

## **Statement on WPW Syndrome**

**By Dr. Robert Campbell, SADS Board Member July 8, 2008**

Wolff-Parkinson-White (WPW) Syndrome was first described over half a century ago. The hallmark ECG features include presence of a delta wave (initial slurring of the QRS complex, wide QRS complex, and short PR interval) in patients who are predisposed to clinical tachycardia (fast heart rates). The ECG abnormalities described above are caused by electrical conduction over an accessory-wide pathway (a microscopic, hair-like 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, the only normal electrical conductor between the atria and ventricles. The cause of WPW syndrome is not known. In most cases it is associated with a structurally and functionally normal heart, though occasionally can be associated with congenital heart disease or heart muscle abnormalities. It is generally not considered to be hereditary, except in the very rare instance when associated with genetic mutation predisposing to WPW syndrome and hypertrophic cardiomyopathy (HCM).

Occasionally WPW syndrome is diagnosed by the presence of the classic ECG findings on an ECG obtained for other purposes; patients may be completely asymptomatic, with no clinical palpitations or tachycardia symptoms. The most common presentation is tachycardia presenting with palpitations and occasionally lightheadedness.

The most common tachycardia 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 accessory pathway. In patients with an otherwise functionally normal heart this is rarely a life threatening tachycardia, but may be frequent, sustained, and symptomatic with light headedness, dizziness, and some shortness of breath or chest discomfort. This tachycardia generally starts and stops precipitously.

However, a rare, but serious presentation can be sudden cardiovascular collapse due to potentially life threatening tachycardias associated with WPW syndrome.

Another tachycardia may be potentially life threatening. In this case the accessory pathway short circuits the AV node. In the presence of extremely rapid atrial rates ( $\geq 300$  bpm with atrial flutter or atrial fibrillation), the accessory pathway may conduct these rapid rates to the ventricle, resulting in ventricular fibrillation, leading to cardiac arrest and sudden death.

Cardiac catheterization with diagnostic electrophysiologic testing can be used to assess the electrical properties of the accessory pathway. Studies have defined accessory pathway electrical findings which profile a higher risk for cardiovascular collapse should atrial flutter or fibrillation occur clinically. The absence of higher risk electrical profile generally predicts the absence of catastrophic clinical risk.

In contrast to many other types of tachycardias, WPW syndrome can be regarded as potentially “curable” with catheter ablation techniques. Radiofrequency ablation uses electrical heating to eliminate conduction over the accessory pathway, while in contrast cryoablation uses freezing to destroy the pathway. These pathways are generally considered high success targets for catheter ablation techniques, but risks of the procedure and the ablation must be considered when making the decision to attempt ablation procedures. A primary concern is damage to the normal AV nodal conduction; permanent damage to the AV node results in a complication referred to as complete heart block, which generally will mandate permanent pacemaker implantation. Other complications including bleeding, heart perforation, and valve damage can occur.

Indications for electrophysiologic testing vary among physicians and from center to center, but many electrophysiologists now recommend consideration for diagnostic electrophysiology study to risk stratify the accessory pathway electrical properties. Ablation can be performed in patients who have symptomatic recurrent tachycardias and/or high risk accessory pathway electrical profile.