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CORRESPONDENCE

Neurovisual rehabilitation in Balint's syndrome

Further to the excellent review of neurovisual rehabilitation by Kerkhoff¹, we think that it is prudent to communicate our experience in the management of a patient with Balint's syndrome after traumatic brain injury. This was seen in a 41 year old right handed manual worker whose initial cranial CT showed right extradural haematoma. Subsequent scans demonstrated left posterior occipital infarct. Brain MRI 3 months after the injury showed high signal in the right occipitoparietal and left occipitotemporal regions. His physical recovery was satisfactory in that he was fully mobile unaided. However, he presented with simultanagnosia, optic ataxia, and psychic paralysis of gaze.² This had an adverse impact on his functional independence; he had difficulty in route finding—crashing into furniture and walls—and other activities of daily living including dressing and toileting. He failed most subtests in the Rivermead perceptual assessment battery (RPAB).

We agree with the author that effective treatment strategies are poorly developed and evaluated. We have identified three approaches for the rehabilitation of the perceptual deficits including those seen in Balint's syndrome.

- The adaptive (functional) approach,³ which involves functional tasks utilising the person's strengths and abilities, helping them to compensate for problems or altering the environment to lessen their disabilities
- The remedial approach,⁴ which involves restoration of the damaged CNS by training in the perceptual skills, which may be generalised across all activities of daily living. This could be achieved by tabletop activities or sensorimotor exercises
- The multicontext approach,⁵ which is based on the fact that learning is not automatically transferred from one situation to another. This involves practising of a targeted strategy in a multiple environment with varied tasks and movement demands, and it incorporates self awareness tasks.

In this patient, we used the adaptive approach, practising functional tasks repeatedly with increasing complexity of the tasks as the sessions continued. This approach assumes that treatment has little effect on impairment and that generalisation to other tasks is unlikely. It also assumes that the brain has limited ability to improve and restore itself after injury.³ The remaining abilities are used to offset the deficits. This patient was able to develop his own compensatory strategies, learned to use his hands to acquire tactile feedback, and managed to direct his

gaze to visually locate objects when required. His performance on the RPAB was improved and he was successfully discharged home with little support.

As Balint's syndrome is likely to be seen in clinical conditions such as Alzheimer's disease, multiple strokes, intracranial tumours, brain injury, and CNS complications of HIV infection, we thought it important to outline the possible options for the management of this condition. Further work is required on a larger series of patients.

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Kerkhoff replies:

Al-Khawaja and Haboubi have reported successful neurovisual rehabilitation in a patient with Balint's syndrome due to a right occipitoparietal and a left occipitotemporal lesion, using adaptive practising of functional tasks with increasing complexity. This case shows, together with some of the other published cases,¹ that individually tailored rehabilitation strategies can be adapted successfully for some severely disabled patients. Due to the various aetiologies and subsequent lesion localisations it is likely that most patients with Balint's syndrome will have some outstanding deficits, but also some spared abilities, which can be used for compensatory purposes (for example, intact tactile feedback¹).

In two patients with Balint's syndrome treated for several months in our department, significant improvements could be achieved by systematic treatment so that both patients could live independently at home with only minimal assistance. One patient, who had received traumatic, uraemic, and hypoxic brain damage at the age of 27 years initially was nearly blind. Two years later, when treatment started in our unit, he couldn't perceive more than two visual stimuli simultaneously (simultanagnosia), was almost unable to read, showed optic ataxia, and had severely impaired spatial-perceptual functions in the visual and tactile modality. However, his memory, intelligence, and executive functions were largely preserved, so that he relearned reading partially, learnt to dress himself partially, and was finally able to travel by train. He managed to live alone in his flat, with only minor assistance from others.

The second patient, a 60 year old physician, had bilateral vascular parieto-occipital lesions. She was initially (falsely) considered as blind, although she could well see and describe faces and correctly identify the colour of one's eyes. She presented with severely disturbed depth and horizontal distance perception, simultanagnosia, and optic ataxia as well as a peculiar deficit in identifying spatial directions and locating sound sources. For instance, it proved difficult for her to identify the direction in which someone pointed when describing a particular route, or to decide in which direction a train would move when looking at the railway track. However, as in the first patient, she had some spared abilities—that is, excellent introspection and awareness of her disorder, preserved cognitive abilities, and she was highly motivated to relearn route finding in her town district. After intensive training for reading and route finding she could be discharged, living independently at home. She continues to use public transport to go shopping, visit friends, see her neurologist, the pharmacist, or going to a concert hall.

I think that systematic treatment in both cases helped to improve basic visual abilities and activities of daily living so that both patients could live independently at home—which was hardly expected when seeing them at the onset of treatment. To conclude, I am convinced that many patients with Balint's syndrome can learn to compensate for at least some of their visual deficits by systematic and individualised treatment. The search for spared functions will undoubtedly disclose multiple ways for compensation and will increase our understanding of some unresolved aspects of this fascinating syndrome—for example, the tactile or auditory-spatial abilities of patients with Balint's syndrome.

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Arnold Chiari malformation and nystagmus of skew

I enjoyed Pieh and Gottlob's article¹ pointing out the association of a Chiari malformation with a "unique" form of nystagmus that they call "the nystagmus of skew." The distinctive feature of this nystagmus is a disjunctive vertical oscillation in which the fast phase of one eye moves upward while, at the same time, the other eye moves downward.

The authors¹ state that their second patient had a "rotatory component" by which I assume they mean torsional; this pattern of nystagmus is already established in the literature and is known as "jerk-waveform see-saw nystagmus".^{2,3} In their first patient they point out that the amplitude of the vertical nystagmus was so small that they were unable to confidently exclude a torsional component. It would have been most interesting to obtain recordings looking for a torsional component using the modified

scleral search coil technique⁴; I suspect that it would have shown a torsional component and that this patient also had jerk-waveform see-saw nystagmus.

Jerk-waveform see-saw nystagmus occurs with unilateral mesodiencephalic lesions, presumed due to selective unilateral inactivation of the torsional eye velocity integrator in the interstitial nucleus of Cajal⁵; during the fast (jerk) phases the upper poles of both eyes rotate toward the side of the lesion. With lateral medullary injury the fast phases of the torsional component jerk away from the side of the lesion.³ In both situations the torsional component is always conjugate. With mesodiencephalic lesions the vertical component is always disjunctive, but with medullary lesions it may be either conjugate (usually upward) or disjunctive.²

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The authors reply:

We thank Lavin for his interesting comments.

We stated in our article that the possibility of a fine see-saw nystagmus could not be excluded. We did re-evaluate our patients with a torsional coil and did not record a torsional component. However, because of the fast improvement in both patients, all the eye movement abnormalities on re-evaluation were minimal. Clinically, even in the stage of maximal abnormalities, in one patient we did not detect any torsional component, which suggests that if there was an element of see-saw nystagmus, it was subclinical.

We did not state that the type of nystagmus associated with the Arnold-Chiari malformation was unique, precisely because we could not rule out with total certainty a see-saw nystagmus, which has been reported in one patient with the malformation. We did, however, point out that this association is unusual.

Because of the lack of strong evidence of a torsional component to the dissociated vertical nystagmus, we preferred the term, kindly suggested by a reviewer, "nystagmus of skew". This would represent a more inclusive, descriptive term, of which both the pendular and the jerk see-saw nystagmus forms and the dissociated vertical nystagmus without demonstrable torsional component would represent subvariants.

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Figure 1 Scintigraphy of the head of patient 1 showing technetium 99m uptake one day before (left) and 7 days after (right) retrograde transductal BoNT/A injection into the salivary glands, showing significant reduction of tracer uptake after the BoNT/A injection (quantification: parotid gland left -59%, right -48%; sublingual gland left -10%; right -56%).

Botulinum toxin for the treatment of sialorrhoea in ALS: serious side effects of a transductal approach

We have read with interest the article by Giess *et al.*¹ which showed that botulinum toxin A (BoNT/A) might be a new treatment option for sialorrhoea in patients with bulbar palsy. We have recently conducted a similar study which was interrupted due to serious side effects.

In September 1998 we injected 25 MU Botox into the parotid glands of a 59 year old woman who had ALS with pronounced bulbar palsy. We noticed a reduction of the sialorrhoea but facial weakness on the left side worsened significantly.

After this experience we developed a protocol for the treatment of sialorrhoea in patients with ALS with bulbar palsy by retrograde injection of BoNT/A through the salivary duct into the salivary glands. We chose the retrograde way of administration of BoNT/A for this pilot study because we thought that this technique would avoid facial weakness.

After informed consent the patients received 12.5 mouse units (MU) BoNT/A (Botox[®]) retrogradely into each parotid and sublingual gland from a small catheter inserted into the salivary duct. Neurological examination and quantification of saliva production were performed before the BoNT/A injection and on days 1, 3, 7, 14, and 28, as well as after 2 and 3 months. Technetium 99m scintigraphy was performed before and 7 days after the injection. Quantification of saliva production was performed with a simple method: the patients were asked to expectorate as much saliva as possible into a paper handkerchief for 10 minutes. This procedure was repeated twice. The mean of the difference in weight of the handkerchief before and after these procedures was taken as the maximum expectorated saliva production (MESP). Quality of life and the clinical effect of the treatment were evaluated by a questionnaire.

We treated a 60 year old woman (patient 1) and a 69 year old woman (patient 2) with this new technique. Both had certain ALS according to the El Escorial criteria, with severe bulbar palsy with durations of 23 and 28 months respectively. Both patients had a significant reduction of MESP seven days

after the injection (76% and 58%; from 5420 mg to 1301 mg, and 4365 to 1829 mg) which lasted for 4 to 8 weeks. Technetium 99m scintigraphy showed a significant reduction of radiotracer uptake into the injected salivary glands in both patients (figure). Both patients estimated the injection procedure as painful. Patient 1 developed a severe swelling of the right sublingual salivary gland and base of the tongue 3 days after the injection which was treated with antibiotics and corticosteroids. Patient 2, who was able to swallow with difficulty before the injection, mentioned impairment of swallowing between days 4 and 21. Both patients had a "moderate" improvement of sialorrhoea but did not want the injections to be repeated. After these experiences we decided to stop the pilot study.

The injection of BoNT/A through the salivary duct reduces the activity of the salivary glands significantly for several weeks but has serious side effects. Local and systemic effects of BoNT/A are probably pronounced in ALS.² Subclinical EMG abnormalities distant to the injection sites have been described in therapeutic doses, but also systemic weakness has been found.³ As there are some reports that BoNT/A injections, even in low doses, may exaggerate pre-existing neuromuscular diseases,⁴ careful monitoring of neurological symptoms, which is difficult in a progressive disease, is needed to exclude side effects of BoNT/A. The drug is effective in reducing drooling⁵ but we need more data about the safety of BoNT/A before it can be used safely for the treatment of sialorrhoea in ALS. The transductal approach in particular seems to have unacceptable side effects.

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