Familial sinuatrial disorder

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A family is reported in which the diagnosis of sinuatrial disorder was present in both the original patient and his father. With a typically varying PP' interval, it could be provoked in both of the other two children by stimulation of the vagus. The mother was normal.

Greenwood and Finkelstein (1964), in a review of 223 patients including their own and those reported by other authors in the preceding 50 years, listed the aetiological factors resulting in sinuatrial block as increased vagal tone, infections in childhood, including rheumatic carditis, diphtheria, and influenza and, in adults, ischaemic heart disease and digitalis intoxication. However, in their series 20 per cent of the patients were reputed to be otherwise normal.

It can be very difficult clinically to differentiate between sinuatrial block due to increased vagal tone, from that which is thought to be due to disease affecting the sinuatrial node and the specialized conducting tissue. Many patients with sinuatrial block present with symptoms associated with a severe bradycardia or transient cardiac asystole. In these the junctional atrioventricular specialized conducting tissue does not usually add its own rhythmic activity to maintain cardiac function. This suggests that during attacks of pronounced bradycardia the activity of the entire specialized conducting tissue is depressed. Since such attacks are commonly associated with intermittent bouts of tachycardia of all types originating from foci within this same conducting tissue, terms which have been used such as sinuatrial block and 'lazy sinus syndrome' (Ginks, 1970) are not satisfactory. We suggest that a more suitable description of this syndrome is sinuatrial disorder.

It is known that the vagus controls the electrical activity of the specialized conducting tissue in the dog (Brockman, 1965). In man, the effect of vagal activity can vary in degree and, in our experience, is not the sole determinant of the depression of activity of the specialized

conducting tissue, since in many patients with sinuatrial disorder the response of the conducting tissue to atropine is not normal. In these cases, however, an increase in heart rate can be obtained by ventricular pacing and in some patients, but not all, also by exercise. In older patients with associated ischaemic heart disease the syndrome of sinuatrial disorder may explain the condition previously described as bradycardiac angina (Fowler, Ikram, and Maini, 1970). The existence of isolated congenital sinuatrial disorder has been regarded as unproven (Muller and Finkelstein, 1966). Previous accounts of familial paroxysmal supraventricular dysrhythmias (Bacos, Eagan, and Orgain, 1960; Rokseth *et al.*, 1970) are probably descriptions of a part of the spectrum of sinuatrial disorder. To our knowledge this is the first report of its familial occurrence and provides evidence of the usually benign and asymptomatic nature of this condition.

Subjects

Case 1 was the original patient referred to the outpatient clinic because of a bradycardia which had been detected at a life assurance medical examination. He was aged 18 years.

Case 2 Aged 50 years, he accompanied his son to the clinic. He was examined initially because of our interest in the possibility of the presence of familial cardiac disease.

Case 3 Aged 20 years, the married sister of Case 1.

Case 4 Aged 14 years, the younger brother.

Case 5 Aged 40 years, the mother.

Case reports

Case 1 He had been well until 12 years of age when he had been treated for well-documented Sydenham's chorea. One year later he had become

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FIG. 1a Electrocardiogram, lead II, showing sinuatrial disorder recorded in 1965 during Sydenham's chorea.

FIG. 1b Bipolar chest lead, recorded in 1970, showing persistence of the irregularly variable PP' interval.

unconscious for some 10 minutes while taking part in a cross-country race. Since then he had been asymptomatic. On examination his heart rate was 42 a minute and irregular in rhythm. The heart sounds were normal, as was the chest x-ray. The electrocardiograms at this time, and those of six years previously, showed his heart rhythm to be typical of sinuatrial disorder with an irregularly variable PP' interval (Fig. 1a and b).

Case 2 His resting heart rate was noted to be 70 a minute and rose only to 96 a minute after he had been running on the spot until breathless. Like Cases 3, 4, and 5 he had no significant past medical history and clinical examination was normal.

Investigations

All the patients had their heart rate and rhythm monitored continuously using radiotelemetric electrocardiography. The paper speed was checked by introducing a 50 cps mains artefact onto the record. The patients were stressed by a modification of a whole-body maximum tolerance exercise test (Kaltenbach, 1968). While the patient was resting in the supine position, vagal stimulation was effected using carotid sinus and eyeball compression. The Valsalva response was then measured using as a standard the inflation of a sphygmomanometer mercury column to 60 mm for 10 seconds. Finally, atropine was given intravenously in aliquots of 0.6 mg until there was no further rise in heart rate. The results of these tests are given in the Table.

Discussion

In Fig. 1a and b, Case 1 is seen to have sinuatrial disorder as shown by a varying PP'

TABLE Summary of changes in heart rate and rhythm following exercise and vagal stimulation and inhibition

	Case 1	Case 2	Case 3	Case 4	Case 5
Control rate	50-70*	70	92	60	80
Resting PR interval (sec)	0.16	0.20-0.24	0.12	0.16	0.14
At peak exercise	125	140	155	145	165
Valsalva 0 sec	60	70	85	46	70
Valsalva 10 sec	90	83	140	98	95
Valsalva reflex stage	52	59	55*	50-70*	52†
Carotid/eyeball compression	Sinuatrial block	58	60	SAB	SAB‡
Atropine	88	88	—	120	130

* Varying PP' interval.

† Sinus arrhythmia.

‡ Sinuatrial block followed by atrioventricular junctional rhythm.

interval. His mean heart rate of 60 a minute was higher than the 42 a minute recorded at the clinic, and at this higher rate the varying PP' interval is not as obvious. After the maximum tolerance exercise his peak heart rate was 120 a minute with no obvious variation in the PP' interval. After recovery and 0.6 mg atropine intravenously, the PR interval varied irregularly from 0.14 to 0.30 sec (Fig. 2a). With a further 0.6 mg atropine the PR interval was 0.16 sec and there was a negligible PP' variation with a heart rate of 88 a minute (Fig. 2b). Complete atropinization was confirmed by the patient experiencing dryness of the mouth and throat and failure to obtain the sinuatrial block produced previously by eyeball compression.

In Case 2, the PR interval at rest varied

FIG. 2a After 0.6 mg atropine the PR interval is variable.

FIG. 2b After 1.2 mg atropine the PR interval is normal and the PP' interval is regular.

between 0.20 and 0.24 sec. Vagal stimulation by eyeball compression and the Valsalva manoeuvre did not produce a significant bradycardia. Neither was there a significant rise in heart rate after intravenous atropine to a total dose of 3.6 mg (body weight 82 kg). In Cases 3 and 4 increase of vagal tone after the Valsalva manoeuvre produced the varying PP' interval typical of sinuatrial disorder (Fig. 3). In Case 3 transient sinus arrest and a coronary sinus beat preceded the sinuatrial disorder. Atropine was not given to Case 3 since she was afraid of injections and we considered that a severe vagal response might have had serious consequences. The heart rate of Case 4 rose to 120 a minute after atropine. Case 5, the mother, developed sinus arrhythmia after the Valsalva manoeuvre, and sinus arrest occurred after eyeball compression but was then associated with atrioventricular junctional rhythm until recovery after a few seconds.

Conclusion Disorder of sinuatrial rhythm can be familial and asymptomatic. It is probably inherited as an autosomal dominant with varying degrees of penetration since the condition was present, to various degrees of severity, in the male and female children of the reputed father who was similarly affected.

Sinuatrial disorder can be easily missed if only short mounted electrocardiogram strips are studied, since only a longer tracing may show the irregular variability of the PP' interval which is one of the characteristics of this condition.

Though usually benign, in our experience the severe form of sinuatrial disorder can be associated with such disabling attacks of sinoatrial block that treatment by ventricular pacing may be required. Some patients may, in addition, give a history of attacks of supraventricular tachycardia, atrial fibrillation, and varying degrees of atrioventricular block. Sinuatrial disorder may only become clinically significant either when there is concurrent cardiovascular disease, such as cerebral or

FIG. 3 Sinuatrial disorder showing typical variable PP' interval, which occurred in Case 3 after the Valsalva manoeuvre.

coronary atherosclerosis resulting in attacks of syncope or bradycardiac angina, or when there is digitalis intoxication. The aetiological relation of persistent sinuatrial disorder with rheumatic carditis, diphtheria, and influenza, particularly in childhood and in the absence of familial studies, is not yet proven.

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