Syncope in hypertrophic obstructive cardiomyopathy due to asystole

Simon Joseph, Raphael Balcon,1 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; Meerschwam, 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; Meerschwam, 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 angiographic 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.
Syncope in hypertrophic obstructive cardiomyopathy due to asystole. 

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 surface of the heart, and then cardiopulmonary bypass 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 vertical 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 incision of the cusp. When bypass was withdrawn the aortic valve was competent, but the right atrium remained distended and non-contractile in spite of stimulation. Satisfactory cardiac action could not be restored despite all attempts 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 syncope (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 dysrythmias, appear to have no electrocardiographic recordings at the time of syncope or sudden death (Braunwald et al., 1964; Marriott, 1964; Penther et al., 1966; Meerscham, 1969).

Johnson (1971) suggested that, in aortic stenosis, baroreceptors in the left ventricular wall may respond to a severe rise in ventricular pressure by initiating a depressor reflex, with bradycardia, peripheral systemic vasodilatation and dilatation of the splanchnic bed. Thus severe hypotension and syncope may be induced. Our patient with hypertrophic obstructive cardiomyopathy developed, possibly 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 dysrythmias, including asystole, may cause syncope or death, either with or without an increase in

FIG. 2 Separate frames from the left ventricular 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.
Syncope in hypertrophic obstructive cardiomyopathy due to asystole.

S Joseph, R Balcon and L McDonald

Br Heart J 1972 34: 974-976
doi: 10.1136/hrt.34.9.974

Updated information and services can be found at:
http://heart.bmj.com/content/34/9/974.citation

Email alerting service

These include:
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/