

# Wolff-Parkinson-White Syndrome In Children: A Single-Center Experience

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## ABSTRACT

This study aims to evaluate the clinical characteristics and outcomes of children with intermittent and persistent Wolff-Parkinson-White (WPW) syndrome.

Children followed up with the diagnosis of intermittent WPW and WPW syndrome between 2014 and 2022 were reviewed retrospectively. The age, gender, duration of follow-up, admission reasons, electrocardiography (ECG), echocardiography, and ambulatory ECG findings, electrophysiological study and ablation results were analyzed.

The study included 99 children with intermittent WPW and 38 children with WPW syndrome. The most common complaints on admission were palpitation (28.5%), murmur (22.6%), and chest pain (19%). Seven of the cases had a history of documented supraventricular tachycardia (SVT) and one of them had SVT on admission. On initial ECG 40.9% and 27.7% of the cases had intermittent and persistent WPW, respectively. Ninety-five (69.4%) cases had a structurally normal heart, whereas congenital heart disease, valvular disease, and mild septal hypertrophy were found in 25 (18.3%), 16 (11.7%), and 1 case, respectively. In the whole study group, 49 (35.8%) cases underwent electrophysiological study (EPS). Five cases had a diagnostic procedure and catheter ablation was performed in 39 (28.5%) cases. Five cases were lost from follow-up after referral for EPS. Recurrence was observed in only one case and none of the cases had any adverse event. Early diagnosis, risk stratification, and appropriate management of patients with WPW syndrome is crucial. The electrophysiological study is used as a risk stratification tool and catheter ablation is a safe curative procedure in high-risk patients.

**Keywords:** Electrophysiological study; intermittent; preexcitation; Wolff-Parkinson-White syndrome

## Introduction

Wolff-Parkinson-White (WPW) syndrome is the most common preexcitation syndrome and it is estimated that there are more than 30000 children with asymptomatic WPW pattern in the U.S.A.(1) The prevalence of WPW syndrome in children aged 6-20 years old was reported as 0.07%.(2)

The diagnosis of the WPW pattern is usually incidental and most individuals are diagnosed during evaluation for another illness or routine follow-up. The classical ECG pattern, - a short PR interval, a wide QRS complex, and a slurred onset of the QRS complex (delta wave)- is adequate for diagnosis. If there is an intermittent loss of preexcitation, then it is called intermittent WPW syndrome.(3)

Most children with WPW pattern are asymptomatic but various types of

supraventricular tachyarrhythmias (SVT) can occur in children with WPW syndrome and very rarely sudden cardiac death (SCD) may be the first presentation of this syndrome. Sudden cardiac death is thought to be due to rapid conduction of atrial fibrillation to ventricles via accessory pathways.(1) The risk of SCD is estimated as 0.25% per year or 3-4% over a lifetime in symptomatic patients with WPW syndrome but it is quite low in asymptomatic patients in comparison to symptomatic patients.(4)

The indications of electrophysiological study (EPS) in patients with symptomatic WPW patients are rather clear, however, there are some debates on the management of patients with asymptomatic and intermittent WPW syndrome.(5)

In this single-center study, we aimed to evaluate the clinical characteristics and outcomes of

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children diagnosed with intermittent WPW and WPW syndrome.

## Material and Methods

Medical records of all children that were diagnosed and followed up with intermittent WPW and WPW syndrome at the Pediatric Cardiology Department of Van Training and Research Hospital between 2014-2022 were reviewed retrospectively. The local ethics committee approved the study.

The age, gender, duration of follow-up, reasons for admission, electrocardiography (ECG), echocardiography and 24-hour Holter ECG findings, and results of EPS and catheter ablation were analyzed.

The Wolff-Parkinson-White pattern was defined according to the typical three main features: A short PR interval, a wide QRS complex, and a delta wave.<sup>(3)</sup> Patients with persistent preexcitation present on ECG and 24-hour Holter ECG were defined as WPW syndrome, whereas patients with an intermittent pattern on ECG and patients with normal ECG but with an intermittent pattern on 24-hour Holter ECG were defined as intermittent WPW.

Echocardiographic examination, 12-lead ECG, and 24-Hour ECG Holter monitoring were performed and analyzed by experienced pediatric cardiologists. The PACES/HRS expert consensus statement on the management of the asymptomatic young patient with a Wolff-Parkinson-White electrocardiographic pattern (5), and PACES/HRS Expert Consensus Statement on the use of Catheter Ablation in Children and Patients with Congenital Heart Disease (6) were used for deciding to EPS and catheter ablation. If the EPS procedure was not scheduled on a close date, beta-blocker treatment was started, and it was ceased 5 days before the procedure. Electrophysiological study and catheter ablation procedures were performed in various other tertiary heart centers by pediatric electrophysiologists.

Statistical analyses were performed using the statistical package program SPSS for Windows 22.0 (IBM Corp., Armonk, NY, USA). The mean, standard deviation, and frequency were used for descriptive statistics.

## Results

The study included a total of 137 pediatric patients with intermittent WPW (n:99) and WPW syndrome (n:38). There were 61 (44.5%) female and 76 (55.5%) male patients. The mean age of the cases was  $11.03 \pm 4.14$  years and the mean duration of follow-up was  $29.31 \pm 32.4$  months. One of the cases had Leighs syndrome, and another one had Down syndrome. None of the patients had a familial history of sudden cardiac death.

At the time of diagnosis, 108 (78.8%) of the cases had admitted with a complaint and 18 (13.1%) cases were already being followed up by the pediatric cardiology department for congenital heart disease, rheumatic heart disease, etc. The most common reasons for admission were palpitation (28.5%), murmur (22.6%), and chest pain (19%). Seven of the cases who admitted with palpitation had a history of documented SVT and one of them had SVT at the time of admission. Eleven (8%) cases were referred from other departments and institutions. Four of these cases were referred with suspected ECG anomaly: 2 were referred with suspected WPW syndrome and 2 with suspected arrhythmia. On admission, ECG revealed a WPW pattern in 3 of them and all underwent EPS and had catheter ablation. An interesting point was that 3 asymptomatic cases were referred just for evaluation for eligibility to sports, and 2 of them had WPW pattern on ECG. Both were treated successfully with catheter ablation. The causes of the initial admission are shown in Table 1.

On the initial 12-lead ECG, 40.9% of the cases had intermittent WPW pattern while the remaining 27.7% had persistent WPW pattern. The ECG and 24-hour ECG Holter findings of all cases are shown in Table 2. The mean heart rate on ECG on admission was  $91.15 \pm 28.06$  beats/minute. Intermittent WPW pattern (60.5%) was the most common finding on 24-hour Holter ECG monitoring. The mean maximum, average, and minimal heart rates on 24-hour Holter monitoring were  $158.64 \pm 20.39$ ,  $86.93 \pm 13.86$ , and  $49.62 \pm 15.3$  beats/min, respectively. Sixteen cases, who had intermittent WPW pattern on initial ECG, had normal 24-hour Holter ECG findings. None of the cases had supraventricular or ventricular tachycardia or atrial fibrillation on 24-hour Holter monitoring.

Echocardiography revealed a structurally normal heart in 95 (69.4%) cases. A total of 25 (18.3%)

**Table 1.** Reasons For Admission At The Time of Diagnosis

Admission Reasons	N (%)
<b>Admitted with complaints</b>	108 (78.8)
Palpitation	39 (28.5)
Murmur	31(22.6)
Chest pain	26 (19)
Syncope	4 (2.9)
Chest pain + palpitation	3 (2.2)
Shortness of breath	3 (2.2)
Dizziness	1 (0.7)
Rheumatic heart disease + palpitation	1 (0.7)
<b>Asymptomatic and being followed-up</b>	18 (13.1)
Follow-up for congenital heart disease	12 (8.8)
Follow-up for rheumatic heart disease	4 (2.9)
Follow-up for post-myocarditis	1 (0.7)
Follow-up for chemotherapy side effects	1 (0.7)
<b>Referred for other reasons</b>	11 (8)
With suspected electrocardiogram anomaly	4 (2.9)
With suspected WPW	2 (1.4)
With suspected arrhythmia	2 (1.4)
Eligibility to sports	3 (2.2)
Asthma	1 (0.7)
Brucellosis	1 (0.7)
Elevated Creatinine Kinase	1 (0.7)
Chest Wall Deformity	1 (0.7)
<b>Total</b>	137 (100)

WPW: Wolff-Parkinson-White syndrome

cases had congenital heart disease (operated/unoperated), 16 had a valvular disease and one had mild septal hypertrophy. The most common echocardiographic abnormality was the secundum type atrial septal defect. The echocardiographic findings of all cases are shown in Table 3.

In the whole study group, EPS was performed in 49 (35.8%) cases. Catheter ablation was performed in 39 cases and 5 cases had diagnostic EPS whereas 5 cases were lost from follow-up after referral for EPS. Radiofrequency ablation (RFA) was the preferred choice of treatment in 33 cases and the rest 6 cases had cryoablation. (Figure 1) The most common causes of admission in these cases were palpitation (28.6%), murmur (26.5%), and chest pain (14.3%). Except for 3 cases, all cases that underwent EPS had WPW or intermittent WPW on ECG on admission. 24-h-Holter ECG monitoring revealed intermittent WPW pattern in 3 cases with normal ECG on admission. These 3 cases were referred for EPS because of a history of previously documented

SVT and all had successful catheter ablation. Recurrence of the WPW pattern after catheter ablation was observed in only one male patient who had chest pain on initial admission and a structurally normal heart. The causes of admission and electrocardiography, echocardiography, and 24-h Holter ECG findings of all cases that were referred for EPS are shown in Table 4.

When the cases with WPW syndrome (n:38) were analyzed in means of EPS, all cases except one were referred for EPS. The only exception was a 3-year-old female patient who had admitted with a murmur and was being followed up with the diagnosis of SVT. One case had diagnostic EPS, 33 had catheter ablation (27 RFA and 6 cryoablation) and 3 were lost from follow-up after referral for EPS. The diagnostic procedure was performed in a case who underwent surgery for subaortic ridge and stenosis, and the electrocardiographic findings were attributed to the previous cardiac surgery.

There were 99 cases with intermittent WPW syndrome, and 77 cases were defined as low risk

**Table 2.** The Electrocardiogram and 24-h Holter ECG Findings of The Whole Group

Electrocardiogram and 24-h Holter ECG findings	Number (%)
<b>12- Lead Electrocardiogram findings on admission</b>	
Intermittent WPW pattern	56 (40.9)
Intermittent WPW pattern	54 (39.4)
Intermittent WPW pattern with supraventricular extrasystoles	1 (0.7)
Intermittent WPW pattern with incomplete right bundle branch block	1 (0.7)
WPW pattern	38 (27.7)
Normal	20 (14.7)
Short PR interval	9 (6.6)
Sinus arrhythmia	3 (2.2)
Sinus tachycardia	2 (1.5)
Supraventricular extrasystoles	2 (1.5)
Ventricular extrasystoles	2 (1.5)
Junctional rhythm	2 (1.5)
Supraventricular tachycardia	1 (0.7)
Incomplete right bundle branch block	1 (0.7)
Sinus bradycardia	1 (0.7)
Total	137 (100)
<b>24-hour Holter Electrocardiogram findings</b>	
Intermittent WPW pattern	83 (60.5)
Intermittent WPW pattern	71 (51.8)
Intermittent WPW pattern with ventricular extrasystoles	8 (5.8)
Intermittent WPW pattern with supraventricular extrasystoles	3 (2.2)
Intermittent WPW pattern with incomplete right bundle branch block	1 (0.7)
WPW pattern	38 (27.8)
WPW pattern	36 (26.3)
WPW pattern with ventricular extrasystoles	2 (1.5)
Normal	16 (11.7)
Total	137 (100)

WPW: Wolff-Parkinson-White Syndrome

because their complaints were not attributed to the presence of intermittent WPW pattern. As a result, 12 cases were referred for EPS and 6 of them were being followed up with the diagnosis of SVT by other institutions. The rest 6 cases referred for EPS had complaints consistent with cardiac palpitations. A total of 6 cases with intermittent WPW had catheter ablation, 4 had a diagnostic procedure and 2 were lost from follow-up after referral for EPS. Radiofrequency ablation was the preferred choice of treatment in all cases, and all ablations were performed in children with a history of the previous SVT.

## Discussion

Wolff-Parkinson-White syndrome is a clinical entity characterized by the presence of  $\geq 1$  accessory pathways between the atria and the

ventricles. Conduction of electrical impulse from atria to ventricle via an accessory pathway bypassing the A-V node results in earlier activation (preexcitation) of the ventricles and predisposes patients to arrhythmias.(7, 8) It is the most common preexcitation syndrome with an estimated prevalence of 1-3 in 1000 individuals and is more common in males.(9) Similarly, in our study, there was a male predominance (55.5%).

The prevalence of WPW pattern is estimated as 0.13 to 0.25% in the general population.(5, 10, 11) The prevalence of WPW syndrome was reported as 0.07% in children aged 6-20 years old.(2) The true prevalence of intermittent preexcitation is unclear because of its intermittent nature but it is estimated as 13-40% in children with WPW syndrome.(12, 13) In a cohort of 295 patients with preexcitation, 13% had intermittent preexcitation and 10% of all patients had a loss of

**Table 3.** Echocardiography Findings of All Patients

<b>Echocardiography findings of all patients</b>	<b>N (%)</b>
<i>Normal</i>	95 (69.4)
<b><i>Congenital Heart Disease</i></b>	25 (18.3)
<i>No history of surgery or catheter intervention</i>	16 (11.7)
Atrial septal defect	8 (5.8)
Mitral valve prolapse	6 (4.4)
Mitral valve prolapse with mild mitral insufficiency	5 (3.7)
Mitral valve prolapse with moderate mitral insufficiency	1 (0.7)
Bicuspid aortic valve	1 (0.7)
Ventricular septal defect	1 (0.7)
<i>History of surgery or catheter intervention</i>	9 (6.6)
Operated secundum atrial septal defect	2 (1.5)
Operated transposition of great arteries	1 (0.7)
Operated atrioventricular septal defect and mitral cleft	1 (0.7)
Ebstein anomaly (Glenn procedure)	1 (0.7)
Double inlet left ventricle (Fontan circulation)	1 (0.7)
Operated subaortic ridge and stenosis	1 (0.7)
Pulmonary stenosis (percutaneous valvuloplasty)	1 (0.7)
Percutaneously closed ventricular septal defect	1 (0.7)
<b><i>Other</i></b>	17 (12.4)
Trivial mitral regurgitation	7 (5.1)
Mild mitral regurgitation	3 (2.2)
Mild mitral regurgitation and aortic regurgitation	2 (1.5)
Mild aortic regurgitation	2 (1.5)
Mild pulmonary stenosis	1 (0.7)
Moderate mitral regurgitation and trivial aortic regurgitation	1 (0.7)
Mild septal hypertrophy	1 (0.7)
<b><i>Total</i></b>	137 (100)

preexcitation on ambulatory monitoring or exercise test.(14) In our study there were 99 (72.2%) cases with intermittent WPW. Only 56 of the cases had intermittent WPW pattern on admission ECG and the rest 43 were diagnosed according to the intermittent WPW pattern on 24-h Holter ECG monitoring. Twenty of the 43 cases diagnosed as intermittent WPW by 24-h Holter ECG monitoring had normal ECG on admission. Interestingly, 16 cases with intermittent WPW pattern on admission, had normal 24-h Holter ECG monitoring. As a result, 11.7% of cases with preexcitation (intermittent and persistent) had a loss of preexcitation pattern on ambulatory ECG monitoring.

Most individuals with WPW syndrome are asymptomatic. Many are diagnosed during investigation for other diseases, routine screening, and evaluation for eligibility to sports.(1) When symptomatic, the symptoms are related to

dysrhythmia. Most symptomatic individuals present with palpitation, dizziness, lightheadedness, syncope/presyncope, chest pain, and very rarely with sudden cardiac death. Kiger et al.(14) reported that palpitations, chest pain, and syncope were the most common symptoms in patients with WPW syndrome. In our study, 21.1% of the cases had no complaints on admission and they were referred for cardiac examination or were already being followed up for other cardiac diseases. The rest of the cases had admitted with a complaint that should be attributed to dysrhythmia. Palpitation followed by murmur and chest pain were the most common reasons for admission both in the whole group and in cases who were referred for EPS. It must be taken into consideration that all cases admitted with a complaint should not be considered as symptomatic patients. We preferred the term "complaint" rather than "symptom" because it is

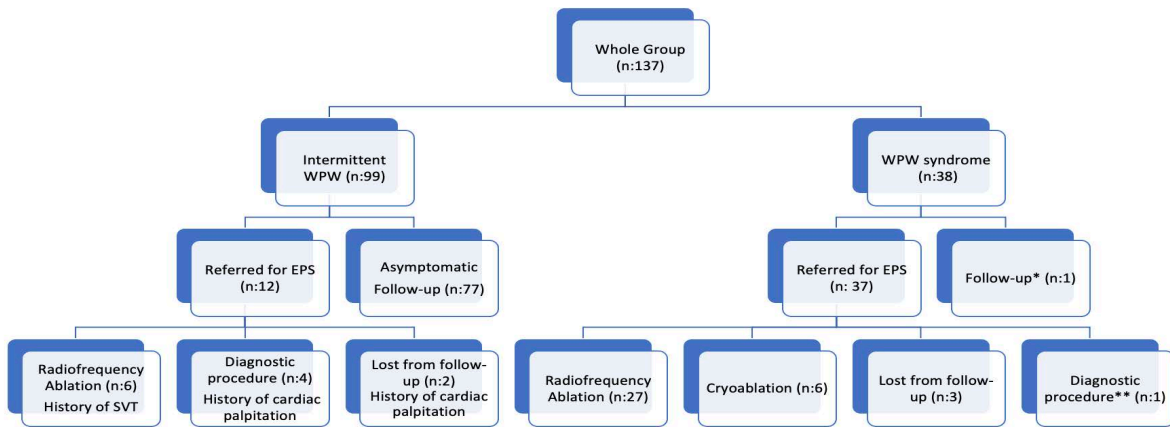
**Table 4.** Findings of Patients Referred For Electrophysiological Study

	Number (%)
<b><i>Reasons for admission at the time of diagnosis</i></b>	
Palpitation	14 (28.6)
Murmur	13 (26.5)
Chest pain	7 (14.3)
Follow-up for congenital heart disease	4 (8.2)
With suspected electrocardiogram anomaly	3 (6.2)
Syncope	2 (4.1)
Eligibility to sports	2 (4.1)
Chest pain + palpitation	1 (2)
Shortness of breath	1 (2)
Chest wall deformity	1 (2)
Follow-up for rheumatic heart disease	1 (2)
<b><i>12- Lead Electrocardiogram findings on admission</i></b>	
WPW pattern	37 (75.5)
Intermittent WPW pattern	9 (18.4)
Normal	3 (6.1)
<b><i>Echocardiography findings</i></b>	
Normal	33 (67.3)
Trivial mitral regurgitation	4 (8.2)
Mitral valve prolapse	2 (4.1)
Mitral valve prolapse with mild mitral insufficiency	1 (2)
Mitral valve prolapse with moderate mitral insufficiency	1 (2)
Operated secundum atrial septal defect	1 (2)
Ebstein anomaly (Glenn procedure)	1 (2)
Double inlet left ventricle (Fontan circulation)	1 (2)
Operated subaortic ridge and stenosis	1 (2)
Atrial septal defect	1 (2)
Ventricular septal defect	1 (2)
Mild mitral regurgitation	1 (2)
Mild mitral regurgitation and aortic regurgitation	1 (2)
Mild pulmonary stenosis	1 (2)
Mild septal hypertrophy	1 (2)
<b><i>24-hour Holter Electrocardiogram findings</i></b>	
WPW pattern	37 (75.5)
WPW pattern	35 (71.4)
WPW pattern with ventricular extrasystoles	2 (4.1)
Intermittent WPW pattern	12 (24.4)
Intermittent WPW pattern	9 (18.4)
Intermittent WPW pattern with ventricular extrasystoles	2 (4)
Intermittent WPW pattern with supraventricular extrasystoles	1 (2)

WPW: Wolff-Parkinson-White syndrome

not always clear-cut and nonspecific symptoms such as chest pain, palpitation, or syncope require clinical correlation which should be more subjective.

Most of the patients with WPW syndrome have structurally normal hearts but if an abnormality is present, it tends to be right sided.(5, 15) Ebstein anomaly is the congenital heart disease that is most commonly associated with WPW syndrome,



**Fig. 1.** Flowchart of Patients That Underwent Electrophysiological Study And Catheter Ablation

EPS: Electrophysiological study, SVT: supraventricular tachycardia

\*: Three years old patient with SVT, and follow-up decision due to the age of the patient

\*\* : History of surgery for subaortic ridge and stenosis, and the electrocardiographic findings were attributed to the previous cardiac surgery

and 10-20% of patients with Ebstein anomaly have WPW syndrome.(14, 16, 17) In our study secundum type atrial septal defect, mitral valve prolapse and mitral regurgitation were the most common cardiac anomalies. This is not surprising because cardiac defects without hemodynamic significance were also included in our study (except patent foramen ovale and very small patent ductus arteriosus) and these defects are also the most prevalent diseases in the general population. Congenitally corrected transposition of great arteries, pulmonary stenosis, and cardiac rhabdomyoma are the other cardiac anomalies commonly associated with WPW.(9) In our study, there existed one case with Ebstein anomaly and one case with pulmonary stenosis.

Patients with preexcitation have a tendency to develop supraventricular tachyarrhythmias and the overall incidence of arrhythmia is reported as 1% per year in patients with WPW pattern.(18) Another study reported the prevalence of SVT in patients with WPW syndrome as 20-30%.(9) In our study, in the whole group 8 (5.8%) cases had documented SVT. One of them had admitted with SVT and six of them were being followed up by other institutions with diagnosis of SVT. The other case was a 3-years old female patient who admitted with a murmur and had SVT during follow-up by our department.

As previously stated, WPW syndrome is characterized by the presence of one or more accessory pathways that lead to preexcitation and dysrhythmia. Although the most common type of

arrhythmia is AV reentrant tachycardia (%80); atrial flutter (<%5) and atrial fibrillation (%15-30) may also occur.(4, 19) Patients with WPW syndrome have a risk of SCD due to atrial fibrillation and subsequent ventricular tachycardia and ventricular fibrillation via accessory pathways. Although symptomatic patients have a higher risk of SCD, SCD may be the first presentation of asymptomatic and intermittent WPW syndrome. Because of the longer refractory period and a decreased conductive accessory pathway, patients with intermittent WPW are thought to have a lower risk of SCD. The risk of SCD is estimated as 0.1% in asymptomatic and 0.3% in symptomatic patients per year. It also accounts for 1.6% of all SCD among young athletes in the U.S.A.(3) Thus, early diagnosis, risk stratification, and appropriate management of patients with WPW syndrome are crucial. The EPS is safely used as a risk stratification tool and when necessary, catheter ablation is performed as a curative procedure. Herein, the point of debate for some authors is when to perform EPS in asymptomatic patients and intermittent WPW patients. The 2012 expert consensus statement by the Pediatric and Congenital Electrophysiology Society (PACES) and Heart Rhythm Society (HRS) on the treatment of asymptomatic, intermittent WPW recommends against routine EPS.(5) Due to the relatively high prevalence of asymptomatic and intermittent WPW and relatively low risk of SCD, catheter ablation is not recommended for everyone with intermittent

WPW. Although at first glance it may seem reasonable to perform catheter ablation to everyone with intermittent WPW; even if it is low, catheter ablation has its own risk of complications.(19) Kiger et al.(14) reported that although patients with intermittent WPW had longer accessory pathway effective refractory period, the frequency of high-risk accessory pathways was similar with persistent WPW. In a study, which retrospectively evaluated 60 children with WPW syndrome, 35% of children that were considered as low risk by presentation were found to have high-risk pathways by EPS.(20) It has also been reported that there is no significant difference in the frequency of high-risk pathways in children with WPW syndrome who presented with syncope, documented SVT, or without symptoms. (21) In our study all cases with persistent WPW had also persistent WPW on 24-h Holter ECG and all except one – because of young age- were referred for EPS. A total of 37 patients with persistent WPW were referred for EPS and 3 were lost from follow-up after referral for EPS. Of the remaining 34 patients, 33 had catheter ablation. The ECG findings of one case were attributed to the previous cardiac surgery for congenital heart disease and that was the only case with a diagnostic procedure and persistent WPW. Only 12 of 99 cases with intermittent WPW syndrome were referred for EPS because the remaining 77 patients were defined as low risk. Two of twelve cases referred for EPS were lost from follow-up after referral. Six of the cases were referred for EPS because of previously documented SVT, and all had catheter ablation. Four cases, who were referred because of complaints consistent with cardiac palpitation had a diagnostic procedure and were considered low risk. Radiofrequency ablation (RFA) was the preferred choice of treatment in 33 cases and the rest 6 cases had cryoablation. Recurrence of WPW pattern after catheter ablation was observed in only one male patient who had chest pain on initial admission and a structurally normal heart.

**Study Limitations:** This study is subject to the usual limitations of a retrospective study. The electrophysiological study was performed in various institutions and some of the patients were lost from follow-up after referral for EPS.

Wolff-Parkinson-White syndrome is the most common form of preexcitation. Although it is low, both persistent and intermittent WPW possess the risk of sudden cardiac death. Early diagnosis, risk stratification, and appropriate management of patients with WPW syndrome are crucial. The

electrophysiological study is used as a risk stratification tool and catheter ablation is a safe curative procedure in high-risk patients.

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