Atrial Fibrillation in WPW Syndrome

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Background: In large-scale general population studies involving children and adults, the prevalence of WPW is estimated to be 1–3 in 1000 individuals. At present, it is estimated that approximately 65% of adolescents and 40% of individuals over 30 years with WPW pattern on a resting ECG are asymptomatic. An incidence of 4.5 episodes of sudden death, including resuscitated sudden cardiac death (SCD), per 1000 patient-years was recently reported. The mechanism of sudden cardiac death in patients with WPW is thought to be associated with atrial fibrillation.

Case Illustration: We reported a case of 45 year old female came with unstable irregular wide complex tachycardia in the form of pre-excited AF. Cardioversion successfully terminate the tachyarhythmias. ECG in sinus rhythm revealed WPW pattern ECG. Patient then referred for catheter ablation. Successful ablation was done resulting a normal pattern ECG.

Conclusion: AF in WPW syndrome could lead to devastating events such as cardiac arrest. ECG recognition at the first place is very important for early management. Early stratification and management in patients with WPW is important to diminished the risk of AF occurrence and SCD.

Keywords: atrial fibrillation, WPW syndrome, accessory pathway, ablation

Ilustrasi Kasus : Sebuah kasus wanita usia 45 tahun datang dengan takikardia QRS lebar disertai tanda instabilitas hemodinamik. EKG menunjukan amban fibrilasi atrial dengan preeksitasi. Kardioversi elektrik sukses menghentikan takikardia. EKG setelah kardioversi menunjukkan pola WPW dengan irama sinus. Ablasi kateter lalu dilakukan pada pasien dan sukses menghilangkan gelombang delta pada EKG.

Kesimpulan : Fibrilasi atrial pada sindrom WPW dapat mengakibatkan henti jantung. Identifikasi EKG saat takikardia sangat penting untuk menentukan manajemen awal. Stratifikasi resiko dini dan manajemen pada pasien WPW sangat penting untuk mengurangi resiko terjadinya kematian jantung mendadak.

Kata kunci : fibrilasi atrial, sindrom WPW, jalur aksesori, ablasi

Introduction

Since Wolff, Parkinson, and White published their sentinel paper in 1930, much has been learned about the anatomy, electrophysiology, and natural history of accessory connections exhibiting antegrade conduction.⁴ Wolf Parkinson White (WPW) pattern refers to the constellation of ECG abnormalities related to the presence of an atrioventricular accessory pathway (short PR interval, delta wave) in asymptomatic patients. WPW syndrome refers to a WPW ECG pattern associated with tachyarrhythmias. In large-scale general population studies involving children and adults, the prevalence of WPW is estimated to be 1–3 in 1000 individuals. Familial studies have shown an incidence of 5.5 in 1000 among first-degree relatives following an index case of WPW. At present, it is estimated that approximately 65% of adolescents and 40% of individuals over 30 years with WPW pattern on a resting EKG are asymptomatic.³ Most worrisome are the uncommon presentations of syncope or aborted sudden cardiac arrest as the first manifestation of WPW syndrome.⁴ The possible mechanisms are presented that may produce ventricular fibrillation in patients associated with atrial flutter or fibrillation.³ An incidence of 4.5 episodes of sudden death, including
resuscitated sudden cardiac death (SCD), per 1000 patient-years was recently reported in a prospective study of asymptomatic adults with WPW followed for a mean of 38 months.4

**Case Illustration**

A 45 year old female came to NCCHK outpatient ward with a chief complain of history of palpitation. Forty days before admission patient felt sudden palpitation while resting. Palpitation accompanied with dizziness with no syncope, chest pain, or dyspnea. She never felt this kind of symptoms before. Patient then came to RSUD Sanglah. The ECG monitor showed irregular wide complex tachyarrhythmias (Figure 1) with rate of 250 x/min without signs of hemodynamic instability. Twelve lead ECG (Figure 2) showed atrial fibrillation, QRS rate 250-300 x/min QRS axis left axis deviation 60 deg, QRS duration 0.14", and the presence of delta wave. Then, patient suddenly loss of consciousness. Electrical cardioversion was performed and the rhythm converts to sinus rhythm. At sinus rhythm, 12 lead ECG showed WPW pattern. Patient was discharged after 16 days of hospitalization and referred to NCCHK with diagnosis of WPW syndrome. She was given verapamil 2 x 40 mg, aspirin 1 x 80 mg, and clopidogrel 1 x 75 mg.

At the outpatient clinic, patient did not felt any palpitation again since the first day at Sanglah. History of syncope, near syncope, or seizure was denied. No history of dyspnea on effort, orthopnea, paroxysmal nocturnal dyspnea, and leg swelling. History of anginal chest pain was also denied. No risk factor for coronary artery disease was noted. Physical examination at the outpatient clinic showed blood pressure of 108/54 mmHg with heart rate of 71 x/min. Non anemic conjunctiva, not icteric sclerae. Normal JVP. Cardiac examination shows normal S1S2, no additional heart sounds nor murmur. Pulmonal auscultation shows vesicular sounds without rales, crackles nor wheezes. Extremities were warm, no edema. ECG (Figure 3) showed normal sinus rhythm, QRS rate 71x/minutes, QRS axis deviated 55 degree to the left, normal p wave with shortened PR interval (0.10”). QRS duration is normal (0.08”). Delta wave was present, biphasic T wave was noted at I, avL, V5-6. Echocardiography showed normal findings with EDD 48 ESD 26, ejection fraction of 76 %, normal diastolic function, and no anatomic defects. The patient then planned to have accessory pathway (AP) ablation. Patient then underwent AP ablation. During programmed atrial stimulation, atrial fibrillation occurs and the patient became unstable. In cardioversion preparation, the rhythm spontaneously converted to sinus rhythm. Electrophysiological studies showed shortened AV interval with posterior RV region as the shortest interval. AV nodal effective refractory period (AVN ERP) measured at 340 ms, atrial ERP at 230 ms, AP ERP retrograde of 310 ms, RV ERP of 220 ms. Multiple radio frequency ablation (RFAs) at right posterior successfully eliminate the delta wave (Figure 4). Retrograde block was also seen post RFAs.

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**Figure 1.** ECG monitor showed wide complex atrial fibrillation

**Figure 2.** Twelve lead ECG showed atrial fibrillation, with pre-excitation pattern

**Figure 3.** ECG showed normal sinus rhythm, with WPW pattern
Patient was discharged at the second day post ablation without palpitation and diminished delta wave (Figure 4).

Discussion

Atrioventricular accessory pathway (AP) is strands of working myocardial cells connecting atrial and ventricular myocardium across the electrically insulating fibrofatty tissues of the AV junction bypassing the atrioventricular node–His-Purkinje system (AVN-HPS). From a developmental aspect, accessory pathway are the result of an incomplete regression of the embryonic muscular continuity between atrial and ventricular myocardium and can occur at different locations around the tricuspid and mitral anulus. The genetic basis of this abnormalities has not been elucidated but it has an association with hypertropic cardiomyopathy or congenital heart defect such as Ebstein anomaly or tricuspid valve defects.

Accessory pathway are usually very thin muscular strands (rarely thicker than 1 to 2 mm) but can occasionally exist as broad bands of tissue. The atrioventricular (AV) AP can run in an oblique course rather than perpendicular to the transverse plane of the AV groove. As accessory pathway is working myocardial cells, impulse conduction is caused by the rapid inward sodium current, similar to normal His-Purkinje tissue, atrial, and ventricular myocardium. Thus, APs generally exhibit “all or none” conduction behavior during electrophysiological evaluation. Rapid non-decremental conduction up to the point of refractoriness is the norm and is exhibited during antegrade and retrograde conduction, especially when competing AV nodal conduction is absent. In contrast to the AVN, which depends on the slow inward calcium current for generation and propagation of its action potential, exhibits what has been called decremental conduction in which the conduction time of the impulse propagating through the AVN increases as the atrial cycle length (CL) shortens. Thus, AV conduction is more rapid through the AV AP than through the AVN, a difference that is increased at a fast heart rate. This difference has potentially great clinical importance. A primary function of the AVN is to limit the number of impulses conducted from the atria to the ventricles, which is particularly important during fast atrial rates (e.g., AF or atrial flutter) when only a fraction of impulses are conducted to the ventricles, whereas the remainder are blocked in the AVN. However, in the presence of non-decrementally conducting APs, these arrhythmias can lead to very fast ventricular rates that can degenerate into VF. The majority (approximately 60%) of AV APs conduct both anterogradely and retrogradely. In some instances, when the antegrade refractoriness of the accessory pathway is particularly long or if the pathway fails to permit such conduction, it may remain concealed and only manifest when retrograde propagation occurs over echo beats or reentrant circuit. APs that conduct only in the retrograde direction occur more frequently, with an incidence of 17% to 37%. Thus, AV conduction is more rapid through the AV AP than through the AVN, a difference that is increased at a fast heart rate. This difference has potentially great clinical importance.

WPW pattern in Minesotta code of ECG is listed as 6-4-1 and characterized by normal P-waves, short P-R interval <0.12 second, prolonged QRS duration ≥0.12 second with a slurred upstroke to the QRS complex, and R peak duration ≥0.06s. All these conditions must coexist in the same beat and also be
present in the majority of beats in any of the following leads: I, II, aVL, V4, V5, or V6. Those findings occurred in a patient that fully pre-excited. The duration of the PR interval is affected by the degree of preexcitation. In fully preexcited complexes, the duration of the PR intervals equals the duration of the P wave or its initial portion. In most instances it is 0.06–0.11 second. When the ventricles are depolarized entirely by the impulse conducted through the accessory pathway (AP), the QRS duration is increased, usually to 0.11 to 0.16 second. In some series, however, the QRS duration was less than 0.10 second in almost one third of the cases, presumably due to fusion between preexcited and normally conducted complexes. The sum of the PR and QRS intervals usually remains within the normal range. The delta wave is the most important finding in the WPW pattern. It depicts slow conduction through the AP and ventricular myocardium between the site of AP insertion and the site at which ventricular activation proceeds via the rapidly conducting Purkinje system. The duration of the delta wave varies between 0.02 and 0.07 second. Theoretically, the delta wave should be present in all leads, but it may become isoelectric and be easily overlooked in the leads with the lead axis, which is nearly perpendicular to the initial QRS forces. The altered sequence of ventricular activation in patients with the WPW pattern results in secondary repolarization abnormalities. Most commonly, the direction of the ST segment displacement and the T wave polarity are opposite to the direction of the delta wave and the major deflection of the QRS complex.

Several algorithms have been developed for localization of the AP using the delta wave polarity. The delta wave vector is helpful, especially when maximal preexcitation is present. However, during NSR, the QRS is usually a fusion and total preexcitation is not present, which limits the accuracy of ECG localization of the AP. In using any localization algorithm, one must be aware of the portion of the QRS complex on which the algorithm is based. Some algorithms use only the first 20 to 60 milliseconds of the delta wave, whereas others are based on the morphology or polarity of the entire QRS complex. ECG algorithms for AP localization are most accurate for the diagnosis of left free wall APs compared with all other locations, achieving at least 90% sensitivity and almost 100% specificity. As opposed to left free wall APs, the ECG diagnosis of right free wall APs is the least accurate and least consistent among algorithms, with a sensitivity of 80% to 90% and a specificity of 90% to 100%. One of the algorithm that is used is displayed below.

The patient came to our patient clinic of NCCHK with history of palpitation. She also had a history of syncope and was admitted to the hospital before. The surface ECG showed a sinus rhythm with shortened PR interval. Delta waves are clearly noted on this ECG but the QRS complex was less than 120 ms. We can conclude that this patient has pre-excitation through the accessory pathway but the excitation wave front is a fusion between AP conduction and normal AV node conduction. From the ECG was also shown that the location of APs closely to right posterior or right posterolateral wall of the RV. But due to its less accuracy of detecting right sided AP, the exact location of the AP could be misleading.

The presence of conducting AP and AV node that act as 2 conduction path potentially induce reentry circuit around the atrium and ventricle. AVRTs (Figure 7) are the most common (80%) tachycardias.
associated with WPW syndrome. AVRT is divided into orthodromic and antidromic according to the direction of conduction in the AVN-HPS.

In orthodromic AVRT, the AVN-HPS serves as the anterograde limb of the reentrant circuit whereas an AV BT serves as the retrograde limb. Approximately 50% of APs participating in orthodromic AVRT are visible in surface ECG and 50% are concealed. The ECG showed a narrow QRS complex due to normal ventricular excitation wave front. Orthodromic AVRT accounts for approximately 95% of AVRTs and 35% of all paroxysmal supraventricular tachycardias. Permanent junctional reciprocating tachycardia (PJRT) is an orthodromic AVRT mediated by a concealed, retrogradely conducting AV AP that has slow and decremental conduction properties. This tachyarrhythmias is usually incessant.

In antidromic AVRT, an AV AP serves as the anterograde limb of the reentrant circuit. Consequently, the QRS complex during antidromic AVRT is fully preexcited. During classic antidromic AVRT, retrograde VA conduction occurs over the AVN-HPS but in some cases it can use another AP. Antidromic AVRT occurs in 5% to 10% of patients with WPW syndrome. Up to 50% to 75% of patients with spontaneous antidromic AVRT have multiple APs whether or not they are used as the retrograde limb during the tachycardia.

Atrial tachycardia (AT), atrial flutter (AFL), AF, and atrioventricular nodal reentrant tachycardia (AVNRT) can coexist in WPW syndrome. In these settings, the AP serves as a bystander route for ventricular or atrial activation, and is not required for the initiation or maintenance of the arrhythmia. Dual AVN physiology has been reported in 8% to 40% of patients with WPW syndrome.

AFL is the most common (60%) regular preexcited tachycardia in patients with WPW syndrome. This relationship can be mediated by contraction-excitation feedback into the atria during the AVRT. Depending upon the various refractory periods of the normal and pathological AV conduction pathways, AFL potentially can conduct 1:1 to the ventricles during a preexcited tachycardia making the arrhythmia difficult to distinguish from ventricular tachycardia.

Atrial fibrillation is not uncommon in patients with WPW and has been noted to occur in 11.5% to 39%. This dysrhythmia is usually precipitated by an episode of AV reentrant tachycardia, but they may also occur alone. Atrial fibrillation in the presence of WPW (Figure 8) is potentially dangerous in that a rapid ventricular response can be generated from nondecremental conduction down the AP and can degenerate into ventricular fibrillation. The mechanism of sudden cardiac death in patients with WPW is thought to be associated with atrial fibrillation or atrial flutter due to that mechanism. An incidence of 4.5 episodes of sudden death, including resuscitated SCD, per 1000 patient-years was recently reported in a prospective study of asymptomatic adults with WPW followed for a mean of 38 months. In two separate combined adult and pediatric studies involving 386 patients followed for 10 years, 15% developed spontaneous AF, with 4 having SCD (ages 20, 31, 34, 71 years). A recent study of 709 patients (age 34-16 years) with WPW undergoing electrophysiologic (EP) testing, SC AF was the initial presentation in 44 patients, AF was inducible in 17% (42/248) of asymptomatic patients compared to 25.5% (24/94) with a history of syncope and spontaneous AF was observed in 10% of patients. The mechanisms by which AVRT precipitates AF are not well understood. Intra atrial conduction disturbances, creating an electrophysiological substrate conducive to AF initiation, but the diminished rate of AF in patients after AP ablation could show that AP also play a role in AF initiation. In recent review by Osmar et al, stated that the existence of a retrograde multiple or multifibre AP, abnormally prolonged and fractionated atrial endocardial electrograms suggest an intrinsic atrial vulnerability that could induce AF.

There are several characteristic findings on the ECG in patients with AF conducting over a AP, so-called

Figure 8. Schematic figures shows AF in WPW.
preexcited AF. The rhythm is irregularly irregular, and can be associated with very rapid ventricular response caused by the nondecremental anterograde AV conduction over the AP. Although the QRS complexes are conducted aberrantly, resembling those during preexcited NSR, their duration can be variable and they can become normalized. This is not related to the RR interval, but rather is related to the variable relationship between conduction over the AP and AVN-HPS. Preexcited and normal QRS complexes often appear. This can result from concealed retrograde conduction into the AP or the AVN. The QRS complex during preexcitation is a fusion of the impulse that preexcites the ventricles caused by rapid conduction through an AP and of the impulse that takes the usual route through the AVN. The number of impulses that can be transmitted through the AP and the amount of preexcitation depend on the refractoriness of both the AP and AVN. The shorter the ERP of the AP, the more rapid is anterograde impulse conduction and, because of more preexcitation, the wider the QRS complexes. Patients who have a AP with a very short ERP and rapid ventricular rates represent the group at greatest risk for development of VF. The patient’s ECG as shown in Figure 1 and Figure 2 clearly demonstrate a irregular wide complex tachycardia. Several differential diagnosis can be made such as AF with bundle branch block, polymorphic VT, torsades pointes, and AF with pre-excitation syndrome. The absence of typical bundle branch block patterns exclude aberrantly conducted AF. Stierer et al. analyzed the morphologic differences between polymorphic VT and supraventricular tachycardia with anterograde conduction over an accessory pathway (preexcited tachycardia) with a QRS complex > 0.12 second in 149 consecutive VTs and 149 consecutive preexcited regular tachycardia. They found that the following characteristics were specific for VT but were absent with preexcited tachycardia: (1) predominantly negative QRS complexes in leads V4–V6; (2) presence of a QR complex in one or more leads V2–V6; and (3) more QRS complexes than P waves (when AV dissociation was present during VT). The sensitivity and specificity of these three markers of VT were 75 percent and 100 percent, respectively. The 3 characteristics of VT did not encountered in the ECG.

In AF with bundle branch block, the presence of delta waves in some of the complex, differ morphology at each complex, and an unusually high ventricular rates indicating AF with pre-excitation pattern.

The electrophysiology study can play a major role in characterizing the significance of bypass tracts and developing appropriate therapy. The electrophysiology study can be used to determine the characteristics of the pathways of a macroreentrant circuit and can help to select effective therapy for macroreentrant arrhythmias. In addition, the electrophysiology study can help to determine the potential of the bypass tract to mediate lethal arrhythmias (the potential for lethal arrhythmias is not an issue in patients with concealed bypass tracts, since it depends on efficient antegrade conduction). Finally, and most importantly, the electrophysiology study can be used to precisely localize and ablate the bypass tract. Preexcitation is associated with a short His bundle–ventricular (HV) or H-delta interval during NSR. The HV interval can even be negative or the His potential can be buried in the local ventricular electrogram. Incremental rate atrial pacing and progressively premature AES produce decremental conduction over the AVN (but not over the AP), increasing the degree of preexcitation and shortening the HV interval, until the His potential is inscribed within the QRS. The localization of AP from EP study can be done by finding the earliest atrial or ventricular potential relative to delta wave, the shortest AV or VA interval at the site localize the AP.

The ECG during preexcited AF affords a “true” assessment of the anterograde characteristics of the accessory pathway. The measurement of the Shortest Pre-Excited R-R Interval (SPERRI) has been used to determine accessory pathway properties; however, much of the literature is based on invasive studies. A SPERRI of 220–250 ms and especially less than 220 ms is more commonly seen in patients with WPW who have experienced cardiac arrest. The sensitivity and negative predictive value of a SPERRI <250 ms is high, but the specificity and positive predictive value are appreciably lower. Thus, SPERRI measurement along with history of symptomatic tachycardia, multiple bypass tract, and Ebstein anomaly confers a high risk profile for WPW developing SCD. During EP study, this patient has induced AF and become hemodynamically unstable. This classifies the patient in high risk patient to SCD. The SPERRI from the presenting ECG while AF shows interval < 250 ms. PACES/HRS consensus statement advise the use of new algorithm (Figure 9) to screen asymptomatic patient with WPW syndrome.

For acute management of AF with WPW, caution is advised against AVN blocking agents for
the treatment of preexcited tachycardias occurring in patients with AT, AFL, or AF with a bystander AP. AV nodal blockage can cause selective conduction to the AP resulting a very fast QRS rate. When drug therapy fails or hemodynamic instability is present, electrical cardioversion should be considered. Procainamide (30 mg/min, maximal dose 17 mg/kg) has traditionally been the treatment of choice for patients who are stable with AF WPW. By blocking fast inward Na current and outward K current, procainamide has been shown to prolong the ERP of atrial, ventricular, and AP tissue as well as slow antegrade and retrograde conduction in the AP. Because of the potential for severe hypotension with rapid IV administration, procainamide requires a somewhat slow rate of infusion and also has a relatively slow onset of action, not reaching therapeutic blood levels for 40 to 60 minutes. Amiodarone (150 mg IV over 10 minutes) is another agent used by practitioners for chemical conversion of patient’s with a wide complex tachycardia and is quoted in the 2005 American Heart Association Advanced Cardiac Life Support guidelines as the antiarrhythmic to consider in WPW AF. The consequences of acute rapid IV amiodarone administration modifies sinus and AV node properties with little, if any, effect on fast-channel tissues (ie, APs). This observation may be explained by the pharmacokinetic fact that accumulation of amiodarone’s desethyl metabolite is responsible for much of the long-term effects on fast-channel tissues. Administration of IV amiodarone to patients in AF has been shown to cause acceleration of the ventricular rate and degeneration into ventricular fibrillation. Taking these factors into consideration, the use of IV amiodarone for the treatment of patients identified as having WPWAF should be made with caution. Ibutilide is a reasonable agent for management of AF in patients with WPW. As a class III antiarrhythmic agent, ibutilide prolongs the action potential duration and refractoriness by enhancing the slow inward sodium current and blocking delayed-rectifier outward K current, resulting in QT interval prolongation. It is given at a dosage of 1 mg (0.01 mg/kg for patients < 60 kg) over 10 minutes and can be repeated once after a 10-minute period. It has a very short half-life of 4 hours and its dosing requires no concern for hepatic or renal function, safe in elderly patients, and it is very rapid in action, with a mean conversion time of approximately 20 minutes. Several case reports have had excellent results with ibutilide in treating wide complex AF and AF WPWAF. With a faster onset of action, a better conversion rate in patient’s with AF/ flutter, prolongation of the AP refractory period, and stable blood pressure profile, ibutilide may be superior to procainamide for chemical conversion of AF WPW. The primary concern with ibutilide use is the development of torsade de pointes due to prolongation of the QT interval. Patients who present with AF WPW, however, usually are young and have normal ventricular function, therefore placing them at a lower risk for ibutilide-induced arrhythmias. For chronic management, catheter ablation is also considered first-line therapy (class I) for patients with WPW patterns that symptomatic or develop AF. This patient came to ED with unstable wide complex tachyarrhythmias. The cardioversion successfully converts the rhythm. The chronic management chosen to this patient is catheter ablation that was successfully done.

**Summary**

We presented a case of 45 year old female with WPW syndrome have coincides atrial fibrillation. An EP study revealed an inducible AF during stimulation with unstable hemodynamic. Catheter ablation was performed successfully. AF in WPW syndrome could
lead to devastating events such as cardiac arrest. ECG recognition at the first place is very important for early management. Early stratification and management in patients with WPW is important to diminished the risk of AF occurrence and SCD.

References