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Case Report

Recurrent Orthodromic SVT AVRT with Multiple Accessory Pathway in WPW Syndrome: Ablate All or Not?

Indra Jabbar Aziz[1*], Ardian Rizal[2]

[1]Brawijaya Cardiovascular Research Center, Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Brawijaya, Malang, Indonesia.
[2]Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Brawijaya, Malang, Indonesia.

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ABSTRACT

Background: Patients diagnosed with Wolff-Parkinson-White syndrome (WPW) have a relatively low, nevertheless persistent risk of unexpected mortality. However, this risk can be effectively mitigated by applying radiofrequency catheter ablation targeting the accessory pathway. It is difficult to accurately study predictors because only a minority of patients develop potentially malignant arrhythmias (MA) or passed away, despite essential risk variables being relatively well-known.[1] This study aimed to describe ablation in WPW syndrome.
Case presentation: A 16-year-old male presents with symptoms characterized by palpitations during intense physical exertion, followed by chest pain episodes. The physical examination, CXR, laboratorium, and echocardiography were all within the normal range; his ECG showed shortened PR interval and a delta wave with a positive delta wave at V1 and mostly positive at the inferior lead. He also has recorded ECG when the patient has a tachycardia event with SVT AVRT orthodromic pattern. He underwent EP Study and had several accessory pathways at the posteroseptal mitral annulus and anteroseptal tricuspid. The decision was made to perform ablation on the posteroseptal accessory pathway located at the mitral annulus. An electrophysiology (EP) investigation was conducted at the anteroseptal tricuspid annulus, which did not induce tachyarrhythmia.
Conclusion: Wolff-Parkinson-White syndrome is a congenital cardiac pathway formation, with not all accessory pathways causing tachyarrhythmias. Ablation therapy is necessary for patients with multiple pathways, with pathways with an ERP less than 250 being the only option.

1. Introduction

Wolff-Parkinson-White syndrome, often known as WPW syndrome, is characterized by symptomatic tachycardia, a short PR interval, and delta waves on the electrocardiogram (EKG). Although extremely rare, the disorder can be fatal if not diagnosed and treated promptly. The WPW pattern on an electrocardiogram can conceal ischemia alterations and may raise the risk of arrhythmia and, consequently, mortality.[2]

Wolff-Parkinson-White (WPW) syndrome is a congenital condition that impacts roughly 0.1% of individuals worldwide. During the early heart developmental stages, the atrial and ventricular myocardium become interconnected. Typically, as the atrial and ventricular septa invaginate and form the annulus fibrosus, they terminate all atrioventricular (AV) links with the exception of the AV node and the His bundle. The anatomical pathways causing ventricular preexcitation are termed persistent AV connections. These pathways can either entirely or partly circumvent the AV node and His bundle system.[3] In individuals with this syndrome, electrical impulses bypass the typical AV node pathway due to an anomalous conduction pathway. Diagnosis hinges on distinct ECG waveform patterns indicative of this irregular conduction. Yet, this alternative pathway can sometimes exhibit accelerated impulse transmission, resulting in occasional bouts of rapid heart rhythms and palpable heartbeats.[4] The Wolff-Parkinson-White syndrome, frequently abbreviated as WPW, represents a prevalent etiology of supraventricular tachycardia in the pediatric population. Distinctive electrocardiographic (EKG) manifestations associated with WPW encompass the presence of a delta wave, an expanded QRS duration exceeding 110 milliseconds, a PR interval less than 120 milliseconds, and inversed T wave configurations.[6]

2. Case Presentation

A 16-year-old male presents with symptoms characterized by palpitations during intense physical exertion, which are followed by chest pain episodes. The described chest pain appears within 3 to 5 minutes after the onset of the palpitations. Over the past several months, there has been a noticeable increase in the frequency of reported episodes of chest palpitations. He had no history of febrile illness,

* Corresponding author at: Brawijaya Cardiovascular Research Center, Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Brawijaya, Malang, Indonesia.
E-mail address: indra.jabbar.a@gmail.com (I.J. Aziz).

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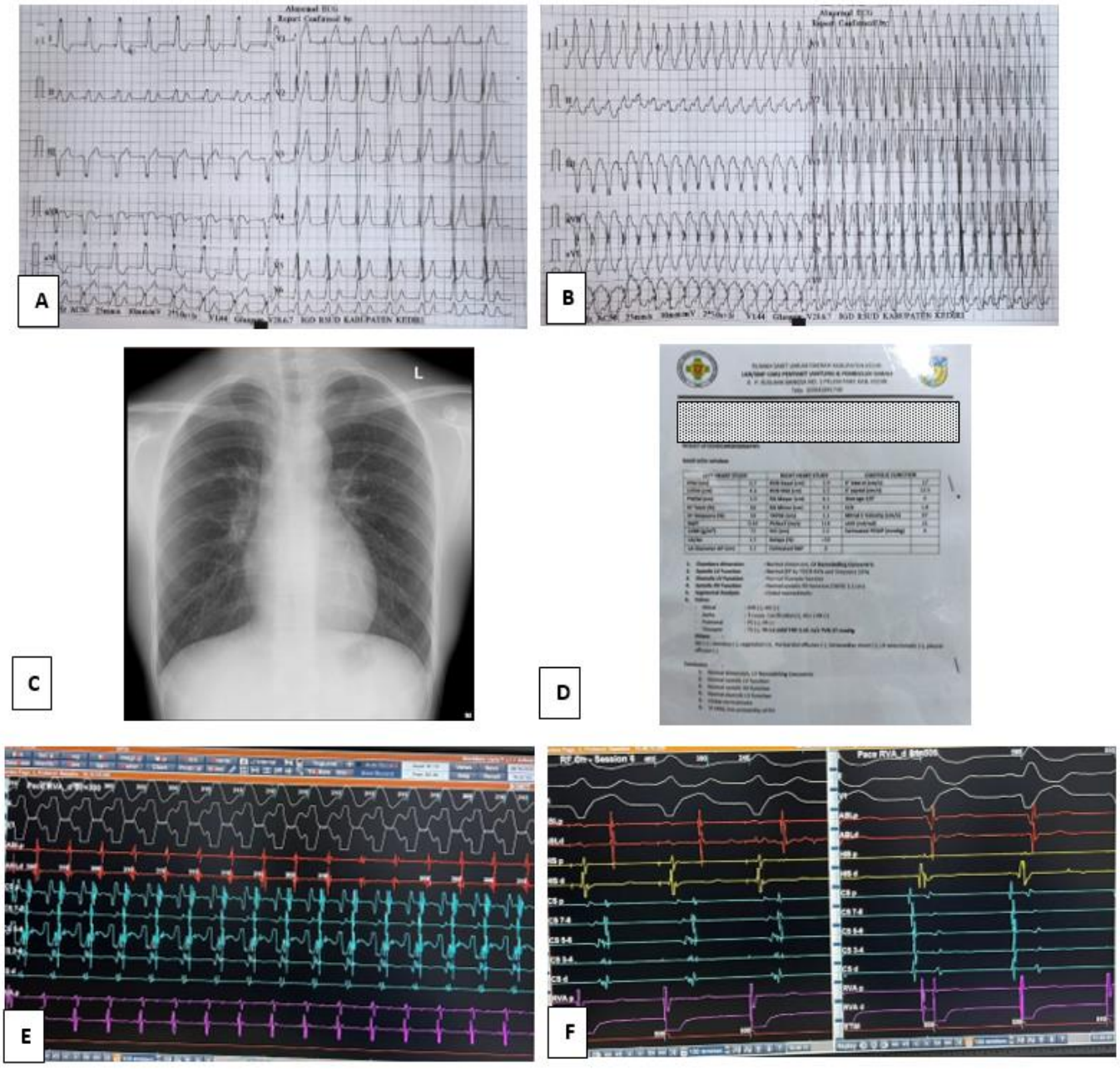

Figure 1. Patient Data : a. ECG at rest with delta wave, b. SVT AVNRT orthodromic ECG, c. Chest X-Ray, d. Echocardiography of Patient, e. EGM of SVT with eccentric pattern, f. Accesory Pathway ablation

dehydration, alcohol consumption, or any medication before having syncope. The patient's family did not have a medical history of a similar condition. The physical examination, CXR, laboratorium, and echocardiography were all within the normal range, and his ECG showed shortened PR interval and a delta wave with a positive delta wave at V1 and mostly positive at inferior lead. He also has recorded ECG when the patient has a tachycardia event with SVT AVRT orthodromic pattern (Figure 1). He underwent EP Study and had several accessory pathways at the posteroseptal mitral annulus and anteroseptal tricuspid. The study of electrophysiology (EP) revealed that eccentric ventricular activation with the right ventricular (RV) catheter had the most rapid activation of the V wave. Eccentric activation was observed during right ventricular pacing, with the fastest A wave detected in the lead CS 5.6. This finding suggests the presence of retrograde conduction in the posteroseptal accessory pathway. The decision was made to perform ablation on the posteroseptal accessory pathway located at the mitral annulus. Additionally, an electrophysiology (EP) investigation was conducted at the anteroseptal tricuspid annulus, with incremental pacing from HRA showing antegrade AP ERP 250 ms, which did not result in the induction of tachyarrhythmia; it was decided not to do ablation at this accessory pathway. The conclusion of the electrophysiology (EP) investigation was obtained, multiple accessories pathway with right accessories pathway antegrade only conduction and concealed left posteroseptal accessories pathway, SVT due to AVRT using left posteroseptal accessories pathway, successful ablation at left posteroseptal accessories pathway, right accessories pathway with ERP 250ms (Benign) did not induce SVT.

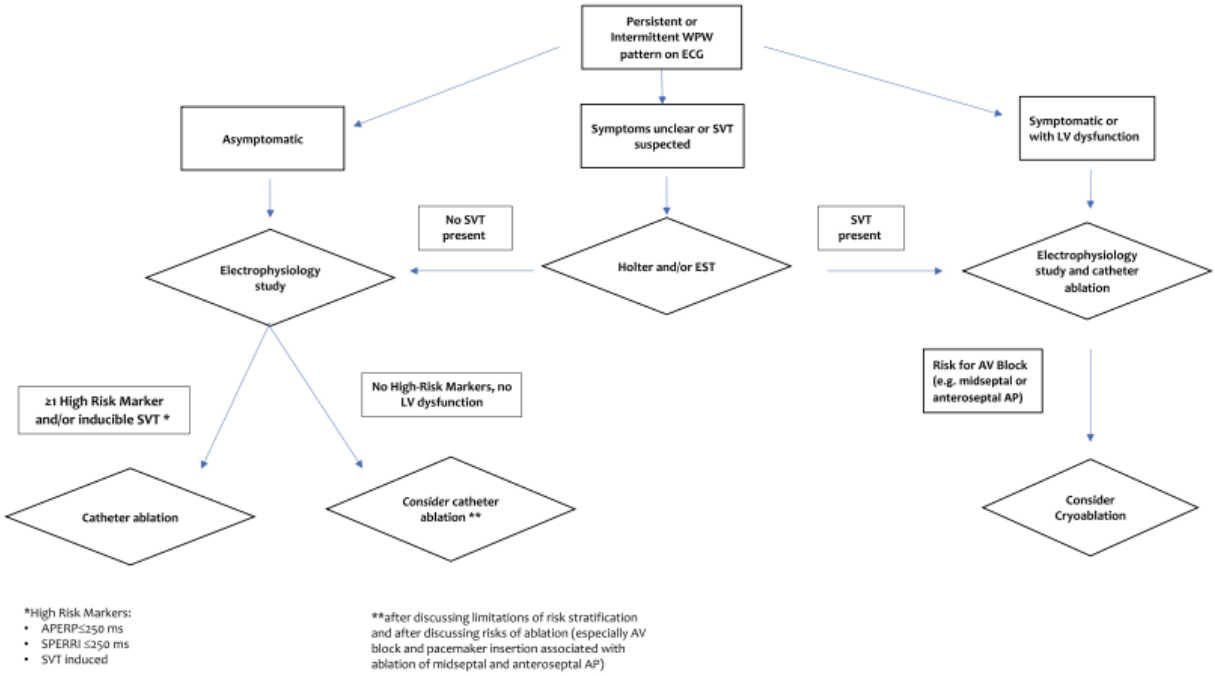

Figure 2. Flow diagram for management of a young person with WPW. [12]

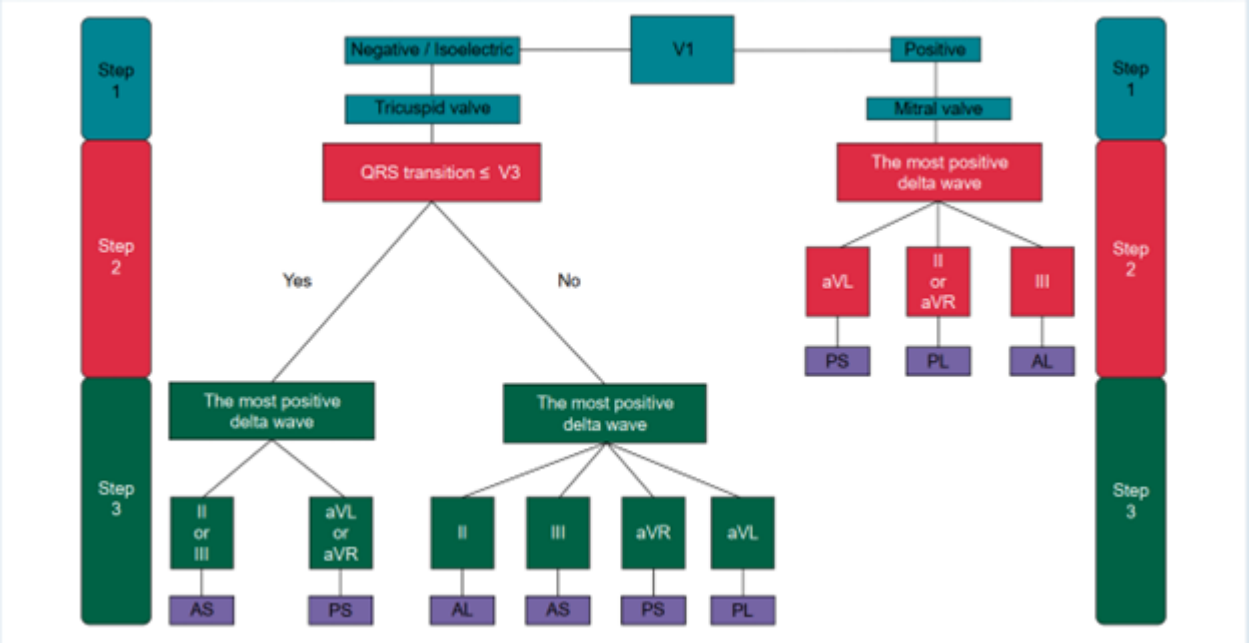

Figure 3. Flowchart for stepwise AP-identification.[15]

3. Discussion

The Wolff-Parkinson-White (WPW) syndrome is characterized by ventricular pre-excitation due to the existence of an extraneous conduction pathway. Clinical manifestations of this syndrome denote both symptomatic presentations and distinct electrocardiogram (ECG) deviations. Conversely, those with only the WPW electrocardiographic pattern might not display clinical symptoms. This syndrome was first elucidated in 1930 by Drs. Louis Wolff, John Parkinson, and Paul White from Boston, Massachusetts, and London, UK. Their characterization related to a syndrome typified by a diminished PR interval, derived from observations in eleven athletically inclined, ostensibly healthy subjects. Further insights in 1932 by Holzmann and Scherf pinpointed the syndrome's origin to an accessory conduction pathway.[7]

Accessory conduction tracts denote myocardial conduction channels traversing the atrioventricular (AV) groove at distinct positions along the annulus fibrosus on both cardiac aspects. Crucially, these tracts are absent within the mitral valve annulus sector between the right and left fibrous trigones. These pathways predominantly inhabit specific cardiac zones. The left lateral wall is frequently involved, representing around 58% of instances. Following this, the posterior septal zone accounts for nearly 24% of cases. The right lateral wall is involved in about 13% of cases, while the anterior septal area is relatively rare, constituting roughly 5% of instances. Unlike the AV node, which typically manifests gradual, incremental conduction, these accessory tracts display swift, non-decremental conduction coupled with an extended effective refractory period (ERP).[8]

Within a sinus rhythm, the electrocardiographic depiction reveals a hallmark configuration. This is delineated by (i) a diminished PR interval (sub-120 milliseconds), (ii) a nuanced initial segment of the QRS complex, often referred to as the 'delta wave', and (iii) a prolonged QRS duration (exceeding 120 milliseconds). Notably, most accessory pathways (APs) resulting in the WPW electrocardiographic

pattern arise in anatomically regular hearts. Infrequent familial iterations of pre-excitation linked with left ventricular (LV) hypertrophy and systemic disorders have also been documented.[9]

The presence of numerous accessory pathways within pediatric groups is an aspect scarcely illuminated in contemporary studies. An intensive analysis was conducted on medical records of 317 pediatric individuals diagnosed with Wolff-Parkinson-White syndrome, who underwent electrophysiological assessment and radiofrequency catheter ablation. Of these, 28 patients (constituting 9% of the aggregate) revealed multiple conduits, collectively summing to 64 pathways. Specifically, 21 patients had two pathways, six had three, and a lone case exhibited four conduits. In terms of distribution, the left lateral wall encompassed 22 pathways, the right lateral wall housed 19, the posteroseptal area had 17, and the anteroseptal region contained 6. Optimal treatment requires meticulous mapping of the principal accessory conduit and recognition of any supplementary tracts. Failing to pinpoint multiple pathways might lead to sustained tachycardia post-ablation. While multiple conduits have been acknowledged in various research undertakings, a focused investigation on the traits of these pathways in a younger population is yet to be undertaken.[10]

Approximately 10% of patients diagnosed with Wolff-Parkinson-White (WPW) syndrome present with antidromic supraventricular tachycardia (SVT), where the accessory pathway functions as the anterograde conduit. Conversely, the retrograde conduit might be represented by either the typical conduction mechanism or an additional accessory pathway. This clinical presentation is frequently termed a manifest accessory trajectory, delineated by evident pre-excitation on the electrocardiogram (ECG) and an augmented predisposition for the onset of antidromic supraventricular tachycardia (SVT) in the implicated cohort.[8] In scenarios involving pre-excited atrial fibrillation (AF), prudence is essential to preclude its transformation into atrial flutter and subsequently deter the potential initiation of 1:1 conduction. Beyond class IC antiarrhythmic agents, therapeutic options such as beta-blockers, diltiazem, or verapamil might be considered, especially when there's an absence of evident pre-excitation on the baseline electrocardiogram (ECG).[9]

In the recent two decades, catheter ablation has emerged and fortified its role as a widely accepted treatment modality for Wolff-Parkinson-White syndrome (WPW) patients. The procedure has continually shown commendable efficacy, characterized by high success rates and minimal complication occurrences. Contemporary guidelines issued by the European Heart Rhythm Association in conjunction with the American College of Cardiology Foundation/American Heart Association/Heart Rhythm Society emphasize catheter ablation as the recommended therapeutic approach for those exhibiting Wolff-Parkinson-White syndrome symptoms.[11]

Catheter ablation stands as the endorsed therapeutic intervention for individuals manifesting symptoms of recurrent atrioventricular reentrant tachycardia (AVRT) or pre-excited atrial fibrillation (AF). For those patients presenting with asymptomatic and sporadic episodes, it remains imperative to judiciously weigh the benefits against the potential risks of the invasive ablation procedure relative to sustained pharmacologic management in therapeutic decision-making (Figure 2). Procedures aimed at the atrioventricular pathway (AP) have showcased a commendable immediate success rate, while also maintaining a comparatively reduced complication frequency, contingent upon the pathway's precise location. Noteworthy complications post-ablation of septal accessory pathways (APs) encompass cardiac tamponade, which is seen in a range of approximately 0.13% to 1.1% of instances, and complete atrioventricular (AV) block, noted in about 0.17% to 2.7% of the patient population. When septal accessory pathways (APs) are proximal to the atrioventricular node (AVN), the electrocardiogram (ECG) typically reveals a positive delta wave in leads avF and AVL, combined with a slender positive delta wave in lead V1, juxtaposed with a predominantly negative QRS complex.[9]

The evolution and efficiency of catheter ablation, complemented by cutting-edge innovations like 3-dimensional (3D) mapping, cryothermal modalities, and intracardiac echocardiography (ICE), have substantially reshaped the electrophysiologists' perspective on Wolff-Parkinson-White syndrome (WPW). When executed by proficient specialists, the curative rate for Wolff-Parkinson-White syndrome (WPW) in the pediatric cohort stands at an impressive 95%. This shifts the risk-benefit equilibrium decidedly in favor of ablation as the therapeutic modality of choice. Within the electrophysiology community, there's a converging view that catheter ablation is warranted for symptomatic WPW patients meeting specific criteria in age or anatomical dimensions. However, the management of asymptomatic individuals remains a topic of active debate and scholarly scrutiny.[12]

Incomplete ablation of the accessory pathway (AP) can often stem from the existence of multiple pathways that elude detection during the electrophysiological study. When spaced by an interval of 1-3 cm, the presence of several accessory paths (APs) has been documented. Within a cohort of 250 patients, the most commonly identified tandem of multiple APs was the coexistence of right posteroseptal and right free wall conduits. The incidence of these multitudinous accessory pathways (APs) fluctuates between 3% and 13%. Afflicted individuals might harbor a heightened predisposition to conditions like supraventricular tachycardia (SVT), antidromic reentrant circuits, expedited conduction during atrial fibrillation (AF), and ventricular fibrillation.[13] It's well-acknowledged within the medical community that ventricular fibrillation (VF) can manifest due to sustained preexcited atrial fibrillation (AF), even in the absence of underlying structural heart abnormalities. This occurrence becomes especially pertinent when multiple accessory pathways (APs) coexist. Detecting the presence of numerous accessory pathways (APs) can pose diagnostic intricacies and is deemed a significant risk determinant for ventricular fibrillation (VF) in patients diagnosed with Wolff-Parkinson-White (WPW) syndrome.[14]

The EASY-WPW algorithm hinges on the scrutiny of QRS polarity and transition to pinpoint the most positive delta wave, or, in its absence, the most pronounced QRS complex. Operationally, the delta wave is perceived as the inaugural 20-40 milliseconds of the earliest QRS deviation (Figure 3).

In the context of a left-sided AP:

- Initial focus is on the V1 lead's polarity; a positive polarity signifies a left-sided action potential.
- Subsequent identification of the most salient delta wave comes next.
- Specific delineation of the atrioventricular pathway (AP) hinges on observing the dominant positive delta wave across leads II, III, aVR, and aVL. For instance, a pronounced positive delta wave in lead aVL indicates a posteroseptal locale, whereas its presence in leads II or aVR points towards a posterolateral orientation. Furthermore, a lead III positive delta wave alludes to an anterolateral position of the AP.

Considering a right-sided AP:

- Either a negative polarity or an isoelectric waveform in V1 earmarks a right-sided atrial potential.
- The consequent step, reliant on the QRS transition in precordial leads concerning the V3 lead's polarity, zeroes in on QRS transitions either ≤ or > V3.
- The AP's precise location is discerned by the predominant positive delta wave across leads II, III, aVR, and aVL. For instance, a positive delta wave in either lead II or III suggests an anteroseptal orientation, while its observation in leads aVR or aVL leans towards a posteroseptal pathway. If the QRS transition exceeds V3, the most pronounced delta wave in aVL hints at a posterolateral AP positioning, whereas in lead II, an anterolateral orientation emerges. Likewise, lead III suggests an anteroseptal placement, and finally, aVR insinuates a posteroseptal AP location.[15]

The clinical and electrophysiological features that are correlated with an increased probability of sudden cardiac death include:

- a younger age group
- the manifestation of AV-reciprocating tachycardia during electrophysiological investigations,
- the presence of multiple accessory pathways
- the capability of the accessory pathway to promote rapid conduction to the ventricles.

In evaluating the electrophysiological dynamics during atrial fibrillation (AF), two principal parameters emerge as pivotal: the Shortest Pre-excited RR Interval (SPERRI), which manifests an initial duration of less than 250 ms, and the Antegrade Effective Refractory Period (ERP) of the atrioventricular node (AVN) that is demarcated by a duration not exceeding 250 ms. The prevailing consensus within the cardiological community advocates for the prophylactic utilization of catheter ablation targeting the accessory pathway, especially under defined clinical scenarios. Such scenarios encompass instances where the SPERRI during AF does not surpass 250 milliseconds, the ERP during programmed atrial stimulation remains within 240 milliseconds or shorter, the accessory pathway is a tangible contributor to the manifest arrhythmic episode, the presence of any high-risk clinical markers, or the detection of multiple accessory pathways. [8] During exercise testing or subsequent to the administration of agents such as procainamide, propafenone, or disopyramide, the abrupt normalization of the PR interval and concomitant absence of the delta wave have been interpreted as surrogate markers of a reduced risk profile in non-invasive electrophysiological evaluations.[9]

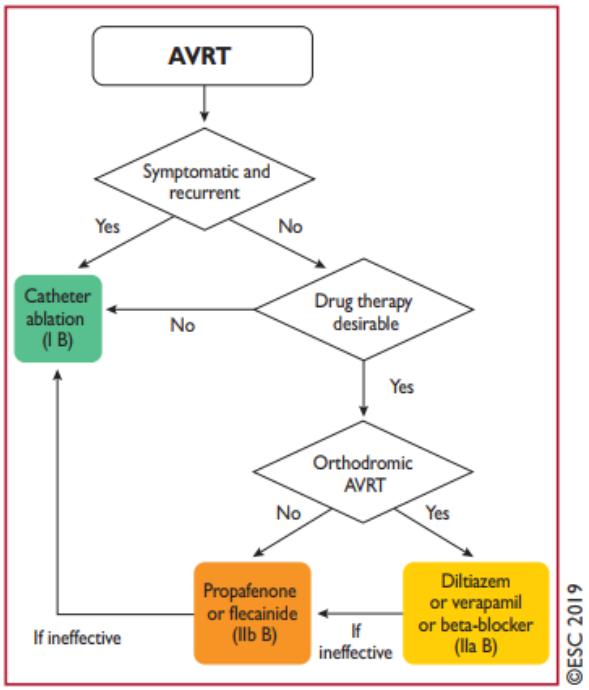

Figure 4. Chronic therapy of atrioventricular re-entrant tachycardia. atrioventricular re-entrant tachycardia.[9]

In assessing the approach to radiofrequency catheter ablation (RFCA) for Wolff-Parkinson-White (WPW) syndrome, it is pivotal to weigh the potential complications of the intervention against the risks of life-threatening arrhythmias if WPW remains unaddressed. The consensus guidelines from the American College of Cardiology (ACC), American Heart Association (AHA), and European Society of Cardiology (ESC) opine that the inherent risks of untreated WPW pose a greater threat than those tied to RFCA. Given its commendable efficacy and minimal risk profile, RFCA is thereby endorsed as the primary therapeutic choice for symptomatic WPW patients by these guidelines (Figure 4). However, a clear stance on prophylactic RFCA for asymptomatic WPW cases is still lacking. Pertinent literature suggests that a notable fraction, about 15%, of those with asymptomatic WPW might confront arrhythmic events later in life. It is paramount to note, though, that only a subset of this population is susceptible to ventricular fibrillation (VF) culminating in cardiac arrest. A substantial majority of asymptomatic individuals remain free of any clinical manifestations. Hence, electrophysiological studies (EPS) play a quintessential role in risk stratification for these asymptomatic WPW individuals, with RFCA being reserved for those discerned to harbor an elevated risk of detrimental arrhythmias.[16]

4. Conclusion

Wolff-Parkinson-White (WPW) syndrome is a variant of ventricular pre-excitation resulting from an extraneous conduction tract. This is demarcated by a reduced PR interval, a nuanced QRS segment, and an expanded QRS complex. Predominantly, accessory pathways (APs) in WPW are present in anatomically regular hearts. Unsuccessful ablation of these pathways can sometimes be ascribed to undetected conduits. The prevalence of multiple APs oscillates between 3% and 13%, elevating the risk factors like tachycardia, antidromic reentry, heightened conduction during atrial fibrillation, and ventricular fibrillation. For those manifesting with recurrent atrioventricular reentrant tachycardia or pre-excited atrial fibrillation, catheter ablation is advocated. Parameters signaling an augmented risk of sudden cardiac demise encompass a younger demographic, AV-reciprocal tachycardia, the presence of severalaccessory conduits, and the pathway's capability to expedite conduction to ventricles. Pertinent metrics encompass SPERRI, the minimal preexcited RR interval, and the effective refractory period (ERP) of the atrioventricular node, both falling below 250 ms.

5. Declaration

5.1 Ethics Approval and Consent to participate
Patient has provided written informed consent prior to involvement in the study.

5.2. Consent for publication
Not applicable.

5.3 Availibility of data and materials
Data used in our study were presented in the main text.

5.4 Competing interests
Not applicable.

5.5 Funding Source
Not applicable.

5.6 Authors contributions
Idea/concept: JAB. Design: JAB. Control/supervision: AR. Data collection/processing: JAB Analysis/interpretation: AR. Literature review: JAB, AR. Writing the article: JAB. Critical review: AR. All authors have critically reviewed and approved the final draft and are possible for the content and similarity index of the manuscript.

5.7 Acknowledgements
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References

1. Pappone C, Vicedomini G, Manguso F, et al. The natural history of WPW syndrome. *Eur Hear Journal, Suppl.* 2015;17:A8-A11. doi:10.1093/eurheartj/suv004

2. Iqbal AM, Ghazni MS, Mubarik A, Zubair N, Jamal SF. The Conversion of Wolff-Parkinson-White (WPW) Pattern into WPW Syndrome in the Presence of Ischemia: A Case Report. *Cureus*. 2019;11(2). doi:10.7759/cureus.4147

3. CAIN ME, LUKE RA, LINDSAY BD. Diagnosis and Localization of Accessory Pathways. *Pacing Clin Electrophysiol*. 1992;15(5):801-824. doi:10.1111/j.1540-8159.1992.tb06847.x

4. Ibrahim Ali Sherdia AF, Abdelaal SA, Hasan MT, et al. The success rate of radiofrequency catheter ablation in Wolff-Parkinson-White-Syndrome patients: A systematic review and meta-analysis. *Indian Heart J*. 2023;75(2):98-107. doi:10.1016/j.ihj.2023.02.001

5. Dalili M, Kargarfard-Jahromi MR, Kamali F. Successful catheter ablation in infants with multiple accessory pathways and incessant arrhythmia. *Iran Hear J*. 2021;22(2):119-123.

6. Chen M, Gomaa H, Cortez-resendiz A, Martin C, Francis A, Bellon A. Case Report Quetiapine and Wolff-Parkinson-White Syndrome. 2020;2020(Table 1).

7. Balta A, Ceasovschih A, Șorodoc V, et al. Broad Electrocardiogram Syndromes Spectrum: From Common Emergencies to Particular Electrical Heart Disorders. *J Pers Med*. 2022;12(11). doi:10.3390/jpm12111754

8. Vijay Y, Sanjeev T, Ratna Mani G, et al. A Wolff-Parkinson-White (WPW) Electrocardiographic Pattern in Asymptomatic Patient – State-of-the-Art-Review. *J Cardiol Cardiovasc Med*. 2022;7(2):045-052. doi:10.29328/journal.jccm.1001132

9. Brugada J, Katritsis DG, Arbelo E, et al. 2019 ESC Guidelines for themanagement of patients with supraventricular tachycardia. *Eur Heart J*. 2020;41(5):655-720. doi:10.1093/eurheartj/ehz467

10. Weng KP, Wolff GS, Young ML. Multiple accessory pathways in pediatric patients with Wolff-Parkinson-White syndrome. *Am J Cardiol*. 2003;91(10):1178-1183. doi:10.1016/S0002-9149(03)00263-7

11. Fujino T, De Ruvo E, Grieco D, et al. Clinical characteristics of challenging catheter ablation procedures in patients with WPW syndrome: A 10 year single-center experience. *J Cardiol*. 2020;76(4):420-426. doi:10.1016/j.jjcc.2020.05.003

12. Etheridge SP, Gakenheimer-Smith L, Asaki SY, Niu M. Asymptomatic Wolff-Parkinson-White Syndrome: An Ounce of Prevention Is Worth the Risk of Cure. *Curr Cardiol Rep*. 2023;25(6):543-551. doi:10.1007/s11886-023-01879-6

13. Helm RH, Varkey SC, Karnik AA. Differential effective refractory period as a useful marker of multiple accessory pathways. *J Arrhythmia*. 2019;35(2):296-299. doi:10.1002/joa3.12162

14. Pappone C, Santinelli V, Rosanio S, et al. Usefulness of invasive electrophysiologic testing to stratify the risk of arrhythmic events in asymptomatic patients with Wolff-Parkinson-White pattern: Results from a large prospective long-term follow-up study. *J Am Coll Cardiol*. 2003;41(2):239-244. doi:10.1016/S0735-1097(02)02706-7

15. El Hamriti M, Braun M, Molatta S, et al. EASY-WPW: a novel ECG-algorithm for easy and reliable localization of manifest accessory pathways in children and adults. *Europace*. 2023;25(2):600. doi:10.1093/EUROPACE/EUAC216

16. Liu A, Pusalkar P. Asymptomatic Wolff-Parkinson-White syndrome: Incidental ECG diagnosis and a review of literature regarding current treatment. *BMJ Case Rep*. Published online 2011:2-5. doi:10.1136/bcr.05.2011.4192