Visual and Somatosensory Evoked Potentials and F-Wave Latency Measurements in Hereditary Neuropathy with Liability to Pressure Palsies

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Summary. Pattern shift visual evoked potentials (VEPs), cervical and cortical somatosensory evoked responses (SEPs) and motor conduction velocities studied by F-wave latency measurements were investigated in two family members with hereditary neuropathy with liability to pressure palsies (HN-PP). In both cases the VEPs and SEP conduction times N 13–N 20 were normal. A bilateral pathological increase of latencies of early SEP components, N 9–N 13 transit times and F-wave latencies revealed a lesion in the proximal parts of the median nerves close to the spinal cord in the older patient. These abnormalities emphasize the close relationship of HN-PP with hereditary polyradiculopathy (Mayer 1975).

Key words: Hereditary neuropathy with liability to pressure palsies – Visual evoked potentials – Somatosensory evoked potentials – F-wave latency measurements


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Introduction

Hereditary neuropathy with liability to pressure palsies (HN-PP) has been described as an entity with characteristic clinical and distinctive neurological features (Davies 1954; Staal et al. 1965; Behse et al. 1972; Bosch et al. 1980). However, the question of its relation to other types of polyneuropathy, especially to the heredofamilial mononeuritis multiplex with brachial predilection HN-MM (Mayer 1975) and the hereditary polyneuropathy of the Charcot-Marie-Tooth type (Mayer 1975; Dietz and Schenck 1975), is still unanswered.

Analyses of visual and somatosensory evoked potentials and F-wave latency measurements introduce the possibility of detecting additional subclinical involvement of the optic nerve and most proximal and central segments of the somatosensory system. Hence they may be useful tools with which to gain further insight into the nature of the disorder and contribute towards a proper classification.

The procedures mentioned above were applied to two patients with HN-PP: a man suffering from recurrent episodes of peripheral nerve palsies for 22 years and his clinically unaffected 11-year-old daughter.

Patients

Patient 1, D.K. This 40-year-old man had his first episode of weakness of the ulnar-innervated muscles of the right hand at the age of 18. At 24, weakness and paraesthesiae occurred in the right foot, caused by a common peroneal nerve affliction with complete recovery after some weeks. Neurological examination at the age of 33 revealed muscle weakness in the distribution of both ulnar nerves. Routine laboratory studies were normal. Nerve conduction studies showed that the velocities were reduced not only in the ulnar nerves, but also in the clinically unaffected nerves such as the median and common peroneal. On needle electromyography the abductor digiti V showed fibrillation potentials on both sides.

When the patient was admitted 2 years later for evaluation of a typical ulnar nerve palsy on both sides, there was slight atrophy of the supraspinate and infraspinate muscles. At the age of 37, anterior transposition of the right ulnar nerve was undertaken. A sural nerve biopsy at the left ankle was obtained; teased nerve fibre preparations showed focal sausage-shaped thickenings of the myelin sheath, or tomacula, in association with chronic demyelination-remyelination. On cross-section, some nerve fibres again showed the enormous enlargement of the myelin sheath accompanied by a large decrease of the axon diameter\(^1\). No abnormality of serum or spinal fluid lipoproteins or globulins could be found; cervical spine radiographs were normal.

Three years later when evoked potentials were studied, the neurologic examination revealed a slight palsy of the left abductor digiti V and sensation disturbance involving the palmar and dorsal sides of the fourth and fifth fingers and the ulnar side of the palm up to the wrist. The knee jerks were depressed, the ankle jerks absent.

Patient 2, K.K. The 11-year-old daughter of D.K., had no history of signs or symptoms referable to the nervous system. At the age of 9, motor nerve conduction velocities along the distal segments of peroneal nerves were moderately reduced; a pronounced reduction by 43% was found across the sulcus segment of the left ulnar nerve. The neurologic examination was consistently completely normal.

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