Wolff-Parkinson-White Syndrome (WPW Syndrome)

Introduction

Wolff-Parkinson-White Syndrome is the second most common cause of regular supraventricular tachycardia in adults, and by far the most common cause of supraventricular tachycardia in the babies and children. The syndrome is caused by a residual congenital muscle fiber that crosses the normally-insulated valve rings that separate the upper and lower chambers of your heart (the atria and ventricles) called an "accessory pathway." The syndrome was named after doctors Wolff, Parkinson and White who noticed an abnormal electrocardiogram ("ECG" or "EKG") finding in 19 athletic males who all had racing heart rhythms. This abnormal EKG finding (called a "delta wave") allows doctors today to diagnose some individuals by their EKG alone. Patients who have a "delta wave" on their EKG but no symptoms to NOT necessarily require treatment of this finding, but those that have symptoms of palpitations or a racing heart rhythm are often recommended to undergo a heart electrical study called an "electrophysiology study" (or "EPS" or "EP Study" for short). It is important to realize that not all patients have a "delta wave" on their EKG. Sometimes, the accessory pathways are hidden on the EKG, and can only be detected by an electrophysiology study. During the electrophysiology study, if the doctor can localize where the accessory pathway is located, then one additional wire can be placed over the accessory pathway to cauterize (or "ablate") it. Because more and more pediatric cardiac electrophysiologists perform catheter ablation on children, this mechanism for supraventricular tachycardias has become less prevalent in adults.
What is an accessory pathway and where are they typically located?

An accessory pathway is nothing more than a small muscle fiber about 1-2 mm in size that crosses from the upper chambers (atria) to the lower heart chambers (ventricles). About 65% of these connections are located between the left atrium and left ventricle. The other 35% or so are located on the right side of the heart between the right atrium and right ventricle. While the EKG can sometimes assist in localizing the accessory pathway, an electrophysiology study can definitively localize the accessory pathway. Left-sided accessory pathways require the doctors to place a wire into the chambers of the heart that pump directly to the brain (either the left atrium or left ventricle), and therefore require a brief period of blood thinning (anticoagulation) during the catheter ablation procedure to prevent clots from forming on the burning wire (ablation catheter) that might cause a stroke.

If my doctors find a left-sided accessory pathway, how will they get to it?

There are two different ways for doctors to place the burning wire (ablation catheter) on the accessory pathway to destroy it. One involves placing the wire into the high pressure artery of the leg and passing it backwards into the left ventricle and then steer the catheter beneath the left-sided heart valve (the "mitral valve") to reach the connection between the atrium and ventricle. This is called the "retrograde aortic" approach. With this approach, the doctor has to manipulate the ablation catheter in the vigorously beating ventricle under the mitral valve. Serious risks with this approach include stroke, cardiac perforation, damage to the aortic or mitral valves, but these risks are not common.

The second approach is called the "transseptal approach." With the transseptal approach, a needle is passed through the wall between the right atrium and left atrium (called the inter-atrial septum) and a long plastic tube placed over the needle into the left atrium from the right groin vein. Through this tube, the burning wire ("ablation catheter") is passed and positioned above the mitral valve to burn the accessory pathway. Serious risks with this approach include stroke and cardiac perforation, but this approach provides unobstructed access to the mitral valve.

Efficacy with either approach is similar. The technique used will primarily be mandated by the familiarity of a particular approach by the cardiac electrophysiologist.

What Therapies Exist?

As mentioned above, catheter ablation is now considered the therapy of choice in symptomatic patients with supraventricular tachycardias, especially Wolff-Parkinson-White Syndrome. This does not mean that all patients MUST undergo the procedure, but if catheter ablation is successful, you are effectively "cured" of the rapid heart rhythm caused by this disorder. Alternatives to catheter ablation are outlined below.

Alternative therapies include medical therapy with a beta blocker medication (like
propranolol (Inderal®), atenolol (Tenormin®), metoprolol (Lopressor®), etc. Beta blocker medications lower adrenaline levels and prevent the rhythm from occurring and slow the rhythm if it does recur.

Alternatively, calcium channel blockers are rarely prescribed to slow conduction through the AV node. Verapamil can paradoxically accelerate accessory pathway conduction and is usually avoided. Diltiazem (Cardizem®) is occasionally used.

Other drugs like digoxin have been used, but are not as effective. Antiarrhythmic drugs such as Amiodarone (Pacerone®) and sotalol (Rhythmol®), can be used as well, but generally have more potential dangerous long-term side effects, and must be monitored closely.

Finally, like AV nodal reentry, some people have recurrent rapid rhythms that are infrequent and easy to self-control by using various methods to increase neural slowing to the AV node (so-called, "vagal maneuvers"). Using these maneuvers, one can sometimes stop the arrhythmia. These maneuvers include:

- Bearing down forcefully like you're having a bowel movement for 5-10 seconds, then slowly exhaling.
- Rubbing the carotid artery in the neck while lying down for approximately 5 seconds
- Placing very cold (soaked in ICE water) cloth on the face abruptly.
- Coughing forcefully

If one of the maneuvers is effective and the rhythm occurs infrequently (e.g., once a year) or is self-limited, then further therapy might not be required, but unlike AV nodal reentrant tachycardia, there are rare circumstances where evaluation of symptomatic patients, even those slightly symptomatic, is advisable (see below).

If these maneuvers are ineffective and you are symptomatic, you should go to an emergency room. Doctors there usually use a very short acting medication called "adenosine" to correct the rhythm. This medication is injected rapidly in your vein though an IV and one feels a very uneasy, warm sensation that passes quickly (usually in seconds) and the arrhythmia usually will stop. On rare occasions the adenosine may not work at low doses, and higher doses are needed. (Transplant patients are the exception, these patients usually need one tenth of the usual dose of adenosine, since they are VERY sensitive to the heart rate slowing effects of this drug on their transplanted heart.)

**My Doctor Mentioned WPW Syndrome Can be Life Threatening. Is this true?**

On very rare occasions, accessory pathways can conduct VERY rapidly from the upper to lower chambers of the heart. This becomes particularly important in people prone to develop a chaotic rapid atrial rhythm disturbance called "atrial fibrillation." Atrial fibrillation is caused multiple circulating wavefronts of electrical activation within the
upper chambers of the heart (the "atria") that effectively makes them beat 200-500 times per minute. When this occurs in a patient with a rapidly-conducting accessory pathway, these impulses can travel down the accessory pathway so rapidly that it causes the lower chambers (the ventricle) to also fibrillate (no longer pump effectively) and a person can die. This problem is extremely rare (1 in 10,000) but has been reported. It is for this reason that symptomatic patients are encouraged to have an electrophysiology study and possible catheter ablation.

**How is Catheter Ablation Performed?**

First of all, you'll be asked to eat NOTHING after midnight the evening before the procedure. Not uncommonly, your doctor might ask you to also NOT take some of the heart rhythm medications before the procedure so your rhythm can be initiated easier during the electrophysiology study (heart electrical study).

You will first to to a pre-procedure holding area, where and IV will be started and blood taken for analysis to assure the procedure is performed safely. Women of reproductive age commonly have pregnancy test the morning of or the day before the procedure. Next you will be connected to a series of EKG leads and a large patch electrode on your back and chest (these serve two purposes: (1) to apply energy to your heart during the "ablation" procedure, and (2) to shock the heart back to normal rhythm in the rare event an irregular or dangerous rhythm were started during the testing of your heart.) The leg area near the groin will be cleaned, possibly shaved, and prepared for the procedure. You will then be transported into the electrophysiology laboratory and asked to lie on a thin table under an xray tube. You'll be secured to the table gently. The legs will again be prepped and a large drape placed over your chest and legs. You will be sedated with medications that help relieve anxiety and discomfort during the procedure. The doctor will then use a local anesthetic to numb the legs at the crease where your leg attaches to the pelvis near the groin. After this, a small needle will be painlessly placed into the vein of your right and left legs and a series of small tubes placed in the veins that have one-way valves on them to permit the passage of wires, but no bleeding from your leg. A series of wires are then passed to your heart under xray guidance. There are no nerves inside the blood vessels leading to your heart, so this is typically completely painless. Once in the proper place, your doctor will measure conduction times within the heart, and then pace the heart in the upper and lower chambers to study the conduction properties of the heart chambers. Skipped heart beats will be applied using a special computer and attempts will be made to induce the abnormal heart rhythm you have been experiencing. You may be able to perceive heart skipping or racing as this portion of the testing is performed. Occasionally, an adrenaline-like medication is required to start the heart rhythm. When this medication is administered, you also might notice your heart pounding more than usual. Do not be concerned, stopping the infusion of this medication will stop the pounding sensation in about 15-20 minutes.

Once the heart rhythm is started, the doctors will study what it takes to initiate and terminate the rhythm. After this is performed, preparations are made to perform catheter
A ablation. A special steerable wire with a slightly enlarged tip is passed from the legs to the heart and positioned to the critical portion of the conduction pathway required to sustain the rhythm (this is determined during the earlier pacing studies). If the accessory pathway is found on the left side of the heart, the ablation wire is placed into either the left ventricle or left atrium using the "retrograde aortic" and transseptal approaches described above. A blood thinner called heparin is administered after the wires are in place to prevent clots from forming on them. One or more applications of radiofrequency energy applications are then applied to the accessory pathway (the name "radiofrequency" comes from the fact that it is a 500kHz alternating current electricity that is applied, and happens to be at the same frequency as 500 on your AM radio dial!). This alternating current passed from the patch on your back, through your body, and received at the ablation wire in your heart. This causes the tip of that wire to become hot (like the filament of a lightbulb) and cauterizes (burns) the tissue beneath the wire. If placed correctly, this should interrupt the reentrant circuit and no further rapid heart beats occur.

After the ablation procedure, additional skipped beats are applied to the heart to be sure no further heart rhythms occur. Once the doctor is satisfied that no abnormal rhythms can occur, you are often monitored in the lab an additional 15-30 minutes and repeat testing performed one more time. If the abnormal heart rhythms STILL cannot be re-initiated, then the procedure is considered complete. If the blood thinner heparin was used, the drug will be "reversed" using another compound. All catheters (wires) will be removed from your heart, the small tubes placed in your legs will be removed, pressure will be applied to the areas of the catheters, then band-aides applied over the wounds. You'll then be removed from the laboratory and observed for at least 4 hours to assure you have tolerated the procedure well.

**What Are the Risks With Catheter Ablation?**

If the accessory pathway lies adjacent to the AV node, then the greatest risk with catheter ablation of the accessory pathway will be accidentally destroying the normal AV node, that is, of developing "complete heart block." If this were to occur after the accessory pathway is ablated, then NO impulses can travel from the atrium to ventricle, rendering the patient pacemaker-dependent (that is, a permanent pacemaker would have to be installed). Depending of the proximity of the accessory pathway to the AV node, the risk of development of this complication is less than or equal to 3 percent. Other risks include, but may not be limited to, bleeding (especially at the sites where the catheters enter the leg bleed vessels), development of a communication between the vein and artery of the leg (called a "fistula") that has to be surgically corrected, development of a large blood clot in the veins of the legs (called a "deep venous thrombosis or DVT"), damage to the heart itself, including perforation of the heart requiring emergent drainage of blood from the lining around the heart (called the pericardium), stroke, infection, nerve injury to the leg nerve, and a reaction to the medications or contrast agents that might be used during the procedure. On occasion, some electrophysiologists place a catheter in the vein in the upper chest area just beneath the collar bone to go to the heart. It is possible to inadvertently puncture the lung while placing that catheter (called a "pneumothorax"). Radiation (xray)
or electrically-induced skin damage can also occur. In total, collective risks in experienced centers are less than or equal to approximately 1.5%. Death is an extremely rare complication.

What Happens After the Catheter Ablation

If your procedure is uncomplicated, most people return home in 4-6 hours. Your doctor will often stop at least some of your heart rhythm medications after the procedure. Have a family member or friend drive you home from the hospital, and normal activities should be able to resume in one to two days after the procedure. Heavy exertion should resume only after the leg sites have completely healed. It is common to feel a small bump or note a small bruise at the sites where catheters were inserted into the legs. These usually completely resolve within a few weeks.

When Should I Call the Doctor After My Procedure?

Call your doctor if:

- The catheterization sites in your legs begins to bleed or the site becomes increasingly painful or swollen
- You feel shortness of breath or chest discomfort
- You have a fever over 100.5
- You feel the symptoms of your heart rhythm problem return

Where can I get additional information?

Additional information can be found at the Heart Rhythm Societies Public and Patients Webpage at: http://www.hrspatients.org/.

Or you can post a question on the MedTees.com Forum section.

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