ment abnormalities seen in progressive supranuclear palsy have not been described.^{3,4} Although ocular findings in our patient suggested progressive supranuclear palsy, the atypical findings of caloric and optokinetic nystagmus, rather than tonic slow eye deviation, suggest preservation of the saccadic generating centres in the brainstem, enabling the production of reflex saccades, but disconnection of these reticular centres from the frontal and parietal regions controlling volitional saccades. These findings are in contrast to those usually noted in progressive supranuclear palsy.

There are various diagnostic possibilities in this case. Increased neurological disabilities in later life attributable to basal ganglia dysfunction are described in patients with postencephalitic parkinsonism.⁵ However, this hypothesis could not explain the loss of responsiveness to levodopa and the emergence of severe gaze palsies in this patient. A further possibility is that he had postencephalitic parkinsonism and then developed unrelated progressive supranuclear palsy. This, however, would seem rather unlikely in view of the rarity of the two conditions. Our patient could have progressive supranuclear palsy, but in the light of his history of an encephalitic illness, oculogyric crises, a resting tremor and, in particular, the very long duration of disease, together with the atypical neuro-otological findings, this diagnosis alone would seem unlikely. Finally, there are reports in the medical literature of other conditions such as diffuse Lewy body disease with considerable impairment of volitional saccades, but relative preservation of reflexive saccades, together with sparing of smooth pursuit function.⁶ Similarly, supranuclear palsies mimicking progressive supranuclear palsy have been documented in vascular disease.⁷

We are unable to classify this patient within one of the currently accepted categories of parkinsonian syndromes. The case raises some unanswered questions regarding the range of pathological effects of the encephalitis lethargica virus, the natural history of postencephalitic parkinsonism almost 60 years after the original infection, and the effect of environmental or other infectious agents on the natural history of this condition. Thorough clinical, neuro-otological, and neuropathological investigation of similar cases may clarify these issues.

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Failure of presumed hepatic myelopathy to improve after liver transplantation

Hepatic myelopathy is a rare complication of chronic liver disease characterised, clinically, by a spastic paraparesis with minimal sensory involvement.¹ It is associated with extensive portosystemic shunting of blood, most commonly due to surgical portocaval shunting but also, occasionally, due to spontaneous shunting through collateral vessels.¹ The pathogenesis is unclear but shunting may allow a neurotoxin to bypass the liver and damage the spinal cord. Pathological studies, which have only been reported in 15 cases, have consistently shown symmetric demyelination, sometimes associated with axonal loss, of the lateral pyramidal tracts beginning in the cervical spinal cord and becoming more intense at lower levels. Occasionally, demyelination has also been found in the ventral pyramidal tracts and in the posterior columns and spinocerebellar tracts.¹ The outcome is variable but most cases progress to severe disability. There is no known treatment: some patients may respond to a regimen of protein restriction, lactulose, and antibiotics but this is not consistent. We report a further patient who we think is the first to have received a liver transplant.

A 52 year old man was referred to our department in April 1993 with progressive difficulty walking. He had a known history of alcoholic liver disease, having presented to his local hospital in 1987 with ascites and in March 1992 with hepatic encephalopathy. At that time he was drinking 30 units of alcohol (mainly spirits) per day, although subsequently he had stopped. In August 1992, over a period of about one month he developed stiffness and weakness of both legs, and his walking became unsteady. Arm, bowel, and bladder function remained normal. Investigations at that stage included MRI of his spinal cord, which was normal, and he was started on a trial of oral steroids although no diagnosis was made. His condition continued to deteriorate such that he was only able to walk a few steps with the help of a Zimmer frame and he was otherwise wheelchair bound. There was no family history and no risk factors for HIV infection. His medication included prednisolone (20 mg per day), diuretics, and quinine and diazepam for leg cramps. On examination, he had ascites, peripheral oedema, and splenomegaly. Neurological examination was normal apart from the lower limbs, in which there was severe bilateral spasticity (Ashworth grade 4-5²), and bilateral pyramidal weakness (MRC grade 2-3) associated with brisk reflexes and bilateral extensor plantar responses. Sensory examination was normal apart from a minor reduction in vibration sensation at the toes. He could stand without support but was not able to walk without help. He had a low platelet count ($67 \times 10^9/l$) compatible with splenomegaly, and deranged liver function

tests: bilirubin 26 mmol/l (normal < 17), alkaline phosphatase 202 u/l (normal 30-140), albumin 28 g/l (normal 35-50), international normalised ratio for prothrombin time 1.4. The following tests were normal or negative: B12, folate, thyroid function, copper, caeruloplasmin, iron studies, antinuclear antibodies, syphilis serology, viral titres (including HTLV-1, hepatitis B and C), CSF biochemistry, oligoclonal bands and microscopy, MRI of spinal cord and brain, and myelography. The patient did not wish to have an HIV test performed. Serum very long chain fatty acids were slightly raised but the skin fibroblast assay was normal, which excluded adrenoleucodystrophy. A liver biopsy confirmed micronodular cirrhosis consistent with alcohol related liver disease, and an angiogram showed severe splenomegaly and a major portosystemic shunt from the left gastric vein to the azygous vein.

There was no response to the combination of a protein restricted diet and lactulose or to attempts to reduce the flow through his shunt with oral nitrates or to symptomatic treatment with physiotherapy and small doses of diazepam. In August 1993 he had a further episode of reversible encephalopathy, and, since there was no evidence of ongoing alcohol intake, he was accepted for liver transplantation on the basis of liver failure alone. A suitable donor organ became available in December 1993, and a successful transplant was performed without major complications. His liver function returned to normal within about six months. Despite this, however, there has been no change at all in the function of his lower limbs in the 18 months since his transplant. He continues to be bothered by spasms at night despite increasing doses of diazepam and baclofen, and is largely wheelchair dependent.

We think that this is the first patient with hepatic myelopathy who has received a liver transplant. The transplant was performed because of liver failure and from that point of view it has been a success. Disappointingly, there has been no appreciable improvement in his neurological symptoms or signs despite normalisation of his liver function. This implies that the spinal cord damage is irreversible, which is compatible with the axonal loss seen at necropsy in some cases.¹ It is unlikely that there will be any significant neurological improvement at this stage. However, it is unclear whether successful transplantation earlier in the course of the disease when, perhaps, there may only have been demyelination of the cord, would have been beneficial. It is also possible that without transplantation he would have continued to deteriorate neurologically. Finally, it is conceivable that the myelopathy is nothing to do with this patient's liver disease but, within the limits of modern investigation, we can find no other explanation.

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