

Short reports

Hand and ECG tremor in spinal muscular atrophy

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SUMMARY The presence of hand and electrocardiogram (ECG) tremor was studied in 31 children with severe, intermediate, or mild form of spinal muscular atrophy. Clinical tremor of the hands was seen in 16 (59%) of 27 patients, all with benign forms of the disease. Nineteen patients had ECG tremors, of whom 17 had the mild or intermediate form. With the exception of one, all patients in the mild or intermediate group had hand or ECG tremor. Hand and ECG tremor are valuable in the diagnosis of the benign forms of spinal muscular atrophy; they are of no value in diagnosis of the severe infantile type.

The clinical presentation of patients with the more benign forms of spinal muscular atrophy (SMA) may be similar to that of patients with myopathies. The presence of muscle fasciculation (especially of the tongue) and tremor of the hands will distinguish the two.¹ In addition a 'muscle tremor' on the electrocardiogram (ECG) was found to be useful in the diagnosis of the benign forms of SMA.^{2,3} The ECG 'tremor' was thought to be the objective manifestation of the clinically observed hand tremor.² No patient with the severe infantile form of SMA, in whom hand tremor is uncommon, was included in these reports.

We reviewed our patients with SMA to evaluate the value of the ECG and tremor in the three clinical types of SMA.

Patients and methods

Thirty-one cases of SMA were studied. The diagnosis was made on the clinical picture and muscle histology. Particular note was made of the presence of a tremor of the hands and fasciculation of the tongue.

The ages of the patients ranged from birth to 32 years (Table 1). There was no difference between genders. Most patients were African (n=21) but 8 were Indian and 2 were coloured. They were divided into the three clinical categories according to severity of disease at presentation:³ (1) Severe, unable to sit unsupported (n=10). (2) Intermediate, able to sit unaided but unable to walk or stand unaided (n=10). (3) Mild (Kugelberg-Welander), able to stand and walk (n=11).

ECGs were recorded in all patients in the conventional way. Controls consisted of 100 ECGs performed on children with no known neuromuscular problems. ECGs from 10 cases of Duchenne dystrophy were also reviewed. The ECGs were examined by us for disturbance of the isoelectric line especially for the presence of irregular spikes occurring between the QRS complexes.

Results

ECG tremor (Table 2). An abnormality of the ECG baseline occurred in 19 of the 31 cases. This abnormality consisted of a very irregular baseline which

Table 1 Clinical data on children with SMA

Type	Number	Age at onset (years)		Age at presentation (years)		Gender M:F
		Mean	Range	Mean	Range	
Severe	10	Birth		5 months	1-15 months	1.5:1
Intermediate	10	0.7	0-1	2.6	1.4-6	1:1
Mild	11	2.8	1-12	12.3	2.5-32	1.75:1

Table 2 Hand and ECG tremor in children with SMA

	Number studied	Severe (%)	Intermediate (%)	Mild (%)	Total (%)
Hand tremor	27	0/8 (0)	7/10 (70)	9/9 (100)	16 (59)
Tongue fasciculation	27	5/7 (71)	5/9 (55)	6/11 (54)	15 (55)
ECG tremor	31	2/10 (20)	8/10 (80)	9/11 (81)	19 (61)

was more marked in the limb leads than the chest leads (Fig. 1). The irregularity was caused by spikes of varying amplitude (0.1–0.2 mV). The frequency could not be assessed because the spikes occurred at random and from different muscles. With the exception of 2 ECGs the abnormality was equally prominent in all the limb leads. Of the 19 ECGs that showed a 'muscle tremor', 8 occurred in the intermediate and 9 in the mild forms of SMA; only 2 of 10 children with the severe form showed this ECG tremor. The ECG tremor was not seen in any of the Duchenne or control patients.

Tremor of hands. The presence or absence of a coarse irregular tremor of the hands was recorded in 27 patients. Of these 16 (59%) had tremors, all of whom were in the intermediate or mild group (Table 2). None of the infants with the severe form had a tremor. Thirteen of the 16 patients with a hand tremor also had a 'muscle tremor' on ECG. Two of the 3 who did not show a muscle tremor on ECG had the mild form, and the other the intermediate form of SMA. One of the patients with the mild form who did not show ECG changes is an older sibling (aged 32 years) of another patient with both ECG and hand tremor.

Among the patients with the intermediate and mild forms of SMA only one patient did not show hand or ECG tremor.

Fasciculation of tongue. Presence or absence of

fasciculation of the tongue was recorded in 27 cases of whom 15 had fasciculation (this was present in 5 of 7 cases with severe form, in 4 of 9 with intermediate form, and in 6 of 11 with mild form of SMA).

Discussion

Our results show that the majority of patients with the more benign forms of SMA have a tremor of the hands and an irregular baseline (muscle tremor) on ECG. The ECG tremor is particularly pronounced over the limb leads and equally prominent in all limb leads; in nearly all patients it was much less prominent over the chest leads. It was present mainly in the chronic benign forms of SMA and seldom in the severe infantile form of the disease.

Thomas and Williams⁴ found discrete spikes in 6 of 550 ECGs performed in one month. Four of their 6 patients were available for detailed study and in each, evidence of anterior horn cell damage was found. Simultaneous recording with needle electromyogram showed perfect synchrony between the spikes on ECG and the typical fasciculation potentials on electromyogram. They concluded that these spikes on ECG represented fasciculation potentials from somatic musculature.

The data from our patients and those of Russman and Fredericks² support this conclusion. In our study ECG tremor was seen in the benign chronic forms of SMA suggesting the presence of progressive

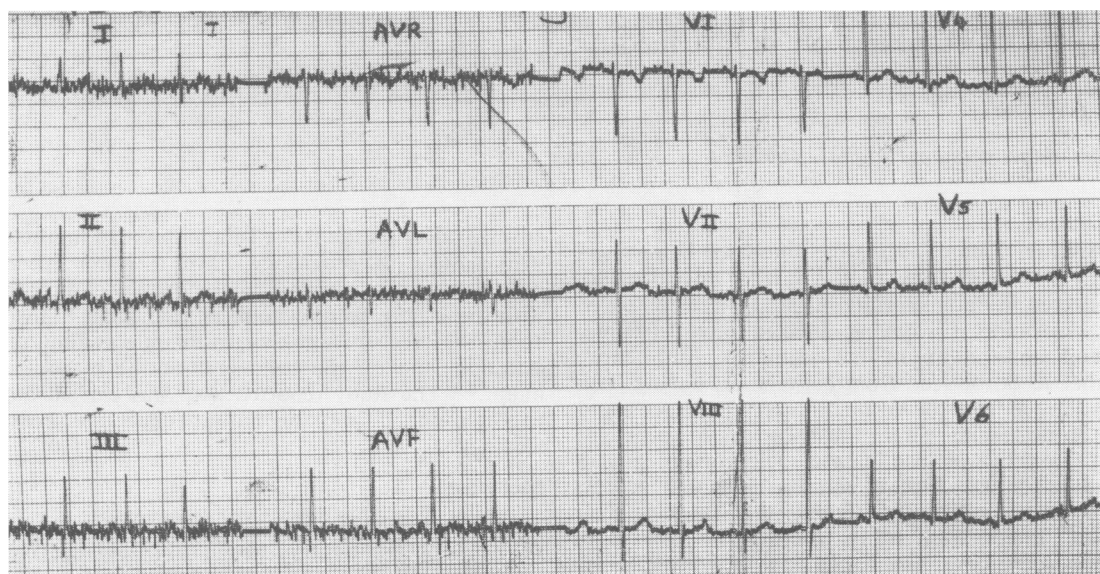


Fig. 1 ECG of a 6-year-old girl with intermediate spinal atrophy. Note spike discharges in all limb leads.

degeneration of the anterior horn cells. Its absence in the severe infantile forms of the disease suggests that fasciculation of somatic muscles is uncommon in this form. Buchthal and Olsen⁵ found fasciculation in only 2 of 30 patients with infantile SMA but in half of 61 patients with the juvenile form culled from reports. Hausmanowa-Petrusewicz *et al.*,⁶ found fasciculation in 10% of muscle samples of type Ia SMA (acute early infantile) compared with about 40% in type III (Kugelberg-Welander) type of SMA.

It is of interest that of two sisters with Kugelberg-Welander syndrome the elder (aged 32 years) showed neither ECG nor hand tremor; the younger sister aged 18 years showed a marked hand and ECG tremor. This suggests that the elder sister may have reached the 'burnt out' stage of the disease while in the younger it was still active. Serial ECGs were not done in any of our patients and this would be of interest in understanding the development of the disease. The close correlation between clinical tremor of the hands and ECG tremor with the type of SMA suggests that the clinical tremor is due to fasciculation of the forearm muscles. However, the presence of ECG tremor in the absence of hand tremor and vice versa indicates that fasciculation detected by ECG does not necessarily emanate from the same muscles causing the hand tremor.

Tongue fasciculation was found in 60% of our patients and this is in accordance with the experience of others.^{7,8} Its presence in a floppy infant with symmetric proximal muscle weakness is diagnostic of

SMA. We found the hand tremor and ECG tremor valuable in the diagnosis of the more benign forms of SMA. They were of no value in the severe infantile form of the disease

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Effects of smoking on breast feeding

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SUMMARY Infant feeding methods used by mothers who smoke and by mothers who do not, who delivered in one maternity hospital, were compared to determine if smoking affected breast feeding. Significantly more of the smoking group chose either to bottle feed from delivery or, having been discharged from the hospital breast feeding, changed to bottle feeding before the baby was 6 weeks old. This pattern was seen in each socioeconomic group and, as the smoking and non-smoking mothers were otherwise comparable, suggests that smoking may have a direct effect on breast feeding.

Smoking during pregnancy is known to have an

adverse effect on the fetus. Infants born to mothers who smoke in pregnancy have a lower mean birthweight and higher neonatal mortality compared with those born to non-smokers.^{1,2} Little is known about the effects of smoking, in the puerperium, on lactation, and on the behaviour of the breast-fed infant.^{3,4} The study reported here was carried out to determine if smoking has an effect on breast feeding.

Methods

The data were collected during a 3-month period in 1981 in a British military hospital in England. The hospital population was made up from service and local civilian mothers.