Natural History of Wolff-Parkinson-White Syndrome Diagnosed in Childhood

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Wolff-Parkinson-White (WPW) syndrome carries a risk for symptomatic arrhythmias and sudden death. The aim of this study was to examine the natural history of patients with Wolff-Parkinson-White syndrome diagnosed in childhood followed longitudinally at a single institution. The study population consisted of 446 patients. The median age of diagnosis was 7 years, and 61% were male. Associated heart disease was present in 40 patients (9%). Modes of presentation included supraventricular tachycardia (38%), palpitations (22%), chest pain (5%), syncope (4%), atrial fibrillation (0.4%), sudden death (0.2%), and incidental findings (26%); data were unavailable in 4%. During the study period, a total of 243 patients (54%) had supraventricular tachycardia, and 7 patients (1.6%) had atrial fibrillation. Of patients who presented at ≤3 months of age, 35% had resolution of manifest preexcitation compared with 5.8% who presented at >3 months of age ($p < 0.0001$). There were 6 sudden deaths (1.3%), with an incidence of 2.8 per 1,000 patient-years. Two of these patients had structurally normal hearts (incidence 1.1 per 1,000 patient-years). Four of these patients had associated heart disease (incidence 27 per 1,000 patient-years) ($p < 0.01$). In conclusion, in a large population of patients with Wolff-Parkinson-White syndrome diagnosed in childhood, 64% had symptoms at presentation, and an additional 20% developed symptoms during follow-up. There were 6 sudden deaths (1.3%), with an overall incidence of 1.1 per 1,000 patient-years in patients with structurally normal hearts and 27 per 1,000 patient-years in patients with associated heart disease. **(Am J Cardiol 2013;112:961−965)**

Wolff-Parkinson-White (WPW) syndrome is a condition characterized by a short PR interval associated with ventricular preexcitation manifested by a delta wave, first described in 1930.[1] The clinical spectrum ranges from no cardiac symptoms to recurrent paroxysmal supraventricular tachycardia (SVT) to, in some cases, sudden cardiac collapse or death. Earlier natural history studies have included varying patient populations, with ages ranging from children to adults, and most have shown a low but finite incidence of sudden death. Concern about the actual risk for sudden death has a tremendous impact on the clinical management of these patients, particularly those who are incidentally found to have preexcitation on an electrocardiogram without associated cardiac symptoms. The purpose of the present study was to examine the natural histories of patients with WPW syndrome diagnosed in childhood followed longitudinally at a single institution.

Methods

This study was a retrospective review. Patients with diagnoses of WPW syndrome who were seen from 1960 to 2010 were identified from the Children's Hospital of Pittsburgh cardiology database. During this time frame, Children's Hospital of Pittsburgh Pediatric Cardiology represented the major cardiology presence in this region, seeing community-based referrals as well as tertiary care referrals. Patient data were obtained from hospital medical records. All patients aged <21 years with available records were included. All electrocardiograms were reviewed by 1 investigator to reconfirm the diagnosis of WPW syndrome. Data collected included patient demographics, hemodynamically significant associated heart disease, clinical presentation, documented arrhythmias, presence of persistent or intermittent preexcitation, spontaneous resolution of manifest preexcitation, and clinical events during follow-up. Patients with bicuspid aortic valves were not considered to have hemodynamically significant lesions unless they had more than mild aortic stenosis or insufficiency or concomitant lesions. The duration of follow-up was defined as time of presentation to last clinical follow-up, time of ablation, or time of death. Ablation was considered to have changed the patient's natural history and thus was considered a primary end point.

Statistical analysis of continuous data was performed using Student's *t* tests. Categorical data were evaluated using chi-square analysis. The incidence of sudden death was calculated from the number of cases observed relative

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See page 965 for disclosure information.

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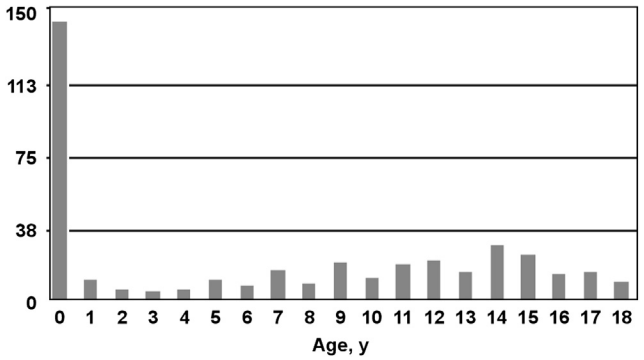

Figure 1. Age distribution at diagnosis of WPW syndrome from 0 to 18 years. Total number of patients = 446.

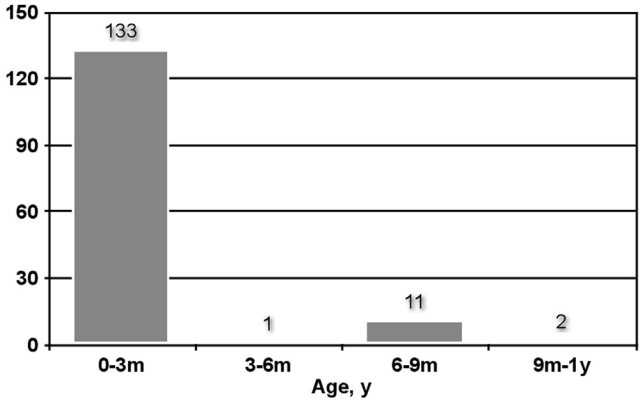

Figure 2. Age distribution of patients aged <1 year at diagnosis of WPW syndrome, with most diagnosed at ≤3 months of age.

Table 1
Associated heart diseases in patients with Wolff-Parkinson-White syndrome (n = 40)

Lesion	n (%)
Ventricular septal defect	10 (25%)
Ebstein's anomaly	8 (20%)
Congenitally corrected transposition of the great arteries	6 (15%)
Coarctation of the aorta	3 (7.5%)
Complete atrioventricular septal defect	3 (7.5%)
Dilated cardiomyopathy	2 (5%)
Hypertrophic cardiomyopathy	2 (5%)
Rhabdomyoma	2 (5%)
Anomalous left coronary from pulmonary	1 (2.5%)
Heterotaxy	1 (2.5%)
Patent ductus arteriosus	1 (2.5%)
Cor triatriatum	1 (2.5%)

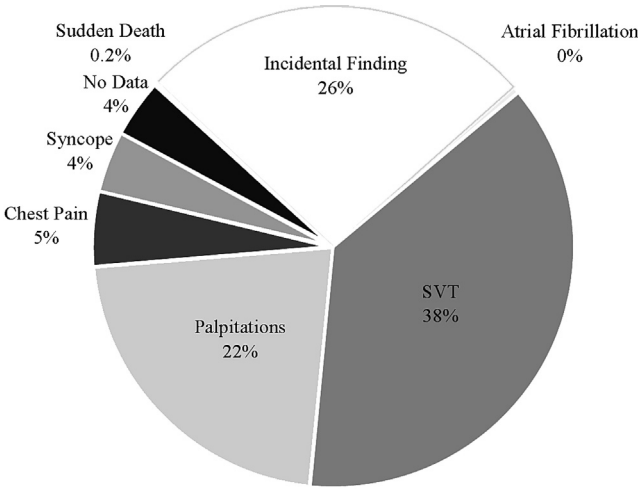

Figure 3. Symptoms at presentation.

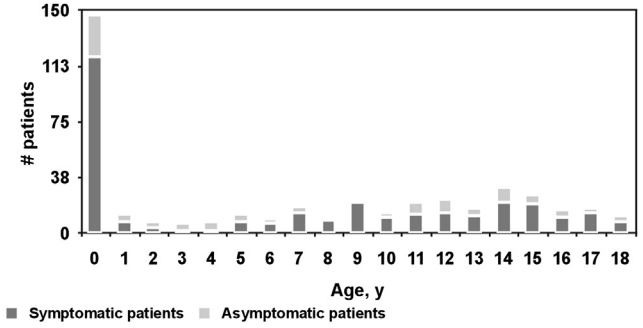

Figure 4. Symptomatic and asymptomatic patients at diagnosis by age. Patients were more likely to be symptomatic at diagnosis across all age ranges.

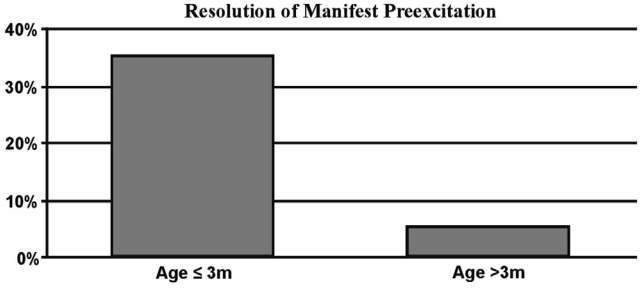

Figure 5. Resolution of manifest preexcitation on the basis of age of presentation. Spontaneous resolution was significantly more likely to occur if WPW syndrome was diagnosed at ≤3 months compared with >3 months of age.

to the total patient-years of observation as derived from the database.

Results

Six hundred patients were identified from the cardiology database search. Medical record patient data were available for 446 patients (75%), who constituted the study population. Some medical records spanning the data collection period were eliminated on the basis of hospital chart storage policy and were not available for review.

There were 274 male (61%) and 172 female (39%) patients. The median age of diagnosis was 7 years (range 0 to 20). The most common age of presentation was ≤3 months, with 133 patients (30%). The remainder of the patients were diagnosed between 4 months and 20 years of age (Figures 1 and 2). The median duration of follow-up was 3 years (range 0 to 23). There was a total of 2,160 patient-years of follow-up. A total of 90 patients had