F response and somatosensory and brainstem auditory evoked potential studies in HMSN type I and II

V Scaioli, D Pareyson, G Avanzini, A Sghirlanzoni

Abstract

To evaluate conduction along the proximal and distal segments of motor and sensory long limb nerves, as well as along the very short acoustic nerve, F response and somatosensory and brainstem auditory evoked potential were studied in a series of patients with hereditary motor and sensory neuropathy (HMSN) types I and II. A diffuse and comparable slowing of conduction in proximal and distal nerve segments, as well as along the acoustic nerve, seems to favour a primary myelin defect in HMSN I. F response and motor conduction velocity showed a similar derangement in both proximal and distal motor segments. Latencies of somatosensory evoked potentials were symmetrically prolonged and correlated with motor nerve impairment. Central conduction times were normal. Studies of brainstem auditory evoked potentials showed a high incidence of acoustic nerve involvement, the most evident abnormality being a statistically significant increase in the latency of the I wave. Our data seem to support the presence of primary myelinopathic damage in HMSN I.

(*J Neurol Neurosurg Psychiatry* 1992;55:1027-1031)

Hereditary motor and sensory neuropathy (HMSN) type I is well characterised by dominant autosomal inheritance, a slowing of nerve conduction and histological findings of nerve hypertrophy with segmental demyelination and remyelination.¹ It has long been disputed, however, whether HMSN I is due to a primary axonal or myelin defect. According to Dyck *et al*,^{1,2} segmental demyelination is caused by a primary axonal derangement. Lewis and Sumner, however, found a uniform slowing of conduction along different segments of the peripheral nerves of patients with HMSN I,³ suggesting a primary myelinic pathogenesis.⁴

A distoproximal gradient in conduction velocities is to be expected in neuropathies due to a primary axonal defect; for the same reason, it is to be expected that primary axonal neuropathies would affect conduction velocity much more in long than in short nerves, whereas any conduction impairment due to primary myelin involvement should affect short and long nerves similarly.

We report the results of a clinical study aimed at evaluating conduction along the

proximal and distal segments of motor and sensory long limb nerves, as well as along the acoustic nerve (the shortest nerve in the body), in a series of patients affected by HMSN type I. A group of patients with HMSN type II, whose axonal pathogenesis is undisputed, were also studied in order to have a reference sample. Neurophysiological studies included the evaluation of motor conduction velocity (MCV), F-response, and somatosensory and brainstem auditory evoked potentials (SEPs and BAEPs).

Materials and methods

Patients

Sixty five patients seen at the "C Besta" Neurological Institute of Milan and diagnosed as having Charcot-Marie-Tooth disease entered the study. All were evaluated according to a clinical, genetic, and electrophysiological protocol.⁵ Briefly, the necessary criteria for a diagnosis of HMSN were the presence of symmetric sensorimotor peripheral neuropathy with a chronic course, associated with skeletal deformities (*pes cavus*); positive family history; and exclusion of other hereditary or acquired chronic polyneuropathies (on clinical and laboratory grounds).⁵

Further subdivision into HMSN type I and type II was based on nerve conduction studies. According to the criteria of Dyck¹ and Harding and Thomas,⁶ 44 patients whose ulnar nerve motor conduction velocity was < 34 m/s (range 7-31 m/s) were diagnosed as being affected by HMSN type I; the 21 patients whose ulnar nerve MCV was > 40 m/s (range 43.3-66.6 m/s) were classified as HMSN type II. The MCV cut off values were chosen to avoid cases of uncertain classification in the so called intermediate group.

Forty one of the 44 HMSN I patients belonged to 24 families with autosomal dominant inheritance, while 18 of the HMSN II patients belonged to 10 families showing autosomal dominant transmission. One HMSN I and 3 HMSN II cases were sporadic. Autosomal recessive inheritance was likely in 2 HMSN I patients.

The mean age of the patients with HMSN I (20 male and 24 female) was 32 years (range 4-72 years); mean duration of the disease was 21 years and mean age at onset was 11 years. Patients affected by HMSN type II (16 male and 5 female) had a mean age of 34 years (range 8-64 years), a mean duration of disease of 17 years and a mean age of onset of 17 years.

Department of
Neurophysiopathology
Istituto Nazionale
Neurologico "C
Besta", Milan, Italy
V Scaioli
G Avanzini

Department of
Neurology
D Pareyson
A Sghirlanzoni

Correspondence to:
Dr Scaioli, Department of
Neurophysiopathology,
Istituto Nazionale
Neurologico "C Besta", Via
Celoria 11, 20133 Milano,
Italy

Received 23 August 1991
and in revised form 17
January 1992 and 11 March
1992

Accepted 16 March 1992

Electrodiagnostic procedures: ulnar nerve MCV and F response

The MCV of the ulnar nerve was studied by recording the compound muscle action potential from the abductor digiti minimi muscle by means of surface electrodes. Needle electrodes were used in the case of severe denervation.

The F response of the ulnar nerve was obtained by means of supramaximal nerve stimulation at the wrist, and recorded using surface electrodes (or needle electrodes in the case of severe denervation) placed on the abductor digiti minimi muscle. The initial latency of at least ten F and M responses was evaluated. Maximal F response motor conduction velocity (FCV) was measured as: $\text{max FCV} = 2D/(F_{\min} - M - 1)$ where D = distance from wrist to the 7th cervical spinous process, $F_{\min}$ = the minimal latency of the 10 F responses, M = the latency of the direct response.⁷

Somatosensory evoked potentials

Somatosensory evoked potentials (SEPs) were obtained by stimulating the median nerve at the wrist. The stimuli were square shaped electrical pulses of a duration of 0.2 ms delivered at 3 Hz through surface electrodes; the intensity, checked by a current probe, was adjusted until only a small thumb twitch was produced (8–25 mA).

Responses were recorded by means of needle electrodes placed at Erb's point for the N9 brachial plexus component, at the 6th vertebral spinous process (Cv6) for the N13 (cervical spinal cord) component, and at C3' or C4' for the N20 cortical component. Cephalic (Fz) and non-cephalic reference electrodes (linked ear and shoulder contralateral to the stimulated arm) were used, and ground electrodes were placed on the forearm of the stimulated side. At least two sets of 1500 artifact free, averaged responses were obtained from each stimulated side by means of a 5–3000 Hz (–12 dB/oct) bandpass filter. Absolute latencies from stimulus onset to the negative peak were measured at Erb's point (N9), at Cv6 (N13), and at the contralateral somatosensory cortex (N20). Interpeak conduction times (N9–N13, N13–N20, and N9–N20) were also measured. Individual scores were regarded as abnormal if absolute or interpeak latencies exceeded the mean control value (45 subjects) by 2.5 SD.

Brainstem auditory evoked potentials

Brainstem auditory evoked potentials (BAEPs) were obtained by means of clicks generated by pulses of 0.1 ms and rarefaction polarity. Stimuli were delivered at a rate of 10 per second and at an intensity of 60 dB above the hearing threshold for each ear determined at the time of testing. The unstimulated ear was masked with white noise (–20 dB). EEG activity was recorded by means of surface electrodes: the active electrode was placed at Cz and the reference electrodes were placed on the ipsilateral and contralateral mastoid process. At least two trials of 2000 single responses each were averaged by means of 50–3000 Hz

(–12 dB/oct) bandpass filter. The absolute latency of waves I, III, and V and the I–III, III–V, and I–V interpeak times were evaluated. The amplitudes of waves I and V and the V/I amplitude ratio were also considered. Individual scores were considered abnormal if absolute or interpeak latencies exceeded the mean control value (58 subjects) by 2.5 SD, or borderline if they were between 2 SD and 2.5 SD above mean control values. Conventional pure tone audiograms were recorded for all patients. Statistical analyses were performed with Student's *t* test for unpaired data and correlation coefficients where appropriate.

Results

F response motor conduction velocity

An ulnar nerve F response was obtained in 36 patients, 23 with HMSN I and 13 with HMSN II. Both max FCV and MCV had a bimodal distribution and were greatly slowed in HMSN I patients, but normal or only slightly slowed in HMSN II subjects (table 1). In the HMSN I group, mean MCV did not significantly differ from mean max FCV (table 1), and there was a high correlation ($r = 0.84$) when the MCV and FCV of the same patients were compared.

Somatosensory evoked potentials

SEPs were obtained in 34 patients, 25 with HMSN I and 9 with HMSN II. The absolute latencies of all SEP components ranged from slightly increased to greatly increased in HMSN I patients, whereas they were normal or only slightly higher in HMSN II patients (table 2; fig). The amplitude of the N20 component was often decreased in both HMSN I and II groups; only in one HMSN I patient could no response be obtained.

The N13 cervical spinal cord component was recorded in 16 HMSN I and in all HMSN II patients; the N13–N20 central conduction time was within normal values in all patients (table 2).

Attempts were made to measure nerve conduction in the most proximal segment of the peripheral sensory pathway through the N9–N13 interpeak time. However, the N9 component was measurable only in eight patients with HMSN I: bilaterally in five, unilaterally in three. In these eight patients, the N9–N13 interpeak time was bilaterally normal (< 4.3 ms) in two patients and increased (from 5 to 15 ms) in six patients.

The N13 and N20 components showed no significant asymmetry in latency: indeed, there was a high correlation between the N13 and

Table 1 Nerve conduction studies

Subjects	Mean (SD) motor conduction velocity (m/s)	Mean (SD) F-wave conduction velocity (m/s)
HMSN I (n = 23)	21.2 (5.2)	22.6 (6.4)
HMSN II (n = 13)	54.9 (6.8)	54.9 (5.3)
Control (n = 27)	56.4 (3.1)	60.1 (2.8)

Table 2 Mean (SD) somatosensory evoked potentials

	Controls (n = 45)	HMSN I (n = 24)	p Value	HMSN II (n = 9)	p Value
N9*					
Right	9.6 (0.8)	15.2 (3.6)	*	10.7 (0.5)	*
Left	9.4 (0.8)	18.7 (5.7)	*	10.9 (1.1)	*
N13†					
Right	12.9 (1.0)	25.1 (7.4)	*	14.4 (1.3)	*
Left	12.8 (1.0)	24.9 (6.9)	*	14.4 (1.2)	*
N20					
Right	18.8 (1.2)	33.3 (7.7)	*	20.4 (1.1)	*
Left	18.8 (1.2)	33.4 (7.7)	*	20.3 (1.1)	*
N13-20					
Right	5.9 (0.5)	5.6 (0.7)	NS	5.9 (0.5)	NS
Left	6.0 (0.6)	5.7 (0.6)	NS	5.9 (0.5)	NS

* = $p < 0.0005$ NS = not significant.

In one HMSN I patient no response could be obtained.

† N13 was obtained in 16 patients and N9 in 8 patients with HMSN I.

Table 3 Mean (SD) brainstem auditory evoked potentials

	Controls (n = 58)	HMSN I (n = 29)	p Value	HMSN II (n = 10)	p Value
Wave I					
Right	1.6 (0.1)	1.7 (0.2)	*	1.5 (0.1)	NS
Left	1.6 (0.1)	1.7 (0.2)	*	1.5 (0.1)	NS
Wave III					
Right	3.7 (0.1)	3.9 (0.3)	*	3.7 (0.1)	NS
Left	3.7 (0.1)	3.9 (0.3)	*	3.7 (0.1)	NS
Wave V					
Right	5.5 (0.2)	5.7 (0.4)	*	5.6 (0.2)	NS
Left	5.5 (0.2)	5.7 (0.4)	*	5.6 (0.2)	NS
Interpeak latency I-III					
Right	2.1 (0.1)	2.1 (0.2)	NS	2.2 (0.1)	< 0.05
Left	2.1 (0.1)	2.2 (0.2)	NS	2.2 (0.1)	NS
Interpeak latency III-V					
Right	1.8 (0.1)	1.9 (0.2)	NS	1.8 (0.1)	NS
Left	1.8 (0.1)	1.9 (0.3)	NS	1.8 (0.1)	NS
Interpeak latency I-V					
Right	3.9 (0.2)	4.0 (0.3)	NS	4.0 (0.1)	NS
Left	4.0 (0.2)	4.0 (0.3)	NS	4.0 (0.2)	NS

* $p < 0.0005$; NS = not significant.

N20 latencies of the two sides in HMSN I patients ($r = 1$ and $r = 0.97$, respectively).

In HMSN I patients, the latencies of N13 and N20 SEP components were compared with the latency of the F response and with MCV. Correlations between motor and sensory latencies and velocities were good, suggesting the reasonably homogeneous involvement of both motor and sensory nerves (ulnar nerve MCV v N20 latency, $r = 0.77$; ulnar nerve FCV v N20 latency, $r = 0.93$; ulnar nerve F wave latency v N20 latency, $r = 0.95$). In HMSN II patients, the absolute latencies of the N9, N13, and N20 components were slightly increased in three patients, whereas the N9-N13 and N13-N20 interpeak times were always normal, suggesting that the decrease in conduction took place distally to the N9 component.

Brainstem auditing evoked potentials

BAEP studies were performed in 31 HMSN I and 11 HMSN II patients. The results of three patients (two with HMSN I and one with HMSN II) were not eligible for data analysis because of abnormalities of audiometric tests, which were consistent with presbycusis in two and pronounced hearing loss due to previous middle ear infection in the third. Table 3 shows BAEP results of the remaining 39 patients. The HMSN I group showed a significant increase in I, III, and V absolute latency. The HMSN II group showed no significant increase in absolute latencies. BAEPs were normal in 7 of 29 (24 %) HMSN I patients.

Wave I latency or I-III interpeak latency, or both exceeded the mean control value by more than 2.5 SD in 10 patients (35%). In detail, wave I latency was higher in five patients (bilaterally in three and unilaterally in two); I-III interpeak latency was increased in four patients (bilaterally in one and unilaterally in three); and in one patient both wave I and I-III interpeak latencies were bilaterally increased. Only one patient had a bilateral increase in all of the interpeak times (I-III, III-V, I-V). In the remaining 12 HMSN I patients (42%), the increase in absolute latencies or interpeak times ranged from 2 to 2.5 SD above the mean control group value. In detail, wave I was increased by 2-2.5 SD in nine patients (bilaterally in three); the latency of wave III was unilaterally increased in two patients; I-III interpeak time was bilaterally increased in one patient; and two patients had borderline values for the III-V interpeak time, as well.

Absolute wave latencies and interwave times were within 2.5 SD for all of the 10 HMSN II patients. BAEPs were normal in 7 HMSN II patients. Borderline values were obtained in three cases: in detail, a unilateral increase in I-III interpeak time was found in two cases and a unilateral increase in wave III was observed in the third patient.

Discussion

The comparable values of FCV and distal MCV in HMSN I patients, both as a group and as individuals, support the hypothesis of a

Figure Latency distribution of the N13 and N20 components of somatosensory evoked potentials in normal subjects and subjects with HMSN I and II.

homogeneous involvement of motor fibres. Increased F latency has previously been reported by Kimura in a group of HMSN patients.^{7,8} Panayiotopoulos *et al* also found a significant delay in lower limb F latency in some patients with Charcot-Marie-Tooth disease.^{9,10} A selective evaluation of type I versus type II was not attempted by these authors, nor was the correlation statistically assessed.

As far as SEPs are concerned, we found a clearcut increase in N20 latency (mean latency 14.5 ms above control value) in all but one HMSN I patient, in whom this component was not detectable at all. Similar results have been reported by Jones *et al*.¹¹ This prolongation in latency was entirely due to peripheral conduction slowing, since no abnormality in central conduction time (N13–N20 interpeak latency) was detected.

The involvement of the peripheral sensory pathway was highly symmetrical. The severity of conduction slowing along the sensory pathway seemed to be roughly correlated with motor nerve involvement. The absence of a distoproximal gradient in conduction slowing along motor nerves, the symmetry of SEP latency delay, and the motor-sensory correlations are in favour of diffuse and comparable myelinopathic damage in HMSN I.

Distal (N9 latency) and proximal (N9–N13 interpeak latency) conduction along the median nerves could be compared in only eight out of 25 patients with HMSN I. The results were conflicting because of the large scatter of conduction in proximal sensory segments, a variability that may be due to nerve potentials originating from the activation of the motor branch of the mixed nerves and impairing the correct recording of N9–N13 interpeak latencies.¹² Additional difficulties in recording the N9 component were probably due to the considerable desynchronisation and reduced amplitude of sensory action potential.

Conduction changes in the short acoustic nerve were also studied. Cranial nerve involvement in HMSN has been previously reported.^{13,15}

Kimura has shown neurophysiological abnormalities in the trigeminal and facial nerves by studying blink reflexes and the direct M response of the VII nerve.¹⁶ Other authors have reported similar findings.^{17,18}

BAEP abnormalities indicating acoustic nerve involvement in HMSN patients have also been reported.^{19,24} However, only a few studies of patient groups systematically deal with the frequency, extent, and pattern of VIII cranial nerve involvement. Given that this is the shortest body nerve (about 20 mm^{25,26}), the acoustic nerve may be particularly informative for pathogenetic investigations. It would be expected to be involved late, if at all, in primary axonal damage with secondary demyelination, whereas it would be expected to be demyelinated early in a primary and diffuse myelinopathy.

Wave I is believed to originate from the distal intracanal tract of the acoustic nerve, and waves II and III from the proximal intracranial acoustic nerve and from the cochlear nuclei,

respectively.^{20,26–28} Therefore an increase in the latency of wave I and in the I–III interpeak time (in the absence of transmission defects) indicates slowing in conduction along the acoustic nerve.^{20,29,30}

Our BAEP studies showed a high incidence of acoustic nerve involvement in HMSN I patients: 22 out of 29 (76%) HMSN I patients showed some change in early BAEP components (latency exceeding mean control values by more than 2 SD) and in 10 of them (34.5%) the early waves were greatly delayed (by more than 2.5 SD). The most evident abnormality was the significantly increased latency of wave I (table 3). These findings are in keeping with a slowing in conduction along the acoustic nerve.

Similar evidence of abnormal acoustic nerve conduction has been presented by Raglan *et al*, who compared audiological and electrophysiological data in 12 HMSN I subjects.²² On the basis of BAEP findings in nine patients, Triantafyllou *et al* have suggested that the involvement of the VIII nerve in HMSN I is more common than clinically expected and is probably related to a demyelinating process.²³

We found an increase in III–V interpeak time (suggestive of brainstem acoustic pathway dysfunction) in only one HMSN I patient, who showed no other sign of central nervous system involvement. A mild and unilateral increase in the latency of wave I or III was found in three HMSN II patients.

The normal BAEP findings in 7 (24 %) HMSN I patients deserve some comment. The acoustic nerve lacks the large myelinated fibres²⁰ that are particularly affected in HMSN I. Their estimated conduction velocity is about 20 m/s^{20,26} and might not be further slowed by the disease. In addition, the proximal segment of the acoustic nerve, close to the brainstem, is ensheathed by central oligodendroglial myelin,³¹ which is not affected by the pathological processes of HMSN I. This tract varies in length in individual patients.³¹ The variability of the ratio of Schwann cells to central myelin may explain the variability of our findings in HMSN I: it is possible that no increase in latency can be detected when the Schwannian tract is too short, but when it is longer an expected increased latency of either wave I or the I–III interpeak time (or both) can be observed.

In conclusion, we found a homogeneous slowing in conduction along the peripheral motor pathways in HMSN I, with both FCV and MCV similarly decreased. Comparison between studies of conduction velocity and somatosensory evoked potential indicates that the motor and sensory peripheral pathways are proportionally involved; no significant asymmetry was found, which suggests an illness homogeneously affecting all of the peripheral sensory and motor nerves. This evidence is further supported by the high incidence of peripheral conduction abnormalities found in the acoustic nerve. Our neurophysiological data, considered as a whole and compared with those of HMSN II patients, seem to support the hypothesis of myelinopathic primary dam-

age in HMSN I.

This conclusion contrasts with the hypothesis of Dyck *et al.* of an axonal primary deficit in HMSN I. On the basis of morphological findings and axonal flow studies, they maintain that demyelination occurs secondarily to axonal atrophy.^{1,2,32} Their evidence partly comes from the finding of a higher frequency and greater degree of distal morphological abnormalities, which is suggestive of axonal atrophy affecting the extremities of the nerve cell first and more severely;¹ they also found that demyelination was clustered on certain teased nerve fibres, as for a primary axonal disease, and not at random, as expected in primary Schwann cell dysfunction.¹

Neurophysiological studies provide only indirect evidence of morphopathogenetic events—in this study, random demyelination or clustering of demyelination on individual fibres cannot be distinguished—but such studies do allow an extensive and reproducible evaluation of many nerve segments and districts in many patients at different disease stages. In our opinion, they seem to reflect the presence of a diffuse and homogeneously distributed demyelinating process, suggestive more of primary than secondary demyelination (depending on axonal atrophy or degeneration). Although our data cannot be taken as definite proof, they do suggest the need to reconsider the pathogenesis of HMSN I in an attempt to reconcile apparently conflicting results.

We thank Eng F Panzica for his skillful technical assistance. This paper was presented in part at the 41st annual meeting of the American Academy of Neurology, Chicago, in April 1989.

- Dyck PJ. Inherited neuronal degeneration and atrophy affecting peripheral motor, sensory and autonomic neurons. In: Dyck PJ, Thomas PK, Lambert EH, Bunge R, eds. *Peripheral neuropathy*. Philadelphia: WB Saunders 1984:1600–55.
- Nukada H, Dyck PJ, Karnes JL. Thin axons relative to myelin spiral length in hereditary motor and sensory neuropathy, type I. *Ann Neurol* 1983;14:648–55.
- Lewis RA, Sumner AJ. The electrodiagnostic distinctions between chronic familial and acquired demyelinating neuropathies. *Neurology* 1982;32:592–6.
- Sumner AJ. Characteristics of the nerve conduction abnormality in HMSN I [abstract]. *2nd international conference on CMT disorders*. New York: Harriam, 1987:9.
- Sghirlanzoni A, Pareyson D, Scaiola V, Marazzi R, Pacini L. Hereditary motor and sensory neuropathy type I and type II. *Ital J Neurol Sci* 1990;11:471–9.
- Harding AE, Thomas PK. The clinical features of hereditary motor and sensory neuropathy types I and II. *Brain* 1980;103:259–80.
- Kimura J. The F-wave. In: *Electrodiagnosis in diseases of nerve and muscle: principles and practice*. 2nd ed. Philadelphia F.A. Davis, 1989:332–55.
- Kimura J. F-wave velocity in the central segment of the median and ulnar nerves. A study in normal subjects and in patients with Charcot-Marie-Tooth disease. *Neurology* 1974;24:539–46.
- Panayiotopoulos CP. F-wave conduction velocity in the deep peroneal nerve: Charcot-Marie-Tooth disease and dystrophia myotonica. *Muscle and Nerve* 1978;1:37–44.
- Panayiotopoulos CP, Scarpalezos S, Nastas PE. Sensory (1a) and F-wave conduction velocity in the proximal segment of the tibial nerve. *Muscle and Nerve* 1978;1:181–9.
- Jones SJ, Carroll WM, Halliday AM. Peripheral and central sensory nerve conduction in Charcot-Marie-Tooth disease and comparison with Friedreich's ataxia. *J Neurol Sci* 1983;61:135–48.
- Halonon JP, Jones S, Shawkat F. Contribution of cutaneous and muscle afferent fibres to cortical SEPs following median and radial nerve stimulation in man. *Electroenceph Clin Neurophysiol* 1988;71:331–5.
- De Recondo J. Hereditary neurogenic muscular atrophies (Charcot-Marie-Tooth disease). In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*. New York: Elsevier, 1975:271–317.
- Killian JM, Kloepper HW. Homozygous expression of a dominant gene for Charcot-Marie-Tooth neuropathy. *Ann Neurol* 1979;5:515–22.
- Testa D, Milanese C, La Mantia L, Mastrangelo M, Crenna P, Negri S. Familial trigeminal neuralgia in Charcot-Marie-Tooth disease. *J Neurol* 1981;225:283–7.
- Kimura J. An evaluation of the facial and trigeminal nerves in polyneuropathy: electrodiagnostic study in Charcot-Marie-Tooth disease, Guillain-Barré syndrome, and diabetic neuropathy. *Neurology* 1971;21:745–52.
- Malin J-P. Trigeminal and facial nerve involvement in Charcot-Marie-Tooth disease. An electrodiagnostic study. *J Neurol* 1981;226:101–9.
- Hausmanowa-Petrusewicz I, Ryniewicz B, Rowinska-Marcinska K, Emeryk B, Kopec A. The subclinical facial nerve involvement in generalized neuropathies. *Electromyogr Clin Neurophysiol* 1987;27:229–34.
- Satya-Murti S, Cacace AT, Hanson PA. Abnormal auditory evoked potentials in hereditary motor-sensory neuropathy. *Ann Neurol* 1979;5:445–8.
- Garg BP, Markand ON, Bustion PF. Brainstem auditory evoked responses in hereditary motor-sensory neuropathy: site of origin of wave II. *Neurology* 1982;32:1017–9.
- David P, Russo G, Pelicano R, Laureati MF, Scoppetta C, Bergonzi P, *et al.* Brain-stem auditory evoked responses in the Charcot-Marie-Tooth disease. *Electroenceph Clin Neurophysiol* 1985;60:85P.
- Raglan E, Prasher DK, Trinder E, Rudge R. Auditory function in hereditary motor and sensory neuropathy (Charcot-Marie-Tooth disease). *Acta Otolaryngol (Stock)* 1987;103:50–5.
- Triantafyllou N, Rombos A, Athanasopoulou H, Siafakas A, Malliara Loulakaki S. Electrophysiological study (EEG, VEPs, BAEPs) in patients with Charcot-Marie-Tooth (type HMSN I) disease. *Electromyogr Clin Neurophysiol* 1989;29:259–63.
- Kowalsky JW, Rasheva M, Zakrzewska B. Visual and brainstem auditory evoked potentials in hereditary motor-sensory neuropathy. *Electromyogr Clin Neurophysiol* 1991;31:167–72.
- Lang J. Facial and vestibulocochlear nerve, topographic anatomy and variations. In: Samii M, Jannetta P, eds. *The cranial nerves*. Berlin: Springer, 1981:363–77.
- Moller AR, Jannetta PJ. Simultaneous surface and direct brainstem recordings of brainstem auditory evoked potentials (BAEP) in man. In: Cracco RQ, Bodis Wollner I, eds. *Frontiers of clinical neuroscience. Vol 3. Evoked Potentials*. New York: Liss, 1986:227–34.
- Curio G, Oppel F, Scherg M. Peripheral origin of BAEP wave II in a case with unilateral pontine pathology: a comparison of intracranial and scalp records. *Electroenceph Clin Neurophysiol* 1987;66:29–33.
- Moller AR, Jannetta PJ. Auditory evoked potentials recorded from the cochlear nucleus and its vicinity in man. *J Neurosurg* 1983;59:1013–8.
- Nelson KR, Gilmore RL, Massey A. Acoustic nerve conduction abnormalities in Guillain-Barré syndrome. *Neurology* 1988;38:1263–6.
- Schiff JA, Cracco RQ, Cracco JB. Brainstem auditory evoked potentials in Guillain-Barré syndrome. *Neurology* 1985;35:771–3.
- Luxon LM. Diseases of the eighth cranial nerve. In: Dyck PJ, Thomas PK, Lambert EH, Bunge R, eds. *Peripheral neuropathy*. Philadelphia: WB Saunders, 1984:1300–36.
- Brimmoin S, Capek P, Dyck PJ. Axonal transport of dopamine-beta-hydroxylase by human sural nerves in vitro. *Science* 1973;180:1295–7.