Syncope in hypertrophic obstructive cardiomyopathy due to asystole

Simon Joseph, Raphael Balcon, and Lawson McDonald
From the National Heart Hospital and the Institute of Cardiology, London

Although faintness, syncope, and sudden death are common in hypertrophic obstructive cardiomyopathy, their cause has not been well documented. The occurrence of ventricular asystole causing syncope after exercise is described in one patient. The elucidation of the disturbance of rhythm may, therefore, be of great importance, as sudden death may be avoided in these patients by the prophylactic use of a pacemaker.

Hypertrophic obstructive cardiomyopathy (Goodwin et al., 1960) is now a well-recognized clinical entity. Its clinical and pathological features have been widely described (Brent et al., 1960; Goodwin et al., 1960; Wigle, Heimbecker, and Gunton, 1962; Maurice et al., 1966; Braunwald et al., 1964; Meerschwa, 1969); symptoms include faintness and syncope which are usually related to effort, and though syncope has frequently been reported (Teare, 1958; Brent et al., 1960; Hollman et al., 1960; Meerschwa, 1969), only once has its cause been documented, when ventricular tachycardia and fibrillation were recorded in two patients (Maurice et al., 1966). A patient with hypertrophic obstructive cardiomyopathy which was proven by haemodynamic and angio-

graphic studies and subsequently confirmed at operation is described; ventricular asystole occurred after effort and was confirmed by continuous electrocardiographic monitoring.

Case report

The patient was a 22-year-old Kenyan male of Goanese origin, who had no relevant history apart from bronchial asthma in early life.

He presented at 15 years with cardiac pain and fatigue on exercise. At 17 he developed episodes of faintness and syncope with effort, and on 12 occasions he had lost consciousness for periods of about one minute. He did not have dyspnoea, palpitation, or epileptic features.

1 Present address: London Chest Hospital, London E1.

Findings of note on clinical examination were normal arterial pulses, blood pressure 120/75 mmHg, normal jugular venous pressure and pulse, and a hyperkinetic cardiac impulse. The heart sounds were normal and there was a soft mid-
systolic ejection murmur at the left sternal edge. The heart was slightly enlarged on chest x-rays and the appearance of the left border was suggestive of septal hypertrophy; the main pulmonary artery was slightly dilated (Fig. 1). The electrocardiogram showed left atrial, left ventricular, and septal hypertrophy. At cardiac catheterization, the mean pulmonary capillary venous pressure was 13 mmHg, with an 'a' wave of 20, and the pulmonary arterial pressure was 26/11 mmHg. There was an 'a' wave of 23 mmHg on the left ventricular pressure trace; the systolic level was 80. There was no systolic gradient between the left ventricle and the aorta in the control state, but after isoprenaline a 30 mmHg difference appeared. Left ventricular cineangiography showed a grossly thickened left ventricle, particularly in the septal region, and the left ventricular cavity was almost obliterated in systole (Fig. 2). The coronary arteries and aorta were normal.

After increasing exercise up to a work load of 500 kpm/minute for 5 minutes on a bicycle ergometer, the pulse rate increased from 80 to 180 a minute. A second exercise test was later carried out; no symptoms occurred at first, but after one to two minutes of rest, the pulse rate slowed and he then developed asystole. Sinus rhythm returned after 30 seconds external cardiac massage and the injection of 0-6 mg atropine sulphate, intravenously. The electrocardiogram was monitored throughout this episode.

In view of the history of recurrent syncope and the documented asystole, it was decided to implant a noncompetitive pacemaker. It was considered that a left ventricular myotomy should be
FIG. 1  Chest x-ray (see text for description).

performed at the same time. At operation, which
was performed by Mr. Donald Ross, pacemaker
electrodes were first sutured to the epicardial sur-
face of the heart, and then cardiopulmonary by-
pass started. On inspection the aortic valve was
normal, and below it there were a number of
vertical columns of hypertrophied muscle in
association with the ventricular septum. Two of
these columns were divided and removed. A ver-
tical myotomy was performed on the remaining
hypertrophied septum, and a wedge resection on
both sides of the myotomy. The non-coronary
cusp of the aortic valve was damaged by retraction,
and it was necessary to perform a pericardial
placement of the cusp. When bypass was
closed the aortic valve was competent, but
right atrium remained distended and non-
contractile in spite of stimulation. Satisfactory
cardiac action could not be restored despite all
treatments at resuscitation.

Discussion

The cause of syncope and sudden death in
hypertrophic obstructive cardiomyopathy has
not been well documented. Goodwin (1964)
considered that ventricular fibrillation was the
usual mode of death, and Braunwald et al.
(1964), while agreeing that dysrhythmias were
the most common cause, emphasized that
sudden increase in the obstructive element
might occur. Ventricular tachycardia and
fibrillation have been recorded in two patients
(Maurice et al., 1966). Atrial tachycardia
(Braunwald et al., 1964) and atrial fibrillation
are also known to occur (Westlake, Cohen,
and Willis, 1962; Shabetai and McGuire,
1963), and atrial fibrillation may cause syn-
ce (Glancy et al., 1970). In obstructive, as

opposed to other forms of cardiomyopathy,
Hollister and Goodwin (1963) found a low
incidence of dysrhythmias, including heart
block, throughout long periods of observation,
though they had no recordings at the time of
syncope or death. Similarly, other workers,
who have noted the low incidence of dys-
rhythmias, appear to have no electrocardio-
graphic recordings at the time of syncope or
sudden death (Braunwald et al., 1964; Mar-
riott, 1964; Penther et al., 1966; Meers-
chwam, 1969).

Johnson (1971) suggested that, in aortic
stenosis, baroreceptors in the left ventricular
wall may respond to a severe rise in ventricu-
lar pressure by initiating a depressor reflex,
with bradycardia, peripheral systemic vaso-
dilatation and dilatation of the splanchnic bed.
Thus severe hypotension and syncope may
be induced. Our patient with hypertrophic
obstructive cardiomyopathy developed, pos-
sibly due to a similar mechanism, bradycardia
and asystole at the onset of syncope. Syncope
after exercise is common in these patients, at
a time when obstruction to left ventricular outflow is greatest (Whalen et al., 1963;
Braunwald et al., 1964; Harrison et al., 1964),
and the abolition of syncope after surgical
relief of the obstruction supports the thesis
that mechanical obstruction may in itself be
responsible for the symptom (Ross et al.,
1966). Alternatively, ventricular dysrhyth-
mas, including asystole, may cause syncope
or death, either with or without an increase in

FIG. 2  Separate frames from the left ven-
tricular cineangiogram in (left) systole, and
(right) diastole (see text for explanation).
obstruction to left ventricular outflow. Therefore, their demonstration becomes of extreme importance in any patient, since sudden death may be avoided by the prophylactic use of a pacemaker.

References


Requests for reprints to Dr. S. Joseph, National Heart Hospital, Westmorland Street, London W1M 8BA.