Reply

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Sirs: Reimers and Pflüger [2] ask whether CNS involvement in acute and chronic neuroborreliosis is HLA related.

As already stated in our publication, the number of patients is too small to give an answer to this question. The comparison with the healthy control group indeed needs some corrections. The chi square test should be used with a so-called "Yates correction" when comparing such a small group of patients and in addition one has to multiply the probability value by the number of alleles which are investigated in this study. The HLA frequencies in the control group are the frequencies of a Caucasian population standardized worldwide in the 1980 Workshop as indicated in the text; but unfortunately this is not mentioned in the table [1]. The number of 100 was used only as a basis for the chi-square test and is indeed an error.

The differences between the two patient groups are preliminary and can only indicate a trend, particularly of a possible involvement for the well-known but rare HLA haplotype A29-B44-DR7. It should be mentioned that these three alleles occur together four times in the CNS group. To verify the involvement of one special HLA allele or of a single haplotype, further extensive investigation of patients and their families is required.

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Recovery from respiratory muscle failure in a sporadic case of Brown-Vialetto-Van Laere syndrome with unusually late onset

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Sirs: The Brown-Vialetto-Van Laere syndrome (BVVLS) is a rare form of amyotrophic bulbo-spinal palsy associated with neurosensory deafness. Up to now, only 26 cases have been described (see [1] for review). It is mainly inherited as an autosomal dominant trait, but sporadic cases have also been reported.

In December 1989 we examined a 38-year-old male patient with negative family history who had slowly developed progressive bilateral hearing loss from the age of 30. Soon after he complained of difficulty in swallowing, facial muscle weakness and impairment of manual dexterity. In the following years muscle weakness became widespread along with step-like progressive worsening of cranial nerve involvement. Neurological examination revealed bilateral deafness, slight bilateral external rectus weakness muscle, severe weakness and wasting of masticatory, velopharyngeal and tongue muscles. There was also diffuse weakness and wasting of axial and limb muscles, mainly affecting the intrinsic hand muscles. Respiration was shallow, involving accessory muscles, but breathing sounds were audible over all the chest and peripheral cyanosis was absent. Deep tendon reflexes were hyperactive in the lower limbs. Plantar responses were flexor and sensation and coordination were normal. Ophthalmological findings were normal.

Extensive laboratory analysis and cerebrospinal fluid examination, including immunological investigations, were normal. Needle electromyography showed fibrillation activity at rest in all involved muscles, with positive sharp waves and surviving motor unit potentials enlarged in amplitude and duration.

Electroneurography of sensory and motor peripheral nerves was normal. Cortical magnetic stimulation of the pyramidal tracts elicited no response. Audiometric examination showed severe bilateral neurosensory hypoacusia. Checkerboard pattern reversal visual evoked potential (EP) examination revealed a markedly delayed P 100 latency with 30 checks in both eyes (131–133 ms, mean normal value ± 3 SD = 115 ms). Somatosensory EPs to from median and tibial nerves were normal. Recording of brain-stem auditory EPs was prevented by the

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severe deafness. Nuclear magnetic resonance imaging of the brain was normal.

Cholinesterase inhibitors had no effect. We administered 1 mg/kg prednisone to the patient daily for 2 weeks, without any benefit. The patient was discharged in an unchanged state. One month later, acute breathing distress led to respiratory arrest. Endotracheal intubation (EI) was performed and mechanical ventilation was started in an intensive care unit (ICU). Neither upper airway obstruction nor clinical and radiological evidence of upper and lower respiratory airway disease was demonstrated. Tracheostomy was performed 10 days after EI and ventilatory support was continued for 2 months, resulting in gradual recovery of spontaneous respiration. Intermittent nocturnal positive-pressure ventilation by tracheostomy was continued. At home, long-term nocturnal ventilatory support allowed the maintenance of a satisfactory diurnal blood gas equilibrium (pH 7.38; pCO^2 44 mmHg; pO^2 86 mmHg; HCO^3 27 mEq/l).

To our knowledge, this is the second sporadic case of BVVLS displaying the complete phenotype. Our case also shows subclinical involvement of the optic nerve, which has rarely been reported [1, 8]. The other peculiar features of our case include very late onset of deafness and relatively short latency between hearing loss and onset of bulbo-spinal involvement. We did not observe improvement following steroid treatment, although it has been anecdotally reported [6]. No treatment is currently available. Descriptions of the illness often include an irregular course, usually resulting in death from respiratory muscle failure [3–6]. ICU procedures were reported as unsuccesful in one case [2]. However, following ICU procedures and continued nocturnal home respiratory assistance our patient has mantained satisfactory diurnal respiratory function. Since long-term nighttime ventilation is known to stabilize pulmonary function for years in patients with severe neuromuscular diseases [7], we suggest that the above-mentioned procedures should also be considered in patients with BVVLS.

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Stroke in an HIV-infected patient

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Sirs: Numerous diseases are usually listed as possible causes of ischaemic stroke in the young adult [4, 8]. HIV infection was not mentioned in some recent series [1, 3, 7].

A 36-year-old Brazilian Caucasian male cook, with bisexual promiscuous habits, had a sudden onset of left-sided weakness. He had no known risk factors for cerebral vascular disease and there was no abuse of intravenous drugs. General physical examination was normal and the neurological examination revealed a left hemiparesis mainly affecting the upper limb that rapidly cleared in a few days. CT showed an infarct in the right middle cerebral artery territory, which was confirmed by MRI. Carotid duplex, transcranial Doppler and two-dimensional echocardiogram were normal. Cerebral digital intra-arterial angiography showed occlusion of a rolandic-parietal branch of the right middle cerebral artery.

A complete coagulation study showed no abnormality. Search for lupus anticoagulant was negative. C protein and free S protein and antithrombin levels were normal. Serum and cerebrospinal fluid (CSF) titres for herpes virus, cytomegalovirus and VDRL were negative and there were no neoplastic cells in the CSF. CD4 count was 450 cells/mm^3 (19%) and sedimentation rate 19 mm (1st h). HIV1 was positive in CSF and serum (Western blot: antibodies against GP160, GP110, GP41, P68, P55, P40, P34, P25 and P18), with negative HIV2 (Western blot and ELISA). AgHBs, anti-HBc and anti-HBe were also positive in the serum.

Stroke is an uncommon event in HIV-infected patients [2, 9, 10, 11], most cases being secondary to endocarditis or haemorrhage in a tumour [5]. In this patient large intracranial and extracranial vessel disease and cardiac embolism were ruled out. There was no evidence of disseminated intravascular coagulation or of a prothrombotic state. There was also no evidence of syphilis [6], herpes or cytomegalovirus infections, which could be responsible for arteritis. Either HIV [11] or hepatitis B virus [6] may be the agent of a cerebral vascular arteritis. Stroke as the presentation form of HIV infection is exceptional, but this case is a reminder of the need to search for HIV infection as a possible aetiology in cases of stroke in young adults.

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