thickened hyaline walls which stained with Congo red, showing the characteristic apple green birefringence under polarised illumination, and were positive with immunostaining for β A4. This immunostaining also showed diffuse plaques, a few containing cores most of which could not be seen on routine staining. No abnormalities were seen in the white matter. Silver impregnation (Bielschowsky) and tau immunohistochemistry did not show neurofibrillary tangles, neuritic plaques, or cortical dystrophic neurites. According to the criteria of the Boston Cerebral Amyloid Angiopathy Group,³ the histological findings and clinical data indicated a diagnosis of "probable cerebral amyloid angiopathy with supporting pathological evidence". DNA was screened by polymerase chain reaction amplification for the presence of mutations in the amyloid precursor protein (APP) gene, which are known to cosegregate with hereditary cerebral haemorrhage with amyloidosis Dutch type (HCHWA-D; codons 670/671 of exon 16 and in codons 692, 693, 713, and 717 of exon 17). No mutation was found.

The patient underwent psychometric reassessment 1 month after the intracerebral haemorrhage and surgical drainage, from which she had made a good physical recovery. In keeping with the location of the haemorrhage, she was found to have significantly impaired visual perceptual functions, with left unilateral neglect, an inability to discriminate shapes, and a severe deterioration in her performance on the visual version of the recognition memory test (verbal version remained within the average range). Additionally, a mild degree of verbal intellectual deterioration was noted (verbal IQ 83).

A wide variety of disorders may produce cortical myoclonus: the differential diagnosis includes anoxic injury, focal CNS damage (vascular or neoplastic lesions of sensorimotor cortex), encephalopathies (metabolic, mitochondrial, viral, paraneoplastic, toxic), degenerations (basal ganglia, spinocerebellar), malabsorption syndromes (coeliac disease, Whipple's disease), storage disorders, and dementias (Creutzfeldt-Jakob disease). Myoclonus may also be encountered in Alzheimer's disease, in which it is associated with increasing severity of dementia. We are not aware of previous reports of cortical myoclonus as a clinical feature of sporadic cerebral amyloid angiopathy. Although this could be a chance concurrence, it is possible that recurrent small intraparenchymal bleeds from angioathic vessels may have been causal, as vascular lesions are a recognised cause for cortical myoclonus. Another possibility which may be worth considering relates to the effects of amyloid β -peptide ($A\beta$) and its metabolic fragments on neuronal membrane ion channels and their associated currents.⁴ Because the pathophysiology of cortical myoclonus is thought to reflect both increased excitability and deficiency of inhibitory processes in the cortex,⁵ the effects of $A\beta$ on a variety of ion currents, both excitatory and inhibitory, might theoretically produce such an imbalance and hence contribute to the development of cortical myoclonus.

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Antiganglioside antibodies in various episodes in a patient with recurrent Guillain-Barré syndrome

Guillain-Barré syndrome is defined as an acute, monophasic polyneuropathy. It recurs in 2%-5% of patients.¹ Both the initial and recurrent episodes are preceded by an infection. Antibodies against several gangliosides are often present in the acute phase in serum samples from patients with Guillain-Barré syndrome.² There have been no previous reports, however, on anti-ganglioside antibodies in patients with recurrent disease. We report finding anti-ganglioside antibodies in a patient with recurrent Guillain-Barré syndrome during the third and fourth episodes.

Ten days after a bout of upper respiratory tract infection, a 26 year old man noted paraesthesias in his fingers and toes, and rapidly developed limb weakness. He had bulbar palsy and areflexic tetraparesis on day 1, and underwent three sessions of plasmapheresis on days 3, 4, and 5. Disability was maximal on day 6. Bulbar weakness disappeared on day 8, and he could walk without support on day 14. He had had similar episodes of acute, monophasic paralytic disease at the ages of 14 and 19. Six years later, he experienced a fourth episode. Three days after an upper respiratory tract infection, he rapidly developed dysarthria, digital paraesthesias, and leg weakness. Neurological examination on day 2 showed bilateral external ophthalmoplegia, facial diplegia, and areflexic tetraparesis. He required mechanical ventilation from days 2 to 19, and underwent five sessions of plasmapheresis on days 2, 3, 4, 6, and 9, then intravenous immunoglobulin therapy from days 11 to 15. His disability began to lessen on day 11 and had disappeared by day 34.

An enzyme linked immunosorbent assay³ showed IgG antibody titres to GM1 (<500), GM1b (128 000), GD1a (128 000), GalNAc-GD1a (<500), GT1a (64 000), GD1b (4000), and GQ1b (32 000) on day 3 during the third episode. No IgM anti-ganglioside antibodies were detected. On day

113, during the recovery phase, the anti-ganglioside IgG titres were <500. Serum on day 3 of the fourth episode had IgG antibodies titres to GM1 (<500), GM1b (256 000), GD1a (128 000), GalNAc-GD1a (<500), GT1a (64 000), GD1b (1:4000), and GQ1b (1:64000). Thin layer chromatography with immunostaining⁴ confirmed that the IgGs in the acute, progressive phase of both attacks reacted with the same gangliosides (data not shown).

Differentiation between Guillain-Barré syndrome and chronic inflammatory demyelinating polyneuropathy (CIDP) relies on the time needed to reach maximum disability. Our patient had four episodes of typical Guillain-Barré syndrome, not of CIDP. Neurological function in each episode reached a nadir within 2 weeks of onset and returned to normal within 5 weeks. The distinction between Guillain-Barré syndrome and CIDP, however, is blurred in the early onset phase in some patients. Protein concentrations in CSF in CIDP tend to remain raised during remission and at the onset of recurrence; whereas CSF protein concentrations are normal within 1 week of the onset of Guillain-Barré syndrome, which may provide a way of distinguishing it from CIDP.¹ Only low anti-ganglioside antibody titres, which were unlikely to have a role in the pathogenesis, were found in some patients with CIDP.⁵ The presence of high anti-ganglioside antibody titres at the onset of a recurrence may be of use in confirming the diagnosis of recurrent Guillain-Barré syndrome.

In recurrent Guillain-Barré syndrome, the nature of the antecedent illness usually tends to differ from episode to episode.¹ Our patient, however, had similar upper respiratory symptoms before each onset of neurological symptoms. Antecedent agents such as *Campylobacter jejuni* and cytomegalovirus have ganglioside epitopes,^{4,5} and therefore may produce the anti-ganglioside antibodies in Guillain-Barré syndrome.

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Urinary retention associated with a unilateral lesion in the dorsolateral tegmentum of the rostral pons

The existence of a brainstem region concerned with micturition has been known since the report of Barrington more than 70 years ago.¹ In animals such a pontine micturition centre has been located in the dorsolateral tegmentum of the rostral pons, corresponding to Barrington's micturition centre,² but such a centre has not been precisely localised in humans. We describe a patient with presumed rhombencephalitis presenting with urinary retention and present his MRI findings. To our knowledge, this is the first MRI demonstration of a circumscribed lesion related to the putative pontine micturition centre in humans.

A 30 year old man with no history of voiding problems developed high fever, light headedness, frequent urination, and voiding difficulty. One day after onset, urinary retention occurred despite his sensation of needing to void. Two days later, in addition to urinary retention, the patient also had left cheiro-oral dysaesthesia and horizontal diplopia. Prostate examination did not show abnormalities. Urinary retention was managed with an indwelling catheter until 7 days after onset of his symptoms. The patient did not have a history of overdistension of the urinary bladder. Even though all his symptoms began to improve, the patient was referred to our hospital for an evaluation of neurological abnormalities.

Examination 9 days after onset showed mild right horizontal gaze paresis with intact vestibulo-ocular reflex, ipsilateral saccadic pursuit, and hypaesthesia around the left mouth angle and thumb and index finger, suggesting involvement of the right pontine tegmentum. Indeed, in addition to scattered amorphous lesions in the pons and cerebellum, brain MRI (1.5 tesla) showed a discrete lesion in the right upper pontine tegmentum

(figure). An MRI of the spinal cord disclosed no abnormal intensity areas. Lumbar puncture yielded CSF with normal cell counts (3/ μ l) and mildly increased protein (52 mg/dl). Neither oligoclonal bands nor myelin basic protein were present in the CSF; intrathecal IgG synthesis was within the normal range. Serum autoantibody and viral antibody tests did not contribute to diagnosis. A CO₂ cystometry with sphincter EMG 3 weeks after onset showed increased bladder volume over 555 ml and atonic cystometrogram (detrusor areflexia), despite the absence of subjective urinary symptoms. Organic obstructive urological disease was radiologically excluded. All neurological and MRI abnormalities cleared by 1 month after onset. One year follow up showed no recurrence of the neurological symptoms.

Based on neurological findings, the patient was thought to have incomplete involvement of the right paramedian pontine reticular formation, smooth pursuit pathway, and medial lemniscus, suggestive of a right pontine tegmental lesion. The association of acute and reversible urinary retention was consistent with a lesion in the rostral portion of the pontine tegmentum.³ Indeed, the MRI with a special focus on the rostral brainstem substantiated our neurological assessment and showed a discrete lesion in the right dorsolateral portion of the upper pontine tegmentum. Other less distinctive lesions in the pons and cerebellum were seen on MRI, but no relevant neurological abnormalities were detected. Simultaneous involvement of the spinal cord was excluded by neurological and radiological examinations. Although the patient was considered to have possible rhombencephalitis, the question of whether it was caused by a direct virus invasion or parainfectious demyelination remains unclear.

In humans, an association between micturition disturbances and brainstem involve-

ment has been suspected on pathological and radiological bases.³⁻⁵ Early in 1926, Holman⁴ documented a relation between micturition disturbance and posterior fossa tumours. Later, a pathological study of brain tumours disclosed a high frequency (63%; 50/79) of voiding difficulties and urinary retention with pontine and fourth ventricle tumours.³ Histological abnormalities were concentrated in the tegmentum of the rostral pons in all such patients.³ The locus coeruleus and adjacent neural tissue were more often involved than other nuclei or regions of the pons.³ However, because of the extensive involvement of these tumours, no localised lesions were noted.³ One recent study with MRI on brainstem stroke disclosed similar results.⁵ Despite numerous reports on the association between multiple sclerosis and micturition disturbances, MRI studies to date have not delineated brainstem lesions specific to the impaired micturition.

In experimental studies, by contrast, Barrington had suggested that in the cat the micturition region was located in the dorsal part of the pontine tegmentum.¹ Recent investigators have reported that it can be located more precisely, in the nucleus locus coeruleus, locus coeruleus alpha, or the dorsomedial part of the dorsolateral pontine tegmentum.² Neurons in the pontine micturition centre may activate the parasympathetic excitatory outflow to the urinary bladder (detrusor), while there also exists a pontine storage centre ventral or lateral to the pontine micturition centre that controls external urethral sphincter function.² Because our patient had atonic bladder and urinary retention, the pontine micturition centre may have been the main site of brain involvement. Atonic bladder may reflect a "shock" state, as has been documented in some stroke patients. However, Sakakibara *et al*⁶ described three patients with atonic cystometrogram 3 months, 6 months, and 3 years after brain-

Brain MRI of the patient. (A) Axial view (2500/103; repetition time/echo time). (B) Sagittal view (2350/103). (C) Coronal view (2920/90). T2 weighted images show a hyperintense focus in the right dorsolateral tegmentum of the rostral pons, just dorsal to the internal edge of the superior cerebellar peduncle including the pontine reticular nucleus, medial parabrachial nucleus, and locus coeruleus. Arrows indicate amorphous lesions in the pons (C). A similar amorphous lesion in the midbrain tegmentum was not confirmed by the axial and sagittal views and is most likely influenced by the cerebral aqueduct (C).