oral chlorambucil resulted in no significant improvement.

All patients had weakness and wasting, with electrophysiological evidence of demer-
vation-reinnervation in one or more extremities that was not attributable to focal nerve or roots lesions. No upper motor neuron signs (bulbar dysfunction, spasticity, brisk reflexes) or cranial nerve dysfunction were seen. No patient had sensory signs or electrophysiological evidence of a peripheral neu-
ropathy involving sensory fibres. Laboratory, enzyme and biochemical, including serum immunoelectrophoresis, CSF and neuroim-
aging studies were carried out to exclude other conditions which may cause motor neuron signs.

Patient 1 was a 62 year old woman with asymmetrical tetraparesis who had progres-
sive weakness, suggestive of a left sciotic neuropathy, for six years. She experienced cramps and fasciculations, right sciotic neu-
ropathy for two years, and radial and median neuropathies for one year. The progression was not slowed by prednisone 1.5 mg/kg per day, for five months. Examination revealed right asymmetrical generalised weakness and muscle wasting, distal in the upper extremities (grade 3 to 4) and distal greater than proximal in the lower extremities (grade 0 to 4). The tendon reflexes were decreased or absent. She was then given oral chlorambucil 0.15 mg/kg per day over a period of six months, without modifying the progression of the disability.

Patient 2 was a 67 year old man who had progressive weakness with wasting and cramps involving the right upper limb for four years. Two surgical explorations of the radial nerve were carried out, without evid-
ence of entrapment. Six months before admission he developed cramps and weak-
ness of his right lower limb. Examination revealed multifocal weakness and cramps and fasciculations in the distribution of the right ulnar, radial and axillary, left radial and right sciatric nerves. Reflexes were decreased or absent. The symptoms and signs were not modified by prednisone, 1.5 mg/kg per day for two months. He was then given oral chlorambucil 0.15 mg/kg per day for six months, without modifying the symptoms.

Patient 3 was a 37 year old man who experienced left hand clumsiness, progressive weakness and wasting with cramps of his forearm for two years. Examination revealed that left deltoid, biceps and triceps (MRC grade 4), wrist extensors and flexors (MRC grade 3), finger extendors and flexors and intrinsic hand muscles (MRC grade 0) were weak and amyotrophic, with active fascicula-
tions throughout. Tendon reflexes were absent in the left upper limb. He was treated with chlorambucil, 0.15 mg/kg per day for 6 months, without improvement.

Strength testing was quantified using a manual muscle score, based on the Medical Research Council scale. Flexion and extension at the ankle, wrist, elbow, hip, knee, and shoulder abduction were tested bilaterally. The patient's ability to perform motor activities of daily living skills was rated using a simplified Barthel index. Motor function was tested at the start of oral chlorambucil and at one, three and six months apiece.

The presence of anti-GM1 antibodies in sera was measured before and after treatment by ELISA using purified ganglioside anti-
gens, as previously reported.5,6 titres greater than or equal to 100 units were considered as "high" in our laboratory.

Multisegmental motor nerve conduction studies were performed with percutaneous supramaximal stimulation while recording the compound muscle action potential with Teca 4 mm disc electrodes. Determination of conduction block and temporal dispersion was made based upon the criteria of Feasby et al. F-wave latencies from at least 10 responses were recorded following stimulation at the wrist or ankle. Using surface electrodes, sensory nerve action potentials were recor-
ded orthodromically. Needle examination was performed using standard Disa concen-
tric electrodes.

The physiological and therapeutic profiles of the 3 patients are presented in the table. All patients showed evidence of a multifocal neuropathy involving motor fibres but spar-
ing of sensory axons. This was shown as asymmetrical and variable conduction block along motor axons in patients 1 and 2, prolongation of F-wave latencies and reduc-
tion of the CMAPs amplitude. Axonal loss was found in all patients, was prominent in patient 1, as shown by denervation potentials at rest, and voluntary motor unit potentials indicative of chronic denervation-reinnerva-
tion.

Relative strength and simplified Barthel index measurements were not improved and both parameters declined or were stabilised over time. By ELISA assay, the levels of anti-
GM1 measured after treatment with chlor-
ambucil were not modified.

Treatment was well tolerated, without haemo-
atological side effects but patients 1 and 2 developed transient haemorrhagic cystitis.

Our study was designed to assess the efficacy of chlorambucil in the treatment of multifocal motor neuropathy syndrome pre-
senting as a progressive, multisegmental weakness with wasting, fasciculations and cramps and anti-GM1 antibodies. Our patients were similar to several previously reported cases of selective motor neuropathy, with or without electrophysiological evidence of conduction block. Recent data have suggested that antibody responses to gangli-
osides are probably T-cell independent responses of B cells and that prednisone is ineffective in suppressing antibodies. Cyclophosphamide has been shown to be effective but in view of the danger of serious toxicity, therapy with this agent is limited.

We therefore elected to use chlorambucil, an immunosuppressive drug with some cell specificity but with fewer side effects.

Clearly, our results demonstrate that chlor-
ambucil is not an alternative drug that can be recommended. Chlorambucil lacked a thera-
peutic effect as judged in the absence of improvement in the manual muscle test score and simplified Barthel index. Moreover, chlorambucil did not reduce significantly the titre of anti-GM1 antibodies. The duration of therapy seems to have been sufficient in our cases, because improvement in strength began within two to five months in the reported improved patients.6,7

In summary, we feel that the use of chlorambucil is not effective in patients with lower motor syndrome and anti-GM1 anti-
bodies. Whether other regimes of immuno-
suppression other than cyclophosphamide or the administration of parenteral immuno-
globulins that have been shown to be effective in other neuropathies, such as chronic inflammatory demyelinating polynuropathy should be used, remains to be investigated.

Table: Physiological and therapeutic response profiles

<table>
<thead>
<tr>
<th>Patients</th>
<th>Motor conduction studies</th>
<th>Anti-GM1 Titer before/after therapy</th>
<th>Manual Muscle Score before/after therapy</th>
<th>Simplified Barthel Index before/after therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>NV</td>
<td>103/120</td>
<td>50/35</td>
<td>78/78</td>
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<td>2</td>
<td>NV</td>
<td>630/612</td>
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<td>78/78</td>
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<tr>
<td>3</td>
<td>NV</td>
<td>132/164</td>
<td>50/35</td>
<td>78/78</td>
</tr>
</tbody>
</table>

NV = at or within 15% normal values.
U = ulnar, R = radial, M = median, T = tibial, P = peroneal.

Values in brackets are the ratios proximal/distal motor action potential area.

Unilateral vestibular paralysis as the sole manifestation of mumps

A 36 year old man noted sudden rotational vertigo with nausea, vomiting and postural imbalance. The next day clinical and electro-
oculographic findings confirmed a left vestibular paralysis. There was spontaneous horizontal-rotatory (counterclockwise) nys-
tagmus to the right enhanced by Frenzel's lenses and increasing on gaze to the right. Stance and gait were unsteady with a tenden-
cy to fall to the left. The left horizontal semicircular canal was unresponsive to stim-
ulation with warm (44°C) and cold (30°C) water. The audiogram was normal.

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Body temperature was normal. White blood cells were elevated (15000/mm³) and bands in normal (85%/16/mm³) within 3 days. CSF analysis revealed 19 white cells/mm³ (with 85% lymphocytes and 3% plasma cells), normal glucose, lactate and protein content without IgG oligoclonal bands. The EEG was normal. On day 2, serum antibody testing (ELISA) revealed positive mumps virus IgM titer up to a dilution of 1:320 and negative IgG (for dilutions >1:40). CSF antibody testing was negative (for dilutions >1:1). Negative mumps virus IgG titers (ELISA) in the serum were positive for dilutions up to 1:640 and negative for IgM (for dilutions >1:40). There was complete opalescence of peaks and thymic stimulation showed improved vestibular function on the left (hyporesponsiveness of 43%). CSF was normal.

Sudden deafness with or without abnormal caloric responses is a possible complication of epidemic parotitis. Several investigators have suggested that subclinical mumps infections without parotitis may produce sudden unilateral hearing loss sometimes with vertigo. Our patient was free of symptoms and did not complain of hearing loss.

In our patient, hearing was spared and only unilateral vestibular paralysis accompanied the serologically proven acute mumps infection. This condition has not been described previously. The Kilham strain of the mumps virus and a neurotrophic mumps strain (isolated from the CSF of a child with mumps meningitis) may infect the endolymphatic structures of the labyrinth. Neurons of the vestibular and cochlear ganglia were regularly infected by the Kilham strain but only occasionally by the neurotrophic strain. Infections of the labyrinthine endolymphatic structures or of the vestibular ganglia may be followed by vestibular paralysis and such a mechanism seems most likely in our patient.

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Ataxic hemiparesis with chorio-oral syndrome in capsular infarction

Ataxic hemiparesis (AH) is defined as an unusual combination of ipsilateral pyramidal and cerebellar signs. AH is ascribed to lacunar or thalamocapsular, supratentorial and brainstem locations. Several reports have established that accompanying hemihypersthesia indicates a thalamocapsular rather than a pontine location for lesions causing the AH syndrome. We describe a variant of AH with sensory impairment restricted to a chorio-oral topography, in association with left hemiparesis.

A 73 year old woman, known to have been hypertensive and diabetic for 10 years, suddenly developed slurred speech and a tingling sensation on one hemiface and in her left thumb, index and middle fingers. Soon after she noticed weakness in her left arm and leg. On admission the next day, general physical examination was normal. Blood pressure was 220/110 mmHg. She was alert and had normal mental status. Her speech was dysarthric. There was slight left central facial weakness and a grade 3/5 left hemiparesis. Reflexes were increased on the left, with Babinski's sign present. Pinprick and light touch sensation were diminished in the left tongue, buccal mucosa, lips and cheek, and in the thumb, index and middle fingers of the left hand. Proprioceptive sensation was unimpaired. The patient was unable to walk alone and fell to the left. ECGs and routine laboratory tests were normal except for a blood glucose level of 207 mg/dl.

By the sixth day, her power improved to grade 4/5. It was at this time that she appeared to have moderate dysmetria and intention tremor of the cerebellar type, with the heel-shin and finger-nose tests on the left. The ataxia was more apparent in the leg than in the arm. CT scan obtained at that time revealed a small hypo-density area in the right posterior limb of the internal capsule with possible involvement of the adjacent lateral thalamus (see below). Parietal-recorded somatosensory potentials following cortical and thalamic stimulation were studied two weeks after clinical onset. The short-latency components from the left side showed decrease in amplitude of N20 to 35% of the normal side and slight attenuation of the subsequent peaks. Latencies were normal.

Eight months after onset, there was no weakness but minimal ataxia of the left extremities persisted. Paresthesia remained in the tips of the left first to third fingers and hypoesthesia for pain and light touch continued in the left perioral region. CT scan again revealed the capsular hypodensity lesion. Somatosensory evoked potentials did not show any asymmetry or abnormal responses.

Originally, persistent sensory disturbances were not considered as part of the AH syndrome. In subsequent reports, the clinical spectrum of AH was widened to include cases with persistent hemisensory deficit ipsilateral to the cerebellar-like ataxia and pyramidal tract weakness. This has been termed ‘hypothetic ataxic hemiparesis’. The sensory loss involves several sensory modalities, more often of the spinohemispheric cortex. The sensory loss is associated with parietal dysmetria and intention tremor of the cerebellar type. The dysmetria was considered as a result of the CNS lesion in the posterior limb of the internal capsule. This suggests that the patient's symptoms are most probably due to involvement of sensory thalamocortical radiation, which occupies the posterior part of the posterior limb of the internal capsule.

Abnormalities in short-latency components of somatosensory evoked potentials, as those observed in our patient, have been reported as being the same in lesions of the thalamus and thalamo-cortical radiations. In summary, atactic hemiparesis associated with ipsilateral chorio-oral syndrome is a previously undescribed symptom complex following capsular infarction. This association should be included in the spectrum of clinical syndromes presumably related to anterior cerebral artery territory infarction.

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Figure CT scan taken at 8 months of onset showing lacunar infarct in the right posterior limb of the internal capsule.