Wolff-Parkinson-White syndrome

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Wolff-Parkinson-White syndrome (WPW-S) is a disorder characterized by presence of an accessory pathway (AP) which predisposes patients to tachyarrhythmias and sudden death (SD). Among patients with WPW-S atrioventricular reentrant tachycardia (AVRT) is the most common arrhythmia, accounting for 95% of re-entrant tachycardias. It has been estimated that ≈ 30% of patients with WPW-S have atrial fibrillation (AF). AF is a potentially life-threatening arrhythmia. If an AP has a short Anterograde Refractory Period (ARP), then rapid repetitive conduction to the ventricles during AF can result in a rapid ventricular response with subsequent degeneration to ventricular fibrillation (VF). The ARP of the AP is the main determinant factor of ventricular rate during AF in the WPW-S.

The paradoxical early recovery of propagation due to anterograde supernormal conduction well inside a prolonged ARP in the AP, may play a relevant role to determine the rate of ventricular response during AF, eventually leading to extremely fast ventricular rates, syncope, and even VF in patients with WPW-S supposed a priori to be exposed to a low risk of SD. This may require very precise conditions, including an enhanced adrenergic influence on the heart. Retrograde supernormal conduction in APs may also participate in the mechanism of paroxysmal supraventricular tachycardias that are not easily induced by invasive EPS anterograde supernormal conduction.

The AP may be located anywhere along the atrioventricular valve.

Most of the patients are young and do not have structural heart disease hence it is important to risk stratify these patients so as to prevent the SD.

***The gamma(2) subunit of AMP-activated protein kinase (PRKAG2) located in chromosome 7 plays an important role in regulating metabolic pathways, and patients with PRKAG2 mutations are associated with familial ventricular pre-excitation, hypertrophic cardiomyopathy and AV block*.**

Management of asymptomatic patients with WPW-S has always remained controversial. Radiofrequency catheter Ablation (RFCA) of APs has become an established mode of therapy for symptomatic patients and asymptomatic patients employed in high-risk professions.

Several recent prospective studies evaluated invasive risk stratification followed by prophylactic RFCA in asymptomatic patients with WPW pattern. Inducibility of arrhythmias in these patients during invasive EPS was shown to predict the development of future symptomatic arrhythmias. Methods used to define the prognosis of WPW are well-defined. At first the maximal rate of conduction through the AP is evaluated; EPS using 1 and 2 extrastimuli delivered at different cycle lengths is then used to determine the AP refractory period and to induce a

supraventricular tachycardia. These methods should be performed in the control state and repeated in adrenergic situations either during exercise test or more simply during a perfusion of small doses of isoproterenol. The induction of an AF with rapid conduction through the AP (> 240/min in control state, > 300/min after isoproterenol) is the sign of a form of WPW at risk of SD. Although ablation of APs performed in 'inducible' patients decreased the incidence of subsequent symptomatic arrhythmias, the studies were not powered to detect a reduction in life-threatening arrhythmias.

RFCA remains the first-line therapy for patients with symptomatic WPW-S. EPS and possible RFCA of AP may be offered to well informed asymptomatic individuals with WPW if they are willing to trade the very small risk of subsequent SD or incapacity for a small immediate procedural risk of serious complications or SD.