usual headache or nausea or both after the procedure. The suspected onset time of intracranial haemorrhage varied from two to seven days after myelography. The site of bleeding varied greatly and was in many cases multifocal: intracerebral, parietal and subarachnoid; intracerebral into basal ganglia; multifocal cortical; subarachnoid; intracerebral or frontal and parietal and temporal; and intracerebral or parietal were all reported. Two of the cases were associated with brain stem herniation.

The high number of such reports led us to investigate the literature for additional information. We found that cases of intracranial haemorrhage associated with myelography had been reported previously and one may be added to the reference list of Dr Satoshi et al. Interestingly, we also found that intracranial haemorrhage after spinal puncture without use of CM (for example diagnostic LP or spinal anaesthesia) was not unknown.1

Schub and Raskin made reference to another 14 cases in their presentation already in 1963.3

At this stage one may only speculate on the possible mechanism for the haemorrhagic incidents: a drop in intracranial pressure associated with CSF leakage may well increase the pressure gradient across fragile venules and capillaries. This in itself may increase the risk of bleeding in predisposed patients. CSF support of the brain may also be affected leading to “sagging” and shearing forces. Heavy vomiting may perhaps precipitate bleeding in some cases. The possible role of the CM is uncertain and harder to explain. At this stage, however, it is important to keep an open mind and to keep looking for explanations. We are grateful to Dr Satoshi et al for publishing their case. Hopefully this will contribute to a renewed interest in this serious but extremely rare complication of the myelographic procedure.

I am personally convinced that the problem has been under-reported as it apparently may occur after a considerable delay thereby hindering the possible association with myelography. A renewed interest may lead to an increased recognition and reporting, thus providing us with a better understanding of incidence, avoidable risk factors, and mechanisms in the future. HP BÖHN

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**Plasma infusion in childhood Guillain-Barré syndrome**

Guillain-Barré syndrome (GBS) is probably the most common cause of acute motor paralysis in children. Although many patients with GBS make a good recovery, there is a mortality of 3–5%, 10–20% require artificial respiration, and 10–22% remain disabled.1 Severity and outcome in adults and children are similar.1 Several studies have shown the usefulness of plasmapheresis in severely affected adults in a specialised setting.1

Technical difficulties and haemodynamic complications have precluded the routine use of conventional plasmapheresis in children. We report four children with GBS and two with chronic inflammatory polyneuropathy who responded well to treatment with plasma infusions.

The criteria used for the diagnosis of GBS were those recommended by the National Institute of Neurological and Communicative Disorders. Irrespective of age, sex, and weight, 250 ml of fresh frozen plasma, tested negative for hepatitis B and HIV, were infused daily through an intravenous cannula into a convenient limb vein for 10–14 days. The clinical progress was assessed in the early stages of the bedridden patient with the MRC sum score: six muscles (deltoit, biceps brachii, extensor carpi, iliopsoas, quadriceps, tibialis anterior) were tested on both sides on the MRC scale to give a score ranging from 0 to 60. We also used the grading scale from the GBS study group: grade 0—healthy; grade 1—minor symptoms or signs of neuropathy; grade 2—able to walk 5 m without assistance; grade 3—able to walk 5 m with assistance; grade 4—confined to bed or chairbound; grade 5—requiring assisted ventilation. None of the children were given steroids or other immunosuppressive drugs.

The nerve conduction velocities (NCV) of the median nerve and the compound muscle action potential (CMAP) from the abductor pollicis brevis were recorded on day one and day 14 of treatment. There were four girls and two boys (age range five to 11 years). Cases 1 to 4 presented acutely, case 5 presented with grade 3 weakness five weeks after onset, and case 6 was seen 40 days after a relapse. None required tracheostomy or assisted ventilation, though the vital capacity reached a critical value of 300 ml in cases 3 and 4. All six showed a good response. The relapsing patient (case 6) improved dramatically from grade 4 to 2 in eight days. Response to plasma infusion is shown in the table. All children required less than 14 plasma infusions to improve one grade. All, including the children with relapse and residual weakness, improved enough to walk unaided by the third week. The duration of hospital stay was reduced to 10–30 days. At the time of discharge from hospital all were ambulant without aid and had only a mild gait abnormality with some difficulty in getting up from a squatting position. NCV of the median nerve and CMAP of the abductor pollicis brevis showed minor improvements in cases 1 to 4, but in the other two there was no measurable improvement after two weeks (table). No haemodynamic complications or allergic reactions were seen and plasma protein measurements twice a week did not show any significant change.

Whether the type of replacement fluid used has any beneficial effect is debatable.4 Improvement seen after high dose gammaglobulin infusion in chronic inflammatory polyneuropathies and in Guillain-Barré syndrome5 prompted us to use plasma infusions in children, in whom only plasmapheresis may be required to produce beneficial results.

Although our study had no control group and consisted of only six patients, the impressive results are worth reporting. The children’s length of hospital stay was reduced to an impressive 10–30 days, much less than the average hospital stay of children managed conservatively, which is around 84 days.6 None of the children required assisted ventilation, though the vital capacity fell to 300 ml in two patients. Possibly the early institution of treatment with plasma prevented their respiratory function deteriorating further. The time taken to improve, and the time taken to reach grade 2 were also remarkably shorter when compared with results from other series of conservative management and plasmapheresis.7–9 High dose gammaglobulin, which is very expensive, is not available in Sri Lanka and plasmapheresis cannot be done as the necessary equipment is not available. Therefore in a developing country like ours the only treatment we can offer these critically ill children is plasma infusion. A larger controlled study is planned.

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**Table Response to plasma infusion in six children with Guillain-Barré syndrome**

<table>
<thead>
<tr>
<th>Case number</th>
<th>Duration of illness before infusion (days)</th>
<th>Grade at infusion*</th>
<th>Time to improve one grade (days)</th>
<th>Time to reach grade 2 (days)</th>
<th>Duration of Hospital stay (days)</th>
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<th>CMAP***</th>
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</table>

*North American trial grading scale/MRC scale arm, leg.
**Nerve conduction velocity (normal for median nerve 51m/s).
***Compound muscle action potential (normal for wrist 10 mv).
Bilateral ptosis, ataxia and areflexia—a variant of Fisher’s Syndrome

Since the original description of the benign neurological syndrome of ophthalmoplegia, ataxia and areflexia (SOAA) by Fisher in 1956, there has been a continuing debate as to whether the syndrome represents a variant of Guillian-Barré syndrome or a form of brainstem encephalitis. Some authors now prefer the hypothesis that SOAA is a unique syndrome combining central and peripheral involvement.1

A 56 year old male was admitted to University Hospital with a history of distal numbness in all four limbs, unsteadiness for four days, and severe bilateral ptosis for one day. He experienced a “flu-like” illness one week before admission. On examination, he was alert and orientated. Nasal tone in his speech was noted. Palpebral fissures were symmetrical and measured 3 mm on resting and 5 mm on maximal opening by using frontal muscles to compensate for the weakness (fig a, b). No limitation of extraocular movement in any direction could be detected. The pupils were 5 mm in diameter on both sides and reacted normally to light. There was no facial weakness or difficulty in swallowing. Normal convergence and Bell’s phenomenon were easily demonstrated. Muscle power to all four limbs was normal. There was marked ataxia on walking and Romberg’s sign was positive. Heel-to-shin test was poorly performed. Finger-to-nose and pronation-supination tests were mildly impaired. There was a generalised areflexia of the limbs. Sensory examination revealed a decrease in vibration and joint position senses in the lower limbs.

Laboratory investigations: CSF examination on the sixth day after onset showed a protein level of 90 mg/dl without pleocytosis. Nerve conduction studies (NCS) on the same day revealed absence of sensory action potential (SAP) in median and ulnar nerves. Somatosensory evoked potentials (SSEP) on the ninth day by stimulation of the median and peroneal nerves revealed prolonged brachial plexus potentials and scalp SSEP from the upper to the lower limbs. Brain stem auditory evoked potentials and patterned visual evoked potentials were all within normal limits. Cranial CT and EEG were normal. No improvement of ptosis occurred with a standard Tension test.

Only supportive treatment was given during admission to hospital. The ptosis started to improve four days after admission and completely resolved on the tenth day. Ataxic gait gradually improved during the same period of time. Only areflexia was recorded at four weeks. Follow up examination revealed no residual symptoms or deficit 11 months after discharge. Repeat NCS and SSEP studies showed marked improvement.

Among the three cardinal features, ophthalmological signs are so varied that they have received a lot of attention in the literature. There have been a variety of clinical presentations documented which raise the possibility that the ocular problems may be supranuclear in origin, notably, a discrepancy between mild ptosis and marked external ophthalmoplegia. The patient, however, presented with isolated and symmetrical ptosis of the upper eyelid of a severe degree without limitation of extraocular movements. Eyelid ptosis is usually explained by weakness of the levator palpebrae superioris muscle due to an oculomotor nerve lesion, or by weakness of Muller’s muscle secondary to involvement of sympathetic innervation, or by intrinsic disorders of the lids and their musculature. Two other possible, but less well-known causes might be ‘cerebral’ and ‘midbrain’ ptosis. The former may be due to failure of some control of elevation of the eyelids exerted cortically. The latter form of ptosis may be due to the anaesthetic management of neurons in the caudal midline of the third nerve nucleus which supply the levators of the eyelids. Clinical observation in this patient could not be explained by the involvement of infra-nuclear mechanism of ocular movement. Transient, symmetrical ptosis in this patient was either due to a self-limiting and inflammatory response in the peri-aqueductal area as proposed by Meineberg2 or to a failure of a corticobulbar influence.

Prolongation of neck or scalp SSEP latencies in this patient with abnormal NCS were in agreement with a sensory neuropathy involving large myelinated fibres as suggested by Gulloiet al. It is likely that sensory polyradiculopathy plays an important role in the ataxia and areflexia of this syndrome. Based on neuro-ophthalmological observations and electrophysiological results in this patient, combined central and peripheral involvement is probable. The clinical manifestations, laboratory findings and clinical course closely resemble those reported cases of SOAA except for the ophthalmological features. Such combinations may be regarded as one of the variations of SOAA.

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Progressive supranuclear palsy as the sole manifestation of systemic Whipple’s disease treated with pexoflaxine

Oculomotoric myorhythmia (OMM) with progressive supranuclear oculomotor palsy has been reported only as a cerebral complication of systemic Whipple’s disease and is thus possibly a unique and pathogonomonic movement disorder in this condition. Whipple’s disease may be confined to the CNS so that when OMM occurs without symptoms, positive evidence for Whipple’s disease may be difficult to find especially when perendoscopic and peroral jejunal biopsy specimens are normal. CSF examination and brain imaging may also be normal. In these circumstances, some authors suggest that cortical brain biopsies have a low sen-