as those separated by changes observed in sural nerve biopsies. The degree of slowing and the reduction in potential amplitude in the sural nerve correlated with the findings for the sensory fibres in the superficial peroneal nerve and the distal sensory branches of the median nerve. It is likely that both types I and II HMSN each display autosomal dominant and autosomal recessive inheritance.⁹⁻¹⁰ Conduction velocity in the recessive type I cases is significantly lower than in the dominant cases.⁵

In the absence of biochemical markers, confirmation as to the existence of more than one gene responsible for a given clinical syndrome can be obtained from genetic linkage studies. This has recently been undertaken for dominantly inherited HMSN. It has been established that type I HMSN is linked to the Duffy locus on chromosome 1,¹¹⁻¹² whereas this is not true for HMSN II.¹² This provides support for the view that there are at least two genetically distinct forms of dominantly inherited HMSN. The clinical features of the two forms, as pointed out by Salisachs *et al*, are generally similar. This merely reflects the fact that both can be classified as Charcot-Marie-Tooth disease. Individual cases cannot always be distinguished on clinical grounds. Nevertheless, statistical analysis has shown significant differences between the two in the incidence of such features as tremor and ataxia, severity of upper limb motor involvement, tendon areflexia and severity of sensory loss.⁵

The concept of Dejerine-Sottas disease is confused as the term has been used widely and imprecisely since its introduction. There is now general agreement that it should be reserved for cases of recessively inherited, childhood onset neuropathy (HMSN III) with hypomyelination in the peripheral nerves as a conspicuous pathological feature.⁴ Although nerve conduction velocity tends to be markedly reduced in Dejerine-Sottas disease, it is not feasible to separate such patients from recessively inherited HMSN in this way. They can be distinguished by nerve biopsy.⁹ It is difficult to comment on the disparity between the two siblings considered to have Dejerine-Sottas disease included in the paper by Dyck and Lambert⁶ and cited by Salisachs *et al*. This is an early report and documentation is incomplete. For example, for the elder sibling, the results of sensory nerve conduction studies are not given and nerve biopsy was not performed. It is certainly possible that these two cases

represent a marked degree of phenotypic variation, which would be unusual for a recessively inherited condition. However, on the evidence presented, it would not be possible to be emphatic that both individuals had the same disorder.

Salisachs *et al* are quite correct in inferring that at present Refsum's disease appears to be a genetically homogeneous disorder. On the other hand, should future observations indicate clustering of conduction velocities within families, the situation would have to be reconsidered, and investigations undertaken to establish whether this was the result of environmental factors, modifying genes or different main genes.

As is true for inherited disorders in general, detailed study frequently discloses genetic complexity. Charcot-Marie-Tooth disease is likely to be no exception. Indeed, the recently described disorder that clinically resembles type II HMSN but which has an earlier onset and a less favourable prognosis, may be genetically distinct.¹³ Classifications can only be provisional, and it is highly probable that the scheme proposed by Dyck and Lambert,¹⁻⁶ which was subsequently supported by the observations of Thomas and Calne,⁷ Buchthal and Behse⁸ and Harding and Thomas,⁹ will require modification in the light of further experience. Nevertheless, at present it still appears to provide the best synthesis of the clinical, electrophysiological and genetic information that is available.

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The Babinski Response

Sir: In "The Babinski Response: A Review and New Observations" published (in your journal 1955;18:250, the authors PW Nathan and MC Smith quoted, with some misunderstanding, Babinski's work. For instance, on page 250 it says: "However, when anatomical evidence presented itself, it began to be obvious that Babinski's statements were wrong. Babinski himself reported cases which did not accord with his previous statements. For instance, in 1899, he reported three cases of paraplegia in flexion. He stated that in all three there was no histological evidence of degeneration of the pyramidal tract. Yet in the first case, the plantar responses were normal, in the second case, the

"phénomène des orteils" was present, and in the third case "tickling the plantar surface of the foot gives reflex movements of the whole limb". As Babinski was concerned in this paper with demonstrating that there is such a thing as paraplegia in flexion without involvement of the cortico-spinal tracts, he seems to have failed to realise that he had recorded different forms of plantar response, one of which was the Babinski response, all present with normal cortico-spinal tract. On his own premises, his Case 2 proved his supposition about the significance of 'le phénomène des orteils' to be wrong".

Babinski reported in 1899 three cases of paraplegia in flexion. Though the title runs "Sur une forme de paraplégie spasmodique consécutive à une lésion organique et sans dégénération du système pyramidal", but actually, the first case is "une tumeur paraît, s'être développée aux dépens de l'extrémité du plexus choroïde du 4^e ventricule", the second case is "dans la région dorsale supérieure une tumeur ovoïde, grosse comme un oeuf de moineau, qui distend le sac de la dure-mère et comprime la moelle" and the third case is "à l'oeil nu on ne voit nettement sur la moelle qu'une large plaque de sclérose siégeant à droite dans le faisceau antérolatéral, aux niveaux des émergences des racines antérieures; cette plaque occupe toute l'épaisseur du manteau blanc et envahit un peu la corne antérieure . . . Au microscope les coupes des régions cervicales inférieure et dorsale supérieure montrent dans le point indiqué, une plaque de sclérose multiloculaire parfaitement typique; les limites de cette plaque sont nettes et la sclérose est absolue en ce sens qu'il n'existe plus aucune trace de myéline dans toute son étendue, mais les cylindres sont conservés, quoique tuméfiés; . . .".

Nathan and Smith stated: "Case 2 proved his supposition about the significance of 'le phénomène des orteils' to be wrong". Apparently that is not what Babinski was trying to say. In the second case-report he said: "La compression est considérable et la moelle a pris à ce niveau la forme d'un croissant; elle n'a d'ailleurs pas subi une réduction de volume notable; elle est surtout déformée. En ce point il existe une sclérose considérable caractérisée par la disparition de la grande majorité des tubes de myéline, par l'épaississement des travées névrogliques et par les altérations hyalines des vaisseaux. Mais on voit une grande quantité de cylindres qui sont conservés

malgré leur dénudation et leur tuméfaction. Il reste même un certain nombre de tubes encore pourvus de leur myéline". In the third case Babinski said: "Le chatouillement de la plante du pied provoque des mouvements très étendus de flexion du pied sur la jambe et d'extension des orteils sur le métatarse".

According to that mentioned above, there is no reason to think that Babinski's case-report in 1899 and his statement on "le phénomène des orteils" are in any way discrepant.

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Nathan and Smith reply: We are pleased to see Dr Jin-yu Hsia's interest in Babinski's work and our paper on the Babinski response. The title of Babinski's paper is Sur une forme de paraplégie spasmodique consécutive à une lésion organique et sans dégénération du système pyramidal.¹ In this paper, Babinski was establishing that with a normal pyramidal system, one could get paraplegia in flexion without increased tendon reflexes and without clonus. His observations on the kind of plantar response obtained in these three cases was incidental. What we drew attention to in our paper² was that the patient whom he reported as Case 2 had the "phénomène des orteils" (Babinski response) with what Babinski wrote was a normal pyramidal system. We pointed out that Babinski did not draw any conclusions from the fact that he had described his response with a normal pyramidal system.

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- 2 Nathan PW, Smith Marion C. The Babinski response: a review and new observations. *J Neurol Neurosurg Psychiatry* 1955;18:250-9.

Sir: I would like to comment on the rheological findings reported by A Brunetti *et al*, Rheological and fibrinolytic findings in multiple sclerosis (*J Neurol Neurosurg Psychiatry* 1981;44:340). In this study whole blood viscosity is found to be significantly increased in patients suffering from multiple sclerosis. Plasma viscosity and haematocrit changes cannot account for this increase. Hence the authors draw the conclusion that the increase in whole blood viscosity is brought about by a loss of the red cells' deformability. This is further supported by the authors' observation that the relative viscosity is also increased in multiple sclerosis.

However the factors affecting whole blood viscosity at shear rate 230 s⁻¹ are not clear enough to allow this conclusion to be drawn without direct measurement. Firstly the patients were on steroids which has some effect on red cell deformability.¹ Secondly an absence of correlation between relative viscosity and red cell deformability recently has been reported.² Therefore it is possible that red cell deformability is decreased in multiple sclerosis—some membrane abnormalities have already been reported³—but this can only be established by a method measuring this factor more directly.

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GIROLAMI *et al* reply:

Sir: We have read with interest the comments by Ernst *et al* about our paper on "Rheological and fibrinolytic findings in Multiple Sclerosis". We agree with the assumption that red blood cell deformability could be determined using a specific method. However there is no sure method to date of evaluating such a parameter. Differential centrifugation and several filter methods have many pitfalls and variables and yield discrepant results.¹