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## Management of asymptomatic ventricular preexcitation

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### ABSTRACT

An incidental discovery of Wolff Parkinson White (WPW) pattern on the electrocardiogram (ECG) is not an infrequent finding facing the physician. Most patients discovered incidentally are asymptomatic and it is hard to justify further management of such patients given the time-honored adage to “first do no harm.” However, this finding does have implications. This article is an attempt to guide clinicians about this important issue that is often faced in the office.

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### 1. Introduction

The classic description of Wolff-Parkinson-White Syndrome goes back to the original paper by the three physicians in The American Heart Journal in 1930. They described a series of 11 patients without structural heart disease with ECG findings of a short PR interval, “bundle branch-block” and paroxysms of supraventricular tachycardia and/or atrial fibrillation [1]. A schematic of a typical conduction system and a corresponding ECG pattern compared to that with an accessory pathway (atrio-ventricular connection) is seen in Fig. 1 [2]. It took decades to understand the implication of this ECG abnormality and the association with an atrio-ventricular connection and its role in the pathophysiology of paroxysmal supraventricular tachycardia (SVT) [3]. As invasive electrophysiology developed and evolved, electrophysiologists came to learn that the extra-nodal atrio-ventricular connection can potentially put patients at risk of sudden cardiac death (SCD) from ventricular fibrillation due rapid antegrade conduction over these electrical connections during atrial flutter/fibrillation [4]. An example of preexcited atrial fibrillation is seen in Fig. 2.

For the purposes of terminology and understanding of this review, the term “ventricular preexcitation” refers to patients with the classic WPW pattern on their ECG without symptoms (also

referred to as “asymptomatic WPW” or “asymptomatic WPW syndrome”). WPW syndrome will refer to patients with WPW pattern and symptoms related to the ECG abnormality (palpitations, pre-syncope, syncope or cardiac arrest).

#### 1.1. Epidemiology

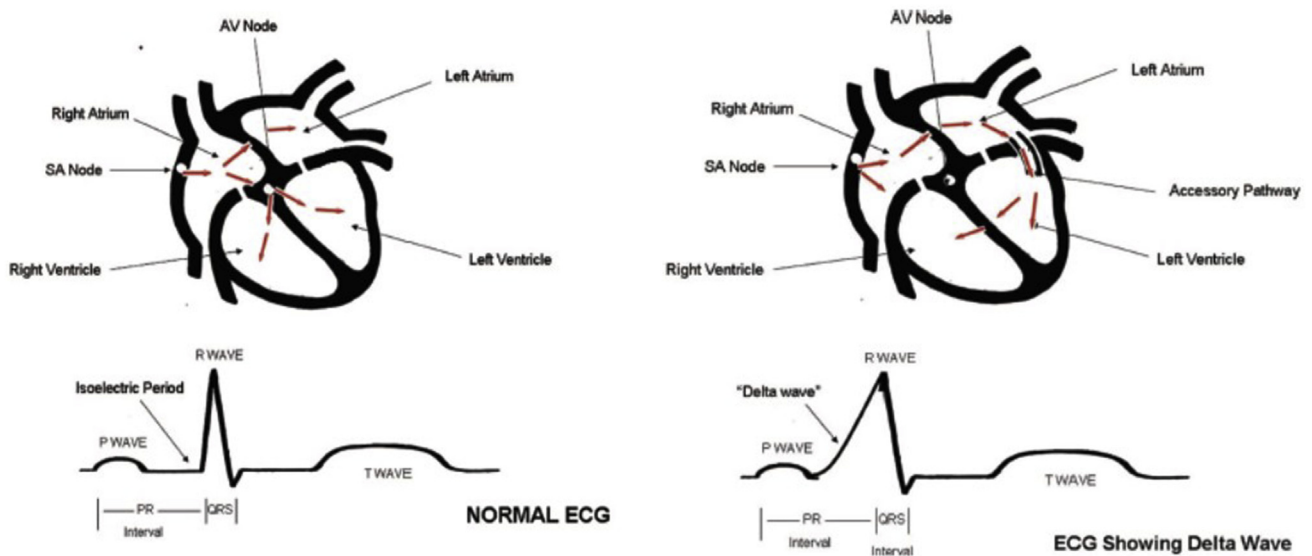
Most patients with ventricular preexcitation do not present with aborted SCD and in fact most can go decades without any symptoms related to their ventricular preexcitation. Population studies using ECGs have attempted to determine the incidence and prevalence of ventricular preexcitation and the incidence of symptoms and arrhythmias secondary to preexcitation. Many epidemiologic studies are fraught with limitations and biases to estimate the true incidence. Studies in the mid to latter 20th century focused on adult males who had ECG screening as part of an entrance health examination before entering the military and that is where the frequently quoted prevalence of 0.1–0.3% is quoted [5]. Smaller community prospective studies have also demonstrated a low incidence rate of ventricular preexcitation [6,7], estimating 4 per 100,000/year. During follow up of community-based studies, patients who were asymptomatic at the time of their diagnosis of ventricular preexcitation were shown to become symptomatic over time. Thirty percent of asymptomatic patients developed symptoms over a 10 year follow up period from the time of diagnosis of ventricular preexcitation [7]. The most common manifestations were documented tachyarrhythmia or intermittent palpitations.

In the pediatric population, the most common manifestation of

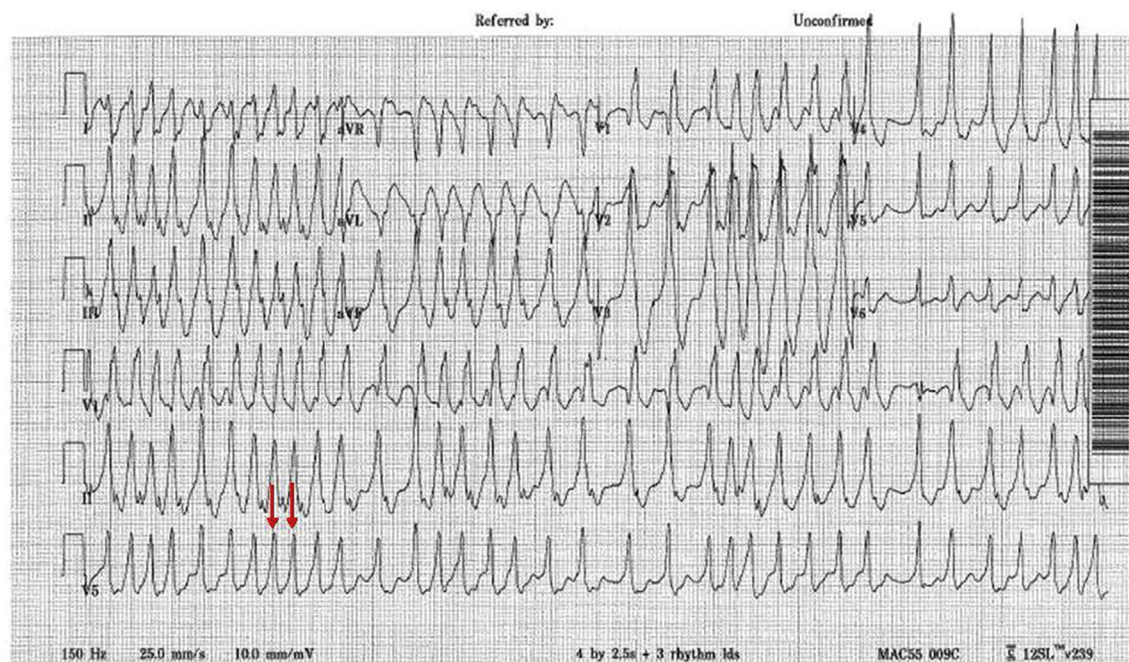
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**Fig. 1.** Left- Schematic of the typical cardiac conduction system in the absence of an extra-nodal atrio-ventricular accessory pathway. On the right is a schematic of the cardiac conduction system in the presence of an extra-nodal accessory atrio-ventricular connection that can conduct antegrade and the corresponding ECG pattern demonstrating ventricular preexcitation. Reprinted from *Circulation*, 122: e480-e483, Kulig J. and Koplan B. Wolff-Parkinson White Syndrome and Accessory Pathways.



**Fig. 2.** Preexcited Atrial Fibrillation- 15yo male presented with sudden onset of palpitations while exercising and was brought by his parents to the local emergency department. He never had a previous history of palpitations, syncope or documented tachyarrhythmias. The patient was promptly DC Cardioverted and admitted for an electrophysiologic study which found a unidirectional antegrade only left lateral accessory pathway. Antidromic AV Reentry was induced and ablated with RF energy. Note the shortest pre-excited RR interval on the ECG during atrial fibrillation is < 200 msec (see arrows).

WPW syndrome is paroxysmal supraventricular tachycardia. The mechanism of SVT with ventricular preexcitation is usually atrio-ventricular reentry tachycardia, orthodromic or antidromic. Orthodromic AV reentry (antegrade down the AVN-His-Purkinje system and retrograde up the accessory pathway) is typically more common than antidromic AV reentry (95% vs 5%) [8]. In the earliest series of infants with WPW syndrome and neonatal SVT, ventricular preexcitation was no longer appreciated on subsequent ECGs in 36% of patients during follow-up after initial diagnosis [9]. Recurrent SVT was not less in these patients compared to those

where manifest preexcitation was persistent. While 90% of infants with AV reentry tachycardia are likely to have less recurrent tachycardia at 1 year of age, nearly 30% will have late recurrence in later childhood.

The estimates of the prevalence of ventricular preexcitation are likely an underestimate, particularly in the pediatric population where there is no clear global consensus or requirement for routine ECG screening. Ventricular preexcitation can also be subtle and therefore there is always the possibility that diagnosis may be delayed in patients who have concealed preexcitation at the time of

an ECG, especially when there are no symptoms suggestive of an accessory pathway or the WPW syndrome. Another caveat to the diagnosis of ventricular preexcitation is that it does not have to be present on all twelve leads of an ECG. It can be present in isolated leads. This is due a number of reasons, including the location of the accessory pathway, the antegrade conduction properties of the pathway relative to the atrioventricular node (AVN) of the patient and the patient's own unique autonomic tone which modulates the conduction of the AVN. Examples of manifest and subtle ventricular preexcitation are seen in Fig. 3.

### 1.2. Risk of sudden cardiac death

The earliest study to evaluate the risk of sudden cardiac death from having ventricular preexcitation was by Klein et al. [10]. In this case-control study of patients with ventricular preexcitation, those with documented ventricular fibrillation were compared to those without and various characteristics were analyzed to determine what features of ventricular preexcitation portended a higher risk of SCD. The study population ranged from 6 to 68 year old patients. In the final analyses, 25 patients with ventricular fibrillation were compared to 73 patients with WPW syndrome but no ventricular fibrillation. The features that were found to be associated with risk of SCD were a shortest preexcited RR interval (SPERRI)  $\leq 250$  msec, having multiple accessory pathways and an individual having had both reentry tachycardia and history of atrial fibrillation (all statistically significant). It should be noted that of the 25 patients who had ventricular fibrillation in the study, 22 had prior symptoms. This has suggested that symptomatic preexcited patients have a different risk of SCD compared to asymptomatic preexcited patients. The most recent multicenter retrospective study of asymptomatic ventricular preexcitation over a 25 year period in pediatric patients through the Pediatric and Congenital Electrophysiology Society (PACES) organization reinforced the use of invasive EP study characteristics in assessment of the risk properties of antegrade accessory pathway conduction (specifically SPERRI  $\leq 250$  msec) [11].

The potential for a sudden cardiac death event from ventricular preexcitation has been well documented but the exact risk in an individual patient with preexcitation is largely dependent on the electrophysiologic properties of the accessory pathway. Knowing that patients with this ECG finding have a risk of SCD, and that SCD alone can be the first manifestation of WPW syndrome [10–12]; there has been considerable effort to determine best course of action in patients with asymptomatic ventricular preexcitation [13]. There is little disagreement on the utility of invasive catheter ablation for the patient with symptoms related to ventricular preexcitation. For the patient with no symptoms, the goal of the clinician is to adequately counsel the patient on their options and lifetime risks, notably for SCD related to their ECG finding. In order to counsel patients and families adequately, it is worth reviewing the available literature on the risk of SCD with ventricular preexcitation.

A recent meta-analysis on the risk of sudden death in asymptomatic preexcitation evaluated the available literature on this complex topic [14]. The analysis included retrospective and prospective studies of asymptomatic patients with ventricular preexcitation. The follow-up data was variable and while there are inherent limitations with meta-analyses, a number of conclusions were made that reinforced the current understanding of risks of SCD with ventricular preexcitation. Twenty studies were included in the final analyses, of which six evaluated pediatric patients and the remaining fourteen in adults with asymptomatic ventricular preexcitation. The overall risk of SCD was determined to be 0.85 events per 1000 person-years. In children, the risk was 1.93 (95% CI

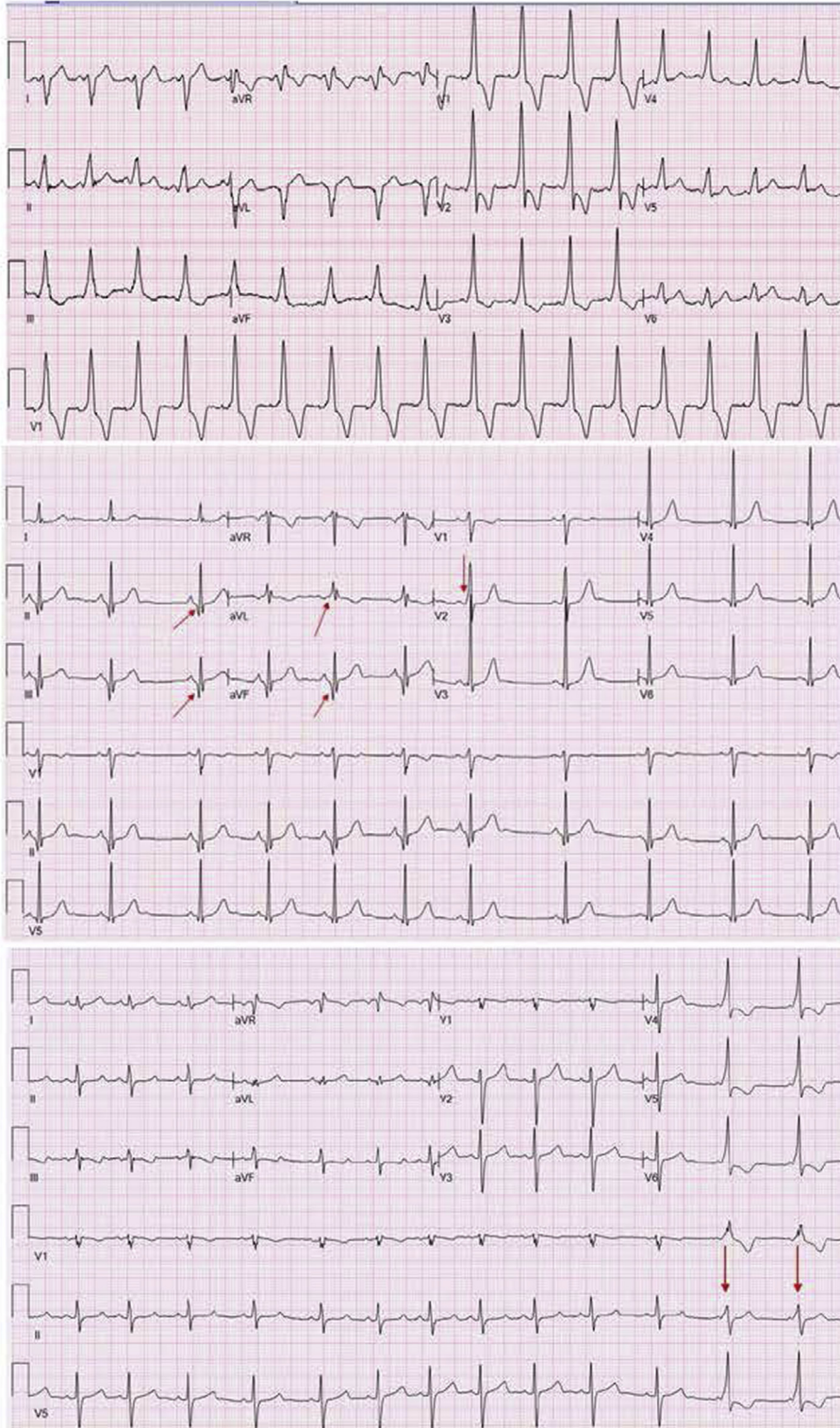
0.57–4.14) events per 1000 person-years and in adults alone the risk was 0.86 (95% CI 0.28–1.75) events per 1000 years,  $P < 0.07$ . Overall, the risk of SCD was found to be low in all patients with asymptomatic ventricular preexcitation, but it also emphasized that children are at increased risk as noted in previous studies. The conclusion was that pediatric patients with ventricular preexcitation should have careful follow-up with close monitoring for arrhythmias.

The current studies have led to the accepted consensus that patients with ventricular preexcitation without symptoms should be counseled on their potential risk of SCD and their risk for arrhythmias such as atrio-ventricular reentry. While the risk of SCD is overall low, it is not zero and so risk assessment is the next step in managing these patients. The adult catheter guidelines for supra-ventricular tachycardia give a Class I recommendation (benefits exceeding risks) for proceeding to an invasive catheter study for patients with symptoms attributed to ventricular preexcitation (WPW syndrome) [8,15]. For patients without symptoms, the recommendation to pursue an EP study is considered reasonable in a patient who has no symptoms. If an invasive study is performed and findings suggest the AV connection is high risk or SVT is induced, then, ablation is also considered reasonable [8,13]. In both pediatric and adult guidelines, risk assessment is recommended once a preexcited patient without symptoms is identified.

## 2. Work-up and evaluation

Once a patient with ventricular preexcitation is identified, it is advisable to start with an echocardiogram to evaluate their cardiac function and anatomy. This helps to rule out congenital heart disease (CHD) and potential cardiomyopathy that may have been subclinical. Early pediatric studies of Wolff-Parkinson-White Syndrome and paroxysmal SVT found that infants with the aforementioned diagnoses have a 20% risk of CHD. The most common lesion was Ebstein's anomaly [9] but other lesions (L-TGA, also known as congenitally corrected transposition of the great arteries) have been found to be associated with WPW and paroxysmal SVT. Ebstein's anomaly in particular has an increased risk for multiple accessory pathways and are an independent risk factor for a life threatening event in the recent published multi-center PACES study [11]. Ventricular preexcitation has also been shown to be associated with certain cardiomyopathies such as hypertrophic cardiomyopathy associated PRKAG2 and AMPK mutations [16]. In this particular cardiomyopathy, there is a strong association between mutations in PRKAG2 and ventricular preexcitation. The autosomal dominant pattern of inheritance mandates the close family members of an index case be screened and evaluated by a cardiologist, even in the absence of symptoms.

A baseline transthoracic echocardiogram also provides a baseline assessment of ventricular function. Ventricular preexcitation can adversely affect systolic function overtime. In a typical heart with normal segmental anatomy and absence of ventricular preexcitation, the bundle of His runs along the left side of the ventricular septum along the membranous septum. Therefore, the proximal left ventricular septum is depolarized first before progressing through the interventricular septum and engaging the distal Purkinje fibers and ventricular myocardium. Accessory pathways with robust antegrade conduction can alter the typical sequence of ventricular activation and in turn lead to dyssynchrony and atypical depolarization through the ventricles. Right free wall and septal accessory pathways have been found to increase the risk of ventricular dysfunction from dyssynchrony. Ablation of these accessory pathways at follow up has been shown to restore normal ventricular function [17–19]. Depressed ventricular function is a reasonable indication for ablation in patients with preexcitation, if



**Fig. 3.** The first ECG is an example of manifest ventricular preexcitation that was found in an 11yo girl who was transported by EMS for respiratory distress due to an asthma exacerbation. She had no symptoms related to her ventricular preexcitation. The preexcitation is apparent in all 12 leads and suggests a left lateral accessory pathway. The second ECG is from a 12yo boy who was diagnosed with an anxiety disorder for years and had an ECG performed for screening before initiation of stimulant medication by his primary care provider. Note the atypical appearance of the QRS complex in the inferior leads and aVL (see arrows). In V2 (see arrow) there is appreciation of ventricular preexcitation. The patient underwent an intracardiac EP study and was found to have inducible orthodromic AVRT through a left posterior accessory pathway. The third ECG is from an 13yo girl with Turner's Syndrome, bicuspid aortic valve and a dilated ascending aorta followed by a pediatric cardiologist. On routine follow-up was found to have intermittent ventricular preexcitation (see arrows) and on review of prior ECGs, never had preexcitation.

the abnormal depolarization from the accessory pathway is felt to be the cause of ventricular dysfunction [13].

After baseline assessment of intracardiac anatomy and ventricular function is performed, a thorough discussion with the patient and family about the life-long consequences of ventricular preexcitation should be taken which includes covering the potential risks of paroxysmal supraventricular tachycardia (atrioventricular reentry and atrial fibrillation) and SCD from preexcited atrial fibrillation.

### 3. Risk assessment/stratification

The assessment of the risk of an accessory pathway is rooted in evaluating for loss of manifest ventricular preexcitation (antegrade conduction through the pathway) during fast heart rates. The most consistent electrophysiologic parameter that has been found to correlate with SCD has been the shortest preexcited RR interval during atrial fibrillation (SPERRI). Outside of the electrophysiology laboratory, this is usually accomplished by evaluating the surface ECG for abrupt loss of ventricular preexcitation at fast sinus rates or during atrial fibrillation as a surrogate for the antegrade effective refractory period of the accessory pathway (APERP). Patients who have intermittent ventricular preexcitation on their surface ECG are assumed to have long antegrade refractory periods and believed to be at a lower risk of SCD due to loss of antegrade conduction during sinus rhythm. In clinical practice, electrophysiologists/cardiologists have defined a patient as “low-risk” or rather “not-high risk” if intermittent ventricular preexcitation is seen on a resting ECG or ambulatory rhythm monitoring (i.e. Holter monitors). Intermittent preexcitation may be due to acceleration dependent block (Phase 3 block) when an impulse reaches a conduction fiber during its repolarization phase. It may also occur when an impulse reaches a conduction fiber during a time of slow diastolic depolarization (automaticity), referred to as Phase 4 block [20]. It is therefore important to characterize the circumstances of intermittent ventricular preexcitation in order to gauge the risk of its antegrade conduction and likelihood to predispose the patient to SCD. In the outpatient setting, risk stratification can be performed with an exercise stress test with continuous ECG monitoring if the patient is ambulatory and able to perform the test adequately.

During exercise and increase in catecholamines, there will be an increase in the sinus rates as well as enhanced conduction through the accessory pathway and atrioventricular node. This may lead to varying degrees of ventricular preexcitation on the surface ECG and mask persistent preexcitation. Abrupt and complete loss of preexcitation during exercise has been shown to correlate to a long antegrade APERP [21]. The caveat is that if preexcitation is subtle but persistent or gradually decreases at faster sinus rates, then the utility of the stress test as a risk stratification tool is limited and another means of risk stratification maybe necessary. An example of abrupt and complete loss of preexcitation during exercise is seen in Fig. 4 [15] which would classify a patient as “not-high risk.” The assessment of abrupt loss of preexcitation and conferring a patient’s risk level as “not-high risk” by exercise stress testing has been found to have low sensitivity when compared to baseline EP testing to detect non-rapid conduction through an accessory pathway but the specificity and positive predictive value were found to be 100% in one retrospective study; however no SCD was reported in their population over a 10-year period [22].

While “intermittent preexcitation” may confer a lower risk of SCD, it does not confer “no-risk” to the patient. Since the publication of the 2012 PACES/HRS guidelines on management of asymptomatic preexcited patients, at least two studies retrospectively analyzed their center’s experience with pediatric patients who were deemed to have intermittent ventricular preexcitation by

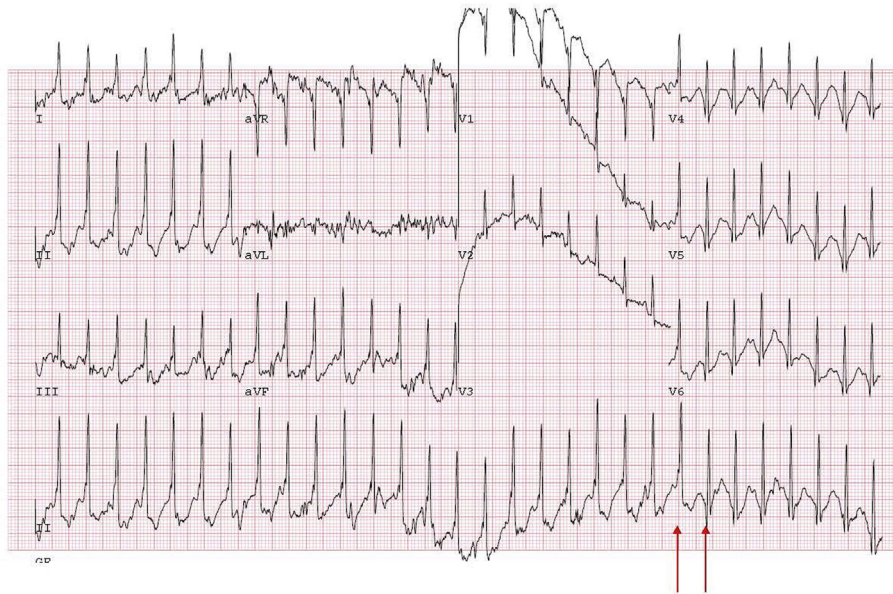
noninvasive means and evaluated their electrophysiologic profiles from invasive EP testing and clinical outcomes were reviewed. Both studies [23,24] found that there was no difference in the prevalence of high risk AP between patients with persistent preexcitation, those with intermittent preexcitation on baseline ECG and those with loss of preexcitation on Holter or exercise. These findings lend support to the recent 2012 guidelines on management of young patients with asymptomatic ventricular preexcitation [13].

The options for noninvasive risk stratification are limited in the current era beyond ambulatory rhythm monitoring and an exercise stress test. Another option for risk stratification in patients is a transesophageal pacing study (TEEPS), which, traditionally, has been useful for pacing infants out of refractory supraventricular arrhythmias. This may be an appealing option if the patient cannot perform an exercise stress test adequately, either from immaturity or the inability to give an adequate effort. TEEPS can be considered a less invasive option for patients if they wish to avoid an invasive catheter study or if an exercise stress test is nondiagnostic as some centers have found the data obtained to be comparable to an intracardiac EP study [25]. It should be emphasized that transesophageal pacing is not without risks as it does require the use of sedation; has the potential for damage to the esophagus and can induce dangerous arrhythmias such as VF in the setting of a high-risk accessory pathway. The choice between TEEPS over an exercise stress test is largely at the discretion of the practicing electrophysiologist and center dependent. One pediatric center demonstrated that TEEPS can be used in concert with exercise stress testing to avoid a transcatheter EP study in those deemed not high risk for SCD (based on SPERRI < 250 ms or APERP < 250 ms if atrial fibrillation could not be induced) [26]. Half of the patients during the study period were able to avoid an intracardiac EP study if classified as low risk based on no symptoms of heart rhythm abnormalities or life-threatening events were found during their follow-up period. It should be noted that follow-up data was available for 88% of the patients classified as low risk. There are no long-term prospective studies evaluating the use of exercise stress testing or TEEPS in risk assessment of asymptomatic WPW.

With regards to ambulatory rhythm monitoring (i.e. Holters), there is no recommendation for their use in the setting of a pre-excited patient without symptoms for the purposes of risk stratification or initial evaluation. A physician always has the option of using ambulatory monitors for screening for asymptomatic arrhythmias. The largest prospective study of children with asymptomatic preexcitation did find atrial fibrillation on biannual Holters after an initial invasive electrophysiology study in 22 out of 184 children [12]. Ambulatory rhythm monitoring could help determine if a patient has multiple accessory pathways (convincing change in preexcitation pattern) but this requires trusting the accuracy of a single or multiple channel monitor with minimal artifact and it may not reflect the same physiologic state as an exercise stress test to appropriately risk stratify a patient.

### 4. Counseling of patient and family

If risk stratification is equivocal from exercise stress testing, or cannot be evaluated with TEEPS then accessory pathway localization may aid in counseling a patient/family on options moving forward to determine if there is benefit from an intracardiac EP study and possible ablation. Specific features on ECGs with manifest preexcitation can be analyzed to determine AP location. These features include the delta wave polarity [27], R/S ratio of the pre-excited QRS complex [28] and QRS polarity. It should be noted that in young patients (with and without CHD), published algorithms are less accurate and there is significant interrater variability [29], however in practice the algorithms are still used by most pediatric



**Fig. 4.** Abrupt loss of preexcitation during exercise stress testing. The ECG demonstrates a patient with a likely right anteroseptal accessory pathway. There is single beat loss of preexcitation (see arrows) upon analysis of the ECG and rhythm strip towards the end with the last several sinus beats without preexcitation. Image courtesy of Ahmad Sami Chaouki, MD, PhD.

and adult electrophysiologists. The caveat to predicting accessory pathway location is that most published algorithms are based on the assumption a patient is maximally preexcited and only one antegrade pathway is present. If the patient's ECG appears maximally preexcited but the preexcitation pattern is atypical (AP cannot be determined) with no evidence of structural or congenital heart disease then the possibility of more than one accessory pathway must be considered. Serial ECGs of the patient, if they exist, may also aid in determining if a patient has multiple accessory pathways or if there are different preexcitation patterns at different sinus rates. Multiple accessory pathways are a risk factor for a life-threatening event/SCD [10,11] and this must be factored in when counseling the patient/family if clinical suspicion exists in the absence of symptoms. Predicting the location of an accessory pathway can help in counseling a patient and family about the risks/benefits of in an intracardiac study and potential ablation. Accessory pathways that are located on the septum have the potential for damage to the conduction system (compact AV node or His Bundle) and pathways within the coronary venous system may have collateral damage to the coronary arteries with radiofrequency (RF) ablation. A manifest preexcitation pattern that is convincingly left sided may require a transseptal puncture which is an added step if the interventional electrophysiologist is likely to ablate the pathway. All these factors should be taken into account when discussing the risks and benefits of an invasive intracardiac EP study and possible ablation.

## 5. Catheter ablation

Catheter ablation has been the gold standard treatment for accessory pathways. The use of RF compared to cryoablation is largely operator dependent with the majority of practitioners opting for RF ablation unless it is felt the risk of RF ablation is unfavorable depending on the patient characteristics (age and size), anatomic location of the accessory pathway (within coronary venous system or paraseptal) and potential risk of damage to the conduction system. The acute success rates for RF ablation in adults and children are quoted to be 93–96% [30,31] and so the potential

for a repeat catheter study and ablation is a remote possibility for patients. If recurrence of the AP is found in follow up, it typically is found within 2 months after ablation. The recurrence rate for accessory pathways varies depending on a number of factors that are largely operator dependent (experience and skill level), but has been found to also be associated with AP location. Right free wall accessory pathways have been found to have a higher recurrence rate compared to left lateral free wall pathways (9–10% compared to 2%) [30,31]. The complication rate overall for RF ablation from the Prospective Assessment after Pediatric Cardiac Ablation (PAPCA) was found to be 4.0% with the incidence of AV block estimated to be 1.2% and occur for pathways in the right and left septal region. No deaths were found in the PAPCA study, however earlier registry studies of pediatric ablation had a low rate of mortality and it is presumed that improved skill, technology and patient selection explains the improved outcomes since the initial RF ablation experience.

Cryoablation is another option for ablation of accessory pathways, especially for those that are anteroseptal, parahisian, mid-septal, posteroseptal or in the coronary venous system. Similar to RF ablation's history, the acute success rates of cryoablation improved over time as experience with it improved and modifications were made to the original techniques. Success rates, recurrence and adverse event estimates with cryoablation vary depending on the center. A recent metaanalysis comparing cryoablation to RF ablation of septal accessory pathways report acute procedural success as 86% vs 89%, respectively [32]. The recurrence rate for cryoablation from the analysis was estimated to be 18.1% compared to 9.9% for RF ablation. No cases of AV block were found for cryoablation while RF ablation accounts for a 2.7% incidence of AV block with septal accessory pathway ablation. Despite the higher recurrence rate, the safety profile is believed to be favorable for ablation targets near the AV node as AV block is often transient with cryoablation after energy is terminated immediately when block is recognized [33].

Taking a patient for an invasive catheter study has the potential for finding unexpected results. Accessory pathway variants maybe found which are not the classic atrioventricular connections that

are synonymous with Wolff-Parkinson-White syndrome. Some of these variants are known to participate in clinical tachycardia (atriofascicular or “Mahaim” pathways) while others are not likely to participate in AV reentry or put a patient at risk of sudden cardiac death. A review of these variants (atriofascicular, nodoventricular, nodofascicular and fasciculoventricular pathways) and how to differentiate them on a surface ECG or in the electrophysiology laboratory is beyond the scope of this review.

Worthy of mention is the fact that all the published data on catheter ablation outcomes (acute success rates, recurrences and complications) for accessory pathways were during the era of fluoroscopy. The advent of 3D electroanatomical mapping has reduced the need to rely on fluoroscopy for most if not all invasive catheter studies and ablations, but has not completely eliminated its need. Most contemporary studies of 3D mapping systems and outcomes have found that ablation success rates have not changed significantly and patients can undergo invasive catheter studies with minimal radiation exposure [34].

## 6. Summary

To “do no harm” is particularly challenging for a patient with asymptomatic ventricular preexcitation as the risk of SCD is low. The current data suggests that pediatric patients are at a higher risk of SCD compared to adult patients with ventricular preexcitation. Catheter ablations in the current era are considered safe overall with a low risk of adverse events and so it is challenging for electrophysiologists not to offer the option to a patient if the risk/benefit profile is favorable. The risk/benefit is unique for each patient as various factors can influence the decision to undergo an invasive catheter study. At this juncture, published guidelines and expert opinion advocate for shared decision making between the patient/family and electrophysiologist. As technology evolves and catheter ablation outcomes improve, it may increase the temptation to ablate. However, practicing electrophysiologists would be wise to remember the words of Mark Twain: “To the man with a hammer, everything looks like a nail.”

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## Declaration of competing interest

The authors declare that they have no know competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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