ECG EDUCATION

Wolff-Parkinson-White syndrome

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Articles in this section are inspired by, but not based on, real cases to illustrate the importance of knowledge about ECGs in relation to clinical situations in general practice. Management is not discussed in detail.

Madeleine, a 5-year-old girl, and her parents have come to see you, her GP, today as instructed for a referral for an urgent cardiology assessment. Madeleine has been feeling faint during dance practice, but has never actually fainted and is usually well with no medical problems. She tells you that she was about to faint and stopped dancing to avoid it. She says that her chest felt heavy and her heart went 'boom'. When the faintness came on it was either during or directly after stopping exertion and it lasted a minute or so. She was taken to hospital by ambulance but not admitted. The discharge information contains the ECG shown below.

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Q1. What does this patient's ECG show (see Figure)?

The ECG shows pre-excitation due to an accessory tract, typical of Wolff-Parkinson-White (WPW) syndrome. The PR interval is short even after considering the patient's age. The QRS complexes do not appear particularly broad; however, there is definite slurring of the upstroke of each R wave, which is most obvious in leads II, III, aVF and V4. The RSR' pattern with T wave inversion in V1 to 2 is a normal variant in children of this age; this is still a type B pattern due to absence of a dominant R wave in V1. There are pseudo-infarction Q waves in lead aVL simulating lateral infarction.

Q2. What is WPW syndrome?

WPW syndrome is a pre-excitation syndrome, a congenital atrioventricular re-entry circuit, which is abnormal conductive tissue between the atria and the ventricles. When the atrioventricular re-entry tachycardia occurs with pre-excitation on an ECG, it is known as WPW syndrome. If there is no pre-excitation on the resting ECG, this is termed a concealed accessory pathway (as it only conducts from the ventricle to the atrium, not from the atrium to the ventricle). In WPW syndrome, the electrical impulse generated in the sinoatrial node passes through the re-entry circuit (which used to be referred to as the Bundle of Kent) instead of the atrioventricular node, and causes premature contractions of the ventricles. The heart rate is not slowed as would normally occur because the re-entry circuit bypasses the atrioventricular node. This results in very rapid tachycardias. These are not true junctional or ventricular tachycardias. If the electrical impulse travels from the atrium to the ventricle via the atrioventricular node but back via the accessory pathway, the QRS complex is narrow and this is referred to as orthodromic tachycardia. If the impulse travels down the accessory tract to the ventricles and back through the atrioventricular node, the QRS complex will be wide and the tachycardia mimics ventricular tachycardia. This is referred to as antidromic tachycardia.

Patients with WPW syndrome present with episodic tachycardia (typically supraventricular tachycardias) and the heart rate will vary from patient to patient due to the nature of the accessory pathway. Sudden death is very rare, but can occur when atrial fibrillation conducts very rapidly over the accessory tract causing an extremely fast and irregular wide complex tachycardia. Patients with wide complex tachycardias should not be treated with

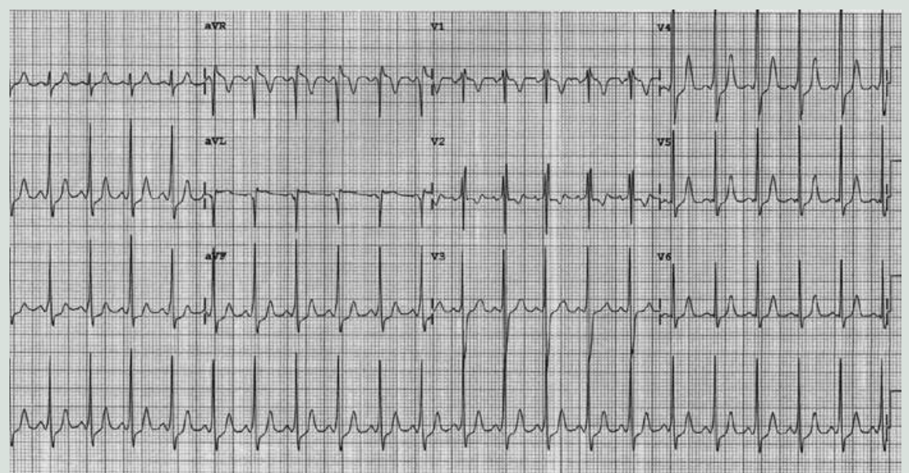

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Figure. The ECG of a 5-year-old girl with Wolff-Parkinson-White syndrome.

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negative inotropes (such as verapamil and flecainide) or atrioventricular node blockers (such as adenosine and digoxin) because of the risk of precipitating sudden death.

WPW syndrome is most commonly diagnosed in children and young adults but may occur at any time of life. It may be associated with cardiac abnormalities, such as hypertrophic cardiomyopathy or Ebstein's anomaly (congenitally abnormal tricuspid valve leaflets, often with an atrioseptal defect). However, usually the heart is normal on examination when the patient is asymptomatic. The incidence of WPW syndrome is less than one in a 1000 people.

Q3. What are the common clinical presentations of WPW syndrome?

The symptoms and signs of WPW syndrome are variable and depend on the ability of the patient to express them as well as the age and cardiovascular health of the patient. Generally, older patients or those with cardiovascular compromise tolerate the arrhythmias less well than younger patients.

When WPW syndrome presents in infants it is harder to diagnose. There may be dusky episodes with increased effort of breathing, lethargy, pallor, irritability, feeding problems and failure to thrive. Older children and adults may complain of a sudden onset, very fast, pounding pulse rate, chest pain, pressure or tightness, palpitations, shortness of breath, sudden fatigue (especially with exertion) or faintness (especially with exertion). Untreated prolonged arrhythmias may cause cardiac failure, especially in infants and the elderly. In the elderly or those with cardiovascular compromise loss of consciousness, acute myocardial ischaemia and cardiac arrest may occur.

Q4. What investigations should be considered in patients with WPW syndrome?

It is important to consider common triggers for arrhythmias, such as electrolyte disturbances, hyperthyroidism, cardiomyopathy, ischaemic heart disease and anaemia. Illicit drug use and alcohol abuse should be considered, especially in younger adults presenting with arrhythmias.

The diagnosis of WPW syndrome is suggested by ECG abnormalities and clinical history and may be confirmed by 24-hour Holter monitoring, telemetry or electrophysiological studies. A cardiac echocardiography should be arranged to exclude structural abnormalities of the heart and to document left ventricular function and wall motion. Stress testing may be arranged, especially if the arrhythmias are associated with exercise, to trial effectiveness of therapy for the condition or if there is concern about possible myocardial ischaemia.

An electrophysiological study allows the assessment of the accessory pathway and whether it is benign or potentially malign. It is safe and effective, with a single procedure cure of 96% and recurrence of 4%. It is the treatment of choice for patients with rapid and symptomatic atrioventricular re-entry tachycardias or drug-refractory tachycardia, and can be personal preference or an occupational requirement. An electrophysiological study also documents the response to medication or ablation.

Q5. How is WPW syndrome managed?

Emergency management of WPW syndrome consists of the Valsalva manoeuvre (if the patient is able to co-operate), carotid massage (on one side only) and urgent medical assistance if the patient is unstable or elderly or if the arrhythmia has continued for more than 20 minutes. In the hospital setting, intravenous adenosine, sotalol, verapamil or diltiazem are used. If the arrhythmia is not responding, the QRS complex is broad or the patient is haemodynamically unstable, cardioversion is the next option.

Nonurgent management may vary with the age of the patient and the frequency and severity of the arrhythmias. Antiarrhythmic medications most commonly include sotolol, verapamil, amiodarone and flecainide.

Radiofrequency catheter ablation is the other common alternative and has largely replaced surgical interventions, which are now of historical interest. Effective ablation will abolish the pre-excitation and prevent any further tachycardias. Any recurrences may then be seen on future ECGs.

Outcome

Madeleine was referred to a paediatric cardiologist and commenced on sotalol 12.5 mg twice daily (1 mg/kg). She has so far remained free of arrhythmias. The dose may need to be increased as she gains weight or if she has breakthrough arrhythmias. Her cardiac echocardiogram was normal. When she reaches 20 kg in weight electrophysiological studies are planned. **CT**

Key points

- **Wolff-Parkinson-White syndrome is a pre-excitation syndrome, a congenital atrioventricular re-entry circuit, which is abnormal conductive tissue between the atria and the ventricles.**
- **Symptomatically, it most commonly occurs in childhood and young adulthood and typically causes supraventricular tachycardias.**
- **The diagnosis is initially suggested by the clinical picture and ECG abnormalities, both of which may be quite variable.**
- **It is important to diagnose or exclude any possible contributing conditions, such as electrolyte disturbance, hyperthyroidism, cardiomyopathy, ischaemic heart disease or anaemia.**
- **Illicit drug use and alcohol abuse should be considered, especially in younger adults presenting with arrhythmias.**
- **Untreated prolonged arrhythmias may cause cardiac failure, especially in infants and the elderly.**
- **Investigations include monitoring, cardiac echocardiography and electrophysiological studies.**
- **Management includes medication to slow conduction through the atrioventricular node or through the accessory pathway, or radio-frequency ablation of the accessory pathway.**
- **Wide complex tachycardias should not be treated with negative inotropes (such as verapamil and flecainide) or atrioventricular node blockers (such as adenosine and digoxin) because of the risk of precipitating sudden death.**

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