Trends in Cardiovascular Medicine 27 (2017) 260–268

Available online at www.sciencedirect.com

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Long term risk of Wolff-Parkinson-White pattern and syndrome

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ABSTRACT

For years, conventional wisdom has held that patients with asymptomatic ventricular pre-excitation (asymptomatic WPW or WPW pattern) were at low risk for adverse outcomes. This assumption has been challenged more recently in a number of observational/natural history studies as well as in prospective trials in which patients were more aggressively studied via invasive electrophysiology study (EPS) and more aggressively treated, in some cases, with pre-emptive catheter ablation, despite the lack of symptoms. In sum, the data do not definitively support one approach (early, up-stream EPS and/or ablation) vs. the other (watchful waiting with close monitoring). The most recent pediatric and adult guidelines reflect this ambiguity with a broad spectrum of approaches endorsed.

Key words: WPW pattern, Ventricular pre-excitation, WPW syndrome, Asymptomatic, Electrophysiology study, Catheter ablation, Sudden death, Cardiac arrest, Ventricular fibrillation, Atrial fibrillation.

Introduction

In 1921, a phenomenon of "intraventricular block and a PR interval of 0.08 ms" in a 19 year-old patient with paroxysms of tachycardia was described by Wedd [1]. In 1930, Louis Wolff, MD; John Parkinson, MD; and Paul D. White, MD described a set of eleven patients with "Bundle-Branch Block with Short P-R Interval in Healthy young People Prone to Paroxysmal Tachycardia" [2]. Thus began our understanding of ventricular pre-excitation. Though a century has passed, our understanding of what is ventricular pre-excitation and the WPW syndrome is still incomplete. While ventricular pre-excitation or the WPW pattern on ECG is not uncommon with an prevalence of up to 0.1–0.3%, [3] a complete understanding of the long-term risk in patients with the WPW pattern vs. the WPW syndrome has not been reached. This review will attempt to summarize our understanding of prognosis to date and outline areas that would benefit from further clarification.

Terminology and pathophysiology

Two terms, the "WPW pattern" and the "WPW syndrome" are often used when describing patients with ventricular pre-excitation (Fig. 1, Panels A and B). The "WPW pattern" refers to ventricular pre-excitation seen on surface ECG while the "WPW syndrome" refers to the presence of ventricular pre-excitation on surface ECG plus the presence of symptoms suggestive of arrhythmia related to the pre-excitation, such as palpitations, episodic lightheadedness, pre-syncope, syncope, or cardiac arrest Ventricular pre-excitation refers to early electrical activation of the ventricle prior to the usual activation of the ventricle through the AV node and His-Purkinje system (Fig. 2). In most cases, the early activation occurs through an additional electrical connection between the atria and ventricles called an atrioventricular (AV) bypass tract, also referred to as an "AV accessory pathway" or "Bundle of Kent". These AV bypass tracts are usually comprised of myocardial tissue, which unlike the AV node, are

The authors have indicated there are no conflicts of interest.

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http://dx.doi.org/10.1016/j.tcm.2016.12.001
1050-1738/

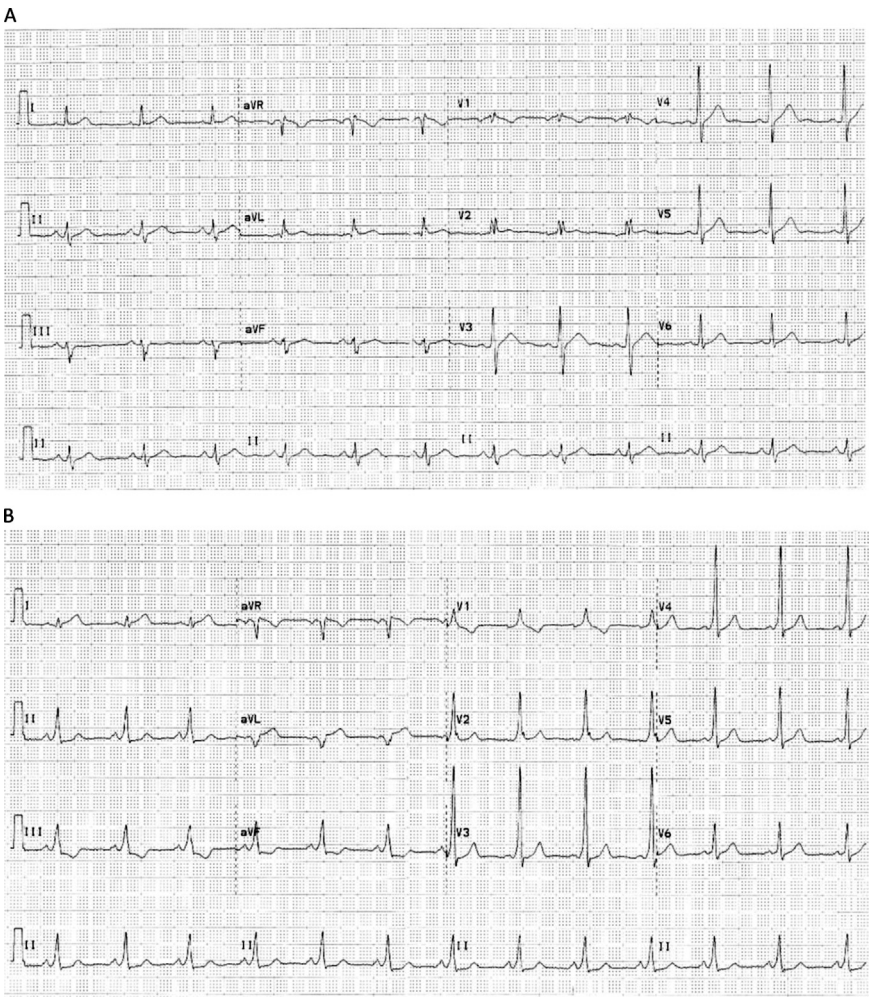

Fig. 1 – Both Panels A and B show 12-lead ECGs in the same patient with a left-sided accessory pathway. In Panel A, ventricular pre-excitation is minimal likely due to more rapid AV nodal conduction causing ventricular activation to be almost entirely over the normal His-Purkinje system. In Panel B, ventricular pre-excitation is much more obvious with clear delta waves seen in all 12 leads.

usually non-decremental or non-delayed in conduction. As such, the ventricle can activate immediately after atrial activation resulting in the appearance of a delta wave which results in shortening of the PR interval and widening of the QRS interval (Fig. 2). This non-decremental property also holds the key to the potential for sudden death in patients with the WPW syndrome. Because there can be a lack of delay in conduction from the atria to the ventricles, atrial fibrillation, where atrial depolarization can occur at rates of >350 beats per minute, can lead to very frequent ventricular depolarization and high rates which could lead to degeneration into ventricular fibrillation (VF). A shortest pre-excited RR interval (SPERRI) <250 ms in AF has been repeatedly shown to be associated with a high risk of VF (Fig. 3).

Past state of knowledge

The prevalence of WPW pattern is thought to be 0.1–0.3% in the general population3 with about 65% of adolescents and 40% of all patients thought to be asymptomatic. In their original paper first describing the WPW syndrome, Wolff, Parkinson, and White included eleven patients ranging from ages eleven to fifty-five at first diagnosis. The patients were described as having symptoms ranging from as few as 10 years and 14 years up to 37 years and 48 years in two patients. That said, while some patients can live for decades with palpitations, it is clear that some patients with the WPW syndrome are at risk for sudden cardiac death (SCD). Indeed, SCD can be the first manifestation of the WPW syndrome, especially in children and young adults [4].

Looking back at the 2003 ACC/AHA/ESC guidelines for SVT, the SCD risk for the WPW syndrome is cited as 0.15–0.39% over 3- to 10-year follow-up [5]. Given this, the guidelines state that “Given the potential for AF among patients with WPW syndrome and the concern about sudden cardiac death resulting from rapid pre-excited AF, even the low annual incidence of sudden death among patients with the WPW syndrome is of note and supports the concept of liberal indications for catheter ablation”. High-risk markers identified retrospectively in patients with WPW syndrome and cardiac arrest were 1) SPERRI <250 ms during spontaneous or induced AF, 2) a history of symptomatic tachycardia, 3) multiple accessory pathways, and 4) Ebstein's anomaly. As such, in these 2003 guidelines, patients with WPW syndrome, whether symptoms were well-tolerated or not, were