syndrome includes drugs, catheter ablation and surgery. RFCA has been the first choice of treatment in patients with WPW syndrome since the 1990s because of its safety and efficacy.

Aggressive treatment for symptomatic patients is strongly recommended, especially when the patients are at high risk for SCD. The possible high risk factors for SCD in patients with WPW syndrome include a very short AP anterograde refractory period (less than 250 ms) during AF, a history of symptomatic tachycardia, multiple APs, and Ebstein's anomaly [4]. The patient reported herein had a history of frequent recurrent episodes of symptomatic tachyarrhythmia, and his shortest RR interval during AF was less than 250 ms. According to these two characteristics, our patient was at high risk for SCD. Timmermans et al reported of 15 SCD in total 690 WPW syndrome patients with an average rate of 2.2 percent in 16-year follow-up period [2]. Another series review showed only two SCD of more than 600 patients with WPW syndrome in the follow-up period of 5 to 20 years [5]. Because of a low incidence of SCD. RFCA for SCD is not recommended for asymptomatic WPW patients unless they are engaged in high-risk occupations. At the same time, we should remember that approximately 10% of SCD survivors with WPW patients, it is the first clinical manifestation of WPW syndrome [6].

Intermittent loss of pre-excitation, sudden loss of AP conduction on exercise stress testing, and loss of AP conduction after treatment of anti-arrhythmic agents have all been suggested as noninvasive makers of low risk for SCD [5]. Pappone et al suggested using invasive EPS to stratify the risk of SCD in patients with WPW syndrome [7]. They found that only 4.3% of patients with non-inducible arrhythmias during EPS developed symptomatic SVT after a mean follow-up of 37.7 months. No cardiac death was noted. In contrast, 25 of 47 patients with inducible arrhythmias developed symptomatic arrhythmias including SVT and AF. Among these patients, two had a resuscitated cardiac arrest and one patient died suddenly. They concluded that EPS can be useful in risk stratification of SCD. SVT inducibility and the presence of multiple APs are associated with high risk of subsequent symptomatic and fatal arrhythmic events. However, Sarubbi et al reported that 58% of healthy children and adolescents with asymptomatic WPW syndrome had sustained SVT and/or AF during EPS. Hence they questioned the value of risk stratification for SCD by EPS [8]. In a randomized study, Pappone et al reported that prophylactic RFCA reduced the number of arrhythmic events by 92% in asymptomatic patients with WPW syndrome [9]. They also found a significant reduction in arrhythmic events in high risk patients aged 5 to 12 years [10]. SCD and VF were found in the control groups, but not in the ablation groups in these two studies. All patients with SCD or VF in these 2 studies had multiple APs.

Aggressive treatment is strongly recommended for all symptomatic patients with WPW syndrome. Patients with the potential risk of SCD should receive EPS and RFCA in experienced centers. Asymptomatic patients with WPW syndrome may need EPS for further risk stratification and prophylactic RFCA for all asymptomatic patients may reduce subsequent arrhythmic events and therefore prevent unexpected SCD in their life time.

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# 沃夫-巴金森-懷特症候群引發猝死合併缺氧性腦病變

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沃夫-巴金森-懷特症候群並非罕見疾病，臨床上極有可能引發猝死，雖然機率很低，但是通常會是一件令人惋惜的事，特別是發生在年輕人身上。我們報告一位32歲男性，因爲猝死被送至急診，一系列的心電圖變化顯示爲沃夫-巴金森-懷特症候群。雖然急救後恢復生命徵象，但仍造成永久性腦病變及呼吸衰竭。本文將探討目前對於沃夫-巴金森-懷特症候群在高頻不整脈燒灼術的處理原則，建議所有病患應接受心臟科醫師詳細評估及追蹤治療。越來越多的證據顯示，積極的不整脈燒灼術不論對於有症狀或無症狀的患者，都可以減少心律不整發生的機會，進而避免猝死的危險性。（中台灣醫誌 **2008;13:216-20**）

**關鍵詞**

沃夫-巴金森-懷特症候群，猝死，心室上頻脈

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