A patient with recurrent syncope

Q1: What does his ECG show?
His admission ECG (fig 1 in questions; p 117) shows sinus rhythm with deep T-wave inversion in leads I, aVL, and V2–V6.

Q2: What other investigations would you consider and what might they show?
A transthoracic echocardiogram showed marked hypertrophy of the left ventricular apex (fig 1). There was no septal hypertrophy, systolic anterior motion of the mitral valve, or evidence of left ventricular outflow tract obstruction. A 24 hour ECG detected asymptomatic episodes of non-sustained ventricular tachycardia. On treadmill exercise testing he managed 10 minutes of the Bruce protocol without exercise induced arrhythmias. Immediately after exercise, he developed profound hypotension and bradycardia (fig 2) with transient loss of consciousness. During tilt table testing he became symptomatic at 14 minutes and dropped his systolic blood pressure from 110 mm Hg to 70 mm Hg followed by a drop in heart rate from 70 to 40 beats/min. The ECG rhythm strip of leads I, II, and III progressively changed from sinus rhythm to sinus bradycardia to nodal bradycardia getting slower and then to a wide complex slow idioventricular type rhythm similar to the postexercise ECG. Changes occurred in the reverse order when tilt was terminated.

Q3: What diagnoses would you consider?
The diagnosis is neurocardiogenic syncope with associated apical hypertrophic cardiomyopathy and asymptomatic non-sustained ventricular tachycardia.

Q4: How would you treat this patient?
He is unlikely to derive benefit from cardiac pacing and medical treatments for neurocardiogenic syncope can be tried. Long term antiarrhythmic treatment is not indicated.

Discussion
Apical hypertrophic cardiomyopathy (HCM) was initially thought to be limited to individuals of East Asian origin, particularly the Japanese population and was originally reported as one of the subsets of hypertrophic cardiomyopathy without gradient.1

The classical condition has been defined initially by the presence of giant inverted T waves (>10 mm) in leads V4 and V5 of the resting ECG, the electrocardiographic sum RV5 + SV1 greater than 35 mm, and a spade-like appearance on left ventricular angiography in end diastole in the right anterior oblique projection.2 In western countries, however, “giant negative T waves” are usually less marked.3

Apical HCM is mainly asymptomatic and has been associated with a benign prognosis.4 It is usually detected by chance as a result of an abnormal ECG. Exertional dyspnoea, fatigue, and chest pain may, however, occur.5 Severe arrhythmias in apical hypertrophy are infrequent but non-sustained and sustained ventricular tachycardia as well as atrial fibrillation have been reported.6 Non-sustained ventricular tachycardia on ECG monitoring has been shown not to be associated with an adverse prognosis in an unselected population with HCM.7 This would suggest that chronic antiarrhythmic treatment is probably not required. While there are no specific studies in relation to the apical HCM variant, rare cases of significant ventricular arrhythmias have, however, been reported. Other rare complications include apical myocardial infarction and the development of an apical aneurysm with normal coronary arteries.8

Exercise induced syncope is part of the spectrum of vasovagal or neurocardiogenic syncope. Syncope episodes related to physical exertion should be evaluated thoroughly because syncope may be the presenting symptom of a serious cardiac disease that may lead to sudden death. Head-up tilt table testing has been found to be effective in reproducing syncope in those who are predisposed to vasovagal reactions. Frequency and severity of events need to be considered when long term therapy is started. The most commonly used drugs are β-blockers.9 Disopyramide has also been used, as well as the selective serotonin reuptake inhibitors,10 and...
Self assessment answers

Digital gangrene: an unusual cause

Q1: What are the cutaneous changes in the hand?
The little finger (fig 1; p 118) shows evidence of gangrene with a proper line of demarcation present. Also there is evidence of splinter haemorrhages, Osler’s nodes, Janeway’s lesions, and clubbing.

Q2: What did the blood culture grow and what is the antibiotic of choice?
The blood culture grew Staphylococcus aureus; on further antibiotic sensitivity it showed that the organism was methicillin resistant. S. aureus (MRSA) positive. S. aureus is a common cause of acute bacterial endocarditis, and is known to cause peripheral embolic phenomena.

Q3: What does the TOE show?
As the initial two dimensional echocardiography did not show any changes we went ahead with TOE; this showed a bicuspid aortic valve with vegetation (fig 2; p 118).

Discussion
Few physicians will be free of the diagnostic challenges posed by endocarditis. Frequently, the presence of endocarditis is disguised, and the prominent organ involvement may vary considerably. Central to the recognition of the disease is the consideration of the diagnosis in any patient who has unexplained fever, who has organ involvement that may be attributed to embolic phenomenon, or who appears to have a multisystem disease.

Digital gangrene in staphylococcal endocarditis is rarely reported, although the incidence of peripheral embolisation is highest with S. aureus endocarditis, the frequency of organs and organ systems involved in the embolic phenomenon in the decreasing order are splenic, cerebral, pulmonary, renal, and coronary followed by the extremities. In a study carried out by Chambers et al in San Francisco General Hospital in patients with S. aureus endocarditis it was reported that systemic arterial embolism was seen in 2%-4% of the patients, and the major episodes were of coronary and bony embolic episodes leading to osteomyelitis. Digital gangrene was seen in only one patient.

Perhaps the most striking feature of endocarditis caused by S. aureus is the extent of embolic manifestations, especially in left sided disease. Left sided lesions cause the phenomenon more frequently than right sided, indicating that this may be due to microvascular septic emboli.

Aortic valve infection, particularly in association with vegetation and congestive cardiac failure, may be an indication for early surgical intervention. The crucial determinant for surgical intervention remains the severity of heart failure and consequently the valve affected rather than the aetiological agent.

TOE has a substantially higher sensitivity (76%-100%) and specificity (94%) than transthoracic echocardiography (TTE) for perivalvular extension of infection because the TOE transducer in the oesophagus is in close physical proximity to the aortic root and the basal septum, where most complications occur. The sensitivity of TOE can be improved by imaging in two or more planes, because incremental planes decrease the number of false negative studies and improve the definition of vegetation extent and mobility. One recent published study comparing TTE and TOE in patients with S. aureus bacteraemia found TOE was essential to establish the diagnosis of infective endocarditis and to detect associated complications.

Final diagnosis
Digital gangrene in a case of bicuspid aortic valve with Staphylococcus aureus endocarditis.

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References
Tachycardia in the presence of a pacemaker

**Q1: What is the electrocardiographic diagnosis?**
Atrial fibrillation in the presence of dual chamber pacemaker.

**Q2: Is the pacemaker malfunctioning?**
No.

**Q3: How do you explain the tachycardia with pacing spikes?**
The heart’s native atrial activity in patients with atrial flutter/fibrillation causes atrial sensing, which triggers the ventricular pacing at the pacemaker’s maximal preprogrammed rate.

**Q4: What is the differential diagnosis for tachycardia with pacing spikes?**
The differential diagnosis of wide complex tachycardia with pacing spikes includes pacemaker mediated tachycardia and runaway pacemaker syndrome.

**Q5: What is the management?**
Deactivation of atrial lead.

**Discussion**
The main purpose of a dual chamber pacemaker is to achieve atrioventricular synchrony. This is achieved by the atrial lead sensing atrial depolarisation and triggering the ventricular lead to depolarise the ventricle after some atrioventricular delay. In patients with no native atrioventricular conduction as in complete heart block, if the native atrial rate increases, the ventricular lead follows suit and discharges at the corresponding rate to maintain atrioventricular synchrony. However, the maximum rate at which the ventricular lead can respond is usually programmed at 120–130 beats/min to prevent very rapid ventricular rates.

Pacemaker mediated tachycardia in those with a dual chamber pacemaker can be due to one of two mechanisms. Firstly, if the patient has a dual chamber pacemaker in the setting of third degree atrioventricular nodal block (complete heart block) develops any atrial tachydysrhythmia like fibrillation, flutter or atrial tachycardia, the atrial lead will sense the atrial activity and trigger ventricular pacing at the pacemaker's maximal preprogrammed rate. The standard 12 lead electrocardiogram (ECG) will reveal a rapid ventricular paced rhythm. A magnet placed directly on the pacemaker switches off the sensing action and induces asynchronous ventricular pacing at the preset demand rate, which is usually 70 beats/min. However, when the magnet is removed and sensing restored, the problem will recur. The pacemaker interrogator/programmer, which is portable and compact, provides data about the pacemaker’s intrinsic functionality and its current functioning. It can generate an intracardiac ECG and atrial and ventricular depolarisation can be easily identified.

Rate controlling drugs will not have any impact on this tachycardia. Pharmacological or electrical cardioversion may be useful. If cardioversion is considered inappropriate, the atrial lead can be deactivated using the interrogator. Anticoagulation should be started. If the underlying rhythm is atrial flutter, it is possible to overdrive the atrium and cardiovert it.

The second mechanism by which pacemaker mediated tachycardia can occur is when premature ventricular contractions (occurring after the atrial refractory period) are conducted retrogradely via the atrioventricular nodal pathway resulting in atrial depolarisation. The pacemaker senses this atrial activity and initiates ventricular pacing, which continues as an endless loop tachycardia or pacemaker mediated tachycardia. Adenosine and Valsalva manoeuvre may or may not terminate this tachycardia.

Both the above problems should be differentiated from runaway pacemaker. Runaway pacemaker occurs when the pacemaker’s pulse generator discharges at a rate above its preset upper limit. The malfunction lies entirely within the pulse generator. It should be suspected if pacemaker dysrhythmias occur at rates greater than 130 beats/min or the upper rate limit if this is known. Magnet application or pharmacological treatment will have little or no effect on the runaway pacemaker but magnet application may be tried. It can be potentially life threatening. Fortunately it is extremely rare now because of improved design. In the setting of haemodynamic compromise, the definitive treatment is emergent removal of pulse generator.

**Final diagnosis**
Pacemaker mediated tachycardia due to tracking of atrial fibrillation by a dual chamber pacemaker.

**Learning points**
- The differential diagnosis for broad complex tachycardia with pacing spikes are pacemaker mediated tachycardia or runaway pacemaker syndrome.
- Development of atrial fibrillation or flutter in patients with a dual chamber pacemaker (in the setting of complete heart block) leads to pacemaker mediated tachycardia. Atrial rhythm is sensed by the atrial lead which triggers ventricular pacing at the maximal preprogrammed rate.
- Deactivation of the atrial lead is often the preferred option to control this tachycardia.

**References**
Tachycardia in the presence of a pacemaker

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