The Wolff-Parkinson-White syndrome

PSVT due to accessory pathways (PSVT. The mechanism of the tachyarrhythmia relates to the presence of two pathways between the atria and the ventricles)

This syndrome is the second most common cause of PSVT. The mechanism of the tachyarrhythmia relates to the presence of two pathways between the atria and the ventricles that have different conducting properties (see figure1).

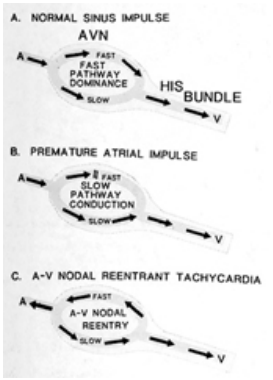

Figure 1
click for big picture

Usually the period of these pathways during which they can not respond to stimulus to conduct (refractory period) exceeds that of the normal AV nodal-His pathway. Thus, a premature atrial impulse may block at the accessory pathway and conduct antegrade down the normal pathway and enter the accessory one in a retrograde direction and reentering the atrium to cause a circus movement tachycardia (orthodromic, see figure 3).

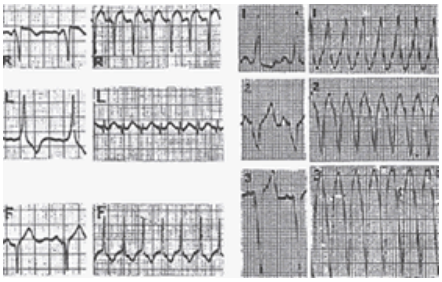

Figure 3
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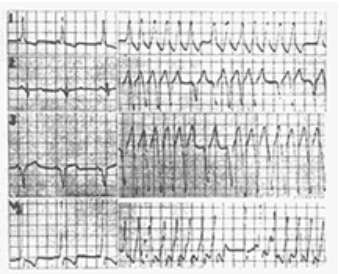

Figure 3a
click for big picture

Since the accessory pathway provides retroconduction to the atria, P waves (if seen) are usually inverted in EKG leads like AVF and V5-6.

Less common is for the accessory pathway to have a shorter refractory period causing a block of an initiating premature atrial impulse in the normal pathway, with antegrade conduction down the anomalous pathway and retrograde invasion of the normal AV node pathway to establish an antidromic tachycardia (see figure 3) with wide QRS's in the EKG. These wide QRS tachycardias may be difficult to distinguish from VT if the existence of WPW was not known prior to presentation with a tachyarrhythmia. In concealed WPW syndrome, only orthodromic tachycardias can occur because of the inability of the bypass tract to conduct in the antegrade direction. Distinction between concealed WPW and AV nodal

reentrant tachycardia may be difficult, although a faster rate (>200 per minute) and a retrograde P wave after, rather than within,the QRS complex favor concealed WPW.When atrial flutter or fibrillation occur in patients with WPW, the risk of potentially lethal arrhythmias due to very rapid conduction across accessory pathways must be considered. The risk is especially treacherous in patients with short-refractory period anomalous pathways, since atrial fibrillation may lead to ventricular fibrillation.

Reference:Myerburg,R.J. and others,Hurst's The HEART,8TH Edition,Recognition Clinical Assessment,and Management of Arrhymthmias and Conduction Disturbances,Ch.36,pp.705-758

Interpretation of PSVT's

PSVT's can be classified into two major groups:short RP and long RP tachycardias, that is the P wave during the SVTs occurs either in the first or second half of the tachycardia cycle.Since the PR interval is inversely related to the RP interval,short RP tachycardias have long PR intervals,and long RP intervals have short PR intervals.

SHORT RP SVTs

These SVTs are defined by having atrial activity:

1) obscured by the QRS complex because ofthe simultaneous inscription of both,
2) occurring in the terminal portion of the QRS complex and often giving the appearance of an R' in lead V1. or
3) present in the STsegment.

Thus, the interval from the onset of the QRS to the P wave is short--"short RP SVT". The most likely SVT for the first and second examples is atrioventricular nodal pathway reentrant tachycardia (AVNRT), using the slow AV nodal pathway anterogradely and the fast AV nodal pathway retrogradely. An SVT traveling to the ventricle over the AV node and back to the atrium over an accessory pathway, called "atrioventricular reentrant tachycardia" (AVRT; WPW), is the most likely cause of the third example, and less commonly the second.

Important data can help refine the diagnosis if a functional bundle branch block (FBBB) also occurs. Prolongation of the SVT cycle length during FBBB is most consistent with an AVRT and the location of the accessory in the same ventricle that gave rise to the FBBB. Thus prolongation of the SVT cycle length during a period of a functional left BBB (LBBB) would be found during AVRT due to retrograde conduction over a left- sided accessory pathway; the same analysis applies to functional right BBB (RBBB) and right-sided accessory pathway.The cycle length prolongs because,during the FBBB,the antegrade impulse must first activate the ventricle contralateral to the ventricle with the FBBB. Failure of the FBBB to prolong the cycle length of the AVRT occurs when the accessory pathway is located contralateral to the ventricle with the FBBB, in many AVRTs due to septal accessory pathways, and non-WPW forms of SVT.EKG algorithms based on the form of the delta wave of the WPW complex can be used to determine the location of the accessory pathway.

LONG RP SVTs

These are characterized by atrial activity locatd "just before" the next QRS complex,so that the P wave is located in the second half of the tachycardia cycle at a conductible PR interal of approximately 300ms. or so. This SVT creates a long interval from the preceding QRS complex to the next P wave. This type is typical of atrial tachycardia and two other SVTs. One is an unusual form of AVRT comprised of a slowly conducting accessory pathway that creates an incessant SVT, which pauses briefly for a few sinus beats and then resumes (called junctional reciprocating tachycardia, PJRT) and because of its incessant nature can cause a tachycardia cardiomyopathy.

The impulse travels over the normal conducting system and then back over the slow-conducting accessory pathway. The third type of long RP SVT is an unusual form of AVNRT, during which the impulse travels to the ventricle over the fast-conduction pathway, and retrogradely over the slow-conducting pathway (the reverse route traveled by the usual type of AVNRT).

Intermittent RP Tachycardia

Several infrequently occurring SVT's can give rise to tachycardias that have PR and RP intervals of about the same duration so that the P wave is found midway in the tachycardia cycle. They include AVNRT when the impulse travels over two slowly conducting pathways, unusual forms of AVRTs, and some atrial tachycardias.

Spontaneous Onset Or Termination of The SVT

A sustained SVT initiated by a premature atrial complex(PAC) causing block in an accessory pathway is most likely an AVRT, where as a sustained SVT started by a PAC that significantly prolongs the PR interval is probably AVNRT. An SVT always stopping with a P wave rather than a QRS complex as the electral event last inscribed in the EKG is unlikely to be an atrial tachycardia, because the atrial focus would always have to block en route to the ventricle at the same time it stopped discharging, unlikely set of coincidences to happen repeatedly. Far more likely is either AVNRT or AVRT, during which atrial activity blocks before reaching the ventricle, thus interrupting the reentrant loop and terminating the tachycardia. Similarly, an SVT that persist uninterruptedly despite blocked P waves is almost certainly an atrial tachycardia,rarely AVNRT, and never the usual forms of AVRT.

Reference:Zipes,D.P.,Clinical Application of the EKG,JACC,Vol.36,No.6,2000:1746-8

Management of PSVT due to WPW syndrome includes: adenosine, verapamil, diltiazem, pronestyl, and quinidine,which may be used to convert the acute tachycardia to normal. Digoxin is to be avoided to prevent shortening of refractory period of accessory pathway as well as atrial muscle. Electrical cardioversion may be used if drug therapy fails. Catheter ablation can be used for life threatening arrhythmias in WPW (atrial fibrillation, see figures 1b, 3b).

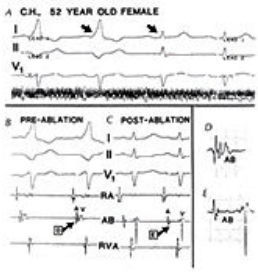

Figure 3b
click for big picture

Surgery can be used if ablation therapy is unsuccessful.

Reference:Myerburg,R.J. and others,Hurst's The HEART,8TH Edition,Recognition Clinical Assessment,and Management of Arrhymthmias and Conduction Disturbances,Ch.36,pp.705-758

Wolff-Parkinson-White Syndrome
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INTRODUCTION

Background: Wolff-Parkinson-White (WPW) syndrome is a congenital abnormality associated with supraventricular tachycardia (SVT). It involves an activation of the ventricles that occurs earlier than expected, called preexcitation, which occurs because of conduction of an atrial impulse not by means of the normal conduction system, but via an extra atrioventricular (AV) muscular connection, termed an accessory pathway, that bypasses the AV node.

Classic ECG findings of WPW syndrome include a short PR interval (<120 ms), a wide QRS complex of longer than 120 milliseconds with a slurred onset producing a delta wave in the early part of QRS, and secondary ST-T wave changes.

Patients with WPW syndrome are potentially at an increased risk of dangerous ventricular arrhythmias due to extremely fast conduction across the bypass tract if they develop atrial flutter or fibrillation. Certain patients with WPW syndrome are at risk for sudden death. In these patients, cardiac electrophysiologic (EP) studies and radiofrequency (RF) catheter ablation may be curative. Other patients have symptomatic SVT, which can also be cured by catheter ablation. Asymptomatic patients may merely need periodic observation.

This review discusses the pathogenesis, clinical presentation, evaluation, and treatment of patients with WPW syndrome.

Pathophysiology: Patients with preexcitation may have SVT due to a reentrant mechanism. The genesis of reentrant SVT involves the presence of dual conducting pathways between the atria and the ventricles. These pathways include (1) the natural AV nodal His-Purkinje tract and (2) an AV accessory tract (ie, AV connection or bypass tract, Kent fibers, or Mahaim fibers.

The 2 pathways usually exhibit 2 different conduction properties and refractory periods that facilitate reentry. The effective refractory period (ERP) of the accessory tract is usually longer than that of the normal AV nodal His-Purkinje tract. Several types of SVT have been described, including orthodromic tachycardias, orthodromic tachycardia with a concealed accessory pathway, and antidromic tachycardia.

Orthodromic tachycardia

When a premature ectopic atrial impulse begins to traverse down towards the ventricle, it may block at the accessory tract but conduct in antegrade fashion down the normal pathway. The impulse then reenters the accessory tract in retrograde fashion to perpetuate a circus movement of the impulse. Such reentrant tachycardia is described as orthodromic. Premature ventricular contractions (PVCs) can also initiate orthodromic tachycardia.

In orthodromic tachycardia, the normal pathway is used for ventricular depolarization and the accessory tract is used for reentry. On ECG findings,

the delta wave is absent, QRS complex is normal, and P waves are inverted in the inferior and lateral leads.

Orthodromic tachycardia with a concealed accessory pathway

Some accessory (bypass) tracts are unable to conduct in the antegrade fashion. These are called concealed accessory pathways (ie, concealed WPW syndrome). Although no evidence of the pathway is present during sinus rhythm (ie, no preexcitation), orthodromic tachycardias can occur.

Differentiation between this type of SVT and usual AV nodal reentrant tachycardia (AVNRT) may be difficult. Nonetheless, if the heart rate is higher than 200 beats per minute and a retrograde P wave is visible following the QRS complex, a concealed accessory pathway may be the diagnosis.

Antidromic tachycardia

Less commonly, a shorter refractory period in the accessory tract may cause block of an ectopic atrial impulse in the normal pathway, with antegrade conduction down the accessory tract and then retrograde reentry of the normal pathway. This type of tachycardia produced is called antidromic tachycardia.

On ECG findings, the QRS is wide, which is an exaggeration of the delta wave during sinus rhythm (ie, wide-QRS tachycardia). Such tachycardias are difficult to differentiate from ventricular tachycardia.

Thus, the mechanism underlying the majority of the tachycardias in patients with WPW syndrome is macroreentry caused by antegrade conduction over the AV node His bundle pathway and retrograde conduction over an accessory pathway (orthodromic). Less common in patients with WPW syndrome is antidromic tachycardia. Even when the accessory pathway conducts only in retrograde fashion, it can still participate in the reentrant circuit and produce an orthodromic AV reciprocating tachycardia.

Frequency:

In the US: The prevalence of ventricular preexcitation is thought to be 0.1-0.3% in the general population. Estimates of arrhythmia incidence in patients with preexcitation vary widely, ranging from 12-80% in several surveys.

Incidence of preexcitation and WPW syndrome varies from 0.1-3 cases per thousand population (average of 1.5 cases per thousand population) in otherwise healthy persons.

In a review of ECG findings from 22,500 healthy aviation personnel, 0.25% exhibited findings consistent with the WPW pattern, with 1.8% incidence of tachycardia.

The location of the accessory pathways, in descending order of frequency, is (1) the left free wall, (2) posteroseptal, (3) right free wall, and (4) anteroseptal.

The presence of concealed accessory pathways accounts for approximately 30% of patients with apparent SVT referred for EP evaluation.

Approximately 80% of patients with WPW syndrome have a reciprocating tachycardia, 15-30% have atrial fibrillation, and 5% have atrial flutter. Ventricular tachycardia is uncommon.

Internationally:

Incidence and prevalence of WPW syndrome worldwide parallels that in the United States.

Mortality/Morbidity:

Patients with WPW syndrome have a very small risk of sudden arrhythmic death. Medical therapy with agents such as digoxin may increase this risk. The risk in asymptomatic patients is extremely low.

Overall, sudden death occurs rarely, with an estimated frequency rate of 0.1%.

Other factors that appear to influence risk are the presence of multiple bypass tracts and a family history of premature sudden death. Sudden cardiac death is unusual without preceding symptoms.

Race: No clear racial predilection appears to exist.

Sex: Prevalence may be higher in males.

Age: Certain factors exist regarding age and the prevalence of preexcitation and WPW syndrome.

WPW syndrome is found in persons of all ages, from those in fetal and neonatal age groups to elderly individuals.

Prevalence decreases with age because of loss of preexcitation. Cases have been described in which electrocardiographic evidence of preexcitation disappears.

In patients with abnormal ECG findings indicative of WPW syndrome, the frequency of SVT paroxysms increases from 10% in people aged 20-39 years to 36% in people older than 60 years.

CLINICAL

History:

WPW syndrome can result in SVT that uses an AV accessory (bypass) tract. The accessory pathway may also act as an innocent bystander and allow conduction during other supraventricular arrhythmias, such as atrial fibrillation or flutter. The possibility of a concealed bypass tract as a mechanism underlying certain types of SVT should be considered because treatment options may vary. Paradoxically, the use of digoxin and perhaps other AV nodal blocking agents may accelerate conduction through the bypass tract, causing potentially lethal ventricular arrhythmias or hemodynamic instability during atrial fibrillation.

SVT in WPW syndrome may begin in childhood or not appear clinically until the patient reaches middle age. In some patients in whom it first presents during childhood, it may then cease for some time, only to recur. In fact, the probability is 75% that the tachycardia will persist if it is still present in patients older than 5 years.

In asymptomatic patients, the probability of losing the capacity for antegrade conduction across the accessory pathway increases with advancing age. This probably results from fibrotic changes at the site of insertion of the accessory bypass tract.

In patients with WPW syndrome, the tachycardia that produces symptoms may be an SVT, atrial fibrillation, or atrial flutter. In a series of 212 patients with tachyarrhythmias and WPW syndrome, SVT alone occurred in 64%, atrial fibrillation alone in 20%, and both occurred in 16% of patients.

Light-headedness and near syncope appear to occur more commonly in persons with WPW syndrome who have paroxysmal SVT (PSVT) or atrial fibrillation than in those with AV nodal reentry.

Syncope can occur because of inadequate cerebral circulation due to a rapid ventricular rate or because the tachyarrhythmia is depressing the sinus pacemaker, causing a period of asystole at the point of tachycardia termination.

PSVT can be followed after termination by polyuria, which is due to atrial dilatation and release of atrial natriuretic factor.

Physical:

During SVT, the rhythm is unvarying and regular, with constant intensity of the first heart sound.

The jugular venous pressure can be elevated, but the waveform generally remains constant.

Clinical features of associated cardiac defects may be present, such as the following:

Mitral valve prolapse

Cardiomyopathy

Ebstein anomaly: Patients with right-sided accessory pathways should be screened for the Ebstein anomaly.

The abnormal QRS complexes of WPW syndrome, when present, may appear similar to those observed in acute myocardial infarction (MI). Repolarization abnormalities are common in patients with WPW syndrome.

Causes: In patients with WPW syndrome, the underlying cardiac structural abnormality consists of accessory conduction tissue that bypasses the normal AV node His-Purkinje system pathway. Such pathways are generally believed to be congenital in nature.

The causes of WPW syndrome can be summarized as follows:

Congenital or hereditary

An accessory pathway is quite likely to be congenital, although its manifestations can be detected in later years and it may appear to be acquired.

Relatives of patients with preexcitation, particularly those with multiple pathways, have an increased prevalence of preexcitation, suggesting a hereditary mode of acquisition.

Associated with congenital cardiac defects

Patients with the Ebstein anomaly may develop WPW syndrome. Patients with the Ebstein anomaly frequently have multiple accessory bypass tracts, mostly right-sided, in the posterior part of the septum or the posterolateral wall. Preexcitation generally occupies the atrialized ventricle. The orthodromic reciprocating tachycardia in such patients exhibits right bundle-branch block (RBBB) and a long ventriculoatrial (VA) interval.

Mitral valve prolapse may be a congenital cardiac defect and may cause WPW syndrome.

Hypertrophic cardiomyopathy may include idiopathic hypertrophic subaortic stenosis or asymmetric septal hypertrophy.

Associated with other acquired cardiac defects - Cardiomyopathies

DIFFERENTIALS

Atrioventricular Nodal Reentry Tachycardia (AVNRT)
Ebstein Anomaly
Lown-Ganong-Levine Syndrome
Syncope

Other Problems to be Considered:

In Lown-Ganong-Levine (LGL) syndrome, patients have a short PR interval and SVT, but no delta wave.

Mahaim fibers connect the atria to the right bundle or the AV node to the ventricle. Such bypass tracts are called atriofascicular. If atriofascicular fibers are present, the ECG findings are a normal or short PR interval and the QRS complex is abnormally wide with a left-bundle appearance. These fibers have decremental conduction properties and can perpetuate clinically significant tachycardias or act as innocent bystanders for other types of tachycardias (eg, AVNRT).

Sometimes, the fibers arise in the His bundle or bundle branches and insert into the ventricular myocardium. These are called fasciculoventricular tracts and, generally, are not involved in tachycardias.

Differential diagnosis of accessory pathway syndromes using EP studies

In patients with LGL syndrome who have an atriohisian tract, the QRS complex remains normal and the short atriohisian interval remains fixed during atrial pacing at rapid rates.

Patients with fasciculoventricular connections show a short His-ventricle (HV) interval and no change in the QRS complex during rapid atrial pacing.

Atriofascicular tract pathways usually represent a duplication of the AV node and the distal conducting system. They occupy the right ventricular free wall. Their proximal end resides adjacent to the lateral tricuspid annulus and exhibits slow conduction, with AV nodelike characteristics. The distal end, which conducts rapidly, inserts into the distal right bundle branch or the apical region of the right ventricle. Preexcitation may not be apparent during sinus rhythm but can be demonstrated with premature right atrial stimulation. Because retrograde conduction is absent, only an antidromic AV reentry tachycardia (ie, preexcited tachycardia) can develop.

Furthermore, concerning atriofascicular tracts, preexcited tachycardia has a left bundle-branch block pattern, long AV interval (due to the long conduction time over the accessory pathway), and short VA interval. If RBBB develops, it can become, by increasing the length of the tachycardia circuit (ie, VA interval prolongs owing to delay in retrograde activation of the His bundle), proarrhythmic and the tachycardia can become perpetual and persistent.

Patients with PSVT usually have narrow QRS complexes. The QRS may become wider owing to aberrant conduction, coexisting bundle-branch block, or involvement of an accessory pathway.

Other forms of tachycardia in patients with WPW syndrome

Patients with WPW syndrome can have other tachycardias during which the accessory pathway is just a bystander, such as AVNRT or an atrial tachycardia that conducts to the ventricle over the bypass tract.

Atrial flutter or fibrillation may also occur in the atrium, unrelated to the bypass tract.

Patients with WPW syndrome who have atrial fibrillation frequently have inducible reciprocating tachycardias. Interruption of the accessory pathway with ablation can prevent recurrence of the atrial fibrillation.

Atrial fibrillation presents a potentially serious risk. At rapid rates, the refractory period of the accessory pathway can shorten, allowing an exceedingly rapid ventricular response. However, such a phenomenon is uncommon, occurring at an estimated frequency of less than 0.1%.

Patients who have intermittent preexcitation or those who lose ECG evidence

of preexcitation with exercise or when injected intravenously with procainamide generally have a long refractory period of the bypass tract. These patients are thought to have a low risk of developing a rapid ventricular rate should atrial flutter or fibrillation develop.

Atrioventricular Nodal Reentry Tachycardia (AVNRT)

Ebstein Anomaly

Lown-Ganong-Levine Syndrome

Syncope

WORKUP

Lab Studies:

Routine blood studies may be needed to help rule out noncardiac conditions triggering tachycardia. These may include the following:

Complete blood cell count

Chemistry panel

Blood urea nitrogen and creatinine to assess renal status

Liver function tests (eg, bilirubin and transaminase levels)

Thyroid panel

Blood levels of antiarrhythmic medications during therapy and monitoring

Imaging Studies:

Echocardiogram may be needed to assess left ventricular function and wall motion and to help rule out valvular disease, Ebstein anomaly, hypertrophic cardiomyopathy (in which the incidence of accessory pathways is increased), or other congenital cardiac defects.

Other Tests:

The diagnosis and management of any cardiac arrhythmia can be accomplished by using findings from ECG and rhythm strip analysis and their relationship to the clinical setting. Recognizing arrhythmias on ECG findings requires a thorough knowledge of atrial and ventricular activation patterns and deductions related to the mechanisms of AV conduction.

The standard 12-lead ECG and additional rhythm strips form a direct and easily accessible resource for analyzing abnormalities of the cardiac rhythm. For many simple arrhythmias, mere recognition of P-wave and QRS morphologies, with their relative timing and their vectors, may be sufficient to confirm a diagnosis.

The location of the accessory pathway using ECG can often be determined by a thorough analysis of the spatial direction of the delta wave in the 12-lead ECG findings by reviewing the maximally preexcited QRS complexes.

Ladder diagram of the ECG

Analysis of more complex arrhythmias may require the use of ladder diagrams or Lewis lines (named after Sir Thomas Lewis,

who first used them).

The ladders usually consist of 3 tiers: A, AV, and V. Additional tiers, such as sinoatrial (SA) conduction, may be added. The A and V tiers correspond to the activation of atrial (A) and ventricular (V) muscle.

AV is used to show conduction in the AV junction. The A line is drawn from the beginning of the P wave, and the V line is drawn from the beginning of the QRS. Time is indicated by the slope of the line.

The site of origin may be represented by a black dot.

A blocked impulse is indicated by a short bar at a right angle to the line, indicating the direction of conduction, and aberrant intraventricular conduction is shown as a pair of slightly divergent lines.

Special ECG leads

When the standard ECG fails to provide adequate information to support a diagnosis, often because of a failure to recognize P waves, certain additional special lead systems can be used to help establish the diagnosis.

A bipolar esophageal lead is used to record left atrial activity, while an intra-atrial electrode during catheterization can be used to record atrial activity from within the right atrium.

Continuous ECG recordings (ie, telemetry, 24-hour Holter monitor, event monitor, implantable loop recorder)

Continuous monitoring of cardiac rhythm can be performed on hospitalized patients in the coronary or the progressive care units with telemetry.

In the outpatient setting, a number of portable recording devices (eg, Holter monitors, event monitors) can be used.

Portable recording systems provide simultaneous 2-lead recording that improves the diagnostic yield tremendously. The 2 most commonly used leads for monitoring are lead II and MCL-I, the latter being similar to V1. These devices have long-term storage capabilities that permit off-line analysis of complex arrhythmias, even if the physician is not available at the time the rhythm disturbance occurs.

For infrequently occurring arrhythmias, a number of event recorders are available. They allow the patient to activate the device by pressing a button when an event occurs, providing internal storage and transmission by telephone or wireless communication to a central station for later review.

Transtelephonic transmitters can be used in real time for somewhat more persistent or frequent events.

A small loop recorder can be implanted similar to a pacemaker and can be removed later for analysis. This can be used in patients with arrhythmias that are difficult to capture.

ECG recognition of reentry over a retrograde (concealed) accessory pathway

A bypass tract that conducts unidirectionally only from the ventricle to the atrium is not detectable on the regular surface ECG findings because the ventricle is not preexcited; thus, the ECG manifestations of WPW syndrome are absent.

Such a bypass tract is described as concealed.

Tachycardia due to the concealed tract should be considered when the QRS complex is normal and the retrograde P wave occurs well after completion of the QRS complex, out in the ST segment or even in the T wave.

Diagnosis of accessory pathways

During ventricular pacing, premature ventricular stimulation activates the atria before retrograde depolarization of the His bundle. This indicates that the impulse reached the atria before it depolarized the His bundle and must have traveled a different pathway (bypass tract).

If the ventricles can be stimulated prematurely during tachycardia at a time when the His bundle is refractory and the impulse still conducts to the atrium, this indicates that retrograde propagation traveled to the atrium over a pathway other than the bundle of His.

If the premature ventricular complex depolarizes the atria without lengthening of the VA interval and with the same retrograde atrial activation sequence, the stimulation site (ie, ventricle) may be assumed to be within the reentrant circuit without intervening His-Purkinje or AV nodal tissue that might increase the VA interval and therefore the AA interval.

In addition, if a premature ventricular complex delivered at a time when the His bundle is refractory terminates the tachycardia without retrograde activation of the atria, it most likely invaded, and blocked in, an accessory pathway.

The VA interval (a measurement of conduction over the accessory pathway) is generally constant over a wide range of ventricular paced rates and coupling intervals of premature ventricular complexes and during the tachycardia in the absence of aberration. Similar short VA intervals can be observed in some patients during AV nodal reentry, but if the VA conduction time or R-P interval is the same during tachycardia and ventricular pacing at comparable rates, an accessory pathway is almost certainly present. The VA interval is usually less than 50% of the R-R interval.

Tachycardia can be initiated easily following premature ventricular stimulation that conducts in retrograde fashion in the accessory pathway but blocks in the AV node or His bundle. Atria and ventricles are required components of the macroreentrant circuit; therefore, continuation of the tachycardia in the presence of AV or VA block excludes an accessory AV pathway as part of the reentrant circuit.

Stress testing

This is an ancillary test and may be used to (1) reproduce a transient paroxysmal arrhythmia, (2) document the relationship of exercise to the onset of tachycardia, (3) evaluate the efficacy of therapy, and (4) assess adverse responses.

A bicycle ergometer or standard treadmill can be used.

Thallium or echocardiographic imaging is not necessary unless an ischemic etiology is considered as a potential cause or trigger of the onset of arrhythmia.

Stress testing may also provide some general insight into the refractory periods of accessory pathways in patients with WPW syndrome.

Procedures:

Intracardiac EP studies

EP studies are performed in a cardiac electrophysiology laboratory. Using multicatheter electrode systems, recordings from many intracardiac sites can be performed simultaneously, facilitating delineation of the sequence of depolarization and impulse conduction in the atria, AV junction, and ventricle.

EP studies can be used in patients with WPW syndrome to determine (1) the mechanism of the clinical arrhythmia, (2) EP properties (eg, conduction capability, refractory periods) of the accessory pathway and the normal conduction system, (3) the number and location of accessory pathways (which is necessary for catheter ablation), and (4) the response to pharmacological or ablation therapy.

Indications for EP studies in patients with WPW syndrome according to the American College of Cardiology/American Heart Association guidelines

Class I indications include (1) patients being evaluated for catheter ablation or surgical ablation of an accessory pathway, (2) patients with ventricular preexcitation who have survived cardiac arrest or who have unexplained syncope, and (3) symptomatic patients in whom determination of the mechanism of arrhythmia or knowledge of the EP properties of the accessory pathway and normal conduction system would help in determining appropriate therapy.

Class II indications include (1) asymptomatic patients with a family history of sudden cardiac death or with ventricular preexcitation but no spontaneous arrhythmia who engage in high-risk occupations or activities and in whom knowledge of the EP properties of the accessory pathway or inducible tachycardia may help determine recommendations for further activities or therapy and (2) patients with ventricular preexcitation who are undergoing cardiac surgery for other reasons.

Class III indications include asymptomatic patients with ventricular preexcitation, except those in class II.

EP features of preexcitation

If a Kent bundle (AV)–type accessory bypass tract conducts in an antegrade fashion, 2 parallel paths can potentially carry the impulse. The first is the natural one, which comes with inherent physiological delay over the AV node. The second is the bypass tract (Kent bundle), which allows the impulse to pass directly without delay from the atrium to the ventricle.

This dual-path mechanism produces a unique QRS complex that is a form of fusion beat due to depolarization of the ventricle from these 2 pathways.The delta wave results from ventricular activation by the impulse traveling over the accessory pathway.

The extent of contribution to ventricular depolarization by the wavefront over each route varies, as follows:

If delay in AV nodal conduction occurs from either rapid atrial pacing or a premature atrial complex, a greater proportion of the ventricle activates via the bypass tract and the QRS becomes more abnormal in shape.

On the other hand, if the bypass tract is far from the sinus node (as in the presence of a left lateral pathway) or if AV nodal conduction is rapid, a larger proportion of the ventricle activates

via the normal pathway.The normal fusion beat during sinus rhythm has a short or negative HV interval. This occurs because the His bundle activation begins later than the ventricular activation from the bypassing impulse, while the impulse traveling over the AV node just reaches the His bundle. Pacing the atrium rapidly at premature intervals accentuates the abnormal ventricular depolarization and further shortens the HV interval.

Recognition and localization of accessory pathways using EP studies

When retrograde atrial activation during tachycardia occurs over an accessory pathway that connects the left atrium to the left ventricle, the earliest retrograde activity is recorded from a left atrial electrode (usually positioned in the coronary sinus). This is a left lateral pathway.

When retrograde atrial activation during tachycardia occurs over an accessory pathway that connects the right ventricle to the right atrium, the earliest retrograde atrial activity is generally recorded from a lateral right atrial electrode. This is a right ventricular free wall pathway.

Participation of a septal accessory pathway creates earliest retrograde atrial activation in the low-right atrium situated near the septum, anterior or posterior, depending on the insertion site.

Mapping techniques with intravenous catheter electrodes placed at the time of surgery may help provide accurate assessments of the position of the accessory pathway. Recording electrical activity directly from the accessory pathway obviously provides the most precise localization.

Retrograde atrial activation over the accessory pathway on EP studies

This can be confirmed by inducing premature ventricular complexes during tachycardia to determine whether retrograde atrial excitation can occur from the ventricle at a time when the His bundle is refractory.

Because VA conduction cannot occur over the normal conduction system because the His bundle is refractory, an accessory pathway must be present for the atria to become excited and most likely is participating in the tachycardia circuit.

The following parameters may be helpful:

Patients with a reciprocating tachycardia due to an accessory AV bypass tract almost always have a VA interval of greater than 70 milliseconds measured from the onset of ventricular activation to the onset of atrial activity recorded on an esophageal lead or greater than 95 milliseconds when measured to the high-right atrium.

In contrast, in most patients with AVNRT, the interval from the onset of ventricular activity to the earliest onset of atrial activity is characteristically shorter than 70 milliseconds.

Intraoperative (multiarray) epicardial mapping and endocardial catheter mapping using EP studies

Mapping of the pathways and sites of origin for both ventricular and supraventricular tachyarrhythmias has led to tremendous improvements in surgical outcomes, which has given way to catheter techniques for ablation procedures.

Multiple electrode arrays allow simultaneous recordings from several intracardiac sites during the same cardiac cycle, generating maps of wave activation. This technology allows the clinical electrophysiologist and surgeon to identify target areas for surgical ablation.

Although quite successful in prior years, intraoperative mapping for WPW syndrome has now been replaced by catheter mapping during EP studies and ablation procedures.

Histologic Findings: Histologic findings of accessory bypass pathways have been described with careful dissection of the AV space.

TREATMENT

Medical Care: Treatment of arrhythmia is directed at the underlying cause and the triggers that perpetuate the arrhythmia. The underlying cause includes primary arrhythmias due to an EP abnormality resulting from definable structural heart disease and occurring independently of hemodynamic or metabolic disturbance. Such arrhythmias include coronary heart disease, ischemia, cardiomyopathy, pericarditis, and WPW syndrome. The triggers that perpetuate the arrhythmia include secondary arrhythmias, such as electrolyte imbalance, metabolic defects, and hemodynamic and hypoxemic abnormalities.

Appropriate treatment of WPW syndrome is based on its likely prognosis. Patients with only ECG evidence of preexcitation, without documented episodes of tachyarrhythmias, generally do not require either aggressive workup through EP studies or treatment with antiarrhythmic agents.

The 3 main treatment modalities for WPW syndrome are drug therapy, electrical (ie, RF) ablation, and surgical ablation. Ablation is the first-line treatment for symptomatic WPW syndrome. It has replaced surgical treatment and most drug treatment. However, drug therapy can be useful in some instances, such as in patients who refuse ablation or in patients in whom ablation fails in one or two attempts. For patients treated longitudinally with pharmacotherapy, consideration should be given to a membrane-active antiarrhythmic drug (class IC or III) with an AV nodal blocker, rather than just an AV nodal blocker, because of the potential for extremely rapid rates during preexcited atrial fibrillation or flutter

Drug therapy (potential antiarrhythmic mechanisms): Antiarrhythmic drugs act on the AV node (ie, AV node blocking agents), myocardial tissue, and/or the accessory pathways. They work by increasing the refractory period or by prolonging the conduction time to prevent perpetuation of an AV reciprocating tachycardia. They may also act to reduce the ventricular response to atrial flutter or atrial fibrillation.

AV node blocking drugs

Adenosine, verapamil, metoprolol, and digitalis all prolong conduction time and refractoriness in the AV node.

Verapamil and metoprolol do not affect conduction in the bypass tract.

Digitalis exhibits variable effects and may even shorten the refractory period.

None of these drugs should be given in an acute phase to a patient with ventricular preexcitation who has atrial fibrillation.

Digoxin is contraindicated in patients with WPW syndrome, although it may play some role in children only. Most deaths from WPW syndrome have been associated with digoxin use.

Propranolol is almost never administered. Metoprolol or atenolol can be useful in some patients.

Agents affecting the accessory pathways

Class IA drugs (eg, procainamide) and class IC drugs (eg, flecainide, propafenone) block conduction in the accessory pathway.

Amiodarone and sotalol influence both the AV node and the bypass tract. They work in similar fashion but affect only the bypass tract.

Class IA and IC drugs that prolong the refractory period in the bypass tract are indicated if drug therapy becomes necessary.

Class IC and IA drugs are best used in conjunction with an AV node blocker, such as metoprolol or verapamil.

Procainamide and quinidine are relics of the past for long-term treatment.

Caution when treating WPW syndrome tachycardia

Digitalis shortens refractoriness in the myocardium and in the bypass tract. Thus, it may accelerate the ventricular response in the setting of atrial fibrillation in a patient with WPW syndrome. Adenosine should not be used in this setting.

Digitalis should not be used in such patients, except perhaps in pediatric or elderly patients. Instead, medicines that prolong the refractory period in the accessory pathway (eg, class IA and IC agents) should be used.

Intravenous verapamil can likewise speed up the ventricular response in patients with WPW syndrome who have atrial fibrillation. This does not appear to happen with oral verapamil. Verapamil is not recommended as a sole agent in patients with WPW syndrome.

Termination of an acute episode

Narrow-complex AV reentrant tachycardia

Such tachycardias manifest with normal QRS complexes, a ventricular rate of more than 200 beats per minute, regular R-R intervals, and a retrograde P wave well beyond the end of QRS.

They should be treated in the same way as AVNRT, by blocking AV node conduction with (1) vagal maneuvers (eg, Valsalva maneuver, carotid sinus massage, splashing cold water or ice water on the face), (2) intravenous adenosine, or (3) intravenous verapamil or diltiazem (ie, if recurrent SVT is present, if adenosine is ineffective, or if the patient is taking theophylline).

Note that atrial fibrillation can occur after drug administration, particularly adenosine, with a rapid ventricular response. An external cardioverter-defibrillator should be immediately available in case it is necessary.

Atrial flutter/fibrillation or wide-complex tachycardia

Atrial flutter/fibrillation can be recognized by the presence of abnormal QRS complexes and irregular R-R intervals. In this setting, drugs that prolong the refractory period of the bypass tract should be used, especially those that also block the AV node (by prolonging refractoriness). Examples of such drugs include procainamide (class IA agent) and propranolol (class II beta-blocker).

If wide-complex tachycardia is present and the diagnosis of ventricular tachycardia cannot be excluded, the drugs of choice are intravenous procainamide or amiodarone (in lieu of cardioversion if the patient is stable hemodynamically). Ibutilide may also be useful in this setting, although data are lacking.

Importantly, avoid lidocaine in this setting. It does not prolong refractoriness in the accessory pathway. Lidocaine may increase the ventricular response if atrial fibrillation is present.

Hemodynamically unstable tachycardia and electrical cardioversion

In patients with a very fast ventricular rate, hemodynamic instability (eg, hypotension, mental status change) may ensue.

The initial treatment of choice in such patients is direct-current synchronized electrical cardioversion.

Electrical cardioversion appears to terminate most effectively the tachycardias due to reentry, such as AVNRT and reciprocating tachycardias associated with WPW syndrome.

The electrical shock depolarizes all excitable myocardium, lengthens refractoriness, interrupts reentrant circuits, discharges foci, and establishes electrical homogeneity that terminates reentry.

Because myocardial damage may occur with increases in applied energy, the minimum effective energy should be used and the energy should be titrated. An energy of at least 100 joules (monophasic or lower biphasic) successfully terminates most SVTs and should be tried initially. If that fails, a second shock with higher energy can be delivered.

Cardioversion can have several adverse effects. It may induce arrhythmias because of inadequate synchronization, with the shock occurring during the ST segment or T wave. Rarely, even a properly synchronized shock can produce ventricular fibrillation. Postcardioversion arrhythmias are generally transient and do not require treatment. Embolic episodes may occur in 1-3% of the patients converted from atrial fibrillation to sinus rhythm if the episodes are longer than 48 hours.

Long-term maintenance treatment

Response to long-term antiarrhythmic therapy for the prevention of further episodes of tachycardia in patients with WPW syndrome remains quite variable and unpredictable. Some drugs may paradoxically make the reciprocating tachycardia more frequent. Dual-drug therapy has been used, eg, procainamide and verapamil (class IA and IV), or quinidine and propranolol (class IA and II). Good reasons exist to avoid quinidine and procainamide; newer drugs that are safer and better are available. Class IC drugs (eg, amiodarone, sotalol) are good choices, but class IC drugs should not be given if the patient has structural heart disease. Class IC drugs are typically used with an AV nodal blocking agent.

The best plan is to not use drugs at all; instead, refer all patients who have symptomatic WPW syndrome for ablation because this cures the tachycardia and eliminates the potential dangerous effects of drugs.

Patients who have accessory pathways with short refractory periods are poor candidates for medical therapy and are best treated with ablation.

Surgical Care: Ablative procedures are the therapy of choice. Electrode catheters can be advanced intravenously to locate and ablate the accessory tract by delivering electrical or RF energy. Cryothermy, lasers, direct current, and microwave energy sources have also been used in the past, but RF catheter ablation has replaced these modalities because it is much more efficacious, safe, and cost-effective.

RF ablation is currently the treatment of choice for most adults and many children with symptomatic WPW syndrome (ie, those who have AV reentrant tachycardia or atrial flutter/fibrillation with conduction of the accessory pathway). Success rates for catheter ablation exceed 90%.

Localization of the bypass tract(s)

First, perform an EP study to (1) determine that the bypass tract is part of the tachycardia reentrant circuit, and (2) locate the optimal site for ablation. Pathways can be located in the left or right free wall or septum of the heart. Multiple pathways may be present in approximately 5% of patients.

Pathways at all the sites in the heart and in persons of all age groups can be ablated successfully. The RF ablation creates conduction block that can be seen on intracardiac electrogram findings (ie, during the EP study) between the atrial activation and the bypass tract potential.

Identification of the ablation site during EP studies

During the EP studies, direct recordings of the accessory pathway indicate the optimal site for ablation.

The ventricular insertion site is indicated by the earliest onset of the ventricular electrogram in relation to the delta wave.

The atrial insertion site is indicated by the region of the shortest VA interval during orthodromic tachycardia (ie, AV reentrant tachycardia) or ventricular pacing.

Successful ablation sites show stable fluoroscopic and electrical features. During orthodromic AV reentrant tachycardia, the time between the ventricular and atrial potentials is short and a pathway potential may be observed.

Generally, a thermistor-tipped catheter is used, which shows a stable rise in catheter tip temperature, suggesting catheter stability and optimal catheter-tissue contact. The tip temperature generally rises above 50°C.

Indications for RF ablation

Patients with symptomatic AV reentrant tachycardia should receive RF ablation.

Atrial fibrillation or other atrial tachyarrhythmias that have rapid ventricular response via a bypass tract is an indication for RF ablation procedures.

Patients with AV reentrant tachycardia or atrial fibrillation with rapid ventricular rates found incidentally during EP studies for unrelated arrhythmia should undergo RF ablation.

Asymptomatic patients with ventricular preexcitation whose livelihood, profession, insurability, or mental well-being may be influenced by unpredictable tachyarrhythmias or in whom such tachyarrhythmias would endanger the public safety should have an RF ablation procedure.

Patients with atrial fibrillation and a controlled ventricular response via the bypass tract are candidates for RF ablation.

Patients with a family history of sudden cardiac death should undergo RF ablation.

Effectiveness of RF ablation: A survey by the North American Society for Pacing and Electrophysiology (NASPE) indicates that ablation is successful. Results are as follows:

For left free wall accessory pathways, 2312 of 2527 patients (91%) were cured.

For septal accessory pathways, 1115 of 1279 patients (87%) were cured.

For right free wall accessory pathways, 585 of 715 patients (82%) were cured.

Complications of RF ablation

In the United States, complications have been reported in 94 of 4521 patients (2.1%). Of the 4521 patients, 13 died (0.2%).

In Europe, the complication rate is reported to be 4.4%. Of 2222 patients, 3 died.

Surgical ablation

Surgical open heart procedures were more common before RF ablation was developed.

Now, RF catheter ablation has virtually eliminated surgical open heart treatments in the vast majority of patients, with the following exceptions:

Patients in whom RF catheter ablation (with repeated attempts) fails

Patients undergoing concomitant cardiac surgery (possible exception)

Patients with other tachycardias with multiple foci who require surgical intervention (very rare)

Consultations: Specific subspecialty consultations are often needed. These may include any of the following:

Cardiovascular specialist

Electrophysiologist

Pediatric cardiovascular specialist

Diet:

The majority of patients presenting with WPW syndrome are not elderly.

Patients presenting with structural heart disease, cardiomyopathy, or heart failure may require a low-salt, low-cholesterol diet.

Activity: Generally, no activity restrictions are recommended in patients with ECG findings of preexcitation but without tachycardias. They should be restricted from high-risk professions (eg, airline pilot) and may be restricted from competitive sports.

Patients presenting with tachycardias and accessory pathways should avoid participating in competitive sports because catecholamines can decrease the refractoriness of the bypass tract and facilitate tachyarrhythmias.

Patients with hypertrophic cardiomyopathy or the Ebstein anomaly should also abstain from competitive sports.

Once a curative procedure (eg, RF ablation of the accessory pathway) has been successfully performed, most patients can return to competitive sports several months later.

MEDICATION

The goals of pharmacotherapy are to reduce morbidity and to prevent complications.

Drug Category: Antiarrhythmic agents -- Prolong refractory period of the conduction tissue, the accessory pathway, or both.

Drug Name

Drug Name	Adenosine (Adenocard) -- Blocks conduction time in the AV node. Can interrupt AVRT by blocking conduction in the AV node to restore normal sinus rhythm in PSVT, including PSVT associated with WPW syndrome. Should not be given to patients with preexcitation.
Adult Dose	6 mg rapid IV bolus over 1-2 s initially; if no response within 1-2 min, give 12 mg rapid IV bolus; repeat 12-mg dose second time prn; not to exceed doses >12 mg
Pediatric Dose	0.1 mg/kg IV; repeat at 0.2 mg/kg if first dose not effective; not to exceed 12 mg Alternatively, 0.05 mg/kg IV; if not effective within 2 min, increase dose by 0.05-mg/kg increments q2min; not to exceed 0.25 mg/kg

Contraindications	Documented hypersensitivity; second- or third-degree AV block or sick sinus syndrome (except in patients with functioning artificial pacemaker); atrial flutter; atrial fibrillation; ventricular tachycardia
Interactions	Coadministration with carbamazepine may produce higher degrees of heart block; dipyridamole may potentiate effects; methylxanthines may antagonize effects; do not administer to patients with a heart transplant
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Adenosine-induced bronchoconstriction possible in patients with asthma May cause prolonged asystole in patients with a heart transplant; may provoke atrial fibrillation
Drug Name	Propranolol (Inderal) -- Class II antiarrhythmic nonselective beta-adrenergic receptor blocker with membrane-stabilizing activity that decreases automaticity of contractions.
Adult Dose	1-3 mg IV under careful monitoring; not to exceed 1 mg/min to avoid lowering blood pressure and causing cardiac standstill; allow time for drug to reach site of action
	(particularly if slow circulation); administer second dose after 2 min prn thereafter, not to be administered sooner than 4 h after initial dose; do not continue doses after desired alteration in rate or rhythm achieved; switch to PO as soon as clinically indicated; 10-30 mg tid/qid (usual)
Pediatric Dose	2-4 mg/kg/d PO divided bid (ie, 1-2 mg/kg bid); IV use not recommended; however, for arrhythmias, dose of 0.01-0.1 mg/kg by slow push has been recommended; not to exceed 1 mg/dose; change to PO as soon as clinically indicated
Contraindications	Documented hypersensitivity; uncompensated CHF; bradycardia; cardiogenic shock; AV conduction abnormalities
Interactions	Coadministration with aluminum salts, barbiturates, NSAIDs, penicillins, calcium salts, cholestyramine, and rifampin may decrease effects; calcium channel blockers, cimetidine, loop diuretics, and MAOIs may increase toxicity; toxicity of hydralazine, haloperidol, benzodiazepines, and phenothiazines may increase
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Beta-adrenergic blockade may decrease signs of acute hypoglycemia and hyperthyroidism; abrupt withdrawal may exacerbate symptoms of hyperthyroidism, including thyroid storm; withdraw drug slowly and monitor closely
Drug Name	Verapamil (Verelan, Calan) -- By interrupting reentry at AV node, can restore normal sinus rhythm in patients with PSVT
Adult Dose	80-160 mg PO tid; alternatively, 5-10 mg IV followed by second dose 15-30 min later if patient does not respond satisfactorily to initial dose; extended-release dosage form may be given qd
Pediatric Dose	Not established
Contraindications	Documented hypersensitivity; severe CHF; sick sinus syndrome or second- or third-degree AV block; hypotension (<90 mm Hg systolic)
Interactions	May increase carbamazepine, digoxin, and cyclosporine levels; coadministration with amiodarone can cause bradycardia and a decrease in cardiac output; when administered concurrently with beta-blockers, may increase cardiac depression; cimetidine may increase levels; may increase theophylline levels
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Hepatocellular injury may occur; transient elevations of transaminases with and without concomitant elevations in alkaline phosphatase and bilirubin have occurred (elevations have been transient and may disappear with continued treatment); monitor liver function periodically

Drug Name	Digoxin (Lanoxin) -- Has direct inotropic effects in addition to indirect effects on the cardiovascular system. However, may shorten refractory period. Most deaths in WPW have been associated with digoxin use.
Adult Dose	NOT RECOMMENDED; has been associated with ventricular fibrillation
Pediatric Dose	5-10 years: 20-35 mcg/kg PO >10 years: 10-15 mcg/kg PO Maintenance dose: 25-35% of PO loading dose
Contraindications	Documented hypersensitivity; ADULT PATIENTS; beriberi heart disease, idiopathic hypertrophic subaortic stenosis, constrictive pericarditis, and carotid sinus syndrome
Interactions	IV calcium may produce arrhythmias in digitalized patients; medications that may increase levels include alprazolam, benzodiazepines, bepridil, captopril, cyclosporine, propafenone, propantheline, quinidine, diltiazem, aminoglycosides, oral amiodarone, anticholinergics, diphenoxylate, erythromycin, felodipine, flecainide, hydroxychloroquine, itraconazole, nifedipine, omeprazole, quinine, ibuprofen, indomethacin, esmolol, tetracycline, tolbutamide, and verapamil Medications that may decrease serum levels include aminoglutethimide, antihistamines, cholestyramine, neomycin, penicillamine, aminoglycosides, oral colestipol, hydantoins, hypoglycemic agents, antineoplastic treatment combinations (including carmustine, bleomycin, methotrexate, cytarabine, doxorubicin, cyclophosphamide, vincristine, procarbazine), aluminum or magnesium antacids, rifampin, sucralfate, sulfasalazine, barbiturates, kaolin/pectin, and aminosalicylic acid
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Hypokalemia may reduce positive inotropic effect of digitalis; hypercalcemia predisposes patient to digitalis toxicity, and hypocalcemia can make digoxin ineffective until serum calcium levels are normal; magnesium replacement therapy must be instituted in patients with hypomagnesemia to prevent digitalis toxicity; patients diagnosed with incomplete AV block may progress to complete block when treated with digoxin; exercise caution in hypothyroidism, hypoxia, and acute myocarditis; adjust dose in renal impairment; highly toxic (overdoses can be fatal)
Drug Name	Procainamide (Procanbid, Pronestyl) -- Class IA antiarrhythmic. Increases refractory period of atria, ventricles, and accessory pathway. Excellent in preexcited atrial fibrillation or flutter.
Adult Dose	30 mg/min IV continuous infusion until arrhythmia suppressed, patient becomes hypotensive, QRS widens 50% above baseline, or maximum dose of 17 mg/kg administered; once arrhythmia suppressed, may infuse at continuous rate of 1-4 mg/min
Pediatric Dose	Not established; suggested as follows: 15-50 mg/kg/d PO divided q3-6h; not to exceed 4 g/d 20-30 mg/kg/d IM divided q4-6h; not to exceed 4 g/d 3-6 mg/kg/dose IV infused over 5 min Maintenance dose: 20-80 mcg/kg/min administered as continuous infusion; not to exceed 100 mg/dose or 2 g/d
Contraindications	Documented hypersensitivity; torsade de pointes; systemic lupus erythematosus
Interactions	Can expect increased levels of procainamide metabolite NAPA in patients taking cimetidine, ranitidine, beta-blockers, amiodarone, trimethoprim, and quinidine; may increase effect of skeletal muscle relaxants, quinidine and lidocaine, and neuromuscular blockers; ofloxacin inhibits tubular secretion of procainamide and may increase bioavailability; when taken concurrently with sparfloxacin, may increase risk of cardiotoxicity

Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Monitor for hypotension; plasma concentrations and active metabolite (NAPA) may increase in renal failure; high or toxic concentrations may induce AV block or abnormal automaticity; toxicity may outweigh benefit long term; do not use as a first-line drug for WPW syndrome
Drug Name	Quinidine (Quinaglute, Quinidex, Cardioquin) -- Maintains normal heart rhythm and converts atrial fibrillation or flutter. Not recommended as first-line drug for WPW syndrome.
Adult Dose	200 mg PO q2-3h for 5-8 doses with subsequent daily increases until sinus rhythm restored or adverse effects occur; not to exceed 3-4 g/d
Pediatric Dose	30 mg/kg/d PO in 5 divided doses
Contraindications	Documented hypersensitivity; complete AV block or intraventricular conduction defects; presently taking ritonavir or sparfloxacin
Interactions	Phenytoin, rifampin, and phenobarbital may decrease concentrations; toxicity increased when taken with ritonavir, sparfloxacin, beta-blockers, amiodarone, verapamil, cimetidine, alkalinizing agents, or nondepolarizing or depolarizing muscle relaxants; may enhance effect of anticoagulants
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Caution in G-6-PD deficiency and those with tendency to develop granulocytopenia; avoid use in myocardial depression, hepatic or renal insufficiency, and myasthenia gravis
Drug Name	Amiodarone (Cordarone, Pacerone) -- May inhibit AV conduction and sinus node function. Prolongs action potential and refractory period in myocardium and inhibits adrenergic stimulation.
Adult Dose	Loading dose: 800-1600 mg/d PO in 1-2 doses for 1-3 wk; decrease to 600-800 mg/d in 1-2 doses for 1 mo Maintenance dose: 400 mg/d PO; alternatively, 150 mg (10 mL) IV over first 10 min, followed by 360 mg (200 mL) over next 6 h, then 540 mg over next 18 h
Pediatric Dose	10-15 mg/kg/d or 600-800 mg/1.73 m2/d PO for 4-14 d or until adequate control of arrhythmia attained
Contraindications	Documented hypersensitivity; complete AV block; intraventricular conduction defects; taking ritonavir or sparfloxacin
Interactions	Increases effect and blood levels of theophylline, quinidine, procainamide, phenytoin, methotrexate, flecainide, digoxin, cyclosporine, beta-blockers, and anticoagulants; cardiotoxicity increased by ritonavir, sparfloxacin, and disopyramide; coadministration with calcium channel blockers may cause additive effect and further decrease myocardial contractility; cimetidine may increase level
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Caution in thyroid or liver disease
Drug Name	Sotalol (Betapace) -- Class III antiarrhythmic agent that blocks potassium channels, prolongs action potential duration, and lengthens QT interval. Noncardiac selective beta-adrenergic blocker.
Adult Dose	80 mg PO bid; increase dose gradually q2-3d to 240-320 mg/d
Pediatric Dose	Not established

Contraindications	Documented hypersensitivity; long QT, history of torsades de pointes
Interactions	Aluminum salts, barbiturates, NSAIDs, penicillins, calcium salts, cholestyramine, and rifampin may decrease bioavailability and plasma levels, possibly resulting in decreased pharmacologic effect; cardiotoxicity may increase when administered concurrently with sparfloxacin, calcium channel blockers, quinidine, flecainide, and contraceptives; toxicity increases when administered concurrently with digoxin, flecainide, acetaminophen, clonidine, epinephrine, nifedipine, prazosin, haloperidol, phenothiazines, and catecholamine-depleting agents
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Beta-adrenergic blockade may decrease signs and symptoms of acute hypoglycemia and clinical signs of hyperthyroidism; abrupt withdrawal may exacerbate symptoms of hyperthyroidism, including thyroid storm; withdraw drug slowly and monitor patient closely; caution in hypokalemia, peripheral vascular disease, hypomagnesemia, and CHF
Drug Name	Diltiazem (Cardizem, Dilacor, Tiamate, Tiazac) -- Slows AV nodal conduction.
Adult Dose	IR: 30-90 mg PO q8h SR: 120-300 mg PO qd IV: 10-20 mg bolus over 10-20 min, followed by continuous infusion at 10-15 mg/h
Pediatric Dose	Not established
Contraindications	Documented hypersensitivity; severe CHF; sick sinus syndrome; second- or third-degree AV block; hypotension (<90 mm Hg systolic)
Interactions	May increase carbamazepine, digoxin, cyclosporine, and theophylline levels; when administered with amiodarone, may cause bradycardia and decrease in cardiac output; when given with beta-blockers, may increase cardiac depression; cimetidine may increase levels
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Caution in impaired renal or hepatic function; may increase LFT levels, and hepatic injury may occur
Drug Name	Ibutilide (Corvert) -- Class III antiarrhythmic agent that may work by increasing action potential duration, thereby changing atrial cycle length variability. Mean time to conversion is 30 min. Two thirds of patients who converted were in sinus rhythm at 24 h. Ventricular arrhythmias occurred in 9.6% of patients and were mostly PVCs. The incidence of torsades de pointes was <2%.
Adult Dose	<60 kg: 0.01 mg/kg IV over 10 min >60 kg: 1 mg IV over 10 min
Pediatric Dose	Not established
Contraindications	Documented hypersensitivity
Interactions	Increases toxicity of quinidine and procainamide; concurrent administration with TCAs and phenothiazines may prolong QT interval; toxicity of digoxin increases when administered concurrently
Pregnancy	C - Safety for use during pregnancy has not been established
Precautions	Caution in renal or hepatic impairment
Drug Name	Dofetilide (Tykosin) -- Increases monophasic action potential duration, primarily due to delayed repolarization. Terminates induced reentrant tachyarrhythmias (eg, atrial fibrillation/flutter, ventricular tachycardia) and prevents their reinduction. No data in WPW syndrome.
Adult Dose	125-500 mcg IV bid; must be started in an inpatient monitored setting
Pediatric Dose	Not established

Contraindications	Documented hypersensitivity; concomitant use of verapamil or the cation transport system inhibitor cimetidine, trimethoprim (alone or in combination with sulfamethoxazole), or ketoconazole; congenital or acquired long QT syndromes; severe renal impairment (CrCl <20 mL/min); prochlorperazine and megestrol coadministration; a baseline QT interval or QTc >440 ms (500 ms in patients with ventricular conduction abnormalities)
Interactions	Verapamil, TMP/SMZ, ketoconazole, potassium-depleting diuretics, digoxin, cimetidine, phenothiazines, triamterene, metformin, prochlorperazine, amiloride, megestrol, and antiarrhythmic agents may increase toxicity
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Maintain potassium levels within reference range prior to and during administration; to minimize risk of induced arrhythmia, calculations of CrCl and continuous ECG monitoring required; cardiac resuscitation equipment and personnel must be present
Drug Name	Flecainide (Tambocor) -- Blocks sodium channels, producing dose-related decrease in intracardiac conduction in all parts of heart. Increases electrical stimulation of threshold of ventricle, His-Purkinje system. Shortens phase 2 and phase 3 repolarization, resulting in decreased action potential duration and ERP. Indicated for the treatment of paroxysmal atrial fibrillation/flutter associated with disabling symptoms and PSVT, including AVNRT, AV reentrant tachycardia, and other SVTs of unspecified mechanism associated with disabling symptoms in patients without structural heart disease. Also indicated for prevention of documented life-threatening ventricular arrhythmias, such as sustained ventricular tachycardia. Not recommended in less severe ventricular arrhythmias, even if patients are symptomatic.
Adult Dose	100 mg PO bid q12h; increase q4d but not to exceed 400 mg/d
Pediatric Dose	3-6 mg/kg/d or 100-150 mg/m2/d divided tid to 11 mg/kg/d or 200 mg/m2/d
Contraindications	Documented hypersensitivity; preexisting second- or third-degree AV block, RBBB associated with left hemiblock (bifascicular block) or trifascicular block, unless a pacemaker is present to sustain cardiac rhythm if complete heart block occurs; concurrent use of ritonavir or amprenavir; recent MI
Interactions	May increase toxicity of digoxin; beta-adrenergic blockers, verapamil, and disopyramide may have additive inotropic effects when administered with flecainide; CYP4502D6 inhibitors (eg, ritonavir, cimetidine, amiodarone) may increase serum levels and cardiotoxicity
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Caution in preexisting sinus node dysfunction, history of CHF, sick sinus syndrome, post MI, or myocardial dysfunction; reserve use for life-threatening arrhythmias only because deaths have been associated with proarrhythmic effects of class IC antiarrhythmics; typically used in conjunction with an AV nodal blocking agent; adjust dose in renal or hepatic impairment
Drug Name	Propafenone (Rythmol) -- Shortens upstroke velocity (phase 0) of monophasic action potential. Reduces fast inward current carried by sodium ions in Purkinje fibers and, to a lesser extent, myocardial fibers. May increase diastolic excitability threshold and prolong ERP. Reduces spontaneous automaticity and depresses triggered activity. Indicated for treatment of documented life-threatening ventricular arrhythmias, such as sustained ventricular tachycardia. Appears to be effective in the treatment of SVTs, including atrial fibrillation and flutter. Not recommended in patients with less severe ventricular arrhythmias, even if patients are symptomatic.
Adult Dose	150 mg PO q8h; increase at 3- to 4-d intervals, not to exceed 300 mg q8h

Pediatric Dose	Not established
Contraindications	Documented hypersensitivity, second- or third-degree AV block, RBBB associated with left hemiblock (bifascicular block) or trifascicular block; concurrent use of ritonavir or amprenavir
Interactions	Rifampin may decrease plasma levels; quinidine may increase pharmacologic effects; may increase plasma levels of beta-blockers, cyclosporine, warfarin, and digoxin; CYP4502D6 inhibitors (eg, ritonavir, cimetidine, amiodarone) may increase serum levels and cardiotoxicity
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Caution in preexisting sinus node dysfunction, history of CHF, sick sinus syndrome, post MI, or myocardial dysfunction; reserve use for life-threatening arrhythmias only because deaths have been associated with proarrhythmic effects of class IC antiarrhythmics; adjust dose in renal or hepatic impairment; typically used in conjunction with an AV nodal blocking agent

FOLLOW-UP

Further Inpatient Care:

Patients with WPW syndrome who are admitted to the hospital after initiation of medical treatment in the emergency department may require further evaluation and management as follows:

Continuous telemetry monitoring to look for resurgence of tachyarrhythmia and the degree of control of the ventricular rate in those with atrial fibrillation

Initiation, dose adjustment, and maintenance of long-term antiarrhythmic drugs for preventing recurrences (However, patients generally undergo ablation.)

Laboratory evaluation and correction of electrolyte and metabolic abnormalities that may have acted as triggers

Evaluation for associated underlying structural cardiac defects, such as Ebstein anomaly and hypertrophic cardiomyopathy, is as follows:

EP evaluation of patients who meet the indications, including the following:

To diagnose and locate accessory pathways and the reentrant pathways or sites of origin of SVTs

To define appropriate therapy

To test the results of therapy

To enable electrocardiographically guided therapy such as RF ablation

RF ablation for patients who are candidates for such therapy, including the following:

Patients with symptomatic tachycardia who cannot tolerate drug therapy or whose conditions are resistant to such therapy

Patients who have atrial fibrillation with a rapid ventricular response via a bypass tract who cannot tolerate drug therapy or whose conditions are resistant to such therapy

Patients who have AV reentrant tachycardia or atrial fibrillation with rapid ventricular rates found during EP studies

Asymptomatic patients whose profession, insurability, mental well-being, or responsibility to public safety may be affected by unpredictable occurrence of tachyarrhythmias

Patients with a family history of sudden cardiac death

Monitor drug use.

Carefully monitor for proarrhythmias, especially when procainamide, quinidine, amiodarone, or sotalol are initiated. A few days of inpatient telemetry monitoring, including determination of QT interval lengthening on ECG readings, is required for these agents. An increase in the QT interval of 25% or greater should be avoided.

Surgical ablation is recommended in certain patients, including the following:

Patients in whom RF catheter ablation fails

Patients who will be undergoing concomitant cardiac surgery

Patients with atrial tachycardias who have multiple foci (sometimes)

Further Outpatient Care:

Patients need to continue antiarrhythmic therapy as prescribed. If symptoms related to tachyarrhythmias recur, patients should inform the physician.

Arrange follow-up visits to assess for the recurrence of arrhythmia, the effectiveness of antiarrhythmic therapy, and adverse effects of medications.

Follow-up ECG or Holter monitoring may be needed to assess for changes in QT duration and the recurrence of arrhythmias or proarrhythmias.

Patients who take amiodarone require careful periodic monitoring for adverse effects and organ toxicity, including thyroid function tests, ophthalmic examination, pulmonary function tests, and hepatic function tests.

Patients who undergo EP studies, RF ablation, or surgical ablation may require monitoring of wound care following hospital discharge. Further follow-up care to assess for the recurrence of arrhythmia is also needed.

Patients with underlying structural heart disease, such as the Ebstein anomaly, may require follow-up care by a specialist pediatric cardiologist.

If a patient with WPW syndrome dies suddenly, siblings and first-degree relatives should be screened for preexcitation.

Unless curative ablation has been performed, patients should refrain from participating in competitive sports.

Routine EP studies are not recommended following RF ablation solely to ensure that the ablation was curative, unless patients become symptomatic.

Asymptomatic patients with only the ECG findings of preexcitation should be seen at frequent intervals and should not undergo any aggressive EP evaluation or pharmacologic or ablative therapy unless they become symptomatic or their profession, insurability, mental well-being, or the safety of the public may be affected by unpredictable occurrence of tachyarrhythmias.

In/Out Patient Meds:

Adenosine

Digoxin (not recommended for WPW syndrome in adults)

Propranolol

Verapamil

Quinidine

Procainamide

Amiodarone

Sotalol

Transfer:

Certain patients with WPW syndrome must be transferred to a tertiary facility for comprehensive evaluation and management by a cardiac electrophysiologist, which may include EP studies or ablative therapy. Such patients include those presenting with any of the following:

Sudden death

Syncope

Significant symptomatic tachyarrhythmias

Uncertain diagnosis in those with wide-complex tachycardia

Associated structural heart disease, eg, Ebstein anomaly, cardiomyopathy, mitral valve prolapse.

WPW syndrome who have a family history of sudden death

Asymptomatic but with WPW syndrome who are in professions in which spontaneous occurrence of tachyarrhythmia may jeopardize public safety, cause much mental anguish, or influence insurability

Atrial fibrillation or flutter

Deterrence/Prevention:

WPW syndrome is largely congenital or hereditary. No particular method exists to eliminate the possibility of developing accessory pathways. In the future, genetic recognition and counseling may become a useful tool.

Fortunately, the majority of patients with ECG findings of preexcitation do not develop tachyarrhythmias.

Patients who present with tachyarrhythmic symptoms require drug therapy to prevent further episodes. Such long-term therapy may include the use of amiodarone, sotalol, quinidine and propranolol, and verapamil and diltiazem on a regular basis.

If the procedure is successful, patients who have undergone ablative treatment are usually cured of the disease and are not at risk for further tachyarrhythmias.

Complications:

Tachyarrhythmia

Palpitations

Dizziness or syncope

Sudden cardiac death

Complications of drug therapy (eg, proarrhythmia, organ toxicity)

Complications associated with invasive procedures and surgery

Recurrence

Prognosis:

Patients with only preexcitation on their ECG findings who are asymptomatic generally have a very good prognosis. Most of these patients do not develop symptoms in their lifetime.

Patients with a family history of sudden cardiac death or significant

symptoms of tachyarrhythmias or cardiac arrest have worse prognoses. However, once definitive therapy is performed, including curative ablation, the prognosis is once again excellent.

Asymptomatic patients should not be evaluated by EP testing unless they are in a high-risk profession. Risk stratification is not generally needed for asymptomatic patients.

Patient Education:

Patient education is of paramount importance in patients with WPW syndrome. This is especially true in asymptomatic young patients who have been told of their abnormal ECG results. Reassurance and periodic follow-up care of such patients is necessary.

Educate patients who are being treated with drug therapy thoroughly regarding the disease and the type of medications they are taking. Such patients must be taught the following:

How to recognize disease recurrence

How to perform vagal maneuvers, when needed

To keep their follow-up appointments

To identify the adverse effects of antiarrhythmic drugs

To avoid competitive sports

To learn about ablative options and the indications for ablation, should they become candidates in future

Patients with WPW syndrome should also educate their family members, and their siblings should be screened for preexcitation.

MISCELLANEOUS

Medical/Legal Pitfalls:

Evaluate patients presenting with symptomatic tachycardia (SVT or wide-complex tachycardia) for the presence of preexcitation on the ECG results.

Evaluate patients with WPW syndrome for the presence of very short refractory periods because these patients carry higher probabilities of developing symptoms or complications. Such patients also respond poorly to drug therapy. Identify these patients, even if asymptomatic, and treat them aggressively using EP evaluations and ablative therapy.

Special Concerns:

Children with symptomatic WPW syndrome who undergo RF ablation sustain myocardial damage or injury. How this damaged myocardium will change as children grow is still not known.

Evaluate patients with Ebstein anomaly for multiple accessory pathways. During EP studies and ablation, all such pathways should be recognized and treated.