

Concealed Accessory Pathway in Late Presentation Wolff-Parkinson-White Syndrome

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Background

During normal electrical conduction of the heart, an electrical impulse begins at the sino-atrial (SA) node and spreads across the right and left atria before passing through the atrioventricular (AV) node. It then passes down to the ventricles via the right and left bundle branches (of His) and finally the Purkinje fibres.

This pattern of electrical conductance creates the familiar tracing on an electrocardiogram (ECG) of a P-wave (representative of atrial depolarisation), followed by a flat section, the P-R interval; representing a delay as the electrical activity passes through the AV node, followed by a QRS wave representing ventricular depolarisation.

The P-R interval is normally 0.12 to 0.20 seconds in duration and the QRS complex is normally less than 0.12 seconds (Figure 1). This is important to note as in Wolff-Parkinson-White (WPW) syndrome, characteristic ECG changes may show a shortened P-R interval and widened QRS complex. These ECG changes may or may not be present at baseline.

In 1930, Louis Wolff, John Parkinson, and Paul Dudley White published an article in the American Heart Journal describing 11

patients with functional bundle branch block, a short P-R interval and arrhythmias (Wolff et al., 1930). To date, the authors now lend their names to a syndrome that encompasses ECG changes revealing of an accessory pathway (AP) of conduction within the heart and tachycardia. If the specific ECG changes associated with an AP occur without tachycardia, this is termed WPW "pattern".

In approximately 0.1% to 0.3% of the population (Ehtisham et al., 2005; Rodday et al., 2012), myocardial fibres connect the atria to the ipsilateral ventricles across the mitral or tricuspid annuli (Wolff et al., 1930).

This acts as a concealed AP for conduction that bypasses the AV node (Figure 2A). Unlike the normal AV nodal function, conduction is not delayed when traveling down these accessory pathways and thus pre-excitation or premature partial-depolarisation of the ventricles occurs, thereby, reducing the P-R interval. This produces the characteristic slurred and slow rising initial up-stroke of the QRS complex, or delta wave, immediately after the termination of the p-wave (Figures 1 & 3).

As described in our case, ECG changes may only occur when the AP becomes electrically active during tachycardia.

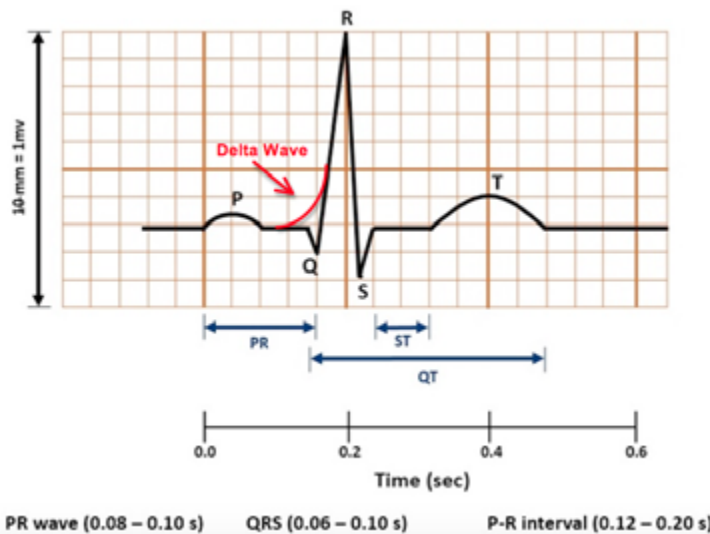


Figure 1. Normal Electrocardiogram (ECG) tracing and Delta wave.

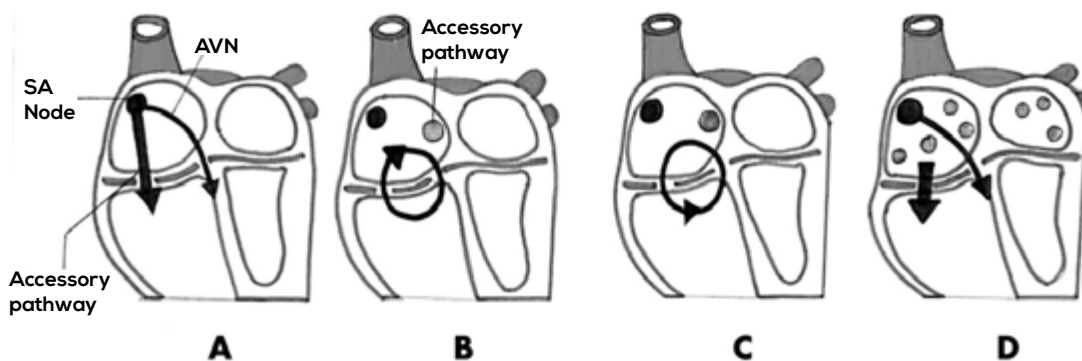


Figure 2. (A) A physiological mechanism of conduction in WPW with accessory pathway conduction (atrioventricular) resulting in the delta wave. (B) Orthodromic conduction in WPW syndrome occurring down AVN and retrogradely up accessory pathway. (C) Antidromic conduction where accessory pathway conducts anterogradely (atrioventricular) and the impulse returns to the atria via the AVN. (D) mechanism of AF in WPW syndrome.

Therefore there are no ECG changes at baseline and this can be described as a concealed AP (Kulig et al., 2010). If ECG changes are present at baseline this is described as a manifest pathway. These APs (Figure 2) may permit atrioventricular re-entrant tachycardia (AVRT), whereby electrical conduction travels as a circuit from the atrium to the ventricles and back to the atrium resulting in repeated AV nodal stimulation, inducing tachycardia. This accounts for 95% of re-entrant tachycardias in WPW patients (Blomström-Lundqvist et al., 2003).

Most commonly, the electrical impulse travels to the ventricles via the AV node and back to the atria via the AP in a retrograde manner. This is classified as orthodromic reciprocating tachycardia, and no delta wave is seen on ECG investigation (Figure 2B).

If the electrical impulse anterogradely travels to the ventricles via the AP and returns via the AV node, this is classified as

antidromic reciprocating tachycardia (Figure 2C). This occurs in only 5-10% of cases (Blomström-Lundqvist et al., 2003). Lastly, the AP may lead to atrial fibrillation (Figure 2D). These arrhythmia experienced in WPW patients are potentially fatal and a summary of risk factors, signs and symptoms, ECG findings, and investigations can be found in Table 1.

Case Presentation

Here we describe a case of a concealed accessory pathway in a 64 year old lady who presented to cardiology in 2015 with acute chest pain, palpitations and shortness of breath on a background of heavy smoking (10 pack years), high cholesterol, type 2 diabetes mellitus and aortic regurgitation.

On admission, the patient was afebrile with a blood pressure of 118/67, a pulse rate of 100/min and respiration rate of 15. The initial symptom of chest pain occurred at 11am in the morning following mild to moderate exertion whilst paint-

ing indoors on a ladder. The pain was continuous, dull and described as a central tightness across the chest radiating to the left neck and shoulder. Her subjective pain score was 8/10. The pain worsened on inspiration and there were no alleviating factors. Other presenting symptoms were pre-syncope, weakness, and numbness on the left side of the tongue.

The patient had slowly walked for nearly an hour to her General Practitioner who called an ambulance and administered sublingual glyceryl trinitrate without symptomatic relief. Of note, the patient's previous medical history revealed two previous admissions to A&E for chest pain in 2006/2007 which had been excluded as cardiac in origin by the cardiology team following negative blood tests, a normal 24 hour Holter ECG, and two non-revealing stress tests achieving 96% of her target heart rate.

Table 1. Summary of most common structural risk factors, signs and symptoms, ECG findings (not always present) and possible investigations in WPW syndrome

Risk Factor	Signs and symptoms	12 lead ECG findings indicative of WPW syndrome	Additional Investigations
Ebstein's Anomaly; The most commonly occurring congenital defect associated with WPW syndrome. It involves malformation of the tricuspid valve and may lead to cyanosis, dyspnoea, fatigue, arrhythmias, and congestive heart failure.	Palpitations	Shortened P-R interval of <0.12 s	Echocardiogram
	Dizziness	A prolonged QRS complex >0.11 s	Treadmill test
	Dyspnoea		Pharmacological testing with procainamide or ajmaline may define duration of anterograde effective refractive period of the accessory pathway.
	Atrial Flutter	Delta Waves (bidirectional AP)	Electrophysiological Study
	Atrial Fibrillation	No delta waves (retrograde AP)	
	Syncope or presyncope		
	Tachycardia		
Sudden cardiac death			

ECG Findings

The appearance of the 12 lead ECG on admission revealed a wide QRS complex and regular monomorphic ventricular tachycardia (VT) (Figure 3). The patient was diagnosed by the clinician on-call as having monomorphic unstable VT and emergency direct current (DC) cardioversion was delivered as a synchronised shock. The patient was then converted to sinus rhythm (Figure 4). A differential diagnosis for wide complex

tachyarrhythmias is given in Table 2 and in this clinical situation, the Brugada Criteria (Figure 5) provides a flow chart to the clinician that may be used to ascertain whether or not the tachyarrhythmia is ventricular or supraventricular in origin.

Investigations

Throughout the patient's admission, blood tests were non-revealing for any indication of ischaemia or other pathology. A coronary angiogram was

performed to rule out coronary artery disease. As represented in Figures 6 and 7, very mild arterial disease within the right and left coronary arteries was found, mainly at the bifurcations and not in keeping with the signs and symptoms experienced by the patient. Both the coronary angiogram and transthoracic echocardiogram did not reveal any structural abnormalities. Following coronary angiography, delta wave ECG changes were detected, allowing for

Table 2. Differential Diagnosis of Wide Complex Tachyarrhythmias

Supraventricular Tachycardia With Bundle Branch Block	Bundle branch block can occur with any supraventricular arrhythmia
Supraventricular Tachycardia With Atrioventricular Conduction Over an Accessory Pathway	May occur during atrial flutter, atrial fibrillation, atrioventricular nodal reciprocating tachycardia (AVNRT), atrioventricular reciprocating tachycardia (AVRT), and atrial tachycardia
Ventricular Tachycardia	May be diagnosed using Brugada Criteria

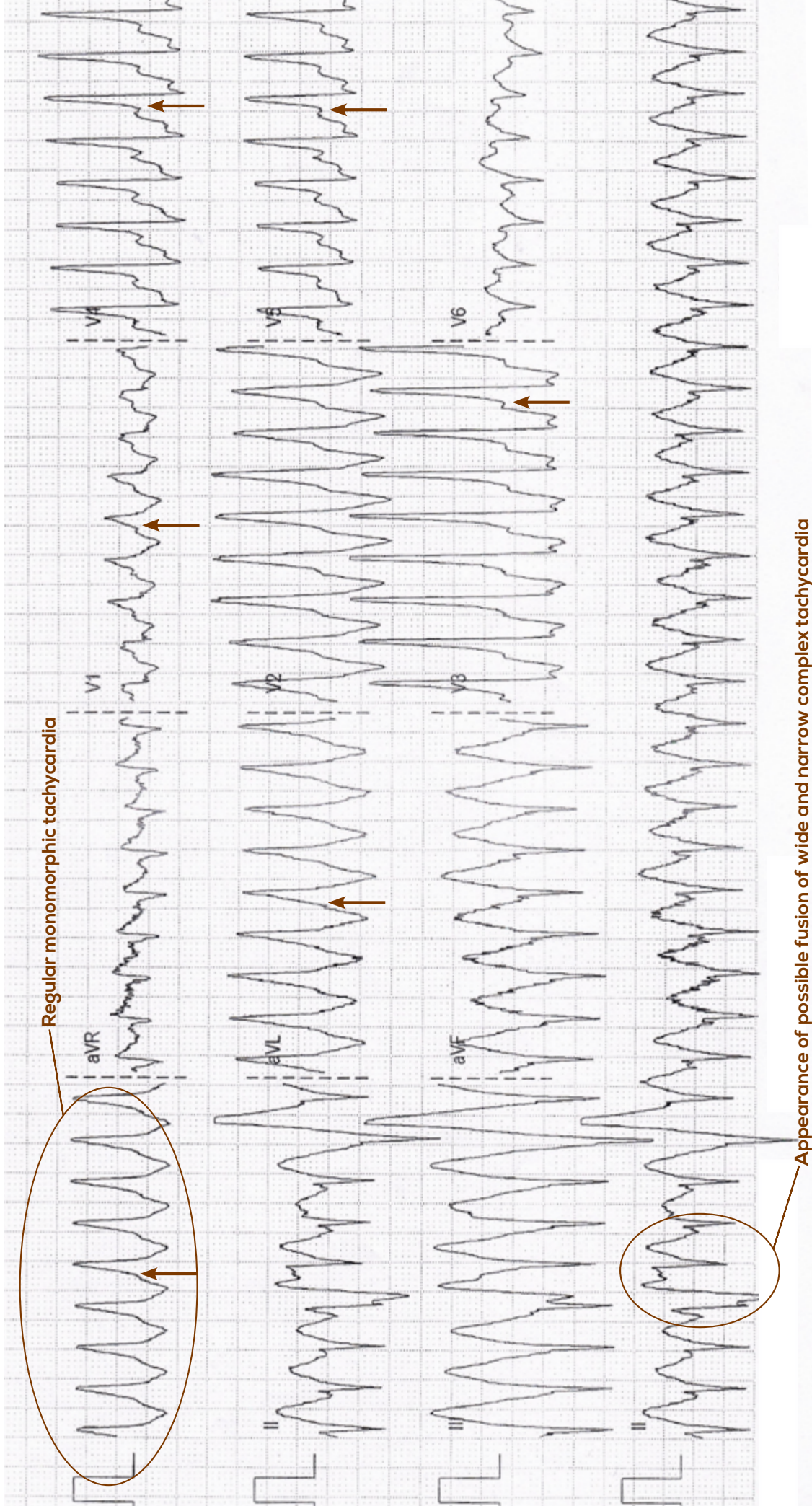


Figure 3. ECG on admission: Wide complex regular tachyarrhythmia showing delta waves (arrows) and right bundle branch block pattern.

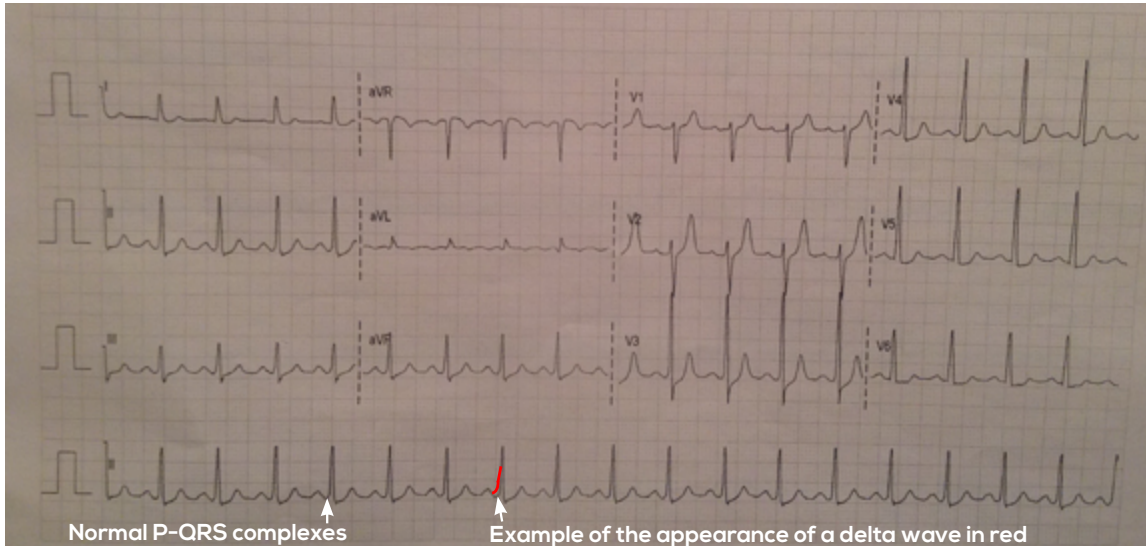


Figure 4. ECG following direct current cardioversion (DCC). Delta waves are not present on the post-DCC ECG which is highly suggestive of a concealed accessory pathway.

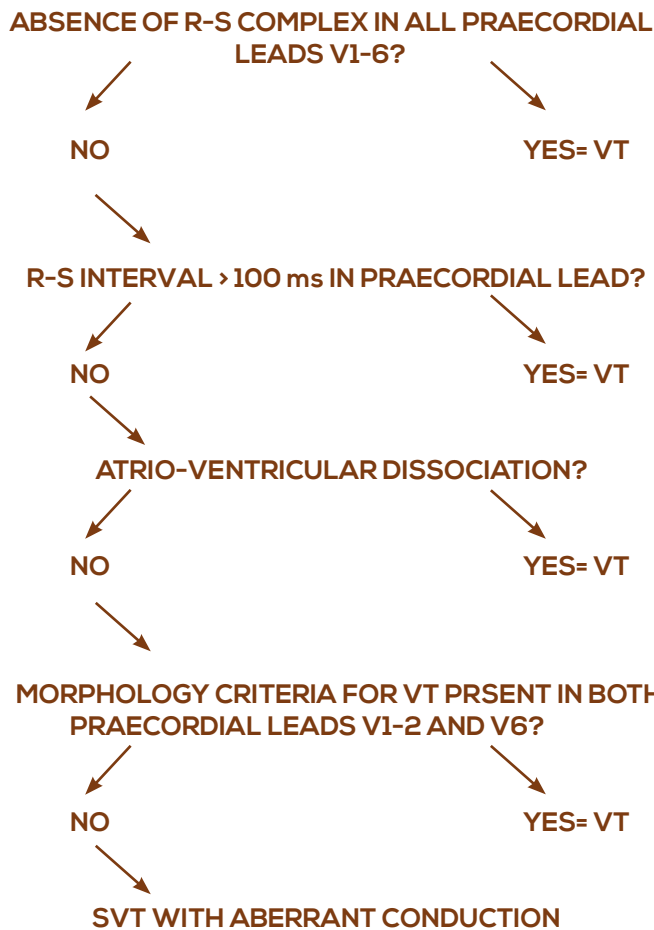


Figure 5. Based on Brugada Criteria (1991). Clinical guideline for a definitive diagnosis of wide complex tachyarrhythmias. In particular this flowchart aids in the differentiation between ventricular and supraventricular tachyarrhythmias.

definitive diagnosis of WPW syndrome. The patient was subsequently sent for electrophysiology in a separate procedure that was successful in locating and ablating a left sided accessory pathway.

Discussion

WPW patients may be asymptomatic or they may experience palpitations, chest pain, syncope, headache, dizziness, nausea, dyspnoea, impaired vision, arrhythmias or even sudden cardiac death in less than 0.1% of cases (Kulig et al., 2010). In a recent 8-year prospective study of 2,169 WPW syndrome patients in Italy, a greater proportion of asymptomatic patients

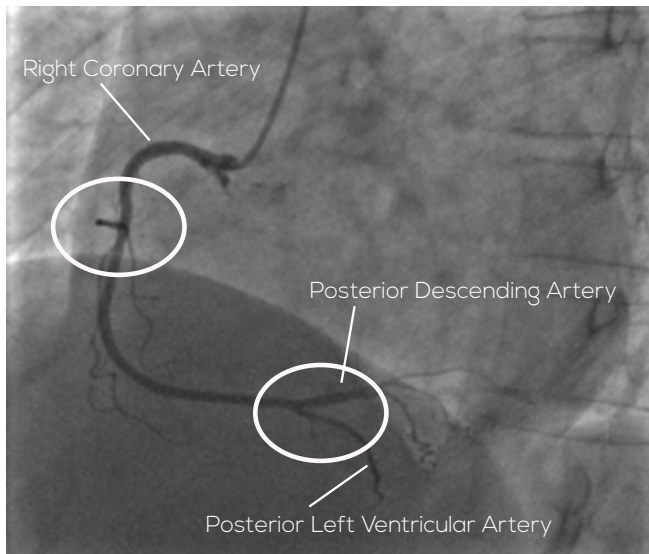


Figure 6. Angiogram of right coronary arteries from patient. Circles indicate areas of mild disease (continuity is slightly irregular in the areas of the bifurcations, as circled, but there is no obvious occlusion or stenosis).

experienced malignant arrhythmias (MA) and ventricular fibrillation (VF) than symptomatic patients. However, this was pertained to be due to symptomatic patients receiving ablation, leading to better long term outcomes. None of the 1168 patients who underwent ablation suffered MA or VF post-operatively compared to 93 of the 1001 non-ablated patients (Papponne et al., 2014). Fortunately, our patient eventually presented with symptoms and ECG changes in keeping with the WPW syndrome diagnosis, which allowed her previously concealed AP to be located and ablated.

In a larger 15 year prospective cohort study of 22,500 healthy aviation personnel undergoing systematic ECG screening, the WPW pattern was seen in 0.25% of subjects, and only 1.8% of those had documented arrhythmia diagnostic of WPW syndrome (Davidoff et al., 1981). For the general population, the risk is considered to be small and screening for WPW syndrome is not recommended at present. However all patients found to have WPW

syndrome should be fully evaluated due to the risk of lethal arrhythmias (Blomström-Lundqvist et al., 2003).

Interesting aspects of this case include the concealment of her ECG findings as baseline and the age of the patient at presentation. Concealed APs may occur in approximately one third of accessory pathway tachycardias and are an important prognostic feature, as accurate diagnosis and ablation should lead to a permanent cure of tachycardia and prevents potentially lethal future arrhythmias (Ross and Uther, 1984). Patients with WPW syndrome may present at any age; however, it more commonly occurs in young people and decreases with age due to loss of pre-excitation (Jung et al., 2011). Our patient was 64 years old when she presented with detectable evidence of cardiac arrhythmia and positive ECG findings. It is therefore important to consider WPW syndrome as a differential diagnosis for cardiac arrhythmias with tachycardia irrespective of age or previously normal ECGs.

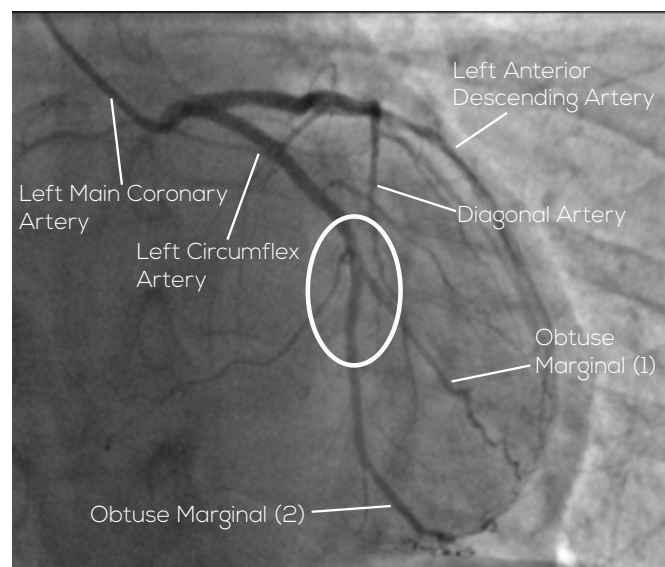


Figure 7. Angiogram of left coronary arteries from patient. Circles indicate areas of mild disease (continuity is slightly irregular in the areas of the bifurcations, as circled, but there is no obvious occlusion or stenosis).

Additionally, the patient presented acutely unwell with ECG findings of complex ventricular tachycardia. The treating clinician suspected the possibility of a fusion of wide and narrow complex tachycardia (Figure 3) and provisionally made a diagnosis of ventricular tachycardia. In actual fact, this was a p-wave followed by a wide complex QRS-complex wave, indicating supraventricular tachycardia. This was only confirmed post coronary angiogram having made a definitive diagnosis of WPW syndrome based on the ECG findings taken throughout the procedure.

The history of this patient was an important clue in helping ascertain a correct diagnosis. A patient with ventricular tachycardia is likely to be extremely unstable and unlikely to walk to her local GP over the course of an hour. However, at the time of presentation the patient was acutely unwell and did not reveal these details until after DC cardioversion.

Certain guidelines (Frankel et al., 2015) will advocate the use of IV adenosine before DC cardioversion in the management of our case. Adenosine is an antiarrhythmic with a half-life of less than 10 seconds and its effects are therefore self-limiting. It is, however, contraindicated in WPW with atrial fibrillation due to the risk of precipitating ventricular flutter and increasing the likelihood of sudden cardiac death. In

these cases, ibutilide, procainamide, or flecainide are the preferred drug choices as they are capable of slowing conduction along the AP (Blomström-Lundqvist et al., 2003). Adenosine used in the presence of unstable ventricular tachycardia may lead to further deterioration and is therefore contraindicated.

As the clinician on call had made the provisional diagnosis of VT during initial presentation, this case highlights the importance of treating the case as ventricular tachycardia until proven otherwise.

Conclusion

This case describes an atypical acute presentation of a concealed AP in a female patient with WPW syndrome, presenting unusually late in life. The ECG appearance was that of a wide QRS complex, regular monomorphic VT, and new delta-waves presenting during an acute episode of tachycardia and post coronary angiogram. This case highlights the need to be vigilant in history taking, ECG analysis, and formulating a differential diagnosis in order to provide the best treatment for our patients.

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