Correspondence

Hemifacial Spasm

To the Editor:

Ronen, Donat and Hill,[1] in their discussion in the case of a child with hemifacial spasm, state that this disorder may be produced by aberrant regeneration of the facial nerve after injury. Same notion has been traditionally held to explain the post-facial palsy synkinesis and other phenomena. However, the weight of the current evidence is against such a mechanism in both conditions, which are clinically and physiologically identical. Although the authors refer to the elegant neurophysiological studies of Nielsen[2,3] in hemifacial spasm, they fail to make any reference to ephaptic transmission, so fervently advanced by Nielsen, as the underlying mechanism of hemifacial spasm. I proposed the same for post-facial palsy phenomena in 1975 and argued for clinical and physiological identity of these two disorders affecting the facial nerve.[4] The weight of the evidence is such now that the notion of aberrant facial nerve degeneration for both hemifacial spasm and post-facial palsy phenomena should be discarded in favour of ephaptic transmission. This same mechanism can be advanced just as well as for phenomena observed after other cranial palsies, (e.g., 3rd nerve palsy).

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Authors' Reply

Two types of hemifacial spasm (HFS) are recognized: cryptogenic, which arises without previous known injury to the facial nerve and is usually caused by compression of the root entry zone of the seventh nerve by blood vessels, and post paralytic, occurring most often after incomplete recovery from Bell's palsy or traumatically induced facial paresis. The two types are often clinically indistinguishable, except for the history of previous facial palsy in the latter. Of the three main hypothesis, one explains post-paralytic HFS on the basis of aberrant regeneration. The other two, ephaptic transmission and abnormal seventh motor nucleus firing, suggest a common pathophysiology for both types[1,2] and are supported by results of recent investigations on patients with "cryptogenic" HFS.[3-7]

The hypothesis which is strongly supported by Dr. Sadjadpour,[1,3] suggests that injury to the nerve results in formation of artificial synapses that facilitate ephaptic transmission between nerve fibers. The other hypothesis advocates altered facial motor neuron firing as the cause of HFS, triggered by antidromic impulses arising at the peripheral lesion.[2,4-7] The notion of aberrant facial nerve regeneration has been disputed in favour of both of these pathogenetic mechanisms,[1,2] but more recently even the notion of ephaptic transmission has been challenged as the sole mechanism in HFS.[4-7]

Since our intention was only to draw attention to the occurrence of HFS in childhood,[8] we did not deal extensively with the main pathogenetic hypotheses and the controversies involved, but in view of the mentioned studies by Møller and Janetta,[4-7] we cannot join Dr. Sadjadpour in supporting the notion of ephaptic transmission as the leading hypothesis for this disorder.

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Multiple Sclerosis Presenting as Third Nerve Palsy

To the Editor:

In the August 1986 issue of CJNS Uitti and Rajput[1] reported what was believed to be the first documented case of multiple sclerosis presenting as a third nerve palsy with pupillary dilatation. We describe here another such case.

Our patient is a 14-year-old girl who developed diplopia in March 1986. She was prescribed glasses and the diplopia disappeared. She noted diplopia again in July 1986 which persisted. Two weeks later she noted that her right pupil was dilated. Past history revealed an episode of numbness in the fingers of both hands lasting several weeks. On examination the right pupil

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was widely dilated and unreactive to light and accommodation. Adduction, elevation and depression of the right eye were decreased. There was no ptosis. Remaining neurological examination and examination of other systems revealed no abnormality.

She was admitted to hospital with a view to doing a carotid angiogram to rule out a compressive lesion of the right third nerve. On the day after admission, examination showed the right pupil to be equal to the left and reactive to light and accommodation. Extraocular movements remained weak in the right eye.

A CT scan with double dose contrast showed multiple enhancing periventricular lesions suggestive of multiple sclerosis. VER testing revealed a latency of 119 ms on the right and 136 ms on the left (normal upper limit: 103). Octupus perimetry revealed a superior paracentral scotoma in the right eye and an inferior paracentral scotoma in the left eye. On Farnsworth-Munsell 100 Hue colour vision testing the gross error score was 28 in the right eye and 63 in the left eye (both within normal limits). The inter-ocular difference in score is abnormal at the 95% confidence level.[2]

Three weeks later the neurological examination was normal. This clinical course was consistent with a diagnosis of probable MS. Isolated third nerve palsy as an initial presentation of multiple sclerosis may be more common than recognized.[3] Invasive procedures like angiograms may be voidable in these cases.

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Age of Onset of Parkinson's Disease

To the Editor:

Teravainen and his colleagues[1] have reported data on the age of onset and age-specific prevalence of Parkinson's disease in 551 parkinsonian patients in Vancouver and Helsinki. They concluded that Parkinson's disease may be starting at an earlier age than in the past. They found the mean age of onset of Parkinson's disease to be 57.8 years in Helsinki and 57.9 years in Vancouver. Other studies quoted have suggested a later age of onset and greater prevalence in later life. However, as early as 1883, Gowers[2] reported a mean age of commencement of 52 years. He stated that onset after 65 was uncommon and that the disease did not belong to "extreme senility." Prior to the encephalitis lethargica era, large series from several countries between 1885 and 1922 found mean ages of onset from 51.8 to 59.6 years.[3-8]

In order to determine whether the age of onset of Parkinson's disease is changing, and to explore the concept that aging plays an important contributory role to the development of parkinsonism (which might suggest that age-specific incidence rates should increase with increasing years) we performed a study similar to that of Teravainen et al involving 1092 patients in six centres located in Canada, Europe and the United States.[9] Similar to the results of Teravainen and colleagues, as well the early studies mentioned above, the mean age of onset in our cases was 57.1 years. 5.4% of our patients had onset before age 40 (in comparison to 7.8% of the Helsinki-Vancouver cases), 27.5% began before age 50 and 62.9% after age 50 years. The peak incidence occurred between ages 60 and 69. Age-specific incidence curves showed a decreased susceptability in the older ages.

We do not believe that our study of over 1000 parkinsonian patients or that of Teravainen et al[1] supports the impression that Parkinson's disease is occurring with an earlier age of onset. Previous studies of the age-specific incidence rates in much smaller numbers of patients have revealed contrasting results. Some show an increase with each advancing decade (for example, [10,11]) while others suggest a decline in late life (for example [12,13]). Although age-related nigral cell loss may contribute to the development of parkinsonism in patients who have previously sustained subclinical damage to this region, the shape of our age-specific incidence curve suggests that individuals who reach later life may be more resistent to the etiological factor(s) which cause Parkinson's disease.

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