Electrocardiographic Clues for Multiple Accessory Pathways in Patients With Pre-Excitation Syndromes*

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Background. Wolff, Parkinson and White (1) in 1930 reported an electrophysiologic syndrome based on their observation of 11 "healthy young" people who had "bundle branch block with short PR interval" and who were "prone to paroxysmal tachycardia." However, substantial progress in understanding the pathophysiology and in devising therapy for the various complex arrhythmias in patients with this syndrome had to wait for the development of clinical electrophysiologic testing, antiarrhythmic drugs and open heart surgery. Electrophysiologic tests have their origin in the recording of the bundle of His electrogram, first documented in the dog by Alanis et al. (2) in 1958. The first successful termination of atrioventricular (AV) reentrant tachycardia (circus movement, AV tachycardia) in a patient with Wolff-Parkinson-White syndrome was accomplished in 1968 by Ryan et al. (3) by asynchronous ventricular capture by an implanted permanent ventricular pacemaker. Most of our current approaches employed for patients with this syndrome are based on the remarkable progress made in the 1970s, as summarized in the now classic review article by Gallagher et al. (4).

This syndrome is relatively common (occurring in about 1 in every 1,000 people) and its clinical manifestations vary greatly, ranging from a totally asymptomatic benign course in some patients to a malignant course in others who die of ventricular fibrillation as a result of an extremely rapid ventricular response in the presence of atrial fibrillation. The rapid ventricular response during atrial fibrillation results from rapid conduction over the accessory pathway and is related to its extremely short anterograde refractory period (5). Because routine use of clinical electrophysiologic tests in all patients with this syndrome is prohibitively costly, electrocardiographic (ECG) clues associated with a benign clinical course might obviate such tests and would thus be of great value. Several such clues have been proposed by Wellens (6). These include an intermittent pre-excitation pattern, loss of a pre-excitation pattern with exercise, absence of extremely short RR intervals during atrial fibrillation and loss of a pre-excitation pattern after administration of ajmaline or procainamide. The first three clues have been shown to be predictors of a relatively prolonged anterograde refractory period of the accessory pathway (5,7,8). However, the predictive value of intravenous procainamide, as suggested in a small initial study (9), was not substantiated by a larger subsequent study (10).

Patients with multiple accessory pathways. The presence of multiple accessory pathways can be diagnosed by electrophysiologic testing and by intraoperative mapping (4,11); in one series they were identified in 52 (1.3%) of 3,880 patients with a pre-excitation syndrome (11). Physicians who care for patients with this syndrome must be aware of the possible presence of multiple accessory pathways for several reasons: 1) Failure to recognize them may result in failure of surgical therapy. 2) There is some evidence to suggest that patients with such pathways may be more likely than patients with a single accessory pathway to develop ventricular fibrillation. 3) Complex cardiac arrhythmias in these patients may be diagnosed incorrectly unless these pathways have been recognized. Thus, the development of ECG clues that accurately predict the presence of multiple accessory pathways would enhance the management of such patients.

In the September issue of the Journal, Wellens and his associates (12) presented six ECG clues as predictors for the presence of multiple accessory pathways. Of 314 patients with a pre-excitation syndrome undergoing electrophysiologic testing, 17 patients were diagnosed as having multiple accessory pathways. The presence of these pathways was validated by intraoperative mapping in 8 of the 17. Twelve lead ECGs from these 17 patients were retrospectively analyzed. In 12 of the 17 patients, one or more of the following six ECG clues to the presence of multiple accessory pathways were observed: 1) two or more different P
wave configurations during orthodromic circus movement tachycardia (four patients); 2) a discrepancy between the estimated site of the ventricular insertion of an accessory pathway during antidromic tachycardia and the estimated site of the atrial insertion of an accessory pathway during orthodromic circus movement tachycardia (seven patients); 3) atrial fibrillation with two or more pre-excitation patterns (six patients); 4) a spontaneous and direct switch from orthodromic to antidromic circus movement tachycardia or vice versa (two patients); 5) a spontaneous and direct switch from one type of antidromic tachycardia to another (two patients); and 6) a change in pre-excitation pattern after the administration of ajmaline or procainamide (three patients).

Although our current understanding of the electrophysiology of pre-excitation syndromes tends to make multiple accessory pathways an appealing mechanism responsible for most of these ECG clues, it is essential to show that these clues are unique to patients with multiple accessory pathways and are either rare or nonexistent in patients with a single accessory pathway. Before these clues are widely accepted, it is also important to demonstrate that these phenomena are directly related to the presence of multiple accessory pathways and not to other abnormal electrophysiologic characteristics of either atrial or ventricular myocardium. For example, two different P wave configurations during orthodromic circus movement tachycardia could theoretically occur because of intermittent intraatrial conduction block (Fig. 1). Similarly, a change in pre-excitation pattern with administration of ajmaline or procainamide could also be a result of intraventricular conduction block induced by these drugs (Fig. 2). With respect to the occurrence of two or more pre-excitation patterns during atrial fibrillation, a rapid ventricular response during atrial fibrillation often makes it difficult to distinguish between varying pre-excitation pathway and rate-dependent bundle branch block.

**Clinical implications.** The clinical spectrum for patients with this relatively common congenital cardiac disorder varies widely from an asymptomatic benign clinical course to a malignant course ending abruptly with ventricular fibrillation. Fortunately, various forms of effective therapy including surgical dissection of accessory pathways are now available for those patients at high risk of sudden arrhythmic death. However, electrophysiologic testing is not cost-effective as a screening test. Furthermore, no firm guidelines for the utilization of electrophysiologic testing have been established to date (13). Thus, it is most important that the theoretically appealing ECG clues to multiple accessory pathways proposed by Wellens et al. (12) be studied systematically and be validated by surgical ablation in future large scale, perhaps multicenter studies. At the same time we need to confirm those ECG clues that are believed to be associated with a benign clinical course, such as intermittent pre-excitation, loss of pre-excitation pattern with exercise, absence of extremely short RR intervals during atrial fibrillation and loss of pre-excitation with procainamide or ajmaline.
References


