Eur Arch Psychiatr Neurol Sci (1987) 236:358–363

European Archives of Psychiatry and Neurological Sciences

Somatosensory Evoked Potentials Following Median and Tibial Nerve Stimulation in Patients with Friedreich's Ataxia

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Summary. The changes in evoked potentials following median and tibial nerve stimulation in nine patients with clinically defined Friedreich's ataxia are reported and discussed. The response originating in the brachial plexus (Erb's point potential) was absent or reduced in amplitude with no prolongation of peak latency, and the response generated in the cauda equina (N18) was absent in all cases. Conduction time from the brachial plexus to cervical spine and medulla oblongata was normal, whereas the central conduction time (N13a/N20, N13b/N20) was delayed. There was moderate to marked attenuation of the primary cortical response to median nerve stimulation. In one patient N20 disappeared during the course of the disease as opposed to the persisting subsequent negative wave, the latter thus simulating a very marked delay in the primary cortical response. Accordingly the cortical response to tibial nerve stimulation, which was only present in two patients and was markedly delayed, might represent a later potential with the primary response absent. The findings are consistent with neuropathological descriptions of a dying back neuropathy with primary axonal degeneration concerning the 1st order sensory neuron. In addition there is evidence either of delayed conduction in 2nd and 3rd order sensory neurones or of abnormal synaptic transmission.

Key words: Friedreich's ataxia – Medianus-SEP – Tibialis-SEP

Introduction

The diagnosis of Friedreich's ataxia (F. a.) has been and still is mainly based on clinical findings. Clinical presentation and course of this progressive degenerative disease are fairly uniform (Lubozynski and Roelofs 1975; Babeau 1978). Nevertheless additional neurophysiological data indicating the site of lesions within the nervous system may be helpful in distinguishing between F. a. and other degenerative diseases with solely peripheral or central manifestation, especially in the early stages of the disorder.

Recording somatosensory evoked potentials after tibial and median nerve stimulation may provide such data as opposed to peripheral neurography, the findings of which are the same in for example F. a. and peroneal muscular atrophy type II (Sauer 1980). We studied nine patients with F. a. in order to establish typical changes in the evoked responses, the presence of which might be an additional diagnostic criterion.

Subjects

Four men and five women with defined F. a. were studied, the diagnosis being based on clinical features. Clinical data are summarized in Table 1. All patients but one fulfilled at least 5 of the primary and 1 of the secondary clinical diagnostic criteria for F. a. proposed by Geoffroy et al. 1976. In one patient with a disease duration of only 18 months, 4 of the primary, and 3 of the secondary clinical features were present. As indicated by Harding (1981) limb and trunk ataxia together with absent tendon reflexes in the legs are the only consistent diagnostic criteria within 5 years of presentation, and both were found in this patient.

A marked decrease or absence of sensory nerve action potentials together with a normal or slightly subnormal nerve conduction velocity was demonstrated in all patients (Table 2), the former being a consistent, but not a pathognomonic neurographical finding in F. a. (McLeod 1971; Peyronnard et al. 1976; Sauer 1980; Oh and Halsey 1973; Jones et al. 1980; Dyck et al. 1971). The disease duration was between 18 months and 15 years in our patients, with a mean duration of 11 years. The age of the youngest patient was 13 years, the age of the oldest 27 with a mean age of 21. In eight cases evoked responses were recorded both after median and tibial nerve stimulation, in one patient only stimulation of the median nerve was performed.

Methods

Median and tibial nerves were stimulated by a bipolar surface electrode (anode distal, cathode proximal), placed at the patients wrist or ankle. A rectangular stimulus with a duration of 0.1 ms was delivered at a frequency of 5 Hz. Stimulus intensity was set 4 mA above the motor threshold of the opponens pollicis and flexor hallucis brevis muscle. Recording was performed using subcutaneous platinum needle electrodes (Disa 25 C 04). Responses after median nerve stimulation were recorded simultaneously at Erb's point, the spinous processes C7 and C2 and on the scalp contralateral to the site of stimulation at C3' and C4' respectively (i.e. 2 cm dorsal to C3 and C4). The reference electrode was placed at Fz. Responses to tibial nerve stimulation were recorded at the spinous processes L5, L1, C2 and on the scalp at Cz' (i.e. 2 cm dorsal to Cz). Reference electrodes were placed at Fz for Cz' and C2 and over the iliac crest for L1 and L5. The bandpass of the amplifier was set to 10–1000 Hz (scalp recording) and to 50–1000 Hz (spinal re-

Table 1. Clinical features of Friedreich's ataxia in patients studied

Patients	Sex	Age (years)	Duration of disease	Relatives affected	Deformity	Deep tendon reflexes		Babinski sign	Ataxia of limbs		Stance gait
						Upper extremities	Lower extremities		Upper	Lower	
H. B.	F	22	9	–	K	A	A	A	+	++	++
S. E.	F	27	10	1 brother	K, PC	A	A	+	+	++	+++
L. R.	F	20	14	–	K, PC	A	A	+	+++	+++	
S. B.	M	23	13	1 sister	K, PC	A	A	+	+	++	++
S. A.	M	25	15	2 brothers	K, PC	A	A	A	++	++	++
S. J.	M	16	8	2 brothers	K	A	A	+	++	+++	++
S. R.	M	26	14	2 brothers	K, PC	A	A	A	+	++	++
S. M.	F	15		–	–	+	+	+	++	++	++
C. S.	F	13	1.5	–	K, PC	+	A	A	++	++	++

K = kyphoscoliosis
PC = pes cavus (claw foot)
A = absent

Patients	Dysarthria	Vibration sense of lower limbs	Position sense of lower limbs	Motor weakness	Abnormal ocular motility	Associated abnormalities
H. B.	A	MR	SR	Distal lower limbs	GEN	–
S. E.	+	A	A	Paraparesis	GEN	–
L. R.	+	A	A	Tetraparesis	DON	Cardiomyopathy
S. B.	+	MR	A	Paraparesis	–	–
S. A.	+	SR	MR	Distal lower limbs	DON	Diabetes
S. J.	+	SR	MR	Paraparesis	DON	Cardiomyopathy
S. M.	A	SR	N	Distal lower limbs	DON	–
C. S.	A	SR	N	A	GEN	Cardiomyopathy

SR = slightly reduced
MR = markedly reduced
A = absent
N = normal
GEN = gaze evoked nystagmus
DON = disturbance of optokinetic nystagmus

Table 2. Sensory nerve conduction velocities and amplitudes of sensory nerve action potentials in the patient group

Patient	Sensory nerve conduction velocity	Amplitude of sensory nerve action potential
H. B.	Normal	Markedly reduced
S. E.	40 m/s	1 μV
L. R.	Slightly reduced	Markedly reduced
S. B.	45.4 m/s	0.3 μV
S. A.	50 m/s	0.4 μV
S. J.	43.2 m/s	0.4 μV
S. R.	50.3 m/s	0.4 μV
S. M.	Slightly reduced	Markedly reduced
C. S.	sensory nerve action potentials absent	

cording). On each side 2000–4000 sweeps were averaged. All patients were clinically evaluated by a neurologist and electroneurography was performed in each case. The technique employed and the normative data of the control groups have been described in detail elsewhere (Stöhr and Riffel 1982; Riffel et al. 1984).

Results

Somatosensory Evoked Potentials after Median Nerve Stimulation

The results are summarized in Table 3. In seven of the patients the Erb's point potential (EP-potential) following median nerve stimulation at the wrist was recorded with normal latency (Fig. 1). In the remaining two patients the response at Erb's point was absent.

In all patients the amplitude of the EP-potential was markedly reduced (mean amplitude 0.2) or absent. The amplitude of the spinal cord potentials N13a and N13b, recorded at C7 and C2, was less reduced as compared to the amplitude of the EP-potential. The mean amplitude ratio EP/N13a in the patient group was 0.6, in the control group 2.3 ± 1.2. In six cases the reduction in amplitude was evident even if one only regards the recording obtained at Erb's point: In these cases the primary response at Erb's point proved to be even smaller than the following negative wave, which corresponded to the cervical potential N13 (Fig. 1). No cervical response was elicited in only one patient, and the EP-potential was also absent.

Table 3. Amplitude and latency parameters of short latency somatosensory evoked potential (SEP) components after median nerve stimulation in patients with Friedreich's ataxia

Patients	Latencies (ms) Erb's point (EP) potential	NSEP 13a	NSEP 13b	N20	Interpeak latencies (ms) EP-N13a	EP-N13b	N13a-N20	N13b-N20	Ratio of amplitudes N20/N13a	EP/N13a
H.B. F	–	17.7	17.3	24.3	–	–	6.6	7.0	0.9	–
S.E. F	–	–	–	–	–	–	–	–	–	–
L.R. F	9.2	13.1	13.2	24.6	3.9	4.0	11.5	11.4	0.2	0.8
S.B. M	11.3	16.1	16.3	27.0	4.8	5.0	10.9	10.7	2.0	0.9
S.A. M	10.8	14.4	14.4	24.0	3.6	3.6	9.6	9.6	0.6	0.2
S.J. M	10.5	14.8	14.6	25.4	4.3	4.1	10.6	10.8	0.5	0.1
S.R. M	10.8	14.8	14.6	24.3	4.0	3.8	9.5	9.7	1.0	0.3
S.M. F	10.0	13.1	13.2	20.8	3.1	3.2	7.7	7.6	0.1	0.9
C.S. F	9.4	13.6	13.8	24.4	4.2	4.4	10.8	10.6	1.1	0.7
Average of patients						4.0	4.0	9.7	9.7	
Control										
$\bar{x}$	10.2	13.5	13.7	19.3	3.4	3.5	5.6	5.6	1.4	2.3
$\bar{x}+2.55$	12.4	15.8	15.9	22.3	4.9	5.4	7.3	7.1		
$\bar{x}-2.55$									0.7	1.1

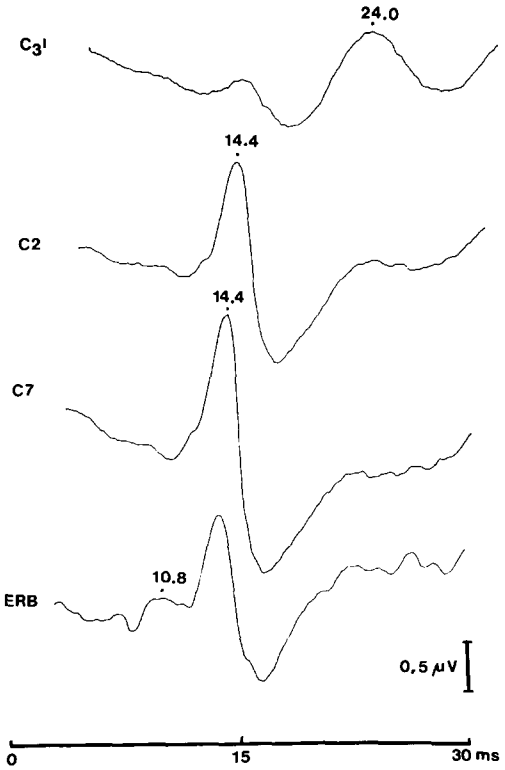

Fig. 1. SEP after median nerve stimulation (patient S.A.) EP-potential recorded with normal latency and markedly reduced amplitude, smaller than the following negative wave. Amplitude of N20 reduced as compared to N13

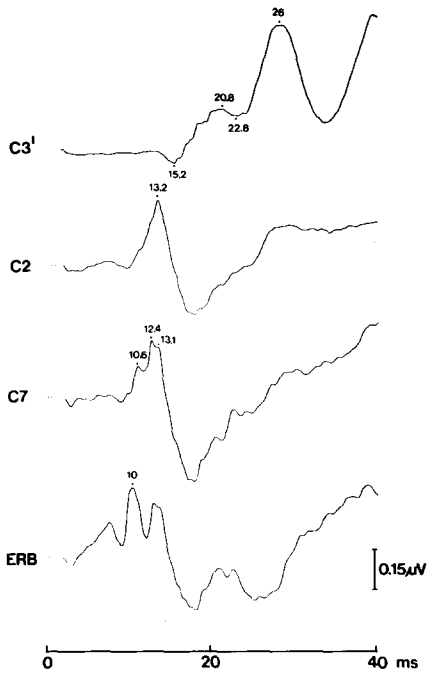

Fig. 2. SEP after median nerve stimulation (patient S.M.) N20 can be identified as a small peak in the ascending slope of the following cortical negative wave

In three cases the latency of N13a and b was within the normal, in three cases within the upper normal range. In two patients there was mild to moderate delay of N13. In all seven patients in whom all three potentials could be identified, the conduction time from Erb's point to C7 and C2 was normal, and in three cases it was within the upper normal range.

The average interpeak latency from EP-potential to N13b (a) was 4.0 (4.0) in the patient group and 3.54 (3.37) in normal subjects. The cortical response N20 was identified in all patients except one, in whom the EP-potential and cervical responses were also absent. The N20/N13a ratio of amplitudes was below the normal range in four patients (Fig. 1), and within the lower normal range in a further three.

In all patients except one there was a delay in conduction from C7 and C2 to the cortex, in one case conduction time was within the upper normal range. In one patient (S.M.) the

disappearance of the primary cortical negative potential N20 was demonstrated in a follow-up examination after several months. In Fig. 2 N20 can be identified as a small, flat peak in the ascending slope of the following negative potential. Several months later the amplitude of this flat peak had become reduced to such a degree, that it could no longer be identified. Only the following negative wave still persisted (Fig. 3).

One of our patients was an early case, the first symptoms having been observed 18 months previously. In this 13-year-old girl the amplitude of the EP-potential was reduced according to the EP/N13a ratio of amplitudes, and the cortical response was moderately delayed with a prolonged interpeak latency N13a to N20.

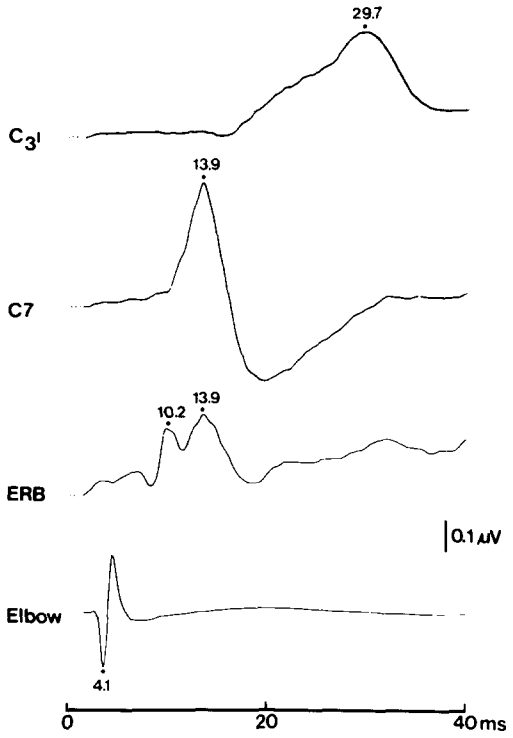

Fig. 3. SEP after median stimulation, same patient as in Fig. 2, follow-up recording. N20 has disappeared, only the following cortical negative wave can still be identified

Somatosensory evoked potentials after tibial nerve stimulation

The results are summarized in Table 4. In three patients no potentials were recorded at spinal or cortical levels following tibial nerve stimulation. In three patients only the negative potential N22 at th L1 vertebral level could be evoked. In two patients there was a N30 response at the C2 vertebral level, whereas N18 at the spinous process L5 was absent in all patients. In these two cases the cortical recording displayed a late positive deflection representing more a P60 wave than a markedly delayed P40 response (Fig. 4). The latency of the cervical response as well as the conduction time from N22 to N30 were within the upper normal range in one case and slightly above the upper limit in the other.

In no patient was the lumbar potential N22 delayed. It's amplitude was comparatively high, so as to allow unequivocal identification. In the early case mentioned evoked responses to tibial nerve stimulation were already absent.

Discussion

Previous pathological and electrophysiological findings in F. a. (Lubozynski and Roelofs 1975; Barbeau 1978; Sauer 1980; McLeod 1971; Peyronnard et al. 1976; Hughes 1968; Dyck and Lais 1973; Greenfield 1954; Kinnier Wilson 1954; Brown 1962; Blackwood et al. 1963; Peters 1970; Cavanagh 1964) were consistent with primary axonal degeneration and secondary segmental demyelination, the neurones in the systems involved dying back gradually from the periphery of the axon to the cell body. In the peripheral nerve a selective loss of large myelinated A-alpha sensory fibres has been described, whereas fine A-delta and C-fibres are spared. The EP-potential, generated in the brachial plexus, was attenuated in the patients, but not delayed. Provided no total degeneration and loss of all large peripheral fibres has occurred, the remaining ones, conducting with normal velocity, will produce electrical activation of the brachial plexus without significant delay. Due to the reduction in number of conducting large fibres, the generated potential is attenuated. N18, the lumbar potential generated in the cauda equina and lumbosacral roots (Delbeke et al. 1978; Dimitrijevic et al. 1978; Stöhr et al. 1982) the amplitude of which is low even in normal subjects, is attenuated to such a degree, that it can no longer be re-

Table 4. Somatosensory evoked responses after tibial nerve stimulation, latencies and interpeak latencies in patients with Friedreich's ataxia

Patients	Latency (ms)			Cortical response	Interpeak latency (ms)			
	N18 at L5	N22 at L1	N30 at C2		N18/N22	N22/N30	N22/cortical response	N30/cortical response
H. B. F	–	–	–	–	–	–	–	–
S. E. F	–	–	–	–	–	–	–	–
L. R. F	–	19.7	–	–	–	–	–	–
S. B. M	–	23.8	–	–	–	–	–	–
S. A. M	–	21.8	31.4	59.3	–	9.6	37.5	27.9
S. J. M	–	22.2	–	–	–	–	–	–
S. R. M	–	23.8	34.6	71.6	–	10.8	–	–
C. S. F	–	–	–	–	–	–	–	–
Control				P40				
$\bar{x}$		21.7	29.5	38.8		7.9		
$\bar{x} + 2.55$		25.8	34.3	43.9		10.4		

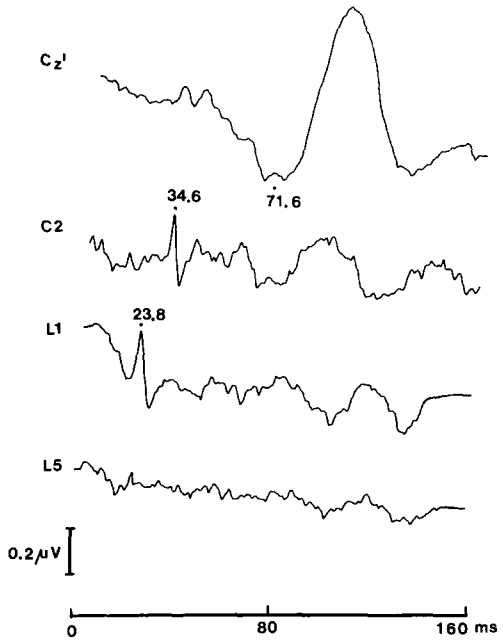

Fig. 4. SEP after tibial nerve stimulation (patient S.R.). Late cortical response representing more a P60 wave than a markedly delayed P40 response

corded using surface electrodes. As proposed by Jones 1977, Allison et al. 1981 and Stöhr et al. 1982, and supported by the findings of Hume and Cant 1978 and Chiappa et al. 1980, N13a recorded at C7 originates in the dorsal horn, whereas N13b recorded at C2 is genereated near the cervico-medullary junction. With the same pathological process affecting both the central and peripheral axons of the 1st order sensory neuron, no delay in conduction from brachial plexus to the cervico-medullary junction is to be expected.

Accordingly in our patients the interpeak latency as measured from EP potential to N13b, was within the normal range. A marked delay of N13 (but not of the EP-potential/N13b interpeak latency) was observed in one of our patients (H.B.). In this case loss of peripheral fibres had already reached such a degree, that no EP-potential could be registered. Advanced degeneration of peripheral or central axons presumably had already produced marked secondary demyelination accounting for the delay. In no case could a reduction in amplitudes equal to the attenuation at Erb's point be observed at C2. This might indicate that the number of intact large central axons of the 1st order sensory neuron is greater than that of large peripheral ones or might be the result of a trans-synaptic amplifying mechanism.

Pathological changes in central sensory fibres in the posterior columns of the spinal cord have been reported to affect the fasciculus gracilis more than the fasciculus cuneatus (Kinnier Wilson 1954). This is reflected by the high percentage of patients in whom no spinal potentials (75%) and no cortical response (75%) could be evoked after tibial nerve stimulation, wheras spinal responses and cortical responses were absent in only 11% after median nerve stimulation. There is also evidence of more serious involvement of peripheral sensory fibres in the legs as compared to the upper extremities. In all patients the response at the L5 vertebral level was absent, the components of which are held to originate in lumbosacral nerve roots. (Delbeke et al. 1978; Dimitrijevic et al. 1978; Stöhr et al. 1982). The EP-potential on the other hand was missing in only two patients out of nine. The recording of evoked potentials after tibial nerve stimulation yielded no sign of marked secondary demyelination up to the C2 vertebral level. However, the response N22 recorded at L1, presumably originating in the lumbosacral dorsal horn, was not delayed in any of the patients, the response recorded at C2 was in time or only very slightly delayed.

Reports concerning the involvement of the 2nd order neuron within the sensory pathway in F. a. are not consistent. In 1954 Greenfield referred to degeneration of cuneate and gracile tracts passed on by trans-synaptic degeneration to the cuneate and gracile nuclei and the medial fillet. Kinnier Wilson (1954) found that degeneration of dorsal columns extended to the nucleus gracilis and cuneatus, but seldom beyond. According to Peters (1970) the degenerative process spreads to the lemniscus medialis not as a rule, but in some cases. Our findings in accordance with the reports of others (Bergamini et al. 1966; Noel and Desmedt 1976; Desmedt and Noel 1973) suggest a high frequency of lesions affecting the 2nd or 3rd order neuron within the sensory pathway, the interpeak latency N13b to N20 being prolonged in 78% of patients.

There have been attempts to explain the delay in conduction from C2 to the cortex (Sauer 1980; Jones et al. 1980). Earlier secondary demyelination than in the periphery due to the different nature of the myelin sheath surrounding the central axon, which is provided by oligodendrocytes and astrocytes, is ruled out by the fact that conduction time from Erb's point to C2 is normal. The fibres giving rise to the response at C2 have a myelin sheath of the "central type" as well as 2nd and 3rd neurones. Another explanation for the discrepancy between normal peripheral conduction velocity and delay of N20 has involved shrinkage of axons and secondary demyelination in a "dying back" form of neuropathy occurring first in the most distal parts of the axons. If pathological changes are mainly located distal to the wrist, were the stimulus is delivered, the segments of fibres involved in the pathological process do not take part in mediating the response at the brachial plexus and cannot cause a delay. If changes at the rostral extremity of dorsal columns are invoked to account for the delay of N20, however, the C2 response, held to be generated near the nucleus cuneatus, ought to be delayed as well.

Evidence of more widespread involvement of the CNS in F. a. exists. Pathological changes within the central auditory pathway and visual pathway are reflected by an increased latency in visual evoked cortical potentials and disappearance of wave V of the auditory evoked response, which is generated within the brainstem, whereas the peripheral response persists (Pedersen and Trojaborg 1981; Pelosi et al. 1984; Taylor et al. 1985). Whether there is abnormal synaptic transmission within the central pathways or whether there are changes in axons and myelin sheaths rostral to the medulla oblongata remains to be resolved. To date no relevant neurohistochemical studies exist. An additional useful tool in assessing this problem might be positron emission tomography.

Bergamini et al. 1966 and Jones et al. 1980 described a very marked delay of N20 in patients with F. a. Our case report of patient S.M. suggested a progressive amplitude reduction and ultimate disappearance of the primary cortical component N20 in the course of disease. If only the second recording in our patient (Fig. 3) would exist one would be inclined to regard the still persisting 2nd cortical negative response as a deformed N20 with a markedly prolonged latency and strik-

ingly flat ascending slope. The reports on a very marked delay of the cortical response might be due to such a misinterpretion of data.

The persisting cortical negative wave was recorded at 29.7 ms. It might represent the 2nd part of a primary cortical response broken down into two peaks, as described by Noel and Desmedt 1976, the 2nd component following the 1st at an average interval of 9.4 ms. The explanation is still hypothetical: a thalomocortical loop is assumed to be formed by re-innervation of synapses in the posterior lateral nucleus, which due to degeneration of fibres within the medial lemniscus lost their afferent neurons. Whether the late cortical responses recorded after tibial nerve stimulation represent the 2nd cortical positive potential P60 or are generated by a similar hypothetical mechanism cannot be decided as yet.

Even if many theoretical problems are still to be resolved, the pattern of changes observed in sensory evoked potentials of patients with F. a. will be helpful in the diagnostic process, as such changes do not occur in other neurological diseases.

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Received December 10, 1986