Survey of Current Practice of Pediatric Electrophysiologists for Asymptomatic Wolff-Parkinson-White Syndrome

Robert M. Campbell, MD*; Margaret J. Strieper, DO*; Patricio A. Frias, MD*; Kathryn K. Collins, MD†; George F. Van Hare, MD‡§; and Anne M. Dubin, MD§

ABSTRACT. Objective. To determine the approach that pediatric electrophysiologists use as they evaluate asymptomatic patients with Wolff-Parkinson-White (WPW) syndrome regarding electrophysiologic testing and radio frequency ablation.

Methods. A 21-question survey was mailed to 66 pediatric electrophysiologists who had voluntarily submitted patient data at any time to the Pediatric Radio Frequency Ablation Registry since its inception in 1990. The survey addressed issues regarding physician experience with electrophysiologic testing and radio frequency ablation, risk assessment, electrophysiologic study, and factors that influence the decision to perform radio frequency ablation in asymptomatic patients.

Results. Returned surveys (43 of 66 [65%]) were analyzed blindly. The 43 physicians who responded were experienced, with 37 reporting >5 years of performing radio frequency ablation and 30 having performed >200 radio frequency ablation procedures. Thirty-six of the 43 electrophysiologists used invasive electrophysiologic study for risk stratification in asymptomatic patients with WPW. Electrophysiologic findings guided selection of patients for radio frequency ablation procedures. Expected radio frequency ablation outcome quotes to the family were consistent with recently published data from the Electrophysiology Society regarding current-era experience with radio frequency ablation.

Conclusion. The majority of responding electrophysiologists use invasive electrophysiologic study both to stratify risk for asymptomatic WPW and to select appropriate patients for radio frequency ablation. This current practice should be communicated to other pediatric cardiologists and pediatricians. Pediatrics 2003;111:e245–e247. URL: http://www.pediatrics.org/cgi/content/full/111/3/e245; Wolff-Parkinson-White syndrome, electrophysiology, radio-frequency ablation.

ABBREVIATIONS. WPW, Wolff-Parkinson-White; SVT, supraventricular tachycardia; EPS, electrophysiology study; RFA, radio frequency ablation.

W olff-Parkinson-White (WPW) syndrome is often associated with supraventricular tachycardia (SVT). However, there is a risk of sudden cardiac death in this syndrome secondary to atrial fibrillation, with rapid anterograde conduction over the accessory pathway to the ventricles, resulting in ventricular fibrillation. Studies in symptomatic patients with WPW syndrome have found that ventricular fibrillation occurred in 2.2% of patients over a 16-year period, whereas natural history data in asymptomatic patients reveal a sudden death rate of 1 per 1000 patient-years of follow-up.1,2 Therefore, the possibility of cardiac arrest or sudden cardiac death must be considered as patient care plans are made. Electrophysiologic testing can determine SVT inducibility, accessory pathway location, and SVT tachycardia mechanism. In addition, several invasively determined conduction characteristics of accessory pathways have been used to try to stratify risk for potential sudden death vulnerability. These have included an effective refractory period of <270 ms for anterograde conduction across the accessory pathway, shortest preexcited R-R interval of <240 ms during atrial fibrillation, and antidromic SVT.3–4

However, the asymptomatic patient with WPW syndrome presents an interesting challenge. Published data report that sudden cardiac death may be the first presenting sign in patients with undiagnosed and/or asymptomatic WPW syndrome.5 As techniques have improved, electrophysiologic study (EPS) and radio frequency ablation (RFA) have become safe in pediatric patients.6 We hypothesized that as a result of this, asymptomatic patients with WPW syndrome are increasingly being referred for electrophysiologic evaluation, with RFA when appropriate. In this study, we sought to elucidate the approach that pediatric electrophysiologists take as they evaluate asymptomatic patients with WPW syndrome in regards to electrophysiologic testing and RFA.

METHODS

A 21-question survey was mailed to 66 pediatric electrophysiologists who had voluntarily submitted patient data at any time to the Pediatric Radio Frequency Ablation Registry since its inception in 1990. These 21 questions addressed issues regarding risk assessment, electrophysiologic testing, factors that influence RFA in asymptomatic patients, and physician experience with electrophysiologic testing and RFA. Individual physicians, rather than institutions, were surveyed, because the intent of the questionnaire was to address patient issues much as they would be dis-
cussed with patients and their parents about diagnostic and management options during the process of informed consent.

RESULTS

A total of 43 replies were received from 66 practicing pediatric electrophysiologists. Responding physicians were experienced, with 37 of the 43 reporting >5 years of performing RFA and 30 of the 43 having performed >200 RFA procedures.

EPS is used routinely by 36 of the 43 electrophysiologists to assess risk of sudden death in asymptomatic WPW syndrome. Intracardiac EPS, which is more invasive but more comprehensive (and allows for RFA, if appropriate), was the preferred approach by 24 respondents; 18 physicians use either intracardiac or esophageal EPS for diagnostic testing. Preferred patient age for EPS ranges from 1 to 13 years (mean: 8.1 years; median: 10 years). Certain comorbidities or issues influenced the decision to recommend electrophysiologic risk stratification. These included the desire to enter military service for a career in which SVT would be contraindicated (38 of 43 respondents), competitive athlete (37 of 43), coexisting congenital heart defect (35 of 43), reactive airway disease (21 of 43), attention-deficit/hyperactivity disorder requiring possible medical treatment (19 of 43), and the need for psychotropic medications (17 of 43). Invasive criteria used for risk stratification are shown in Table 1. Induction of atrial fibrillation during EPS was variable with 24 of 43 always and 10 of 43 sometimes eliciting atrial fibrillation. During EPS, 23 of 43 electrophysiologists routinely use isoproterenol as a provocative agent, whereas 18 of 43 do not.

Two or more pediatric electrophysiologists attend each RFA sometimes (20 of 43) or always (13 of 43) in a pediatric catheterization laboratory (26 of 43) or shared adult facility (14 of 43). The decision to perform RFA was based on several criteria (Table 1). It is interesting that 10 of 43 electrophysiologists would perform an ablation “because you were there.” Only 9 of 43 electrophysiologists would not perform RFA for asymptomatic WPW syndrome. In the absence of a patent foramen ovale for access to left-sided accessory pathways, 31 of 43 physicians preferentially use a transseptal puncture approach to the left atrium, with 7 of 43 using a retrograde arterial approach and 4 of 43 using either. These electrophysiologists (25 of 43) have performed >100 transseptal procedures, 30 of 43 >50 transseptals) quote the risk of transseptal puncture as <1% (27 of 43) and <2% (35 of 43). RFA outcome quotes to the family are summarized in Table 2.

### TABLE 1. Indications for Risk Stratification and RFA

<table>
<thead>
<tr>
<th>Invasive EPS Criteria</th>
<th>Respondents Who Would Use as Risk Stratifier</th>
<th>Respondents Who Would Use to Determine RFA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shortest preexcited R-R during atrial fibrillation &lt;240 ms</td>
<td>37/43 (86%)</td>
<td>33/43 (77%)</td>
</tr>
<tr>
<td>Accessory pathway effective refractory period &lt;240 ms</td>
<td>24/43 (56%)</td>
<td>19/43 (44%)</td>
</tr>
<tr>
<td>Inducible SVT</td>
<td>16/43 (37%)</td>
<td>11/43 (26%)</td>
</tr>
</tbody>
</table>

DISCUSSION

At present, the indication for EPS and RFA in patients who are asymptomatic with WPW syndrome is unclear. The guidelines of the American College of Cardiology and American Heart Association regarding this subject classify this condition as a class II indication, defined as “conditions for which electrophysiological studies are frequently performed, but there is less certainty about the usefulness of the information that is obtained. Experts are divided in their opinion as to whether patients of these conditions are likely to benefit from an electrophysiological study.” This study documents that the majority of these responding experienced pediatric cardiac electrophysiologists are currently recommending invasive EPS to risk-stratify asymptomatic patients with WPW syndrome and to select, from this group, patients for RFA.

The risk for asymptomatic pediatric patients with WPW syndrome is not well defined. In 1979, Klein et al³ raised concerns for asymptomatic patients with WPW syndrome by documenting ventricular fibrillation in 3 pediatric patients (ages 8–16 years) who presented with cardiac arrest. In 1993, Russell et al⁸ described 256 pediatric patients with WPW syndrome preexcitation, 6 of whom had presented with a life-threatening event as the first manifestation of their preexcitation syndrome. Among this group of 256 patients, 60 (23.4%) were asymptomatic. In 1995, Deal et al⁵ representing the Pediatric Electrophysiology Society, reported 42 patients with WPW syndrome who experienced cardiac arrest. Twenty of these patients (mean age: 11 years) had cardiac arrest as their initial presenting symptom. In 1 of the largest reports of children who had WPW syndrome and experienced cardiac arrest, Bromberg et al⁹ in 1996 described 60 patients who underwent surgical correction of WPW in the years before widespread use of RFA. Ten children had experienced a clinical cardiac arrest; of this group, only 1 had a history of

### TABLE 2. RFA Outcome Quotes

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Respondents Outcome Quotes</th>
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<tbody>
<tr>
<td>Acute success</td>
<td>42/43 (98%)</td>
</tr>
<tr>
<td>&gt;90%</td>
<td>32/43 (74%)</td>
</tr>
<tr>
<td>&gt;95%</td>
<td></td>
</tr>
<tr>
<td>Procedural complication rate</td>
<td>27/43 (63%)</td>
</tr>
<tr>
<td>&lt;1%</td>
<td>33/43 (77%)</td>
</tr>
<tr>
<td>&lt;2%</td>
<td></td>
</tr>
<tr>
<td>Recurrence rate</td>
<td>24/43 (56%)</td>
</tr>
<tr>
<td>&lt;5%</td>
<td></td>
</tr>
<tr>
<td>&lt;10%</td>
<td></td>
</tr>
</tbody>
</table>
syncope or atrial fibrillation. In this entire group of patients, a shortest preexcited R-R interval <220 ms during atrial fibrillation was more sensitive than clinical history for identifying those patients at sudden death risk. The absence of life-threatening symptoms did not necessarily connote low risk. Finally, Dubin et al\textsuperscript{10} recently assessed 100 pediatric patients who had WPW syndrome and underwent electrophysiologic evaluation for risk stratification, documented SVT, or syncope. Asymptomatic patients had statistically the same recognized EPS risk profile as the symptomatic patients. These authors theorized that the risk of sudden cardiac death in asymptomatic patients might be higher than previously suspected. These data seem to be reflected in the present clinical practices of the pediatric electrophysiologists queried, with the majority (84\%) using EPS studies in asymptomatic patients for risk stratification.

It is interestingly that risk assessment itself may be problematic. Noninvasive methods (Holter monitoring, exercise treadmill testing, and intravenous procainamide infusions) as well as transesophageal EPS are relatively nonspecific, nonsensitive, and incomplete.\textsuperscript{4} During intracardiac EPS, a shortest preexcited R-R interval during atrial fibrillation >250 ms may be reassuring with a strong negative predictive value >99\%, but a positive test (shortest preexcited R-R interval <250 ms) may have a very low specificity.\textsuperscript{4,11} These findings, along with clinical outcome data, seem to be reflected in the electrophysiologist’s responses to probability of performing an ablation.\textsuperscript{12–14}

CONCLUSION

Pediatric EPS and RFA have become routine and safe. We found the clinical practice of pediatric electrophysiologists to reflect this, with more physicians using EPS as a tool to assess risk and proceeding to ablation when appropriate. This evolution in clinical practice may aid pediatricians and pediatric cardiol-

ogists in determining timing of referrals of these patients.

REFERENCES

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