

(1) OVERVIEW OF THE ECG

There is a regular bradycardia at 36 per minute. The QRS complexes are wide, with a right bundle branch block pattern.

MORE DETAILED ANALYSIS OF THE ECG

P-waves are visible before each QRS complex. The P-wave axis is about $+10^\circ$, so they probably originate in the sinus node. The PR interval is prolonged. However, it is variable, being 400ms for the first complex and increasing progressively to 520ms for the last complex. The QRS complexes, on the other hand, are absolutely regular. This strongly suggests that there is no relationship between atrial and ventricular activity – i.e. complete AV dissociation.

At first glance, the atrial rate also appears to be slow. However, there is a sharp deflection distorting the ST segment in V1 (Figure 1, arrow). It falls exactly between the two visible Ps in this lead. If one tracks it through the remainder of the tracing, it is apparent that the other P-waves are hidden by the T-waves (Figure 2, arrows). The atrial rate is therefore around 72 per minute and there is complete heart block. The escape rhythm is wide and slow and probably arises in the left ventricle or the left anterior fascicle (right axis, RBBB pattern), although origin in the His bundle with RBBB and posterior fascicular block is possible.

The correct answer is therefore (1): sinus rhythm with complete heart block; wide QRS escape.

(2) WHICH INVESTIGATION(S) WOULD BE APPROPRIATE?

As a 29-year-old, previously well man, he is not the usual candidate for degenerative complete heart block. There is nothing in his history to suggest recent myocardial infarction or myocarditis. He admitted to previous consumption of “tik” (metamphetamine) and cannabis, neither of which is known to cause heart block. He also mentioned that he had been told some 10 years ago that he had a very slow pulse. Physical examination did not reveal any abnormalities, other than bradycardia and evidence of AV dissociation (varying pulse volume, cannon waves in the jugular pulse and variation in the first heart sound).

There are many causes of heart block in young people, some of which are listed in Table 1.

In a study of survival after pacemaker implantation at Groote Schuur Hospital, in 232 patients between ages 21 - 50 years, the most common cause of heart block was surgery for valvular heart disease.⁽¹⁾ Surprisingly, 54% had idiopathic conduction disease at the time of presentation. Of these, none developed evidence of structural heart disease over the course of follow up, up to 20 years. Their survival was not significantly different from that of an age-matched group derived from the South African National Life Tables. In a recent North American study of young and middle aged patients with unexplained heart block (≤ 60 years), 34% of patients had had a diagnosis of cardiac sarcoidosis. This study suggested that cardiac sarcoidosis should be entertained in all patients with heart block ≤ 60 years.⁽²⁾ Further study is needed in the South African population, as the diagnosis may have been missed in our earlier study.

Permanent pacemaker implantation is undoubtedly indicated. Before doing this, however, an MRI scan was performed. This showed normal left ventricular function and no evidence of infiltration or other cardiac pathology. An endocardial biopsy was therefore not done. An exercise stress test was not done, but might have elucidated the mechanism of his effort-related dizziness, whether due to chronotropic incompetence or to ventricular tachyarrhythmia. It would not have influenced either the decision to pace or the pacing mode.

The correct answer to (2) is (e): All of the above.

In view of his history of bradycardia as a teenager and lack of evidence for structural heart disease, we assumed that his heart block was congenital.

Congenital complete heart block may be detected in utero, after delivery or later in life. Around 50% of affected infants have associated severe congenital heart defects and the majority of these do not survive. A proportion of the remainder experience few or no symptoms until later in life. While the majority have narrow QRS escape rhythms, in some the QRS is wide.

In view of his symptoms, the decision to pace him is straightforward.⁽³⁾ If he had presented in his teens with asymptomatic complete heart block, presumed to be congenital, the decision to pace would have been more controversial. A prospective observational study of 102 patients, aged 15 years or more,

TABLE I: Causes of heart block in young adults

Idiopathic conduction system disease
Post surgical
<ul style="list-style-type: none"> • Valve repair/replacement • Following surgery for congenital heart disease
Acute myocardial damage
<ul style="list-style-type: none"> • Trauma (e.g. stab) • Infarction • Myocarditis
Chronic infiltrative/inflammatory myocardial disease
<ul style="list-style-type: none"> • Sarcoidosis • Other granulomas (e.g. Wegener's) • Amyloid
Connective tissue disorders
AV node catheter ablation/damage
<ul style="list-style-type: none"> • Inadvertent • Deliberate
Congenital
<ul style="list-style-type: none"> • Isolated • With other congenital heart disease

without symptoms, documented reduced survival and significant likelihood of developing syncope, sudden death or mitral regurgitation.⁽⁴⁾ A wide QRS escape rhythm was not predictive, but a prolonged QT interval was. This study prompted us, at Groote Schuur Hospital, to recommend pacing to older children and teenagers with congenital complete heart block, even in the absence of symptoms.

LESSONS AND CONCLUSIONS

- Complete heart block is unusual in people under the age of 60 and should prompt a search for a cause requiring treatment, such as sarcoidosis.

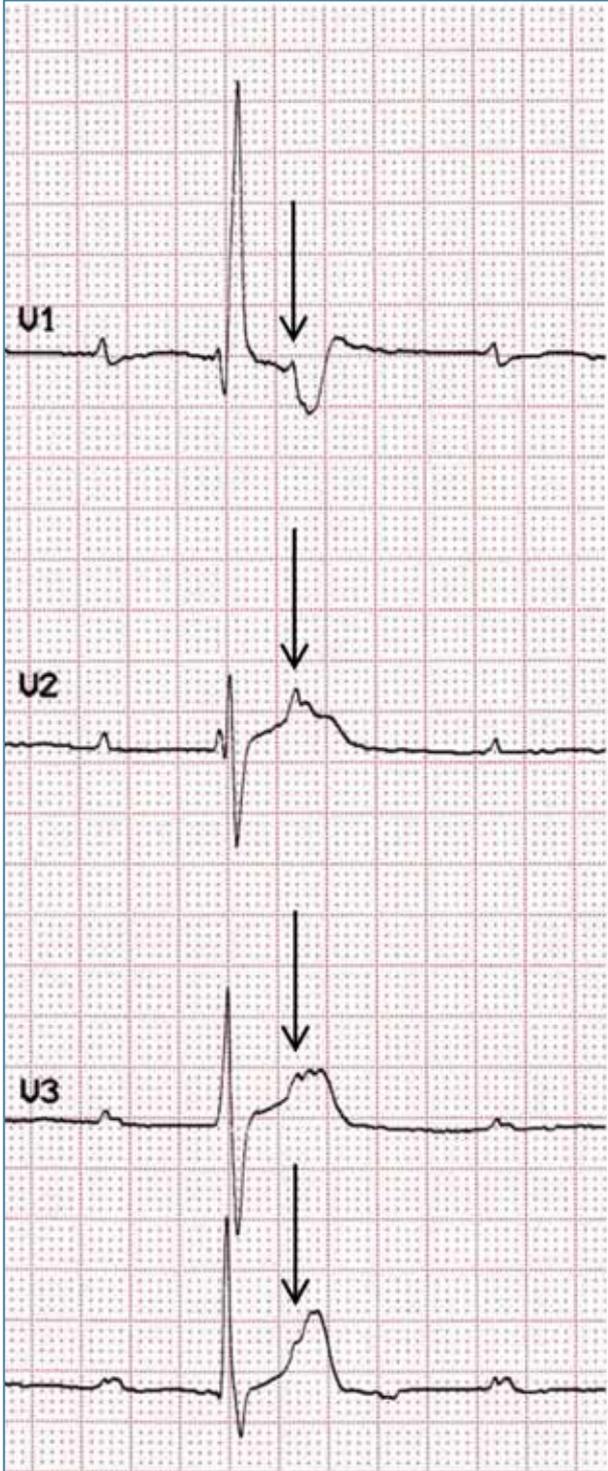


FIGURE 1: Arrows indicate the P-wave distorting the ST segment.

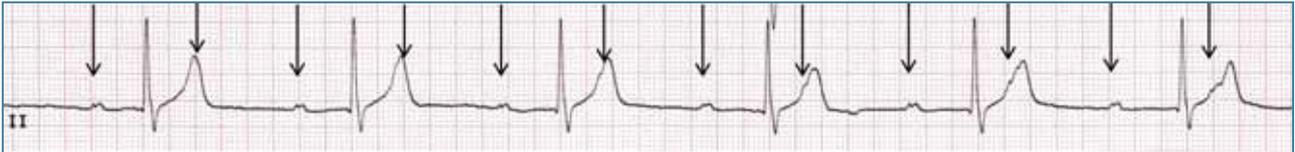


FIGURE 2: Arrows indicate the hidden P-waves.

- Occasionally patients with congenital heart block present as adults.
- In the absence of a reversible cause, complete heart block is an indication for permanent pacemaker implantation.

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