JACC: CLINICAL ELECTROPHYSIOLOGY
VOL. ■, NO. ■, 2017

ISSN 2405-500X/$36.00
PUBLISHED BY ELSEVIER
https://doi.org/10.1016/j.jacep.2017.10.009

Life-Threatening Event Risk in Children With Wolff-Parkinson-White Syndrome

A Multicenter International Study

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ABSTRACT

OBJECTIVES This study sought to characterize risk in children with Wolff-Parkinson-White (WPW) syndrome by comparing those who had experienced a life-threatening event (LTE) with a control population.

BACKGROUND Children with WPW syndrome are at risk of sudden death.

METHODS This retrospective multicenter pediatric study identified 912 subjects ≤21 years of age with WPW syndrome, using electrophysiology (EPS) studies. Case subjects had a history of LTE: sudden death, aborted sudden death, or atrial fibrillation (shortest pre-excited RR interval in atrial fibrillation [SPERRI] of ≤250 ms or with hemodynamic compromise); whereas subjects did not. We compared clinical and EPS data between cases and subjects.

RESULTS Case subjects (n = 96) were older and less likely than subjects (n = 816) to have symptoms or documented tachycardia. Mean age at LTE was 14.1 ± 3.9 years of age. The LTE was the sentinel symptom in 65%, consisting of rapidly conducted pre-excited atrial fibrillation (49%), aborted sudden death (45%), and sudden death (6%). Three risk components were considered at EPS: SPERRI, accessory pathway effective refractory period (APERP), and shortest paced cycle length with pre-excitation during atrial pacing (SPPCL), and all were shorter in cases than in control subjects. In multivariate analysis, risk factors for LTE included male sex, Ebstein malformation, rapid anterograde conduction (APERP, SPERRI, or SPPCL ≤250 ms), multiple pathways, and inducible atrial fibrillation. Of case subjects, 60 of 86 (69%) had ≥2 EPS risk stratification components performed; 22 of 60 (37%) did not have EPS-determined high-risk characteristics, and 15 of 60 (25%) had neither concerning pathway characteristics nor inducible atrioventricular reciprocating tachycardia.

CONCLUSIONS Young patients may experience LTE from WPW syndrome without prior symptoms or markers of high-risk on EPS. (J Am Coll Cardiol EP 2017;■:■-■) © 2017 by the American College of Cardiology Foundation.

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ABBREVIATIONS AND ACRONYMS

APERP = accessory pathway effective refractory period

ART = antidromic reciprocating tachycardia

AVRT = atrioventricular reciprocating tachycardia

CHD = congenital heart disease

EPS = electrophysiology study

LTE = life-threatening event

ORT = orthodromic reciprocating tachycardia

SPERRI = shortest pre-excited RR interval in atrial fibrillation

SPPCL = shortest pre-excited paced cycle length with atrial pacing

SVT = supraventricular tachycardia

WPW = Wolff-Parkinson-White syndrome

Sudden death in Wolff-Parkinson-White (WPW) syndrome is a rare but potentially preventable problem affecting young, otherwise healthy people. Sudden death is usually a consequence of atrial fibrillation with rapid conduction over an accessory pathway resulting in ventricular fibrillation. Because WPW patients develop atrial fibrillation more frequently than the general population, an important question is whether there is a risk of ventricular fibrillation should atrial fibrillation occur. Assessing accessory pathway conduction properties by using electrophysiology study (EPS) is advocated as a preventive strategy against sudden death, as noninvasive risk stratification tools are imperfect (1-4). Inducible atrioventricular re-entrant tachycardia (AVRT) or EPS data suggesting a pathway capable of rapid anterograde conduction are identified as predictors of malignant arrhythmia (5-8). Because catheter ablation can cure WPW syndrome and eliminate risk (9), the small long-term risk of a life-threatening event (LTE) must be balanced with the immediate albeit low risk of an ablation.

The low event rate of WPW syndrome, reduced further by catheter ablation, makes risk assessment a challenge. Data investigating possible risk factors for LTE in children with WPW syndrome, however, remain critical. In this study, we compared children with WPW syndrome who had experienced an LTE with a control population (WPW syndrome without LTE) to identify characteristics associated with sudden death risk.

METHODS

This multicenter, international, retrospective case-control study involved 22 centers from 6 countries (United States, Canada, New Zealand, Cuba, Czech Republic, and Wales [United Kingdom]) solicited through the Pediatric and Congenital Electrophysiology Society (PACES). Data collected encompassed the era of catheter ablation in children, from January 1990 through June 2016. All centers obtained local investigational review board approval, and institutional databases were searched to identify children with WPW syndrome. De-identified data were managed using Research Electronic Data Capture (REDCap), hosted at the University of Utah. REDCap is a secure, Web-based application designed to support data capture for research (10). All data were reviewed by the data coordinating center and statistician for appropriateness for inclusion.

CASE SUBJECTS. Case subjects were children with WPW syndrome who had experienced an LTE at ≤21 years of age. An LTE was defined as sudden death, aborted sudden death, or a clinical episode of pre-excited atrial fibrillation with the shortest pre-excited RR interval (SPERRI) in atrial fibrillation of ≤250 ms, regardless of symptoms or documented pre-excited atrial fibrillation associated with hemodynamic compromise, syncope, or seizure, regardless of the SPERRI. Subjects who experienced pre-excited atrial fibrillation without associated hemodynamic compromise, syncope, or seizure and a SPERRI >250 ms were excluded. Cases of sudden death were included if a pre-mortem electrocardiogram (ECG) and/or EPS proving WPW syndrome was available.

CONTROL SUBJECTS. Control subjects were ≤21 years of age with WPW syndrome who had not experienced an LTE or clinical pre-excited atrial fibrillation and had undergone an EPS. For each case subject, 4 age-matched subjects (±24 months of age at EPS or LTE if no EPS was performed) and 4 non-age-matched subjects were selected by each center. Two sets of subjects were selected to potentially mitigate and investigate influences of age and size on ablation outcomes and risk. Matched subjects were selected from the same institution when possible or from other participating centers. Except for analyses involving age, subjects were evaluated as a single control group. Congenital heart disease (CHD) was noted, but cases and subjects were not matched for this variable.

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Manuscript received August 16, 2017; revised manuscript received October 3, 2017, accepted October 12, 2017.