less than 0.1% of sufferers. I believe the evidence shows that the true figure is nearer 10%. I therefore consider that the absence of a known affected relative should not deter a neurologist from diagnosing Huntington’s chorea in a patient who shows the characteristic clinical features of the disease.

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**References**


**Charcot-Marie-Tooth disease**

**Sir,**

We read the paper by Brooks and Emery with great interest. We agree with the authors that in Charcot-Marie-Tooth disease slightly affected females are easily missed on clinical examination. If the mode of inheritance in a family is in question it is necessary to detect these female carriers.

The importance of examining nerve conduction in unaffected persons was stressed by de Weerdt and Fryns and van den Berghe, who studied families with the hypertrophic (or demyelinating) type of the disease. In these two families X linked inheritance seemed probable.

However, if the neuronal type of the disease is in question, motor nerve conduction studies are of no help, because motor conduction velocity is usually normal, especially in persons who are only slightly affected. Sensory conduction studies probably discriminate better between affected and unaffected persons.

We would like to draw attention to the use of late response studies (Hoffmann (H) reflex and F response) in hereditary polyneuropathies. In patients with chronic renal failure these studies were abnormal at a time when no clinical evidence of peripheral neuropathy existed and conventional motor and sensory nerve conduction studies were normal. Recently we examined a family with the neuronal type of Charcot-Marie-Tooth disease in which X linked heredity seemed likely. H reflex investigation appeared to discriminate well between affected and unaffected subjects.

In future studies, investigation of the H reflex in families with Charcot-Marie-Tooth disease may contribute to detection of carriers with only minor symptoms and possibly to more insight into the pathophysiological backgrounds of the different genetic forms of Charcot-Marie-Tooth disease.

J J Heimans and D Lindhout

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**References**


This letter was shown to Dr Brooks and Professor Emery, who reply as follows:

**Sir,**

We agree with Drs Heimans and Lindhout that full electrophysiological investigation is necessary in the assessment of patients with Charcot-Marie-Tooth neuropathy and their families.