Brain MRI showing clearly defined hyperintense signal alteration in globus pallidus bilaterally (arrows). which led to her admission to our department two months after the acute event. As well as these anterograde memory problems and a retrograde amnesia for about a week she showed normal behaviour and the neuropsychological examination disclosed normal results. In neuropsychological testing she showed an IQ of 117 (WAIS) that was supposed to be somewhat lower than her pre-morbid level, although it was in the normal range. Results of tests of language function and semantic memory (token test: 0 errors, Boston naming test: 82 out of 85), visuospatial abilities (copy of the Rey figure: 36/36), and “frontal” functions (phonetic fluency: 19 words beginning with “s” in two minutes, semantic fluency: 26 animals in two minutes) were all within the normal range. Memory function as assessed by a German version of the Warrington recognition memory test yielded results below the 5th percentile for words (37/50) and faces (36/50). Recall of the Rey figure (7/5/36) and the results of a list learning task (10/15 after five learning trials) were also highly impaired. The patient therefore had a rather pure non-material specific disorder of episodic memory. After nine months formal testing showed only a slight improvement in her memory performance. She is still not able to return to night school or her part time job, but she has learned to make extensive use of a diary and a timetable. She had occupational therapy in an outpatient clinic for several months.

Brain MRI (figure) performed three weeks after onset showed bilateral hyperintense lesions in the globus pallidus. Additional involvement of basal temporal structures was suspected, but could not be shown clearly because of movement artefacts. In a second MRI examination about two months later these hyperintense lesions in the globus pallidus had partly disappeared and the basal temporal structures were normal.

The symmetric lesions in the globus pal- lidus resemble the findings of Squier and colleagues in their neuropathological study, who pointed out that the pallidum is rich in serotonin releasing neurons. They suspected that a local release of serotonin might have led to prolonged vasospasm and necrosis. The hippocampi, essential for episodic memory function, are also rich in serotonin releasing neurons and are known to be targets of MDMA intoxication in animals. Therefore we propose that in the present case MDMA ingestion led to alterations in the globus pallidus (seen in MRI but clinically silent) and in the hippocampi (causing persistent memory problems). In post-mortem studies of patients who died after MDMA ingestion, other mechanisms of damage were suspected. In the present case there was no hypothermia or a water intoxication could be ruled out by normal serum sodium concentration. The sudden onset with a suspected generalised seizure could point to cerebral anoxia due to asphyxia as another possible reason for the patient’s memory disorder, but the fit was observed and no apnoea or even cyanosis was reported.

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Rotated drawing: a mini mental state examination performance with strong lateralising significance

Neuropsychological evidence clearly has a strong bearing on our understanding of the process of object recognition. Of particular relevance are recent reports of a few patients who have drawn dramatically misorientated, although otherwise quite accurately reproduced, drawings when attempting to copy a line drawing. The copies are almost invariably rotated relative to the model by 90°, or in one case by either 90° or 180°. It has been difficult to draw conclusions about the localising significance of this unusual neuropsychological sign. However, it is notable that, with the exception of one report of two instances of transient global amnesia, all of the cases have involved right-sided1-4 or in one case by either 90° or 180°. However, the lesions location within the right hemisphere has varied greatly, often exclusively involving the frontal lobes7 by contrast with the present study. These data suggest that rotated drawing might well have lateralising relevance as a clinical sign—although its localising relevance within one hemisphere is less clear.

One of the most interesting clinical findings in the present study is the co-occurrence of rotated drawing and left visuospatial neglect. None of the previous case reports of rotated drawing have involved left neglect, nor have the actual reproductions of patient performance depicted in these papers shown the classic signs of left neglect.6,7 However, it is possible that the co-occurrence of rotated drawing and neglect in the present study...
study is a result of a sampling bias. The patients in this study represent a series of admissions to a rehabilitation unit—where the referral of patients with right-hemispheric lesions is most common when it involves the obvious and remediable signs of left hemiparesis and hemispatial neglect. It is likely that those such as the patient of Solms et al., whose bilateral frontal pathology was largely asymmetrical in the standard neurological and neuropsychological examination, would never have been admitted to the unit.

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A novel point mutation in the GTP cyclohydrolase I gene in a Spanish family with hereditary progressive and dopa responsive dystonia

The GTP cyclohydrolase I (GTP-CH I) gene is the causative gene of hereditary progressive and dopa responsive dystonia (HPD/DRD) in both Japanese and non-Japanese patients. We report a novel missense mutation in the GTP-CH I gene in a Spanish family with HPD/DRD.

We studied eight members of a Spanish family with HPD/DRD (figure, A), and 30 unrelated controls. The four affected members met the clinical criteria for HPD/DRD.1 Fragments of DNA containing the entire coding region of the GTP-CH I gene were obtained from genomic DNA by polymerase chain reaction (PCR) according to the method of Ichinose.1 Direct nucleotide sequencing of PCR products was performed with an automated DNA sequencer (ALFexpress, Pharmacia Biotech) using the same primers as for amplification.

We found a single base pair change at position 25 of exon 4 in the GTP-CH I gene, which consists of a C → A substitution (figure, B), leading to an amino acid change (Arg178Ser). As the C → A mutation abolishes a restriction site for Mbo II in exon 4, this exon was amplified in all subjects, and then digested by Mbo II, to show linkage between this novel mutation and the four affected members of this HPD/DRD family. The restriction fragment length polymorphism (RFLP) generated by Mbo II consisted of two fragments in healthy subjects (113 and 187 bp) and one additional fragment in patients (300 bp) (figure, A). The restriction pattern in all four affected members was consistent with the heterozygous status. No asymptomatic members of the HPD/DRD family or unrelated normal controls showed such a restriction pattern.

This is the first reported mutation in the GTP-CH I gene in a Spanish family with HPD/DRD. It supports the idea that the GTP-CH I gene is the causative gene of HPD/DRD worldwide. However, it is necessary to take into account that in some 14q-linked cases of HPD/DRD no mutations have been identified yet.1,2 Mutations in some regulatory regions of the GTP-CH I gene may explain these negative findings.

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