minimus (normal >2-8 mV) and 0-66 mV in the extensor digitorum brevis (normal >2-5 mV) (figure). A follow up examination one year later again showed normal motor and sensory nerve conduction. Repetitive stimulation of the ulnar nerve at 3 Hz recording from the abductor digit minimus showed a 17% decremental response and post-tetanic potentiation of 30%. Compound muscle action potential amplitude of the abductor digit minimus was 7-5 mV.

Antibodies to N-type calcium channels were assayed in the Institute of Molecular Medicine laboratories, Oxford, using [3H]-w-conotoxin-labelled human neuroblastoma (SK-N-BE) cells. Titres are expressed as pM, values >30 pM being considered positive. The Figure shows serum anti-VGCC antibody titres at four time points after the diagnosis of SCD. The titre was also measured in a CSF sample taken about six months after presentation. This titre was 42 pM; correcting for the IgG concentration of 0-02 mg/ml gave a normalised titre of 2100 pM IgG. A progressive decline in the serum anti-VGCC antibody titre was noted, concurrent with the increase in amplitude of the abductor digit minimus compound muscle action potential amplitude, over the next 18 months (figure). Two years after initiation of immunotherapy, the patient showed improvement of gait and strength, although diplopia and cerebellar dysfunction persisted.

This patient shows the clinical features of SCD and the clinical and electromyographic features of LEMS. Prior reports have shown a positive effect of plasma exchange and immunosuppressive treatment in patients with LEMS, including a fall in the titre of anti-VGCC antibodies with immunosuppressive treatment associated with improved electrophysiological measures of disease activity. With immunosuppressive treatment, our patient showed improvement in neuromuscular, but not cerebellar, symptoms and this improvement was associated with decreasing serum titres of anti-VGCC antibodies (figure).

The finding of raised antibody titres to VGCC in patients with LEMS, and in patients with Lambert–Eaton myasthenic syndrome (LEMS), raises the possibility that the two disorders are due to autoantibodies of similar specificities. The presence of anti-VGCC antibodies in both serum and CSF in our patient is consistent with a previous hypothesis. The development of SCD in only a subset of patients with LEMS suggests that if both are due to the same autoantibody, the ability of the antibody to enter the CNS might be altered. This could be due to higher serum titres of anti-VGCC antibodies in patients with SCD and LEMS, or to intrathecal antibody production as occurs in multiple sclerosis. The second possibility is supported by the presence of oligoclonal immunoglobulin bands in the CSF.

The lack of effect of immunosuppressive treatment on the symptoms due to SCD, by contrast with those due to LEMS, may be because of irreversable damage to Purkinje cells. Alternatively, the immunosuppressive regimens used to date may be ineffective in penetrating the blood–brain barrier and the CNS. This could be because antibody forming cells making pathogenic antibody have taken up residence inside the blood–brain barrier, thus making it less accessible to immunosuppressive treatment. It is also possible that cerebellar neurons are affected differently by anti-VGCC antibodies than presynaptic terminals at the motor endplate. Neurons express a number of subtypes of VGCCs, some of which are expressed preferentially by cerebellar Purkinje cells.

The pattern of combined central and peripheral nervous system involvement in this patient, together with increased antibody titres to VGCCs and a differential response of neuromuscular symptoms to immunosuppression, suggests that VGCC antibodies may play a differential role in the pathogenesis of the peripheral and central deficits in LEMS and SCD.

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10 Miller RJ. Multiple calcium channels and neuronal function. Science 1987;235:45-52.

Benign thalamic cyst presenting with contralateral postural tremor

Postural or action tremors have several aetiologies, but intracranial masses are rarely implicated as a cause. We present a young girl with a cystic mass of the right thalamus, whose sole presenting complaint was a unilateral postural tremor affecting the left upper and lower limbs.

A 11 year old girl presented with involuntary movements of the left upper limb for seven months and the left lower limb for five months. She had no history of weakness of these limbs, gait disturbance, or symptoms suggestive of raised intracranial pressure. On examination she had no papilloedema or weakness of any of her limbs. There was a coarse postural tremor (6-8 Hz) of the outstretched left upper limb, which was more prominent in the distal muscle groups. On attempting to stand or walk, the left lower limb also manifested the tremor; it was most evident in the ankle and foot. The tremor was accentuated slightly with movement of the limbs and seemed to have a greater amplitude while nearing the target in the finger to nose test. The tremor was abolished when the limbs were at rest and when the patient was asked to clench her max- mus or gaze ataxia and had a normal muscle tone and a normal speech. There was no dystonic posturing or sensory abnormalities.

CT of the head showed a large, well defined, rounded, non-enhancing, high density mass occupying the right thalamus with the same density as CSF. The lateral and third ventricles were normal sized. MRI suggested the presence of fluid in the mass with an intensity similar to that of CSF (figure A) and (B). A CT guided stereotactic aspiration of the cyst yielded 45 ml of clear, colourless fluid. As there was no enhancing wall a target was not chosen for biopsy. Post-operative CT showed pronounced reduction in the size of the cyst with no blood within or outside it. Analysis of the fluid showed 47.1% glucose, 33 mg/dL, and 2 lymphocytes per mm3. The fluid was centrifuged and the sediment examined for cyscercicercus scolices, hydatid sand, and malignant cells, all of which were negative. A diagnosis of a benign thalamic (ependymal) cyst was made. There was a dramatic improvement in her symptoms in the immediate postoperative period with virtual cessation of the tremor.
At follow up three months after surgery, the patient was asymptomatic and had no tremors of the left upper or lower limbs. A small residual cyst was seen on CT but it was not producing any mass effect or ventricular dilatation. No further treatment was offered and she was advised clinical and CT monitoring.

Intracranial tumours are on occasions known to produce a parkinsonian syndrome with rest tremors, bradykinesia, and rigidity. Another type of tremor that has been described in association with intracranial masses is “rubral tremor”. Involvement of the red nucleus and the decussating fibres of the superior cerebellar peduncle by lesions in the midbrain region has been implicated in the production of “rubral tremor”. In both these instances, the tremor is present at rest, although rubral tremors may be aggravated by maintenance of a posture or goal directed movement. Postural tremors can theoretically occur in isolated cerebellar hemispheric lesions including mass lesions, but this manifestation is rarely seen in clinical practice. The cerebellar connections with the brainstem, especially the superior cerebellar peduncle, should be spared for a postural type of tremor to occur. An intention tremor results if these connections are involved. The postural tremor caused by cerebellar lesions is a slow tremor of 4 to 5 Hz and is thought to result from hypotonia of the affected limbs.

The postural tremor in our case was unexpected considering the location of the mass in the thalamus with compression of the adjacent basal ganglia. The tremor is likely to be affected by the cyst include the basal ganglia and their connections, dentatorubrothalamic fibres, and the thalamic nuclei. Lesions of these structures are not known to produce isolated postural tremors. The mass in our patient could be expected to produce a rest tremor. Even an intention tremor is conceivable considering the possible compression of the superior cerebellar peduncle by the cyst. As mentioned, only discrete lesions of the cerebellar hemisphere have been considered to cause postural tremors and our patient had neither hypotonia nor involvement of the cerebellar hemispheres.

The tremor in our case was contralateral to the side of the tumour and responded to aspiration of the cyst contents, suggesting that the pressure exerted by the cyst on adjacent neural structures was in some way responsible for the production of the tremor. To the best of our knowledge there has been no previous report of unilateral postural tremor caused by a thalamic or basal ganglia lesion.

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1 Friede RL, Yasargil MG. Supratentorial intracerebral epithelial (ependymal) cyst: review, case reports and fine structure. 3 Neurosurg Psychiatry 1971;40:127-37.

**MRI of neuromyelitis optica: evidence for a distinct entity**

Neuromyelitis optica was originally described as a distinct demyelinating disorder characterised by visual disturbances and spinal cord signs occurring closely in time. Later studies on patients presenting with this syndrome often reported the subsequent evolution of additional neurological deficits, found pathological evidence for a more widespread demyelination, or disclosed other specific aetiological such as acute disseminated encephalomyelitis, Behcet’s disease, or systemic lupus erythematosus. From these findings it was concluded by many that neuromyelitis optica was inseparable from multiple sclerosis and its existence as a specific entity was questioned. MRI promises to alter this attitude, as we show by the presentation of two cases.

Starting in 1981, case 1, a 28 year old woman, had repeatedly experienced paraesthesiaes of the extremities and episodes of blurred vision. In 1988, during a further bout of visual disturbance MRI of the brain showed swelling and contrast enhancement of the optic chiasm and of the prechiasmatic portion of the optic nerves. An exploratory frontal craniotomy six months later showed gliotic optic nerves that were enucleated by abnormally thick arachnoidea but no tumour.

One year later the patient was first seen at our department for rapid onset of left leg weakness and bilaterally ascending paraesthesia. At that time visual acuity was reduced to finger counting. MRI showed swelling of the upper thoracic cord with irregular intramedullary enhancement after giving gadolinium-DTPA. The brain seemed normal, including the chiasm. Lumbar puncture and a spinal angiogram were uninformative. Laboratory tests were negative for involvement of any other organ, systemic infection, or collagen vascular disease. No specific antibodies except for an intermittently raised titre against Toxoplasma gondii was found.

Over the next four years the patient continued to have bouts of myelitis at times involving almost the entire spinal cord, as shown by MRI. Cell counts in the CSF ranged from 0 to 10/mm^3 and consisted mainly of lymphocytes and activated monocytes. Sometimes fat containing macrophages indicating tissue necrosis were also seen. The protein content of CSF was mostly increased and ranged from 71 to 153 mg/100 ml but there was no evidence of intrathecal immunoglobulin production. Repeated search for oligoclonal bands was negative.

Various therapeutic regimens including high dose steroids, immunoglobulins, immune adsorption, and antibiotic treatments failed to stabilise the patient’s clinical condition. When she was last seen in mid-1992 she was bedridden because of a spastic tetraparesis with a bilateral sensory level at C2 and had unchanged severe visual impairment. MRI of the brain continued to be normal whereas the spinal cord at the cervical and upper thoracic region was very atrophic. The patient refused the administration of contrast material at that time.

For five months case 2, a 22 year old woman, had experienced continuous deterioration of visual acuity associated with increasing bilateral leg weakness and urinary incontinence before she was seen at our department. Visual field testing showed concentric narrowing on the right and a temporal hemianopia on the left with sparing of the fovea. Her right arm distal to the elbow was mildly hypaesesthesia and sensation to pinprick and pain was reduced distally from T7 bilaterally. The plantar response was upgoing on both sides. A spinal tap gave a colourless CSF with 4 lymphocytes/mm^3 and a protein concentration of 50 mg/100 ml. There was no evidence of oligoclonal bands. MRI showed right sided enlargement of the chiasm and of the right optic nerve with uptake of contrast material. The remainder of the brain was normal. There was also minimal swelling of the cervical medulla and patchy, intramedullary enhancement from C1 to
critically significant groups × measures interaction.

The effect that Rosser and Hodges claim may well be present in their data and, if so, would help to contribute to the understanding of the relation between allegedly cortical and subcortical dementias. Unfortunately the analyses actually reported do not properly permit the suggested conclusion to be drawn and it would therefore be useful to know if the relevant interaction really is significant.

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Hodges replies:

We are very grateful for Miller's comments on our paper. We have now performed a split plot analysis of variance with both within subject and between subject effects, which confirmed the presence of a significant interaction (F (df 41,3) = 19.6, P < 0.001). This highly significant group conditions interaction confirms the differential effect of Alzheimer's disease vs Huntington's disease and progressive supranuclear palsy on category and letter based fluency tests, which we hope will convince Professor Miller.

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Jarisch-Herxheimer reaction in a patient with neurosyphilis: non-convulsive status epilepticus?

In their lesson of the month Zifko et al.1 described a patient with neurosyphilis who developed fluctuating consciousness, disor-

Firstly, autonomic symptoms of fever and tachycardia in our patient occurred before onset of confusion and altered consciousness, and were not present during the period of disorientation and psychomotor restlessness, as mentioned in the text. Secondly, although periodic lateralised epileptiform discharges may be a sign of non-convulsive status, this abnormality is non-specific. Thirdly, during CT, psychomotor restlessness was treated with 70 mg intravenous diazepam, which did not affect the confusion. Non-convulsive status of complex partial type usually responds well to diazepam.

Zifko et al reply:

We appreciated the interesting response of Dune and Heye to our article. Although we cannot completely exclude the possibility of non-convulsive status epilepticus in our patient, we have several arguments that make their theory unlikely.

Risperidone in Parkinson's disease

A recent, excellent review of the management of Parkinson's disease called attention to the atypical neuroleptic drug clozapine (an antagonist of dopamine D2 and serotonin 5HT2 receptors) for amelioration of psychotic symptoms derived from dopaminergic treatments, when temporary withdrawal of antiparkinsonian drugs fails. Small doses of the atypical neuroleptic drug risperidone (0.25–1.25 mg/day) can also be used for ameliorating hallucinations induced by levodopa without worsening motor symptoms in Parkinson's disease.2 Risperidone has a strong affinity for 5HT2 receptors and only moderate affinity for D2 receptors. For parkinsonian patients in whom the 1–2% risk of agranulocytosis with clozapine is unacceptable, risperidone is another option.

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