Self-assessment corner

Palpitations in a 43-year-old man

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A 43-year-old man presented to the Accident and Emergency department with sudden onset of palpitations associated with chest dullness, dizziness, and diaphoresis. On examination, the patient was haemodynamically stable, with normal heart sounds and no murmurs. The admission 12-lead electrocardiogram (ECG) is shown in figure 1 below.

Figure 1 Admission 12-lead ECG

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Questions

- 1 What is the underlying rhythm?
- 2 What is the diagnosis?
- 3 What treatment options are available in the acute phase?
- 4 What is the long-term strategy in treating this condition?

Answers

QUESTION 1 UNDERLYING RHYTHM

Atrial fibrillation with pre-excited complexes. The absence of any distinct p-wave activity and the irregularity of the wide QRS complexes supports the diagnosis of atrial fibrillation. The QRS complexes, in any one lead, have a similar morphology.

QUESTION 2 DIAGNOSIS

Wolff–Parkinson–White syndrome. On restoration of sinus rhythm, the resting 12-lead ECG (figure 2) reveals a short PR interval followed by the delta wave, representing pre-excitation of ventricular tissue prior to normal conduction and depolarisation through the AV node and His bundle. Note that the delta wave is noticeable in some, but not all the leads of the standard 12-lead ECG.

QUESTION 3 TREATMENT OPTIONS

Acute treatment of this dysrhythmia depends on the clinical state of the patient, which in turn depends on the rate and resultant cardiac output.

Haemodynamically stable

After assessment of the cardiac status and routine blood tests, the treatment is aimed at controlling the ventricular rate and restoring sinus rhythm.

Controlling ventricular rate This is achieved by various anti-arrhythmic drugs, which either block conduction down the accessory pathway (AP) or prolong the refractory period of the AP. The various drugs available are listed in the table. It is imperative to avoid anti-arrhythmic drugs that ‘block’ AV node conduction (eg,

Figure 2 Resting 12-lead ECG in sinus rhythm

Table Drugs used in the management of Wolff–Parkinson–White syndrome

Directed at accessory pathway	Directed at atrial stability
Flecainide (class Ic)	Flecainide
Sotalol (class II and III, at higher doses)	Sotalol
Propafenone (class Ic)	Amiodarone
Amiodarone (class III)	Propafenone (not as good as the rest)
Quinidine*	
Procainamide*	
Disopyramide*	

* Not very effective, especially in patients with ‘fast’ pathways

digoxin, calcium channel blockers, and beta-blockers) in this situation as the rapid ventricular response may degenerate into ventricular fibrillation. It is generally good practice to avoid these drugs in all broad-complex tachycardias, especially in the acute setting. Fortunately, adenosine has been used safely in this dysrhythmia, due to its short half-life (seconds).

Restoring sinus rhythm This can be achieved either using drugs (chemical cardioversion) or synchronised direct current cardioversion. Fortunately, of the anti-arrhythmics used to achieve cardioversion, the majority are effective in controlling the ventricular rate by a direct influence on the accessory pathway (table).

Concurrent intravenous heparin therapy is recommended while the patient remains in the irregular tachycardia. The likelihood of intracardiac thrombosis increases with the duration of the dysrhythmia, due to impaired atrial contractility. It has been noted that ‘atrial stunning’ following successful direct current cardioversion can occur and, therefore favour thrombosis due to relative stasis in the left atrium and its appendage.

This gentleman was commenced on intravenous heparin, prior to synchronised DC cardioversion, which was successful at 50 Joules.

Haemodynamically unstable (with impending circulatory collapse)

This requires urgent intervention which is best achieved by DC cardioversion.

QUESTION 4 LONG-TERM STRATEGY

Having ‘shocked’ this gentleman electrically, one will have to ‘shock’ him mentally with the diagnosis of Wolff–Parkinson–White syndrome and its inherent risks. The main concern is the small but significant risk of sudden death. In the case of this patient, he is at risk of a future episode of atrial fibrillation inducing ventricular fibrillation, especially as the shortest RR interval on the admission ECG is measured at 180 ms. (This figure is significant. See discussion.)

The long term management of this patient’s condition consists of the following:

Education and explanation This is essential to ensure co-operation and compliance. Explanation of the serious nature of his condition is best dealt by a knowledgeable physician, to allay his fears, and avoid any further shocks!

Anti-arrhythmic therapy This is essential to prolong the conduction through the accessory pathway, hence reducing the risk of a life-threatening arrhythmia, while the patient awaits definitive therapy.

Electrophysiologic studies and ablation The definitive treatment in this case is the localisation of the accessory pathway(s). Once this is done, ablation of the pathway can be performed, usually using radiofrequency energy

delivered by special catheters. Once successfully ablated, the patient does not require anti-arrhythmic therapy.

Discussion

The Wolff–Parkinson–White syndrome was initially described in 1950.¹ At the time, it was noted that several young healthy individuals had short PR intervals with left bundle branch block. It was later on that the abnormal QRS morphology was attributed to pre-excitation of ventricular tissue. This pre-excitation on the ECG was termed the delta wave.

Wolff–Parkinson–White syndrome results from the congenital presence of impulse-conducting fascicles (Kent bundles), otherwise known as accessory pathways. These fascicles connect the atria and ventricles across the annulus fibrosis. These connections can occur anywhere in this fibrous ‘divide’ and hence can give rise to different QRS morphologies and patterns of pre-excitation.^{2,3} An individual may have several accessory pathways. On occasions, the fascicles are not a discrete band, but instead exist as a mesh of fibres. Conduction through the fascicles may occur antegradely (atria to ventricle), retrogradely (ventricle to atria), or in both directions. The pattern of conduction is important, as this would dictate the types of dysrhythmias occurring (see box and figures 3 and 4) and the appropriate drug therapy.

PREVALENCE

Wolff–Parkinson–White syndrome is not an uncommon disorder. The ‘prevalence’ of pre-excitation in the general population is estimated at 0.1% to 0.3%.⁴ Wolff–Parkinson–White syndrome accounts for 20% of cases of paroxysmal supraventricular tachycardia. Patients may be asymptomatic, but the majority present with symptoms ranging from ‘simple’ palpitations to sudden death (see box).

INVESTIGATION

As this condition is associated with sudden death,⁵ there is a need to investigate this condition with a sense of urgency, especially those at risk. Ideally all patients should be considered for electrophysiologic studies, but realistically the demand that this would generate would be enormous and, as yet, not cost effective. It is generally accepted that asymptomatic patients should not be investigated, as the risk of sudden death is extremely low. In the case of symptomatic patients, it is important to try to identify those at risk of

sudden death.^{4,6} Therefore, numerous groups have tried to profile those at risk patients with Wolff–Parkinson–White syndrome (see box). It is generally accepted that patients with a previous history of syncope, recurrent atrial fibrillation, short pre-excited RR intervals dur-

Figure 3 Schematic diagram of possible tachycardias associated with Wolff–Parkinson–White syndrome. Note conduction through both the AV node and accessory pathway can be antegrade or retrograde. The direction of the tachycardia, and hence re-entrant circuit, depends on the conduction velocity and refractory periods of both ‘limbs’ of the circuit

Figure 4 Atrial fibrillation and accessory pathway. The chaotic atrial rhythm due to multiple microcircuits (arrows in atrium) is transmitted to the ventricle via the AV node and accessory pathway. The AV node, unlike myocardial tissue, exhibits decremental conduction (reduction in conduction velocity as the PP interval shortens)

Wolff–Parkinson–White syndrome: associated tachycardias
<ul style="list-style-type: none">• orthodromic tachycardia (re-entrant circuit with antegrade (‘usual’) AV node conduction)• antidromic tachycardia (circuit with retrograde AV node conduction)• atrio-ventricular reciprocating tachycardia (usually in the presence of multiple pathways)• atrial fibrillation (? predisposition due to micro-reentry circuits)

Wolff–Parkinson–White syndrome: attributable symptoms
<ul style="list-style-type: none">• intermittent palpitations• chest discomfort• dyspnoea (sudden)• dizziness• pre-syncope• syncope• sudden death

Wolff–Parkinson–White syndrome: risk factors for sudden death

- age
- history of syncope
- previous episodes of atrial fibrillation
- antegrade AV conduction with an effective refractory period of < 230 ms*
- atrial vulnerability (propensity to develop atrial fibrillation)
- presence of multiple pathways
- family occurrence (not hereditary)
- associated coronary artery disease

*This figure varies from centre to centre.

ing atrial fibrillation (< 230 ms), or fast conducting accessory pathways, should be referred for electrophysiologic studies and pathway ablation. Electrophysiologic studies enable the cardiologist to locate the accessory pathway and look for other pathways that may be concealed. Once this information is available, then ablation can be considered. Other patients that should be considered for electrophysiologic studies and pathway ablation are those with persistent symptoms despite adequate anti-arrhythmic therapy and those with drug-related side-effects.

MANAGEMENT

The management of symptomatic patients with Wolff–Parkinson–White syndrome inadvertently varies from centre to centre, depending on resources and local expertise in electrophysiology. Drug therapy with various anti-arrhythmics is essential to reduce episodes of tachycardia. The ideal agents are those that prolong the conduction time and/or increase the refractory period of the part of the circuit used in sustaining the tachycardia. Therefore, drugs can be directed at the accessory pathway or the AV node (figure 3). Blocking conduction through the AV node, using digoxin, calcium channel blockers, or beta-blockers, is a possible strategy only when the tachycardias that can be induced spontaneously or during electrophysiological studies incorporate the accessory pathway as the retrograde limb (orthodromic AV reciprocating tachycardia). Otherwise, such a strategy can be lethal, especially in the presence of atrial fibrillation. Hence, the decision to use AV ‘blocking’ drugs should only be made by experienced physicians, usually with the benefit of recent electrophysiological data.

DRUG THERAPY

Prolonging the conduction time through the accessory pathway is possible using any of a multitude of anti-arrhythmics (table). The ideal anti-arrhythmic would also have prophylactic properties against the recurrence of atrial fibrillation, as this is thought to be the initiating arrhythmia that precipitates ventricular fibrillation and sudden death. The mechanism for atrial fibrillation in Wolff–Parkinson–White syndrome is poorly understood, but it is debated that the presence

of an accessory pathway may support micro-reentry circuits, which in turn would increase the chances of recurrence of atrial fibrillation.⁷ Currently the effective drugs available are sotalol, flecainide, propafenone, and amiodarone. These drugs prolong the refractory periods of the accessory pathways, especially those at high risk, but this effect can be reversed by excessive circulating catecholamines and during exercise. Again this stresses the need for further investigation and intervention, despite apparent ‘cure’ of symptoms with medication.

SURGICAL ABLATIONS

The curative procedure for Wolff–Parkinson–White syndrome is accessory pathway ablation. Ablation⁴ can be performed using various methods. Surgical ablation was initially performed in the 1960s. At the time this offered the first chance of cure, but was associated with significant mortality and morbidity. Despite considerable improvement in surgical ablation techniques, it is now only considered when a concomitant cardiac operation is necessary, catheter ablation has been unsuccessful, or access to the accessory pathway is technically difficult.

CATHETER ABLATION

The advent of catheter ablation has revolutionised the treatment of symptomatic Wolff–Parkinson–White. Initially, ablation was performed by delivering an high energy direct current shock between a catheter, placed close to the pathway, and a reference electrode. This essentially created excessive tissue damage and as a result this method is associated with numerous rhythm abnormalities. The technique has improved, but the need for general anaesthesia to relieve patient discomfort is a considerable disadvantage. More recently (1987) radiofrequency energy has been utilised with considerable success. The method utilises low energy to induce damage through transference of heat. The success rate is extremely high (> 95%), especially in experienced centres which adopt strict training programmes.⁸ Nowadays, patients can undergo an electrophysiological study followed by radiofrequency ablation in the same session. The various complications associated with radiofrequency ablation are listed in the box. Recurrence of the pathway is well recognised

Wolff–Parkinson–White syndrome: complications of radiofrequency ablation of accessory pathways

• local vascular problems	(1.0%)
• chest pain	(0.8%)
• tamponade	(0.6%)
• thromboembolic	(0.5%)
• aortic valve perforation	(0.4%)
• pericarditis	(0.3%)
• atrioventricular block	(0.3%)
• pneumothorax	(0.1%)
• coronary artery occlusion	(0.1%)

(<5%). This is mainly due to the arrangement of the accessory bundle, which may exist as a diffuse mesh of fibres. These fibres may be stunned during radiofrequency ablation, and later on may recover. Infrequently radiofrequency ablation is not possible due to the position of the accessory bundle, surgical ablation is to be considered.

Conclusion

The Wolff–Parkinson–White syndrome is a relatively common and curable condition. As it is associated with sudden death in a minority of patients, all symptomatic patients require investigation and treatment based on various characteristics of the accessory pathway and the symptomatology. The advent of accessory pathway ablation has made freedom from a lifetime of medication a reality, especially with the introduction of radiofrequency catheter ablation. The success and low complication rates associated with radiofrequency ablation

Wolff–Parkinson–White syndrome: management options for patients with symptomatic disease

- drugs
- anti-tachycardia pacemakers (usually in addition to anti-arrhythmic agents)
- ablation:
 - (a) surgical
 - (b) catheter (intracardiac)
 - direct current
 - radiofrequency energy

should encourage physicians to refer their patients for consideration of accessory pathway ablation, as this currently offers the ideal cure.

Final diagnosis
Wolff–Parkinson–White syndrome

Keywords: atrial fibrillation, Wolff–Parkinson–White syndrome, cardioversion

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