Matters arising

Writing tremor: its relationship to essential tremor

Sir: Kachi and coworkers[1] investigated nine patients with primary writing tremor and suggested that it is a variant of essential tremor. We report a case of primary writing tremor which responded to primidone therapy.

A 63-year-old right-handed black male complained of "shakiness on writing." He noted that his writing had progressively got worse over the past one year and was currently unreadable. He had recently ceased writing. He had no difficulties with other tasks. He had found that alcohol ingestion in small amounts would improve his writing but several hours after alcohol cessation his writing would be worse. There was no family history of neurological disease or tremor. Neurological examination was normal except for a mild bilateral postural hand tremor and a marked tremor consisting of a rhythmic pronation/supination of the right hand when attempting to write. Propranolol therapy was started and the dose was increased to 100 mg/day. No effect was noted. The dose was not increased further because of a cardiac condition. Propranolol was discontinued and primidone, 50 mg at nighttime, was given. After one week of therapy the patient reported that his tremor had "quieted down," and that his writing was almost normal especially in the morning. A mild tremor on writing was noted. The dose of primidone was increased to 250 mg and further improvement in writing occurred. Placebo therapy was then substituted for primidone. One week later the patient reported that this "medicine did not work" and he was again having difficulty with writing.

Kachi and coworkers[1] noted that the frequency of primary writing tremor was 5 to 6 Hz and that the tremor was improved by alcohol and propranolol. These characteristics are similar to essential tremor. Primidone has recently been found to be effective in reducing essential tremor.[2 3] The drug has not been shown to be effective in other movement disorders. The efficacy of primidone in primary writing tremor further indicates that this entity is a subtype of essential tremor.

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[1] Kachi T, Rothwell JC, Cowan JMA, Marsden CD. Writing tremor: its relationship to benign essential tremor. *J Neurol Neurosurg Psychiatry* 1985;**48**:545–50.
[2] O'Brien MP, Upton AR, Toseland PA. Benign familial tremor treated with primidone. *Br Med J* 1981;**282**:178–80.
[3] Findley LJ, Calzetti S, Richens A. Double blind study of primidone in essential tremor: preliminary results. *Br Med J* 1981;**205**:608.

The Flick Sign in Carpal Tunnel Syndrome

Sir: We read with interest the paper by Dr WEM Pryse-Phillips in which the accuracy of the "flick sign" in predicting the presence of carpal tunnel syndrome was assessed. He found that 93% of patients with electrodiagnostically proven carpal tunnel syndrome admitted to flicking the affected wrist and hand in an effort to relieve the discomfort. The false positive rate was under 5%. Therefore, the flick sign was found to be extremely sensitive and specific for the diagnosis of carpal tunnel syndrome. In the discussion section of his paper, Dr Pryse-Phillips indicated that such a reliable clinical diagnostic test could be valuable in areas where electrodiagnosis is not available.

To assess the accuracy of this sign in our EMG lab, patients referred over the last four months with hand pain or dysaethesias were routinely asked what they did with their hand(s) to relieve the discomfort. If they did not volunteer that they flicked their wrist(s), they were specifically asked. The electrodiagnostic criteria for the diagnosis of carpal tunnel syndrome were essentially the same as those used by Dr Pryse-Phillips except that we routinely use the median palmar sensory latency across the wrist. A latency of greater than 2·2 milliseconds over a distance of 8 cm is considered to be abnormal.

The diagnosis of carpal tunnel syndrome was made in 56 patients. Only 14 (25%) demonstrated the flick sign. Nine patients demonstrated the flick sign without evidence of carpal tunnel syndrome. Seven of these demonstrated no pertinent electrophysiologic abnormalities, and two had cervical radiculopathy. The false positive rate was 39%.

We found that the sign was neither sensitive nor specific for carpal tunnel syndrome. We have no explanation for our different results, but the flick sign is not unlike other clinical signs of carpal tunnel syndrome in this regard. Pryse-Phillips notes that a positive Tinel's sign has been reported in 0% to 89% of patients.

It is our feeling that carpal tunnel syndrome can produce a variety of upper extremity complaints, and that no single clinical sign is of sufficient diagnostic accuracy. Surgery is best reserved for patients with electrically proven carpal tunnel syndrome.

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Reference

[1] Pryse-Phillips WEM. Validation of a diagnostic sign in carpal tunnel syndrome: *J Neurol Neurosurg Psychiatry* 1984;**47**:870–2.

Pryse-Phillips replies:

I am both surprised and disappointed that Dr Krendel and his colleagues so significantly failed to verify my observations on the Flick Sign in Carpal Tunnel Syndrome (CTS). Presumably, our different findings must be due to patient populations, patient selection or interpretation of the sign.

Apart from the absence of black people in my patient group, I would suppose our patient populations to be similar. Whether any learned pattern of hand movement is more common in either population is an ethnological question without answer.

Dr Krendel's false positive rate of 39% presumably refers to his nine patients with a positive flick sign but with negative electrophysiological tests for CTS. I included five such patients in my series and am aware that many patents with CTS have clinical symptoms but no obvious EMG findings (8% in Kimura's series[1]). In many such cases, CTS is nevertheless the correct diagnosis and the false positive rate quoted may be fallacious.

The selection of patients with hand pain as a leading symptom may also explain part the discrepancy in our figures since I rely more upon acroparaesthesia in the diagnosis of CTS rather than pain, heaviness and numbness; my clinical impression is that the flick sign correlates well with acroparesthesiae while hand pain (as with rheumatoid arthritis, or old fracture) inhibits movements.

The interpretation of physical signs is a subjective matter. I do not know how much hand movement had to occur for Dr Krendel and his colleagues to record a positive flick sign but presumably it was a good deal

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more than I accepted. Flicking or oscillating the hand (or by extension, making other similar movements) in response to questions about the patient's way of relieving paraesthesiae occurred in 45 of the last 48 patients (94%) with documented CTS seen in my laboratory compared with a positive Tinel sign in 54% and a positive Phalen's sign in 52%; informal feedback from other physicians has confirmed my belief that patients with CTS very frequently move their hands to relieve paraesthesiae while those with other neural lesions of the arm do so only seldom. Why the occurrence of the sign should vary between regions I too am at a loss to explain but will have to accept regretfully that the flick sign is as influenced by some unknown population factor as are other clinical evidences of CTS. I too consider electrodiagnosis necessary before surgery is contemplated for CTS but still consider that a reliable diagnostic sign would be valuable if electrodiagnosis is unavailable. The findings from Duke University make it unlikely that the Flick Sign will qualify.

Reference

[1] Kimura J. *Electrodiagnosis in Diseases of Nerve & Muscle*. Philadelphia, FA Davis. 1983. 495.

Coincidence of Wilson's disease with other movement disorders in the same family

Sir: Dr Parker describes a fascinating Australian family in whom 22 members inherited various manifestations of torsion dystonia, apparently as an autosomal dominant trait.[1] Within the same family, a mother who exhibited blepharospasm, whose own mother and grandmother also were affected, gave birth to one child who developed generalised torsion dystonia, two others who developed spastic dysphonia, and two other siblings who developed Wilson's disease. There was no suggestion of consanguinity in this branch of the family. This is a striking coincidence, but it is not unique. We have encountered a similar family in which Wilson's disease was diagnosed in one individual, but another movement disorder without the biochemical characteristics of Wilson's disease appeared in another family member.

JP, a 36-year-old lady (kindly referred by Dr G Harwood), complained of progressive forgetfulness and slowing of movement over the previous seven years. More recently she had begun to fall, her speech had become quiet, and she had had some difficulty with swallowing. Her parents were unrelated. Examination revealed a Parkinsonian gait, generalised rigidity, a flexed posture, and severe bradykinesia and akinesia of all movement. The optic discs, retinae, eye movements, tendon reflexes, and plantar responses were normal. There was left-sided visual inattention and a left-sided hemisensory impairment for all modalities. She was orientated for time and place, was of average intellectual ability, but exhibited considerable defects of both verbal and visual memory. Extensive investigation failed to reveal the cause for this akinetic-rigid syndrome. Full blood count, routine biochemistry, serology, thyroid function tests, CSF examination (including search for oligoclonal bands), serum B12 and folate concentration, plasma lactate, cortisols and amino acids were normal. Leucocyte enzymes (arylsulphatase A, B-galactosidase, and hexosaminidase) were normal. The urine contained a normal pattern of amino acids and sugars and no excess mucopolysaccharides. CT scan showed mild enlargement of the cortical sulci, but no other abnormality. MRI scan revealed symmetrical lesions affecting the heads of both caudate nuclei and the anterior parts of both lentiform nuclei. The nature of these changes was not clear. An EEG showed generalised disorganisation of alpha activity but no specific abnormality. Visual, brainstem, and sensory evoked potentials showed no definite abnormality. Nerve conduction study (Dr P Payan) showed normal amplitude, form and latency of sensory action potentials, but a raised threshold to sensation of the stimulus to the left leg. A bone marrow aspiration was normal, as was a rectal biopsy. Her serum caeruloplasmin was 1·2 μmol/l (normal range 1·3–2·9), and serum copper was 15·0 μmol/l (normal 12·5–19); urinary copper excretion varied from 0·34 μmol/24 h to 1·0 μmol/24 h (normal range less than 1·25 μmol/24 h). A liver biopsy specimen showed normal histology with no stainable copper, and a copper content of 5·24 mg/100 g dry weight. Slit lamp examination showed no Kayser-Fleischer ring.

JB, her younger brother, had died at the age of 24 years of Wilson's disease. He had presented at the age of 18 years (to Dr KJ Zilkha) with a 5 year history of progressive difficulty with walking and speech. Subsequently he had developed a tremor of his right hand and deterioration in school performance. Examination revealed an expressionless face with a constant facile grin. There was marked generalised bradykinesia and akinesia, severe generalised rigidity and tremor of the right arm. There was no evidence of liver disease, but early Kayser-Fleischer rings were present. Biochemical investigation revealed a low serum copper (7·8 μmol/l) and caeruloplasmin (0·7 μmol/l), and excessive urinary excretion of copper (8 μmol/24 h). Administration of 1 g of penicillamine increased urinary copper to over 12·5 μmol/24 h (normal range less than 4·7 μmol/24 h). He was treated with penicillamine and initially showed some improvement. However, he subsequently deteriorated, despite addition of potassium sulphide and BAL.

While our patient's brother clearly had Wilson's disease, she did not. Although presenting with an akinetic-rigid syndrome and cognitive deficit, biochemical studies of copper metabolism were not diagnostic of Wilson's disease. The slightly low serum caeruloplasmin may indicate that she was a heterozygote for Wilson's disease, but this is not certain.

We have encountered two other patients presenting with movement disorders who have exhibited abnormalities of copper metabolism to suggest that they may be heterozygotes for Wilson's disease.

KM, a 16-year-old girl (kindly referred by Dr R Thomson), developed progressive disturbance of speech from the age of about 10 years, followed by increasing tremor of the hands and head, and muscle spasms affecting the head and left arm and foot. School performance had not deteriorated. There was no family history of any similar disorder and her parents were unrelated. On examination she had severe spastic dysphonia, torticollis with the head deviated to the left, dystonic adductor spasms of the left arm at rest or on action, and dystonic posturing of the left foot on walking. The optic discs, retinae, and eye movements were normal. Muscle power, the tendon reflexes, the plantar responses and sensation also were normal. Investigation failed to reveal the cause of her progressive dystonic illness. Full blood count, routine biochemistry, plasma amino acid content, urinary amino pattern, mucopolysaccharide content, leucocyte enzymes, fasting pyruvate and lactate levels were all normal. Bone marrow examination and jejunal biopsy were normal, as was a rectal biopsy. Electroretinograms suggested an abnormality of cone-mediated responses. Serum copper content (17·7 μmol/l) and caeruloplasmin (2·1 μmol/l) as well as urinary copper excretion (0·42 μmol/24 h) were normal. However, a liver biopsy specimen, while showing no definite pathological change and no stainable copper, contained an excess

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