

A note from the SADS Foundation

We provide this information with the hope that informing physicians, other health care providers, and the public will encourage early and correct diagnosis and proper therapy.

Why do I need to know about WPW?

Published data estimate that 1-3/1000 individuals may have WPW syndrome. WPW-associated arrhythmias may precipitate sudden death in children and adolescents, as the first manifestation of the disorder. However, with increased awareness and effective treatment options, WPW syndrome can be diagnosed early and sudden death prevented. Still, this condition is often undetected prior to death and not recognized as the cause of death. Autopsy will often be “autopsy negative”, with no structural or functional cardiac abnormality detected and no postmortem “footprint” for WPW left behind. WPW syndrome is a treatable disorder, and with correct diagnosis, most deaths should be preventable.

Physicians need to know:

- When to consider WPW syndrome as a possible diagnosis.
- Approaches to risk stratify patients for risk of sudden death.
- Treatment options, including medications and cardiac ablative therapies.

Patients and Parents need to know:

- The warning signs and symptoms of WPW syndrome.
- Who to see for proper testing.
- How clinical arrhythmias may be managed.
- How to risk stratify patients to prevent cardiac risk.
- How to choose the best treatment options.

How is WPW inherited, and who in a known or suspected family should be tested?

WPW is not generally thought to be a genetic or heritable disorder. However, some familial studies have shown an incidence of 5.5/1000 patients among first-degree relatives following an index case of WPW (ventricular preexcitation).

What about genetic testing?

Genetic testing is not indicated for isolated ventricular preexcitation or WPW syndrome.

What is the treatment and who should be treated?

Treatment can include daily medications or cardiac ablation techniques. Patients with symptomatic SVTs or patients identified with high-risk AP profiles during electrophysiological testing could be considered for ablation procedures.

Medication compliance

As with any recurrent tachycardia, SVT may respond to daily medications to prevent recurrent episodes. Medication compliance, medication tolerance, and the presence or absence of side effects requires close monitoring.

How can parents protect their kids?

Consultation with a pediatric cardiac electrophysiologist for expert evaluation and management should be obtained.



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The Sudden Arrhythmia Death Syndromes (SADS) Foundation is a leader in education, research and advocacy. Our Mission is to save the lives and support the families of children & young adults who are genetically predisposed to sudden death due to heart rhythm abnormalities.

Wolfe-Parkinson-White Syndrome

A Guide for Patients and Health Care Providers

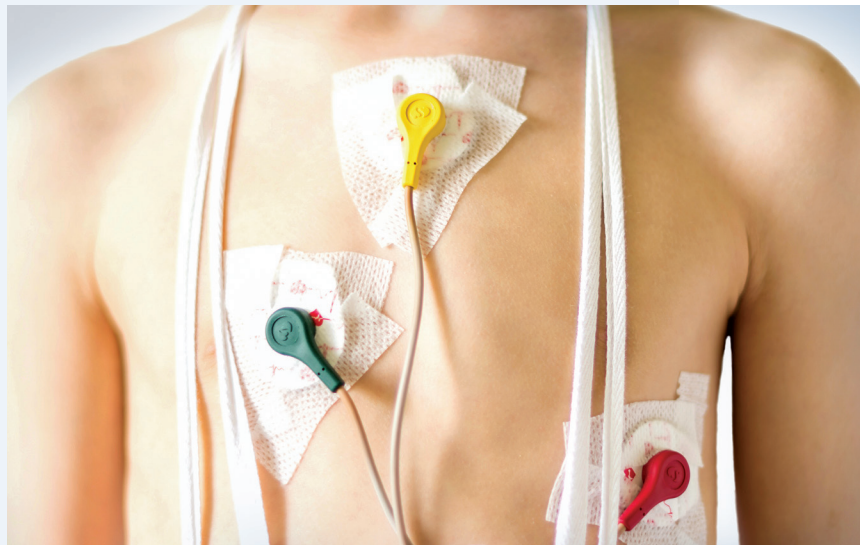


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What is WPW Syndrome?

Wolff-Parkinson-White (WPW) Syndrome was first described in a 1930 publication. As formally defined, WPW syndrome describes patients with unusual EKG pattern (ventricular preexcitation) and either clinical fast heart rates (tachycardias) and/or documented tachycardias. Some patients may manifest the EKG findings of ventricular preexcitation in the absence of tachycardias; often referred to as asymptomatic WPW syndrome, or alternatively, ventricular preexcitation.

The pattern of preexcitation on EKG results from electrical activation of the ventricles (lower pumping chambers) over an accessory pathway (AP), a microscopic electrical connection between the upper and lower chambers of the heart (atria and ventricles), respectively. This accessory pathway is in addition to the AV node, normally the only electrical conductor between the atria and ventricles. The cause of WPW syndrome is thought to be the result of failed regression of primitive muscle connections in the embryo between the atria and ventricles. In most patients ventricular preexcitation is associated with a structurally and functionally



normal heart. However, approximately 10-15% of patients may have associated structural congenital heart defects or heart muscle abnormalities. WPW Syndrome is not generally thought to be hereditary, but may recur in some families. In rare instances, ventricular preexcitation pattern may be present as a feature of a genetic mutation associated

with development of thickening of the heart muscle (hypertrophic cardiomyopathy – HCM).

The presence of an AP predisposes patients to the onset of sudden fast heart rates, called supraventricular tachycardia (SVT). In the presence of a structurally and functionally normal heart, most patients will present only with palpitations or lightheadedness. The most common SVT is called orthodromic reentry tachycardia (ORT). This looping tachycardia travels electrically down the AV node to the ventricles and then back to the atria over the AP. This is rarely a life threatening SVT, but may be frequent, sustained, and symptomatic causing lightheadedness, dizziness, and sometimes shortness of breath or chest discomfort. This tachycardia generally starts and stops precipitously. In children, adolescents, and young adults, an infrequent but serious type of WPW SVT may be associated with cardiovascular collapse due to potentially life threatening tachycardia. During extremely rapid atrial rates (> 300 bpm with atrial flutter or atrial fibrillation), the AP conducts these rapid rates to the ventricle, resulting in ventricular fibrillation, leading to cardiac collapse, cardiac arrest, and possibly sudden cardiac death. The incidence of this potentially life-threatening arrhythmia is thought to be no more than 1-2% of patients with ventricular preexcitation.

Cardiac catheterization with diagnostic electrophysiologic testing can be used to assess the electrical properties of the AP. Studies have defined which AP electrical properties profile a higher risk for cardiovascular collapse. The absence of the higher risk electrical profile generally predicts a low likelihood of any predictable catastrophic clinical risk.

A 2012 expert consensus statement from the Heart Rhythm Society and Pediatric and Congenital Electrophysiology Society describes management of young patients with asymptomatic WPW. This statement recommends risk stratification, using electrophysiologic testing, to identify patients with high-risk AP electrical properties and appropriate catheterization therapies to destroy the AP and thus the risk for sudden cardiac arrest. This 2012 publication now guides management of patients seen in pediatric cardiac arrhythmia programs across the country.



WPW syndrome can be regarded as potentially “curable” with catheter ablation techniques. Using temporary pacemaker catheters inserted during cardiac catheterization, radiofrequency energy (radiofrequency ablation) uses electrical heating to eliminate conduction over the AP. An alternative ablation therapy (cryoablation) uses freezing to destroy the AP. These pathways are generally considered high success targets for catheter ablation techniques but the risk of the procedure and the ablation must be considered when making the decision to attempt ablation procedures. A primary risk is permanent damage to the AV node (a complication referred to as complete heart block), which generally will mandate permanent pacemaker implantation. Other potential complications include bleeding, heart perforation, damage to the coronary arteries, and damage to cardiac valves. X-ray exposure should be considered a potential risk; newer catheterization laboratory imaging techniques may allow for EP diagnosis and ablation treatment without the need for any radiation exposure.

Indications for electrophysiologic diagnostic testing vary, but most electrophysiologists now follow the 2012 guidelines and recommend diagnostic electrophysiologic risk stratification in patients \geq 8-12 years of age to define AP electrical properties. Cardiac ablation can be performed in patients who have symptomatic SVTs and/or in asymptomatic patients with high-risk AP electrical profile at risk stratification.

What are the symptoms of WPW

Patients experiencing WPW-associated SVTs will generally feel palpitations (fast heart rates) and may have transient lightheadedness, chest discomfort, or shortness of breath. Patients with high-risk AP electrical properties may suffer syncope, seizure, or sudden cardiac arrest.

When should the diagnosis be suspected?

When someone presents with palpitations, syncope, or seizure of unknown etiology.

How is the diagnosis made?

Electrocardiogram. If ventricular preexcitation is noted, echocardiography should be performed to assess cardiac anatomy and function and to exclude associated structural congenital heart defects or heart muscle disorders (cardiomyopathy).