Wolff-Parkinson-White syndrome (WPW) is a syndrome of pre-excitation of the ventricles of the heart due to an accessory pathway known as the Bundle of Kent. This accessory pathway is an abnormal electrical communication from the atria to the ventricles.

The incidence of WPW syndrome is between 0.1 and 3% of the general population.

While the vast majority of individuals with WPW syndrome remain asymptomatic throughout their entire lives, there is a risk of sudden death associated with the syndrome. Sudden death due to WPW syndrome is rare (incidence of less than 0.6%, and is due to the effect of the accessory pathway on tachyarrhythmia’s in these individuals.

Pathophysiology

In normal individuals, electrical activity in the heart is initiated in the sinoatrial (SA) node (located in the right atrium), propagates to the atrioventricular (AV) node, and then through the bundle of His to the ventricles of the heart.

The AV node acts as a gatekeeper, limiting the electrical activity that reaches the ventricles of the heart. This function of the AV node is important, because if the signals generated in the atria of the heart were to increase in rate (as they do during atrial fibrillation or atrial flutter), the AV node will limit the electrical activity that conducts to the ventricles. For instance, if the atria are electrically activated at 300 beats per minute, half those electrical impulses are blocked by the AV node, so that the ventricles are activated at 150 beats per minute (giving a pulse of 150 beats per minute). Another important property of the AV node is that it slows down individual electrical impulses. This is manifest on the ECG as the PR interval, the time from activation of the atria (manifest as the P wave) and activation of the ventricles (manifest as the QRS complex).
Individuals with WPW syndrome have an accessory pathway that connects the atria and the ventricles, in addition to the AV node. This accessory pathway is known as the bundle of Kent. This accessory pathway does not share the rate-slowing properties of the AV node, and may conduct electrical activity at a significantly higher rate than the AV node. For instance, in the example above, if an individual had an atrial rate of 300 beats per minute, the accessory bundle may conduct all the electrical impulses from the atria to the ventricles, causing the ventricles to activate at 300 beats per minute. Extremely fast heart rates are potentially dangerous, and can cause hemodynamic instability. In some cases, the combination of an accessory pathway and cardiac arrhythmias can trigger ventricular fibrillation, a leading cause of sudden cardiac death.

**Diagnosis**

One beat from a rhythm strip in V2 demonstrating characteristic findings in WPW syndrome. Note the characteristic delta wave (marked in blue) (subtler here than in some cases), the short PR interval of 0.08 seconds, and the long QRS complex at 0.12 seconds.

WPW syndrome is commonly diagnosed on the basis of the surface ECG in an asymptomatic individual. In this case it is manifested as a delta wave, which is a slurred upstroke in the QRS complex that is associated with a short PR interval. The short PR interval and slurring of the QRS complex is actually the impulse making it through to the ventricles prematurely (across the accessory pathway) without the usual delay experienced in the AV node.
If the patient experiences episodes of atrial fibrillation, the ECG will show a rapid polymorphic wide-complex tachycardia (without turning of the points). This combination of atrial fibrillation and WPW is considered dangerous, and most antiarrhythmic drugs are contraindicated.

When an individual is in normal sinus rhythm, the ECG characteristics of WPW syndrome are a short PR interval, widened QRS complex (greater than 120 ms in length) with slurred upstroke of the QRS complex, and secondary repolarization changes reflected in ST segment-T wave changes. In individuals with WPW syndrome, electrical activity that is initiated in the SA node travels through the accessory pathway as well as through the AV node to activate the ventricles via both pathways. Since the accessory pathway does not have the impulse slowing properties of the AV node, the electrical impulse first activates the ventricles via the accessory pathway, and immediately afterwards via the AV node. This gives the short PR interval and slurred upstroke to the QRS complex known as the delta wave.

Patients with WPW often exhibit more than one accessory pathway, and in some patients as many as eight additional abnormal pathways can be found. This has been seen in individuals with Epstein’s anomaly.

Wolff-Parkinson-White syndrome is sometimes associated with Leber’s hereditary optic neuropathy (LHON), a form of mitochondrial disease.
Treatment is based on risk stratification of the individual. Risk stratification is performed to determine which individuals with WPW syndrome are at risk for sudden cardiac death (SCD). Sudden cardiac death in these individuals is due to the propagation of an atrial arrhythmia to the ventricles at a very high rate.

A good history should be taken to determine whether an individual has factors suggestive of a previous episode of unexplained syncope (fainting) or palpitations (sudden awareness of one's own, usually irregular, heartbeat). These may be due to earlier episodes of a tachycardia associated with the accessory pathway.

Individuals with WPW syndrome in whom the delta waves disappear with increases in the heart rate are considered at lower risk of SCD. This is because the loss of the delta wave shows that the accessory pathway cannot conduct electrical impulses at a high rate (in the anterograde direction). These individuals will typically not have fast conduction down the accessory pathway during episodes of atrial fibrillation.

Risk stratification is best performed via programmed electrical stimulation (PES) in the cardiac electrophysiology lab. This is an invasive procedure, in which the rate of impulse propagation via the accessory pathway is determined by stimulating the atria and by inducing transient atrial fibrillation.

High risk features that may be present during PES include an effective refractory period of the accessory pathway less than 270 ms, multiple pathways, septal location of pathway, and inducibility of supraventricular tachycardia. Individuals with any of these high risk features are generally considered at increased risk for SCD and should be treated accordingly.

It is unclear whether invasive risk stratification (with programmed electrical stimulation) is necessary in the asymptomatic individual. While some groups advocate PES for risk stratification in all individuals under 35 years old,
others only offer it to individuals who have history suggestive of a tachyarrhythmia, since the incidence of sudden death is so low.

Treatment

Acutely, people with WPW who are experiencing a tachydysrhythmia may require electrical cardioversion if their condition is critical, or, if more stable, medical treatment may be used. Patients with atrial fibrillation and rapid ventricular response are often treated with procainamide to stabilize their heart rate. Patients with a rapid heartbeat with narrow QRS complexes (circus movement tachycardias) may also be cardioverted, alternatively, adenosine may be administered if equipment for cardioversion is immediately available as a backup.

The definitive treatment of WPW syndrome is a destruction of the abnormal electrical pathway by radiofrequency catheter ablation. This procedure is performed almost exclusively by cardiac electrophysiologists. Radiofrequency catheter ablation is not performed in all individuals with WPW syndrome because there are inherent risks involved in the procedure.

When performed by an experienced electrophysiologist, radiofrequency ablation has a high success rate. If radiofrequency catheter ablation is successfully performed, the patient is generally considered cured. Recurrence rates are typically less than 5% after a successful ablation. The one caveat is that individuals with underlying Ebstein's anomaly may develop additional accessory pathways during progression of their disease.