Syncope in hypertrophic cardiomyopathy

WILLIAM McKENNA, LOUISE HARRIS, JOHN DEANFIELD

From the Division of Cardiovascular Disease, Department of Medicine, Royal Postgraduate Medical School, London

SUMMARY A patient with hypertrophic cardiomyopathy, syncope, and frequent ventricular tachycardia was monitored during a syncopal episode. An unrecordable blood pressure and the loss of a left ventricular outflow tract murmur without evidence of arrhythmia suggested a primary haemodynamic mechanism such as reduction in left ventricular volume.

Death is often sudden in hypertrophic cardiomyopathy¹ and is associated with syncope² and serious ventricular arrhythmia.³ Though asystole,⁴ complete heart block,⁵ rapid atrioventricular conduction precipitating ventricular fibrillation,⁶ and massive myocardial infarction⁷ have been documented, the cause of death remains uncertain in the majority of patients who die suddenly. Acute haemodynamic deterioration with increased left ventricular outflow tract gradient or tachycardia with decreased stroke volume have been proposed but never documented as the antecedent mechanism.⁸ We present a patient with hypertrophic cardiomyopathy, syncope, and frequent ventricular tachycardia, who was monitored during a syncopal episode, with loss of blood pressure and left ventricular outflow tract murmur without ventricular tachycardia.

Case report

A 52-year-old nursing sister was well until 1978 when she began to experience palpitation. She was found to have a jerky, ill sustained pulse, a palpable left atrial beat with prominent left ventricular impulse, and a mid-systolic murmur. The electrocardiogram showed left ventricular hypertrophy with repolarisation changes, the chest radiograph was normal, and M-mode echocardiogram disclosed a thickened ventricular septum (2.2 cm) with mid-systolic closure of the aortic valve and systolic anterior motion of the mitral valve. The diagnosis of hypertrophic cardiomyopathy was made and propranolol 40 mg daily was started. The patient continued to experience palpitation and developed increasing breathlessness and chest heaviness during effort. On Boxing Day 1979, she had a prolonged syncopal episode. This was preceded by the sudden onset of fast palpitation after

rising from a chair. Subsequently ambulatory electrocardiographic monitoring disclosed frequent short episodes of ventricular tachycardia. The propranolol was increased to 120 mg per day, but she experienced another syncopal episode. This was preceded by tachycardia which persisted after climbing stairs quickly. Mexilitene 600 mg daily was added and the patient was referred to the Hammersmith Hospital.

While she was on mexilitene and propranolol the patient continued to have daily episodes of ventricular tachycardia (Fig. 1). Propranolol and mexilitene were stopped. Treadmill exercise tests were performed and the patient exercised for six minutes, completing stage 2 of the Bruce protocol.⁹ Cardiac catheterisation and left ventricular angiography confirmed the diagnosis of hypertrophic cardiomyopathy. Left ventricular end-diastolic pressure was 16 mmHg, and there was a resting left ventricular outflow tract gradient of 50 mmHg. The mean pulmonary artery pressure was 24 mmHg and there was no right ventricular gradient or mitral regurgitation.

While she was walking in the hospital accompanied by one of us (WM) the patient complained of palpitation and breathlessness. The pulse was found to be rapid and jerky and a systolic murmur was present. Though the patient sat down, the symptoms persisted. Over a period of two minutes the pulse became impalpable, the murmur disappeared, and the patient then lost consciousness. Despite external cardiac massage peripheral pulses were not palpable and signs of anoxia developed. She remained unconscious for 90 seconds, after which she recovered spontaneously with a gradual restoration of the peripheral pulses and the systolic murmur. There were no neurological sequelae. During this episode the patient was wearing an electrocardiographic recorder. This disclosed an increasing sinus rate over three minutes, from a baseline of 70 beats/minute to a maximum of 130

Fig. 1 This is one of 12 episodes of ventricular tachycardia which were recorded during six days of three channel ambulatory electrocardiographic monitoring. The longest episode of ventricular tachycardia was 10 beats and the maximum heart rate never exceeded 160 beats/minute. The peak hourly and daily ventricular extrasystole count was 36 and 163, respectively.

Fig. 2 Three channel ambulatory electrocardiographic monitoring. (a) Sinus tachycardia 130 beats/minute during syncope; (b) atrioventricular dissociation with a junctional rhythm (beats 1, 3, and 5) and varying degrees of fusion with sinus impulses (beats 2, 4, and 6). Ventricular rate 50 beats/minute; (c) restoration of sinus rhythm with pronounced ST segment change; (d) this trace was taken earlier in the day and was identical to the pattern to which she returned three minutes after the episode.

beats/minute at the time that she lost consciousness (Fig. 2). During the recovery period, ST segment depression was more pronounced and she showed atrioventricular dissociation with an accelerated junctional rhythm of 50 beats/minute and occasional fusion beats (Fig. 2); the sinus rate gradually accelerated with a return to normal sinus rhythm (70 beats/minute) but with pronounced ST depression (Fig. 2).

Within three minutes the electrocardiogram had returned to the baseline pattern.

The patient was started on verapamil 120 mg daily. After three days, however, she complained of more frequent episodes of palpitation, breathlessness, chest heaviness, and dizziness and she was found to be in pulmonary oedema. Verapamil was discontinued, the pulmonary oedema cleared, and amiodarone was

started with a loading dose of 1200 mg daily for one week followed by a maintenance dose of 400 mg daily. The patient improved and four months later she remains well and has had no palpitation or syncope. She continues to experience breathlessness and heaviness in the chest on effort. Seventy-two hour ambulatory electrocardiographic monitoring disclosed no ventricular tachycardia and a peak hourly and daily ventricular extrasystole count of three and nine, respectively. Treadmill exercise testing was repeated and she exercised for nine minutes, completing stage three of the Bruce protocol without distress.

Discussion

This patient has hypertrophic cardiomyopathy with rapidly progressive symptoms, syncope, and ventricular tachycardia; all features that are associated with poor prognosis.^{2,3} Though it is logical to assume that syncopal episodes may be caused by arrhythmia this patient demonstrates that this need not be so. Ventricular tachycardia is a marker of severe disease and is often present in patients who have syncope and die suddenly.^{2,3} It is clear, however, that other causes of syncope and sudden death operate.

The precise mechanism causing syncope in our patient is unknown. The episode was preceded by sinus tachycardia without definite ST changes, though in the presence of pre-existing repolarisation abnormalities ST change is difficult to assess. The loss of murmur and peripheral pulse was probably the result of left ventricular cavity elimination and a sudden decrease in stroke volume. Whether this was primarily the result of an acute decrease in afterload or in preload or the result of a reflex or metabolically mediated decrease in compliance or of some other mechanism is uncertain.

Though previous reports have suggested good symptomatic relief and improved exercise tolerance with verapamil,¹⁰ this has not been our experience.¹¹ While on verapamil this patient developed left ventricular failure which cleared and has not recurred since verapamil was discontinued. The selection of the patient may be very important; the identification of those patients with hypertrophic cardiomyopathy who may benefit from verapamil requires further examination.

Amiodarone improved the symptoms and exercise tolerance and abolished serious ventricular arrhythmia in our patient. The potent antiarrhythmic effect of amiodarone in hypertrophic cardiomyopathy has been demonstrated.¹¹ Further work to characterise

the effect of amiodarone on ventricular function and exercise tolerance is in progress.

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Requests for reprints to Dr W J McKenna, Division of Cardiovascular Disease, Hammersmith Hospital, Du Cane Road, London W12 0HS.