in his drawing skills. Handedness was assessed by informal questioning about hand, foot and eye preference for various activities, direct observation and the results of the Handedness Inventory. All measures used indicated strong right preference with no evidence of left hand bias. There was no family history of left-handedness. Left hemisphere language representation was indicated. His IQ was estimated to be above average. Pure tone audiometry was not performed. Bedside testing of hearing was normal. The CT scan, performed 3 months after onset, showed an infarct which was predominantly in the right parietal region, with some extension inferiorly into the posterior temporal lobe (fig). There was no evidence of any abnormality in the left hemisphere.

We postulate that our patient’s word deafness was due to the right temporoparietal lesion seen on CT. Patients with tumours of, or injury to, the right hemisphere may develop “crossed” aphasias, though such lesions may cause aphasias by pressure or distortion of the left hemisphere. Lesions in the non-dominant temporal lobe have been reported to produce an agnosia for non-verbal sounds with preserved comprehension of the spoken word and impaired appreciation of music. Spreen described a man with an agnosia for non-verbal sounds, but intact language comprehension. He had had a stroke causing a marked left hemiparesis and subsequently developed emotional liability, seizures and bilateral pyramidal signs. Handedness was not tested. At necropsy, the patient had bilateral subdural haematomas and a very extensive area of infarction in the right, presumably non-dominant, hemisphere in the fronto-temporo-parietal areas. In contrast, our patient had only a single clinical episode, a more restricted neuropsychological deficit and a smaller lesion. Nonetheless, although the relationship of the lesion to the deficit appears much clearer, other possibilities must be considered: it is possible that there was infarction of the contralateral hemisphere, not visible on the CT scan; it is also possible that the patient was right hemisphere dominant for language (or had bilateral language representation), despite his strong right-handed preference. However, since all these alternative explanations seem unlikely, we conclude that the right hemisphere infarct was the cause of our patient’s pure word deafness.

MARK ROBERTS
PETER SANDERCOCK
Fazakerly Hospital, Liverpool

ERIC GHADIALI
Walton Hospital, Rice Lane,
Liverpool L9 1AE, UK

References
7 Shankweiler DP. Performance of brain-damaged patients on two tests of sound localisation J Comp Physiol Psychol 1961;54:375.

Accepted 26 June 1987

Photosensitivity related to valproate withdrawal

SIR: Photosensitive epilepsy comprises patients in whom seizures are provoked by environmental flicker stimulation. Gener-

ised epileptic discharges are evoked by intermittent photic stimulation, constituting a photoconvulsive response. Sodium valproate is the drug of choice for photosensitive epilepsy. Three patients are reported who developed photosensitivity related to sodium valproate withdrawal.

The first case was a 47 year old man who had had four generalised tonic-clonic convulsions, nocturnal or on awakening, from 18 years to 24 years of age. He was seizure-free until 36 years of age, of phenobarbitone therapy since aged 29. Sodium valproate treatment (15 mg/kg/die, plasma level 73.3- µg/ml) was started at 36 years of age because of a relapse with a morning generalised convolution. EEG initially showed generalised spike and wave discharges at rest and during hyperventilation and then normalised. Intermittent photic stimulation always had no effect. There was gradual sodium valproate reduction at 45 years of age. Six months later, on 5 mg/kg/die of sodium valproate (plasma level 18.5 - µg/ml) for 3 weeks, he suffered from two tonic-clonic convulsions when watching television. EEG showed generalised epileptic discharges, evoked by intermittent photic stimulation. Sodium valproate dosage was increased to 10 mg/kg/die (plasma level 44.0 - µg/ml). He was seizure-free for the following year and photoconvulsive response disappeared.

A 32 year old man had two morning generalised convulsive seizures at 20 and 22 years of age. EEG showed generalised discharges during hyperventilation but no paroxysmal response to intermittent photic stimulation (fig). Sodium valproate therapy (13 mg/kg/die, plasma level 62.7 - µg/ml), started after the second fit, controlled the seizures and normalised the EEG. Sodium valproate was gradually stopped at 30 years of age. Three weeks after drug withdrawal he had a tonic-clonic convolution while using a video-computer. EEG showed generalised epileptic discharges exclusively during intermittent photic stimulation (fig). On phenobarbitone treatment (1-5 mg/kg/die) there was no seizure recurrence and no abnormal response to intermittent photic stimulation for the following 2 years.

The third case was a 16 year old girl who otherwise neurologically normal had two nocturnal clonic seizures involving the right half of the face at 8 and 9 years. CT of the head was normal. EEG showed left centrotemporal sharp waves but no photocconvulsive response. EEGs of the parents and the older sister were normal. Sodium valproate therapy (20 mg/kg/die), started following the first seizure, was gradually
withdrawn at 13 years of age. Photic driving was occasionally seen during drug reduction. One month following sodium valproate withdrawal a generalised tonic-clonic convolution occurred while she was playing a video game at home. EEG showed persistence of the left centro-temporal discharges and generalised spike and wave discharges evoked by intermittent photic stimulation. No further seizure or photovisual response occurred afterwards, while off antiepileptic treatment.

That photosensitivity was temporarily linked to sodium valproate withdrawal is beyond doubt. All patients had generalised convulsions under environmental flicker stimulation and photoconvulsive response a few weeks after sodium valproate reduction or withdrawal. Furthermore, photosensitivity rapidly disappeared off antiepileptic drugs in the third patient and on phenobarbitone in the second patient. A causal relationship is also highly probable in the first two cases. Both patients showed photosensitivity at an age in which it does not usually develop. Also, phenobarbitone withdrawal was not related to photosensitivity in the first case, at an age when photosensitivity may first appear. In spite of age and type of epileptic syndrome, causal relationship between sodium valproate withdrawal and photosensitivity occurrence may be sustained even in the third case. In fact, tonic-clonic convulsions have been reported as a late relapse in benign partial epilepsy with rolandic spikes, not related to drug suspension; generalised spike and wave discharges are quite common on the EEG of children, but they are rarely evoked by intermittent photic stimulation. Photosensitivity characteristically appears around puberty but persists for many years. Thus, it is unlikely that sodium valproate withdrawal triggered underlying previously developed photosensitivity. Finally, a family history of photosensitive epilepsy or of a photoconvulsive response without seizures, usually common in photosensitive epilepsy, is lacking in our patient.

The mechanism of occurrence of photosensitivity following sodium valproate withdrawal is unknown. Photosensitivity is usually rapidly controlled by sodium valproate. A rebound effect related to drug withdrawal may be supposed. Further cases are needed to establish whether this transient photosensitivity is really a consequence of drug withdrawal. In any case, epileptic patients should avoid exposure to environmental flickering light during sodium valproate reduction and shortly after withdrawal.

GIOVANNI AMBROSETTO
CARLO ALBERTO TASSINARI
Istituto di Clinica Neurologica,
Via Ugo Foscolo 7,
40123 Bologna, Italy.

References

Accepted 26 June 1987

Adult metachromatic leucodystrophy: an underdiagnosed disease?

Sir: Metachromatic leucodystrophy is an inherited autosomal recessive disease due to deficient activity of cerebroside sulphatase and is characterised by demyelination and accumulation of galactosyl-sulphatide in nervous and other tissues. It can begin at any time from infancy to late adult age, this probably depending on residual enzymatic activity. Usually two fully distinct forms of the disease are recognised: lat infantile and adult (juvenile cases overlap both, in clinical course, age at onset and duration of the disease).

The adult form is rare and peculiar in its psychiatric presentation, lack of family history and absence of neurological signs. Curiously it was the first to be recognised as such, but until a few years ago was rarely considered and diagnosed. Some authors
Photosensitivity related to valproate withdrawal.

G Ambrosetto and C A Tassinari

*J Neurol Neurosurg Psychiatry* 1987 50: 1709-1710
doi: 10.1136/jnnp.50.12.1709

Updated information and services can be found at:
http://jnnp.bmj.com/content/50/12/1709.citation

**Email alerting service**

These include:

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/