Electrocardiographic Findings in Children with Spinal Muscular Atrophy

Joh-Jong HUANG, MD, MS, Yuh-Jyh JONG,* MD, MS,
Ming-Yii HUANG,** MD, Chi-Hsiun CHIANG,* MD, PhD,
and Teh-Yang HUANG,* MD, PhD

SUMMARY

Most of the patients in this study with spinal muscular atrophy were found to have tremors of the isoelectric line in the electrocardiogram (ECG) tracings. There were a total of 47 cases of SMA (mean age 40.8 months). All three types of SMA had a similar incidence (about 80%) of tremors in the tracings ($p = 0.885$). In 7 cases the ECG tremors had an intermittent pattern. ECG tremors were commonly found in the majority of SMA patients and this finding, though non-specific, may suggest a possible SMA diagnosis. (Jpn Heart J 1996; 37: 239-242)

Key words: Electrocardiogram Spinal muscular atrophy Child

SPINAL muscular atrophies (SMA) are degenerative diseases with progressive loss of the alpha motor neuron of the spinal cord.¹⁾ The incidence of SMA is estimated to be about 5-10/100,000 births, mainly with an autosomal recessive inheritance.²⁾ Three types of SMA with different degrees of severity were recognized as type 1 (severe), type 2 (intermediate), and type 3 (mild) according to the different level of motor function they achieved.³⁾ Although all 3 types present with weakness, the clinical presentation of the milder forms may be quite similar to that of patients with myopathies.⁴⁾ In the past, it has been reported that tremors recorded on the electrocardiogram (ECG) might be a common finding of SMA, although with some inconsistency.⁵⁻⁷⁾ We have reviewed the ECG findings of patients with SMA to evaluate the presentation of the ECG tremors.

MATERIALS AND METHODS

The ECG records of forty-seven children who were diagnosed as having SMA were used. Age ranged from 2 months to 13 years 11 months (mean

From the Department of Family Medicine, *Department of Pediatrics, and **Department of Radiotherapy, Kaohsiung Medical College, Kaohsiung City, Taiwan.

Address for correspondence: Teh-Yang Huang, MD, PhD, Department of Pediatrics, Kaohsiung Medical College, No. 100, Shih-Chuan 1st Road, Kaohsiung City 807, Taiwan.

Received for publication November 24, 1995.

Accepted December 25, 1995.

40.8 $\pm$ 39.2 months). All cases were examined in the nonsedated state. The ECGs were performed at a standard speed of 25 mm per second with 0.1 mV per mm of height as the ordinary voltage calibration. ECGs were measured using an FK-12 electrocardiogram (Fukuda Denshi Co., Tokyo). The tremors of the isoelectric line in the routine electrocardiogram were given special attention.

The SMA patients were divided into the three clinical categories as follows: type 1 (severe SMA, Werdnig-Hoffmann disease); type 2 (intermediate or chronic Werdnig-Hoffmann disease); type 3 (late-onset juvenile SMA, Kugelberg-Welander disease).^{3,8)}

The chi square test was used to compare the incidence of ECG tremors among the three categories of SMA.

RESULTS

Forty-seven cases were included in the study (Table I). The sex ratio was 2: 1, with males predominating. The mean age was 40.8 $\pm$ 39.2 months. There were 7 type 1 cases with a sex ratio of 6: 1 and mean age of 6.7 $\pm$ 7.5 months; 27 type 2 cases with a sex ratio of 1.7: 1 and mean age of 39.8 $\pm$ 36.1 months; and 13 type 3 cases with a sex ratio of 1.6: 1 and mean age of 61.4 $\pm$ 43.4 months.

Table I. Electrocardiological Findings in 3 SMA Subtypes

SMA type	Number of cases	Age (months)	Sex ratio (Male: Female)	ECG Tremors		
				Continuous (%)	Intermittent (%)	Absent (%)
1	7	6.7 $\pm$ 7.5	6:1	6 (85.7)	0 (0)	1 (14.3)
2	27	39.8 $\pm$ 36.1	1.7: 1	16 (59.3)	6 (22.2)	5 (18.5)
3	13	61.4 $\pm$ 43.4	1.6: 1	9 (69.2)	1 (7.8)	3 (23.1)
Total	47	40.8 $\pm$ 39.2	2: 1	31 (66.0)	7 (14.9)	9 (19.1)

Figure 1. Tremors in the 12 ECG leads of a SMA type III patient.

Table 2. Incidence of Electrocardiographic Tremors in SMA Patients

	Year	Number of cases	ECG Tremors in SMA			
			Type 1 (%)	Type 2 (%)	Type 3 (%)	Total (%)
Russman et al ⁵⁾	1979	19	NR	NR	NR	19/19 (100.0)
Dawood et al ⁶⁾	1983	31	2/10 (20.0)	8/10 (80)	9/11 (81.8)	19/31 (61.3)
Coletta et al ⁷⁾	1989	13	3/3 (100.0)	8/8 (100.0)	1/2 (50)	12/13 (92.3)
This study	1995	47	6/7 (85.6)	22/27 (81.5)	10/13 (76.9)	38/47 (80.9)

NR = Not reported.

Figure 2. The intermittent pattern of tremors in lead II and AVL of the ECG of a SMA type I patient.

The pattern of ECG tremors is shown in Figure 1. This finding was noted in 38 cases (80.9%). The remaining 9 cases had no evidence of tremors in their tracings. Tremors were observed in six type 1 cases (85.7%), twenty-two type 2 cases (81.5%) and ten type 3 cases (76.9%) (Table II). All three types of SMA had a similar incidence of ECG tremors ($\chi^2 = 0.243$, $p = 0.885$). Of the 38 cases with ECG tremors, all tremors were noted in limb leads. However, only twelve cases were found to have had tremors in precordial leads: one type 1, eight type 2 and three type 3.

Not all the tremors showed a continuous pattern in the tracings; there were 7 with an intermittent pattern: six type 2 and one type 3 (Figure 2). Most of these were found in limb leads. Only one case with an intermittent pattern was found to have tremors in both the limb and precordial leads.

DISCUSSION

The most common, but non-specific, ECG finding of SMA patients in the study was the tremors, which were present in the tracings of most patients. All tracings with tremors were noted in the limb leads, with only some in the precordial leads. The origin of the tremors was postulated to be from the fasciculation of somatic muscles, which may be due to neuronal reinnervation activity during the course of degeneration of the alpha motor neurons.^{6,7,9-12)} The ECGs of patients who are anxious, cold or tense may show artifacts which are similar to ECG tremors. However, the incidence rate of those artifacts in the general population has been shown to be much lower than that of ECG tremors of SMA

patients.¹³⁾

There was no difference in the incidence of tremor in different SMA types. About 80% of the cases of each type had ECG tremors.⁵⁾ This finding was different from previous studies. Some reported that all or almost all patients had tremors; others reported that the majority of mild or intermediate type patients had the tremors, but they were uncommon in the severe type.^{6,7)} Nevertheless, all of these studies were with a small series with possible bias.

Not all the ECG tremors were present continuously in the tracings. The ECG tremors may group into clusters with no electric activity from the ECG isoelectric baseline between clusters of tremors (Figure 2). Some (7/38) showed this intermittent pattern. Most of the intermittent ECG tremors were noted in the limb leads. Only one case was noted in both the limb and precordial leads. This finding was not present in previous studies, and the mechanism and the meaning of the finding need further study.

Because the ECG tremors were noted in the majority of the SMA patients in this study, though non-specific, this phenomenon may suggest a possible SMA diagnosis.

ACKNOWLEDGMENTS

We thank Miss Chiung-Ping Ou for her kind help during this study.

REFERENCES

1. Dubowitz V. Muscle disorders in childhood. Philadelphia: Saunders, 1978: 146-78.
2. Emery AEH. Population frequencies of inherited neuromuscular diseases: A world survey. *Neuromusc Dis* 1991; 1: 19-29.
3. Munsat TL. Workshop report: International SMA collaboration. *Neuromusc Dis* 1991; 1: 81.
4. Moosa A, Dubowitz V. Spinal muscular atrophy in childhood. *Arch Dis Child* 1973; 48: 386-8.
5. Russman BS, Fredericks EJ. Use of the EKG in the diagnosis of childhood spinal muscular atrophy. *Arch Neurol* 1979; 36: 317-8.
6. Dawood AA, Moosa A. Hand and EKG tremor in spinal muscular atrophy. *Arch Dis* 1983; 58: 376-86.
7. Coletta C, Carboni P, Carunchio A. Electrocardiographic abnormalities in childhood spinal muscular atrophy. *Int J Cardiol* 1989; 24: 283-8.
8. Souidun S. Neuromuscular diseases. In: David RB editor. *Pediatric Neurology for the Clinician*. Norwalk: Appleton and Lange, 1992: 410-1.
9. Dubowitz V. Infantile muscular atrophy, a prospective study with particular reference to a slowly progressive variety. *Brain* 1964; 87: 707-12.
10. Simpson JA, Thomaidis T. Fasciculation and focal loss of nerve accommodation in peripheral neuropathies. *Acta Neurol Scand* 1988; 77: 133-41.
11. Swash M. Motor neuron disease. *Postgrad Med J* 1992; 68: 533-7.
12. Leigh PN, Ray-Chaudhuri K. Motor neuron disease. *J Neurol Neurosurg Psychiatry* 1994; 57: 886-96.
13. Thomas DJ, Williams DO. Detection of somatic muscle fasciculation on electrocardiograms. *Br Med J* 1976; i: 557-9.