Ventricular arrhythmias in syndrome of balloon deformity of mitral valve

Definition of possible high risk group

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Twenty patients clinically identified as having balloon deformity of the mitral valve were studied to assess the incidence of ventricular arrhythmias. Echocardiography and phonocardiography were used to confirm the nature of the mitral valve lesion. Continuous 24-hour electrocardiograms were obtained from all patients and analysed by a computer and 2 observers. One patient had ventricular fibrillation and 3 patients had ventricular tachycardia. There was a high incidence of other less severe forms of ventricular arrhythmias. Eight patients had inferolateral ST and T wave abnormality on the resting electrocardiogram, and were described as having the auscultatory-electrocardiographic variant of the balloon mitral valve syndrome. The occurrence of serious ventricular arrhythmias (ventricular fibrillation and tachycardia) was significantly more frequent in this group. This raises the possibility that the resting electrocardiogram may identify those patients with balloon deformity of the mitral valve who are at risk from sudden death.

Balloon deformity of the mitral valve has been detected with increasing frequency since the characteristic mid-systolic click was first described by Cuffer and Barbillon (1887). In most patients the balloon deformity has little haemodynamic consequence but the condition has been associated with ventricular and supraventricular arrhythmias (Hancock and Cohn, 1966; Kreisman *et al.*, 1971; Gooch *et al.*, 1972; Sloman, Wong, and Walker, 1972; Jeresaty, 1973; Rizzon *et al.*, 1973; DeMaria *et al.*, 1974; Gulotta *et al.*, 1974; Schaal, Fontana, and Wooley, 1974; Winkle *et al.*, 1975), and sudden death has been reported in 9 patients (Hancock and Cohn, 1966; Barlow *et al.*, 1968; Trent *et al.*, 1970; Jeresaty, 1973; Marshall and Schappell, 1974; Rakowski *et al.*, 1975). The incidence of the lesion is not known accurately but in a recent necropsy series the abnormality was detected in 0.33 per cent of the population studied (Rizzon *et al.*, 1973), and clinical reports have suggested incidences from 6 to 10 per cent

(Markiewicz *et al.*, 1975; Brown, Kloster, and DeMots, 1975; Procacci *et al.*, 1975). Reports of the condition are confused by the heterogeneity of pathologies which can cause mechanical dysfunction of the mitral valve leaflets. The incidence of arrhythmias may vary in each group and it is possible that sudden arrhythmic death is more common in a specific population.

The object of the current study was to assess the type and significance of rhythm disorders in patients with idiopathic balloon deformity of the mitral valve.

Patients and methods

Clinical investigation

The subjects were 20 patients attending the department of cardiology in the Royal Infirmary and Royal Hospital for Sick Children of Edinburgh who had the clinical features of balloon deformity of the mitral valve without coexisting cardiovascular disease. There were 13 female and 7 male patients whose ages ranged from 12 to 61 years. Initially the diagnosis was made on the presence of a mid-systolic click or late systolic murmur or both. The

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patients were examined by 12-lead electrocardiography, phonocardiography, posteroanterior chest x-ray film, echocardiography, and 24-hour continuous ambulatory electrocardiographic monitoring. The latter recordings were obtained using the Oxford Instruments Miniature Analogue Tape Recording System which allowed all patients to undertake all normal activities without restriction. The electrocardiographic lead used was a modified V5.

24-hour electrocardiographic analysis

The tape recordings were replayed at 60 times real speed and processed by a Neilson Arrhythmia Computer (Neilson, 1974) programmed to detect ventricular ectopic beats. This produced a chart of the 24-hour ventricular ectopic beat activity. The computer also 'froze' arrhythmias in a solid-state memory from which they could be written out in real time on standard electrocardiographic paper. During the processing, the recordings were observed by two independent observers who scored arrhythmias and ensured accuracy of the computer programme if background noise levels or electrical axis shift necessitated modification. The tape recordings were repeated if because of poor quality less than 22 out of 24 hours were capable of analysis. No attempt was made to analyse the recordings of atrial ectopic beat activity but the system identified supraventricular tachycardia.

Arrhythmia definitions

Ventricular tachycardia was defined as 3 or more consecutive ventricular ectopic beats at a rate greater than 100/min. R-on-T phenomenon was defined as a ventricular ectopic beat falling within 85 per cent of the prevailing QT interval. Coupled ventricular ectopic beats were those occurring in pairs with R-R less than 400 ms. Ventricular ectopic beats were defined as frequent when they occurred more often than 30 an hour.

Auscultatory-electrocardiographic variant

Routine 12-lead electrocardiograms were carefully examined for inferolateral ST and T wave abnormalities and a group of patients with these changes was described as the auscultatory-electrocardiographic variant of the balloon deformity of the mitral valve (Humphries and McKusick, 1962; Barlow and Bosman, 1966; Sloman *et al.*, 1969; Liedtke *et al.*, 1973; Pocock and Barlow, 1970; Ehlers *et al.*, 1970; Stannard *et al.*, 1967).

Results

Clinical presentation

Of the 6 patients who were asymptomatic, 5 had

been referred because auscultatory abnormalities had been found on routine examination; in one patient, ventricular ectopic beats were noted on an electrocardiogram. Of the remaining 14 patients, 12 had complained of symptoms suggestive of an arrhythmia, namely syncope, palpitation, or dizziness; 7 of the 14 also complained of ill-defined chest pain. The recorded duration of symptoms or signs ranged from a few days to 30 years.

Routine electrocardiogram

In 9 of the 20 patients, the routine 12-lead electrocardiogram was normal. In 3, varying degrees of right bundle-branch block were present. In the remaining 8, non-specific ST and T wave changes were present in the inferolateral leads; these patients constituted the group already defined as the auscultatory-electrocardiographic variant of the balloon deformity of the mitral valve. There was no apparent difference in age or sex distribution in this group compared with the other patients, but complaints of lassitude, breathlessness, and chest pain were more common.

Posteroanterior chest x-ray film

Of the 20 patients, 8 showed minimal cardiomegaly on a routine chest x-ray film (cardiothoracic ratio >50%).

FIG. 1 Typical echocardiogram and simultaneous phonocardiogram.

FIG. 2 Numbers of patients (total 16) showing each type of ventricular arrhythmia on 24-hour continuous electrocardiogram.

Phonocardiography

The combined features of a mid-systolic click and late systolic murmur were confirmed in 18 of 19 patients. The phonocardiogram was repeatedly unsatisfactory in one patient; another had only a late systolic murmur: echocardiography confirmed mitral valve prolapse in both patients. Several attempts were required with some patients before a satisfactory record was obtained.

Echocardiography

This showed prolapse of one or both mitral valve leaflets in 17 patients. In 3 patients the mitral echogram was normal, but a midsystolic click and late systolic murmur were confirmed by phonocardiography. Fig. 1 shows a typical tracing with the simultaneous phonocardiogram.

FIG. 3 'Most severe' ventricular arrhythmia shown in 16 patients on 24-hour continuous electrocardiogram. The 'severity' of arrhythmia has been arbitrarily graded from infrequent VEBs to VF. Shaded areas represent patients with the auscultatory-electrocardiographic variant.

Ambulatory electrocardiographic monitoring

Only one patient had no arrhythmia recorded during the 24 hours. Supraventricular arrhythmias alone were found in 3 patients, which in 2 were at a rate of 160/min while the third, a girl of 12, developed atrial flutter with 1:1 atrioventricular conduction and a ventricular rate of 300/min.

The remaining 16 patients all showed ventricular arrhythmias of varying type. Fig. 2 shows the numbers of patients with specific arrhythmias on the 24-hour recording. Multiform ventricular ectopic beats were the most common form of ventricular arrhythmia. We have not attempted to document the numbers of episodes of each arrhythmia in each patient as on repeated recordings these figures were variable. Fig. 3 shows the results when the ventricular arrhythmias are arbitrarily graded in 'severity' from infrequent ventricular ectopic beats to ventricular fibrillation. Each patient is classified on the basis of the most 'severe' arrhythmia detected on their 24-hour electrocardiogram. Ventricular fibrillation was detected in one patient. Her tape recorder had been equipped with an alarm device which, when pressed, produced a pulse on a parallel tape track. The tracing of this event is shown in Fig. 4. This arrhythmia produced syncope but fortunately was self-terminating. Three patients showed ventricular tachycardia and in 4 the R-on-T phenomenon was the most 'severe' arrhythmia detected.

Relation between ventricular arrhythmias and resting 12-lead electrocardiogram

In Fig. 3 the hatched areas represent those patients with the auscultatory-electrocardiographic variant of the balloon mitral valve as previously defined. Thus ventricular fibrillation and ventricular tachycardia were seen only in patients with this abnormality on their electrocardiogram. This difference is statistically significant ($P < 0.05$) (χ^2 + Yates correction). This accounts for 4 of the 8 patients of this group. Two others had R-on-T phenomenon, one had multiform ventricular ectopic beats, and the remaining patient had only a supraventricular tachycardia at a rate of 160/min. The high incidence of 'serious' ventricular arrhythmias in those patients having inferolateral ST and T wave changes on the resting electrocardiogram suggests an association between these observations.

Discussion

Continuous 24-hour electrocardiographic monitoring of 20 patients with the clinical features of apparent idiopathic balloon deformity of the mitral

FIG. 4 Ventricular fibrillation detected in one patient. Patient pressed the alarm button as shown.

valve has shown that the incidence of arrhythmias, and in particular of 'serious' forms of ventricular arrhythmia, may be higher than has been widely recognized in the past. The coexistence of rhythm disorders and mechanical dysfunction of the mitral valve in the condition has been attributed to a myocardial disorder (Scampardonis *et al.*, 1973; Gulotta *et al.*, 1974) possibly secondary to an initially abnormal mitral valve apparatus (Nutter *et al.*, 1975). Whether or not the non-specific ST and T wave changes are taken as evidence of a disorder in the myocardium, it is possible that they may identify a group of patients who are at high risk from arrhythmias. In support of this suggestion are the 11 reported patients with balloon deformity of the mitral valve who died suddenly or had ventricular fibrillation, and for whom electrocardiographic data are available: 8 had the auscultatory-electrocardiographic variant of the syndrome and 3 showed QT prolongation (Hancock and Cohn, 1966; Barlow *et al.*, 1968; Trent *et al.*, 1970; Jeresaty, 1973; Marshall and Shappell, 1974; Rakowski *et al.*, 1975).

These results and the previous reports of sudden death suggest that it is important to diagnose this condition. Unfortunately the recognition of the syndrome is made difficult by the variability of symptoms and signs. Often the presenting complaints of vague chest pain, lassitude, and light-headedness suggest a functional diagnosis. The auscultatory features of a midsystolic click or late systolic murmur or both may not be constantly present in an individual and it is often difficult to obtain an echocardiographic record of the posterior leaflet of the mitral valve. Left ventricular cine-angiography carries a small but definite risk. For these reasons the auscultatory signs remain the

simplest and most useful diagnostic feature. The expense and personnel involved in long-term ambulatory monitoring prohibit its use in an undefined population, but our results suggest that it is possible that the 12-lead electrocardiogram can define a 'high risk' group for whom this technique could be of value.

Allen, Harris, and Leatham (1974) report a good prognosis for the condition of balloon deformity of the mitral valve but their exclusion of patients with electrocardiographic abnormalities may have biased the results in favour of those less likely to develop arrhythmias. In contrast, our experience confirms that ventricular arrhythmias are a part of the syndrome of balloon deformity of the mitral valve and suggests that all patients with an abnormal resting electrocardiogram by virtue of inferolateral ST and T wave changes should be investigated by long-term ambulatory monitoring even in the absence of symptoms. In the 20 patients studied all episodes of ventricular tachycardia and fibrillation were seen in individuals with such electrocardiographic abnormalities. It is not yet known whether ventricular arrhythmias associated with balloon deformity of the mitral valve are well tolerated or if they carry the long-term risk of sudden death. At present it is our policy to treat those patients who are symptomatic because of their arrhythmias and those in whom we have detected ventricular tachycardia or fibrillation. The choice of antiarrhythmic agent requires care as it is likely that long-term medication will be necessary. Some patients will respond to beta-adrenergic blocking drugs but others are resistant (Gooch *et al.*, 1972), and in this situation, the orally active antiarrhythmic drug, mexiletine (Talbot *et al.*, 1973; Campbell *et al.*, 1973, 1975) given three times daily has been well tolerated and effective in abolishing

ventricular tachycardia and fibrillation in our 4 patients with these rhythm disorders. Ambulatory electrocardiographic monitoring has proved useful in evaluating the effect of therapeutic intervention and has allowed us to use the minimal drug dosage consistent with arrhythmia control. It is clearly important to establish whether long-term antiarrhythmic therapy in these patients can reduce the risk of sudden arrhythmic death.

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