

copper content of 51.5 mg/100 g dry weight (normal less than 30 mg%). There were no Kayser-Fleischer rings on slit lamp examination of the eyes. No explanation for the high liver copper content was established. She was not taking drugs when the biopsy was undertaken, and there was no clinical or biochemical evidence of liver disease.

SW, the other patient, is a 14-year-old boy (kindly referred by Dr J Pilling). The product of a normal birth from unrelated parents, he was well until the age of 11 yr, when his mother noticed that his head turned slightly to one side when he ate. Over the next 3 years his dystonia progressed until, when examined by us at age 14, the following features were present: marked laterocollis and also thoraco-lumbar scoliosis concave to the right, intermittent blepharospasm and oromandibular dystonia, dystonic writer's cramp of the right hand and mild dystonia of the left hand and both legs. The remainder of the neurological and general examination (including slit-lamp examination of the eyes) was normal. Full blood count, plasma and urinary amino acid screen and routine biochemistry were normal, with the exception of a raised alkaline phosphatase of 402 IU/l (normal range below 16 years 40-280 IU/l); the patient was taking primidone for his dystonia with no benefit). CT scan showed slight symmetrical dilatation of the lateral ventricles with some widening of the Sylvian fissures. Levels of serum copper and caeruloplasmin were low (8.4 $\mu\text{mol/l}$ and 0.4 $\mu\text{mol/l}$) respectively. However, 24 hour urinary copper excretion was only 0.4 and 0.5 mmol/24 h. A previous liver biopsy at Addenbrooke's Hospital (Dr JM Walshe) had shown normal histology and a liver copper content of 41.4 $\mu\text{g/g}$ wet weight. Although this was above the normal range (10 $\mu\text{g/g}$ wet weight) for that laboratory, it was not clearly in the range (more than 60 $\mu\text{g/g}$ wet weight) expected in their series of neurological Wilson's disease with preserved liver function.

It seems possible that non-Wilsonian movement disorders may occur in relatives of patients with proven Wilson's disease, or in individuals with abnormalities of copper metabolism perhaps suggesting that they are heterozygotes for Wilson's disease, more often than one would expect by chance. At present, the evidence does no more than hint at this possibility. However, if this suggestion is confirmed by more reports of such an association, then some explanation may be required. Perhaps the inheritance of one gene for Wilson's disease, while insufficient to cause the neurological complications of that illness by itself, may do so if another

gene for neurological illness also is inherited.

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Reference

- ¹ Parker N. Hereditary whispering dysphonia. *J Neurol Neurosurg Psychiatry* 1985;48: 218-24.

Delayed neuropathy after trichlorfon intoxication

Sir: We have read with interest the studies of delayed neuropathy after organophosphorus poisoning by Vasilescu *et al*¹. We now report nine cases of delayed neuropathy after trichlorfon (Flibol-E) intoxication.

We observed 70 cases of trichlorfon intoxication (mainly suicide attempts) between 1971 and 1983. Twenty five of them were re-examined in 1984. In 12 cases (48%) no sign of delayed polyneuropathy developed. Four former patients had complaints (paraesthesiae, weakness of hands) after 2-3 months of poisoning but at the time of the re-examination they were healthy. Eight patients had serious delayed polyneuropathy. All of them, except one, consumed alcohol prior to or at the same time as the poison. After 1.5-9 years from the time of intoxication seven patients showed footdrop, difficult gait, distal muscle atrophy of the peroneal and small foot muscles, abolished Achille's reflex. The peroneal nerve was unexcitable in all seven patients; electromyography revealed marked distal denervation of the lower limb muscles. One patient had very brisk knee jerks, patellar clonus and Babinski's sign 2 years after poisoning. His lower limbs were so spastic that he was nearly unable to walk. One patient had severe gastrointestinal haemorrhage and pneumonia ten days after the organophosphorus poisoning. (Therefore he was excluded from the study. (He has muscular rigidity and Parkinsonian tremor beside the distal polyneuropathy.)

In conclusion, 36% of our re-examined patients had severe residual signs of delayed polyneuropathy mainly distal motor type. In one case signs of CNS lesions have persisted.

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Reference

- ¹ Vasilescu, C, Alexianu, Marilena, Dan A. Delayed neuropathy after organophosphorus insecticide (Dipterex) poisoning: a clinical, electrophysiological and nerve biopsy study. *J Neurol Neurosurg Psychiatry* 1984;47:543-8.

Ineffectiveness of phenoxybenzamine in essential tremor

Sir: Mai and Olson¹ reported that the alpha-adrenergic blocking drug, thymoxamine, given intravenously, significantly suppressed essential tremor in four patients and suggested that alpha-adrenergic blockers may be useful in the treatment of essential tremor. We investigated the effect of phenoxybenzamine, an alpha-adrenergic blocker, in five patients with essential tremor. Average age was 59.6 years and average tremor duration was 18.5 years. The drug was administered orally with an increase in dose of 10 mg a week to 30 mg/day. Tremors were measured with an accelerometer and amplitude and frequency determined by spectral analysis. Tremorgrams were recorded before treatment and after two weeks on the 30 mg/day dosage. The patients reported no subjective improvement in their shakiness or in their functional abilities. Tremor amplitude increased by an average of 8% with therapy as compared to pretreatment. Tremor frequency was unchanged. One patient complained of dizziness and confusion while taking phenoxybenzamine. The results from this study did not support a role for alpha-adrenergic blockers in the treatment of essential tremor.

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Reference

- ¹ Mai J, Olson RB. Depression of essential tremor by alpha-adrenergic blockage. *J Neurol Neurosurg Psychiatry* 1981;44:1171.