LETTERS TO THE EDITOR

Acute upside down reversal of vision in vertebrobasilar ischaemia

Acute upside down reversal of vision is an uncommon and little known phenomenon consisting of transient complete 180 degree inversion of the visual image. The pathogenesis and the anatomical sites of this dysfunction are unknown. Lesions involving cortical areas, mainly the parieto-occipital region, or the vestibulocerebellar system have occasionally been documented.¹⁻⁵

We observed two patients who experienced this bizarre visual illusion, both revealing features of vertebrobasilar ischaemia.

Patient 1, a 69 year old woman, was admitted because two weeks earlier she had experienced sudden malaise, sweating, nausea, vomiting, right occipital headache, followed by a 180 degree vertical inversion of the visual image, lasting about 20 minutes. Two similar episodes had occurred the day before admission. On admission the patient was alert, cooperative and well-oriented. The neurological examination was normal. In particular, no neurophthalmological abnormalities were found on clinical examination. Blood parameters, urine, chest radiograph and ECG proved normal. Cervical radiographs revealed osteoarthritis with osteophytes and narrowing of disc space C6-C7. EEG was normal. Brainstem auditory evoked potentials (BAER) revealed increased latency of V wave on right stimulation. Cerebral CT and MRI (figure) showed an ischaemic-like lesion, 2 cm diameter, in the right cerebellar hemisphere in the territory of the medial branch of the posterior inferior cerebellar artery (PICA), without mass effect. Moderate periventricular white matter abnormalities coexisted. Four vessel cerebral angiography revealed a right vertebral artery stenosis (50%) and two small arteriovenous malformations on the course of the right ascending cervical artery; a decreased flow in the basilar artery

was noted. Ticlopidine 250mg daily was given and the patient was discharged. No further attacks or other neurological disturbances occurred during the next two years.

Case 2, a 52 year old woman, with a 40 year history of bilateral chronic otitis with residual deafness, had recent recurrent episodes of sudden sweating, nausea, occipital headache, dizziness, sometimes followed by a transient loss of consciousness. The whole episode usually lasted about 30-40 minutes. Frequently, at the height of dizziness, the patient experienced a 180 degree vertical visual inversion of images. These episodes occurred monthly. On admission, the neurological examination was normal. Rare, isolated, left-sided jerks of horizontal nystagmus were revealed by ENG. A 20 mmHg difference between right and left brachial arterial pressure (right > left) was noted. Ultrasound vascular investigations (Doppler cortico-vertebral echotomography and cerebral transcranial Doppler) revealed a left subclavian artery stenosis with a steal syndrome. Cerebral SPECT, CT and MRI proved normal. BAER was unavailable due to the peripheral deficit caused by chronic otitis. Cerebral angiography was refused. Flunarizine 10mg daily was given and the patient was discharged with a warning to avoid strenuous physical activities, especially those involving upper limbs and neck. No further episodes were reported in the subsequent six months.

These two women presented episodes of vertically inverted vision—upside down phenomenon—associated with clinical signs and symptoms of vertebrobasilar insufficiency. Both reported transient visual inversion of 180 degrees, which was bilateral, of sudden onset and lacking subjective impression of movement (rotatory or torsional). In the first patient, neuroimaging revealed a right hemispheric cerebellar infarction. In the second, a vertebrobasilar failure due to a left subclavian stenosis was detected. The pathogenetic mechanism underlying upside down visual inversion is unknown. Since the visual images enter the retina inverted, it may be assumed that the upside down phenomenon results from a transient failure of the mechanisms mediating reinversion, even though the anatomical structures involved are unknown. In earlier observations,² parietal and/or occipital lesions were sometimes found, suggesting a cortical origin of the dysfunction, probably affecting the integrative control of spatial vision. More recent cases,^{1,3-5} documented with neuroimaging techniques, revealed an association with vestibular/cerebellar lesions, that is, vertebrobasilar TIAs, Wallenberg's syndrome and also cerebellar infarct in two cases.^{1,5} In our patients, the relationship between the upside down visual inversion and the signs and symptoms of vertebrobasilar insufficiency, without evidence of cerebral damage, supports the idea that a transient inactivation of infratentorial structures may cause this unusual phenomenon. Besides the integrity of the visual system, space visual perception needs a flow of extraretinal information, mediated by the vestibular and cerebellar systems.^{6,7} It has been suggested that damage to such structures may cause tilt and complete inversion of the visual space.⁸ The upside down phenomenon may occur following dysfunctions at various levels of the complex vestibulo-cerebellar-ocular system mediating the stabilisation of the visual function so that

cortical involvement is not indispensable.

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- 1 Ropper AH. Illusion of tilting of the visual environment. Report of five cases. *J Clin Neuroophthalmol* 1983;3:147-51.
- 2 Solms M, Kaplan-Solms K, Saling M, Miller P. Inverted vision after frontal lobe disease. *Cortex* 1988;24:499-509.
- 3 Steiner J, Shahin R, Melamed E. Acute "upside down" reversal of vision in transient vertebrobasilar ischemia. *Neurology* 1987;37:1685-6.
- 4 Mehler MF. Complete visual inversion in vertebrobasilar ischaemic disease. *J Neurol Neurosurg Psychiatry* 1988;51:1236-7.
- 5 Charles N, Froment C, Rode G, et al. Vertigo and upside down vision due to an infarct in the territory of the medial branch of the posterior inferior cerebellar artery caused by dissection of a vertebral artery. *J Neurol Neurosurg Psychiatry* 1992;55:188-189.
- 6 Hornsten G. Constant error of visual egocentric orientation in patients with acute vestibular disorder. *Brain* 1979;102:685-700.
- 7 Gloning K, Hoff H. Cerebral localisation of disorders of higher nervous activity. Clinical, anatomical and physiological aspects. In: PJ Vinken and GW Bruyn, eds, *Handbook of clinical neurology*, Vol 3, Amsterdam: North Holland, 1969, 22-47.

Subcortical environmental reduplication: SPECT findings in a patient with a right thalamocapsular haemorrhage

Recently Nighoghossian *et al*¹ reported the case of a patient with a previous history of a left fronto-basal haemorrhage, who developed environmental reduplication following an infarction of the retrolenticular portion of the right internal capsule. SPECT revealed right fronto-parietal cortical hypoperfusion. A similar delusional misidentification syndrome was described previously in a patient with a right thalamic haemorrhage, but its functional correlate using SPECT was not studied.² We describe the neuroimaging and cognitive functioning of a case of environmental reduplication associated with a right thalamocapsular haemorrhage.

A 71 year old ambidextrous man suddenly developed a left-sided weakness and mild dysarthria. He had had hypertension but no history of previous cerebrovascular events. Neurological examination revealed a dense left hemiplegia, and a left sensory loss affecting all modalities. Visual fields were full, and there was no evidence of visual or auditory extinction on double simultaneous stimulation. He showed left hemispatial neglect on drawing, and on a letter cancellation task he only crossed targets on the right side of the paper. He did not deny his left hemiplegia, but he had a tendency to attribute it to previous "chest problems". He reported a feeling of nonbelonging of his paralysed left arm, and also said that he had three left legs and a strange left arm crossed over his chest. The patient said that he could walk almost normally and repeatedly tried to walk unaided despite his dense hemiplegia. He was alert and oriented to time and person, but not to place. While he

Figure Axial MR T2-weighted image showing a high signal area in the territory of the medial branch of the right PICA.

gave the correct name of the hospital, he located it in another city. Despite being repeatedly informed that the hospital was in Málaga, he denied it and was convinced that it was a new branch in the city he named. Although he remained in the same room during his admission to hospital, he insisted he had been moved daily to identical hospitals (all with the same name) that were located in neighbouring cities (Torremolinos, Fuengirola, Marbella).

One week after onset, the patient had a detailed neuropsychological examination. He was found to have a WAIS verbal IQ of 94 and a performance IQ of 51. On the Wechsler Memory Scale (WMS), his memory quotient was normal (99 points); his scores on immediate history recall and associated learning were average though his performance on the visual reproduction subtest was below average. There was no evidence of confabulation in response to items of the Mercer's confabulation battery, but he did poorly on tests thought sensitive to frontal lobe dysfunction (Wisconsin Card Sorting Test (categories achieved = 0; normal range = 4–6), Trail-making test (part A) (below percentile 10)). He also showed a severe impairment on visual-perceptual tests (Visual Form Discrimination (14 points; normal range 23–32), Facial Recognition (15 points; percentile rank = 1). His language was almost intact, except for a mild visual naming impairment (Boston Naming Test, 38 points (maximum = 60)).

CT and MRI scans showed a right thalamic haemorrhage with extension into the posterior limb of the internal capsule, corona radiata and ventricular system. Mild symmetrical periventricular white-matter changes compatible with leukoaraiosis were also observed. Regional cerebral blood flow was studied with ^{99m}Tc -HMPAO and SPECT, using an Elscint Apex 609 RG gamma camera. Focal blood flows were analysed semiquantitatively in twelve circular regions of interest which were placed over the cortical mantle in three successive slices. Asymmetry indices (AI) for each lobe were calculated using the following formula: $(R - L)/(R + L) \times 200$.³ A marked decrease of perfusion was observed in the right thalamus and basal ganglia as well as in the left cerebellum. Hypoperfusion was also noted in widespread cortical regions of the right hemisphere affecting mainly the frontal lobe (AI = -29.6), and to a lesser extent the parietal (AI = -10.8) and temporal (AI = -10) lobes (negative AI values indicate left sided hyperactivity relative to the right sided activity).

The assessment of neuropsychological functions in our case of subcortical environmental reduplication revealed more pervasive deficits than those observed in the patient reported by Nighoghossian *et al.*,¹ but similar to those of previous cases showing evidence of environmental reduplication and cortical involvement. Moreover, the combination of deficits on nonverbal memory, awareness, visual-perceptual skills and reasoning abilities supports the view that environmental reduplication is a complex, multifactorial delusional misidentification syndrome.^{4,5}

Some previous reports emphasised the association between environmental reduplication and the superimposition of bilateral frontal-lobe and right hemisphere cortical involvement, while others suggested that unilateral (right) lesions of either the fronto-

parietal or parieto-temporal cortices are sufficient to cause it.^{4,5} In our patient, right thalamocapsular damage may have induced functional depression of various distant but anatomically connected cortical areas. Data from SPECT, however, revealed secondary cortical deactivation affecting mainly the right frontal cortex. In this context, we suggest that functional deactivation of the right cortical mantle, in addition to thalamocapsular injury, may underlie environmental reduplication and its associated neuropsychological deficits. In addition, given that environmental reduplication probably requires preexisting brain pathology (for example, cortical atrophy) besides the specific sites of brain damage in the right hemisphere,^{1,2} the presence of leukoaraiosis in the case by Nighoghossian *et al.*¹ and in our own patient might be another risk factor for developing it after acute stroke.

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- 1 Nighoghossian N, Trouillas P, Vighetto A, *et al.* Spatial delirium following a right subcortical infarct with frontal deactivation. *J Neurol Neurosurg Psychiatry* 1992;55:334–5.
- 2 Leiguarda RC. Environmental reduplication associated with a right thalamic haemorrhage. *J Neurol Neurosurg Psychiatry* 1983;46:1154.
- 3 Pozzilli C, Passafiume D, Bernardi S, *et al.* SPECT, MRI and cognitive functions in multiple sclerosis. *J Neurol Neurosurg Psychiatry* 1991;54:110–5.
- 4 Patterson MB, Mack JL. Neuropsychological analysis of a case of reduplicative paramnesia. *J Clin Exper Neuropsychology* 1985;7:111–21.
- 5 Kapur N, Turner A, King C. Reduplicative paramnesia: possible anatomical and neuropsychological mechanisms. *J Neurol Neurosurg Psychiatry* 1988;51:579–81.

Evidence for presynaptic inhibition on trigeminal primary afferent fibres in humans

In a recent study we have shown that a conditioning electrical stimulus applied to the trigeminal afferent fibres of intensity below the reflex threshold (Th) (0.90–0.95 times the perceptive Th) produces unexpected changes of test trigemino-facial reflex responses.¹ In particular, for the R2 response, there is a monotonic depression starting at 20–30 ms of interstimulus interval, reaching a maximal value at 50–100 ms and recovering within 300–400 ms (figure 1). Based on the similarity of the time-courses of the R2 inhibition and of the trigeminal primary afferent depolarisation described in the cat,² we propose that presynaptic inhibition from low-Th trigeminal afferent fibres was the primary factor contributing to the depression of the test response. We describe experimental evidence to support this.

Two complementary experimental

designs, approved by the Local Ethical Committee, have been employed: 1) the time-course of the R2 inhibition was compared, in the same subject, with that of the soleus Ia presynaptic inhibition; 2) the effect of intravenous administration of thyrotrophin releasing hormone (TRH), a substance shown to increase presynaptic inhibition in humans,³ has been tested in parallel on both spinal Ia presynaptic inhibition and R2 reflex inhibition. In addition, spinal Ia presynaptic inhibition and trigeminal R2 inhibition were studied in a patient affected by chronic progressive spino-bulbar spasticity, a rare disease presenting with spastic paralysis of facial, laryngeal and lower limb muscles, due to a progressive and parallel involvement of corticobulbar and cortico-spinal projections.⁴ Many patients with spasticity show a reduced or absent presynaptic inhibition on the soleus Ia primary afferent fibres, probably due to the loss of a tonic supraspinal excitatory drive on interneurons acting presynaptically on Ia terminals. If inhibition of the R2 reflex is of presynaptic origin and it is subjected to such a descending control, then it could be expected that a lesion of the corticobulbar and cortico-spinal projections would also result in a parallel reduction of presynaptic inhibition on both spinal Ia and trigeminal primary afferent fibres.

Figure 1 shows the time-course of presynaptic inhibition of the soleus H reflex in a normal subject. We used a method introduced by Morin, Pierrot-Deseilligny and Hultborn in 1984,⁵ which we briefly describe. The test response was a soleus H reflex evoked by stimulating the posterior tibial nerve. To elicit Ia afferent discharge from pretibial muscles, a short lasting vibration (10 ms; 3 cycles; 300 Hz) was applied over the tibialis anterior tendon by an electromagnetic hammer, 10–400 ms before the test stimulus. Under this experimental condition, evidence shows that the resulting long-lasting depression of the soleus H reflex is due solely to presynaptic inhibition from pretibial Ia afferent fibres to those mediating the afferent test volley.⁶ In fig 1 the time-course of the inhibition of the trigemino-facial R2 reflex following a weak conditioning stimulation (0.95 times the perceptive PTh) of the first trigeminal branch is also reported. There is a clear strict similarity of the curves of the R2 and soleus H reflex inhibition. Identical findings have been observed in two other normal subjects. By exploring the upper curve in fig 1 it is apparent that the long-lasting inhibition of the soleus H reflex is preceded by an early short-lasting facilitation (see also the upper curve in fig 2). This early facilitation is due to mechanical spread of the vibration to soleus spindles causing homonymous Ia facilitation in the soleus motor neurons.⁶ This explains why this early facilitation is lacking on the R2 reflex curve (lower curve in fig 1).

The histograms on the right in fig 1 show the enhancement of the R2 and H reflex depressions after acute intravenous administration of TRH. It has been demonstrated that a single subcutaneous injection of high-dose TRH (1.2–2.5 mg/Kg) produces dramatic and long-lasting (1–2 hours) increase of the soleus presynaptic inhibition in patients with amyotrophic lateral sclerosis.³ We have observed a similar but short-lasting (10–20 minute) enhancement of the soleus Ia presynaptic inhibition in normal