HeartRhythm

The Official Journal of the Heart Rhythm Society, The Cardiac Electrophysiology Society, and The Pediatric & Congenital Electrophysiology Society

Heart Rhythm Society SM

Top Stories: Pediatric Electrophysiology

Top stories in Wolff-Parkinson-White syndrome (2022–2023)

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(Heart Rhythm 2024;21:357–358)

Incidence and outcomes of the WPW patterns in neonates

The Copenhagen Baby Heart Study, a multicenter, prospective, population-based cohort study of 17,489 neonates younger than 30 days, identified 17 with a WPW pattern, a prevalence of 0.1%.[1] This was more frequent in boys and was not associated with structural heart disease. Accessory pathways (APs) were primarily left-sided. At follow-up, the WPW pattern had disappeared in most, suggesting either an intermittent nature or that normalization occurs.

Life-threatening events in children with WPW syndrome in a large contemporary representative population

Identifying that estimates of life-threatening event (LTE) risk in WPW are limited by selection bias inherent to tertiary-care referral-based cohorts, a large claims database (IBM MarketScan Research database) was used to evaluate subjects with WPW (age 1–18 years).[2] Those with WPW syndrome were compared 3:1 with matched controls. "The prevalence of WPW syndrome was 0.03% (8733 per 26,684,581). Excluding congenital heart disease/cardiomyopathy, 6946 subjects were analyzed."[2 p.642] An LTE occurred in 49 (0.7%), including ventricular fibrillation (VF) in 20 (0.3%). The incidence of VF was 0.8 events per 1000 person-years, and an LTE was 1.9 events per 1000 person-years. There were no occurrences of VF in controls; the rate of LTEs was 70 times higher in subjects with WPW syndrome.

Myocardial work in children with WPW syndrome

WPW syndrome has been associated with dyssynchrony, and when left ventricular (LV) dysfunction is present, catheter ablation (CA) may be required, regardless of symptoms. To evaluate the value of noninvasive myocardial work in predicting abnormalities in myocardial performance in children with WPW syndrome, 75 children (age 8.7 ± 3.5 years) were recruited: 25 with manifest WPW syndrome and 50 age- and sex-matched controls.[3] Global myocardial work index, global myocardial constructive work, wasted work, and work efficiency were estimated as were standard parameters of LV function. A QRS duration of >110 ms was sensitive and specific at identifying worse values in work indices. In children with WPW syndrome, myocardial work indices were significantly reduced, even with normal LV ejection fraction and global longitudinal strain. This study supports the use of myocardial work during follow-up in children with WPW syndrome as a sensitive measure of LV performance and can aid in treatment decisions.

Association of weight with CA outcomes in pediatric WPW syndrome

A multicenter National Cardiovascular Data Registry IMPACT Registry study of 4456 subjects with WPW syndrome who had undergone CA from 2016 to 2019 reported an association between patient weight and procedural outcomes.[4] Subjects with weight < 30 kg (14%) were more likely to have preprocedural supraventricular tachycardia (45% vs 29%; $P < .001$) and less likely to have right septal AP (25% vs 33%; $P < .001$). Adverse events were rare, although with a higher incidence (transient atrioventricular block being the most common) in the <30 kg cohort (0.3% vs 0.05%; $P = .04$). Success was higher in the <30 kg cohort: 95% vs 92% ($P = .009$). Although these data reflect operator/center practices, higher rates of acute CA success were reported at the cost of higher rates of adverse events in patients with weight < 30 kg. These data may further inform the dialogue regarding the risk/benefit of CA in pediatric WPW syndrome.

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Wolff-Parkinson-White (WPW) syndrome is "the Rosetta Stone" of electrocardiography. In the 93 years since the initial description, debates on prevalence, risk, and management have peppered the scientific literature.

https://doi.org/10.1016/j.hrthm.2023.11.031
1547-5271/$-see front matter

Surgical treatment of WPW syndrome: Resuscitation of a forgotten technique

Ablation of APs is strongly recommended in patients with WPW syndrome and preexcited atrial fibrillation to prevent sudden cardiac death. There is an inherent assumption, supported by large studies, that all patients with WPW syndrome can be cured by CA. Vigneshwar et al[5] reported a case of a 27-year-old man with WPW syndrome and VF who underwent multiple failed CA attempts using endocardial and epicardial approaches and was eventually cured with surgery. This case is a reminder that certain APs are in the subendocardial posteroseptal area—the "pyramidal space" wherein a fat pad insulates the pathway from ablation energy. In this case the accessory pathway must be surgically divided. Before CA, surgery was the criterion standard for curing WPW syndrome. CA has replaced surgery, and hence surgical knowledge of this technique has faded. This report is a reminder of ongoing need for surgeons to have knowledge of operative techniques to appropriately care for rare high-risk patients.

Funding Sources: The authors have no funding sources to disclose.

Disclosures: Dr Shah is a consultant to Medtronic and Tenaya Therapeutics.

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