

Visual restoration therapy

Disappointing results from Nova Vision's visual restoration therapy

J C Horton

We should refocus our search for new treatments in a more fruitful direction

Lesions posterior to the optic chiasm produce homonymous visual field loss—overlapping scotomas in the nasal field of one eye and the temporal field of the other eye. Patients retain normal acuity, but find their lives changed forever. A homonymous hemianopia, when complete, makes safe driving impossible and reading a chore. Although some patients experience partial, spontaneous improvement following the acute phase of an event, most remain handicapped by permanent field loss. No treatment was available before the recent advent of visual restoration therapy.

In a sensational series of reports, Sabel and colleagues (Kasten *et al*) have described partial recovery of homonymous visual field defects by intensive computer based rehabilitation therapy.¹⁻³ Their approach is remarkably simple. Patients practise perimetry at home for an hour a day, 6 days a week, for 6 months, using a software program loaded on their personal computer. A chin support is used for head stability and a monitor is placed 30 cm away. Stimuli are white, suprathreshold lights measuring 0.15° in diameter shown against a dark background. Protocols are tailored for each patient to present most stimuli near the border of the field defect (“transition” zone) to maximise potential therapeutic benefit. Sabel has founded a company (NovaVision) that offers visual rehabilitation therapy for about €5000.

The idea behind visual restoration therapy is that after stroke or traumatic brain injury, a region of salvageable vision exists between areas of the visual field served by normal and damaged brain tissue. Visual stimulation in this zone with more than 1000 trials a day is postulated to resuscitate its functional potential. After treatment, homonymous field defects have been reported to show a mean azimuth reduction of 4.9° (nine patients).¹ Individual patients have shown up to 30° of field recovery. These are dramatic results for patients suffering from post-chiasmal visual field loss. A wildly optimistic commentary

accompanying the findings in a scientific journal carried the title, “Those that were blind can now see.”⁴

A major problem with the data reported by Sabel *et al* was that the same software program used for visual restoration therapy was also used to show improvement in the visual fields. Obviously, the data would be more compelling if visual field improvement could be demonstrated with any standard clinical perimeter. When patients with post-chiasmal lesions were tested before and after visual restoration therapy with the Tübinger automatic perimeter, no benefit of treatment could be detected.¹

To resolve doubts regarding the efficacy of visual restoration therapy, Sabel teamed up with scientists in Tübingen, a centre renowned for leadership in the field of perimetry. In the resulting study, published in this issue of *BJO* (p 30), 17 patients with stable homonymous field defects were treated according to the visual restoration therapy protocol. Independent visual field testing was done before and after treatment at Tübingen to assess the outcome. A crucial innovation was that perimetry was performed using a scanning laser ophthalmoscope, which allows the examiner to control fixation assiduously by simultaneous visualisation of the retina, fixation cross, and stimulus. Under such conditions, invalid trials as a result of inadequate fixation (for example, saccades) can be disregarded. Unfortunately, the study found no significant improvement in visual field defects, although most patients had the subjective impression that they had benefited from visual restoration therapy. This discrepancy underscores a limitation of outcome satisfaction surveys: patients can be swayed by placebo effects.

Regrettably, it still remains true that no therapeutic intervention, prosthesis, or prism can correct effectively the underlying visual field deficit

How can one reconcile Sabel's original findings with this latest study? Patients

with homonymous field defects compensate by making frequent saccades towards their scotoma in an effort to maintain surveillance of blind regions in their visual fields.^{5,6} It is notoriously difficult to control fixation in such subjects. During visual restoration therapy, fixation is monitored by randomly changing the colour of a 0.75° fixation light from bright green to yellow, whereupon the subject is required to respond within 500 ms by pressing a button. The problem with this technique is that the colour transition is so easy to detect that it does not require foveal vision. Patients soon learn that they can sneak 5° saccades into their blind hemifield, and still detect a change in the colour of the fixation monitoring light. Hence, the mean 5° improvement in the visual field defect.

Several aspects of the original report describing visual rehabilitation therapy should have raised doubts earlier. Firstly, no information was provided regarding false negative, false positive, and fixation loss rates for patients. Perimetric data purporting to show improvement in visual fields are difficult to interpret without these indices. Secondly, the proposed mechanism for partial field recovery in patients with complete hemianopia was flawed. In such subjects the normal occipital lobe and the affected occipital lobe are physically separate—no fringe of injured but salvageable tissue exists that represents the border of the visual field defect. In this situation, why should visual rehabilitation therapy produce field recovery along the vertical meridian? Thirdly, visual rehabilitation therapy was reported to be effective for both monocular optic nerve diseases and homonymous, post-chiasmal lesions. It is difficult to conceive of a physiological mechanism that could explain improvement from the same treatments at different levels of the visual system. It is so unlikely, in fact, that an artefact such as poor fixation control should have been suspected immediately. Fourthly, why should an artificial stimulus applied for an hour a day be more effective than the incredibly rich repertoire of natural light patterns that stimulate the retina under normal, everyday circumstances?

If physical therapy is helpful in patients who are partially paralysed by a stroke, why shouldn't visual rehabilitation therapy work too? The difference, of course, is that the former involves motor systems that can be retrained to compensate for deficits. Through therapy, patients learn how to use new motor strategies with still functional muscle groups to accomplish a physical act. In contrast, lesions of the retino-geniculocortical pathway produce a purely

sensory deficit. No credible evidence exists to suggest that the adult visual cortex can be revived after injury by training exercises or visual therapy. Patients with homonymous hemianopia can benefit from counselling to assist with safe travel, obstacle avoidance, and career planning. Regrettably, it still remains true that no therapeutic intervention, prosthesis, or prism can correct effectively the underlying visual field deficit.

This is not the first time that hopes for visual field recovery by rehabilitation training have been raised and dashed. Twenty years ago, Zihl and von Cramon reported improvement in field deficits in patients with post-geniculate damage by visual training.⁷ The findings were later shown to be an artefact of poor fixation control.⁸ Sabel is due great credit for

submitting visual rehabilitation therapy to independent scrutiny. What distinguishes medicine from "alternative therapies" is that it strives to be evidence based. Here, a proposed therapy has been retested scientifically and found to be ineffective. This information allows us to refocus our search for new treatments in a more fruitful direction.

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Correspondence to: J C Horton, UCSF Department of Ophthalmology 10 Kirkham Street, K301 San Francisco, CA 94143, USA; horton@itsa.ucsf.edu

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Editorial

A work out for hemianopia

G T Plant

A rigorous study finds no evidence of improvement in homonymous visual field defects with training

At the recent meeting of the International Neuro-ophthalmology Society in Geneva one of the most interesting presentations was given by two collaborators who disagreed on the interpretation of their joint findings. The atmosphere was more than usually stimulating. The disputed results are published in this issue of *BJO* (p 30).

It was Josef Zihl from Munich who in 1979, with von Cramon, put forward evidence that it might be possible with practice to extend areas of residual vision in cases with homonymous visual field defects secondary to occipital damage. I have always been sceptical of these findings.

In recent years the work of Erich Kasten and Bernhard Sabel and others in Magdeburg has raised interest again in the prospect of using training methods to bring about a reduction in the extent or density of visual field defects in such patients. The method is referred to as visual restitution training (VRT). However, it is well established that patients with homonymous hemianopia develop eye movement strategies that are adaptive and can potentially improve performance on conventional perimetric

tasks unless eye movements are rigidly controlled (see for example Pambakian *et al*¹). Patients will make an involuntary exploratory saccade into the blind field more frequently than into the sighted field. On the next saccade the fovea is returned to the fixation target. Methods of monitoring fixation, which rely upon the patient reporting a change at the fixation target, may not be fully sensitive to such eye movements and furthermore the authors here suggest that the fixation target used in the previous studies from Kasten's group may have been detectable eccentrically away from the fovea.

This is a very different situation from the type of eye movement artefacts that are controlled for in conventional perimetry, where the patient's eye may wander for seconds of time. Rather, we are dealing here with eye movements the duration of which are not much more than two saccadic latencies. These eye movements will also defeat the strategy of presenting targets for less than the latency of saccades. That is fine to prevent patients from shifting gaze towards a target detected in the periphery but if the patient is making frequent

exploratory saccades throughout the testing period some targets will be detected.

In the present study the authors have used the "gold standard" for controlled perimetry using the scanning laser ophthalmoscope to monitor the fundus. The conclusion is incontrovertible that using these detection targets there is no expansion of the seeing field as a result of VRT. It remains possible that improvement may have been in the nature of relative defects which would not have been detected by the method employed in this study to detect absolute defects.

This is not to say that there is nothing to be gained from attempts to rehabilitate patients with homonymous hemianopia by encouraging the development of eye and head movement strategies as Zihl himself later reported. These may be tailored for specific tasks—for example, navigation (Christopher Kennard's group at the Charing Cross Hospital in London) and reading (Richard Wise and Alex Leff at the Royal Free Hospital in London). These strategies do not claim to improve the bare perimetric results but may enable the patient to make better use of his or her residual vision.

Collaborations of this type require a degree of courage and trust and we should suitably applaud these researchers.

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Correspondence to: G T Plant, The National Hospital for Neurology and Neurosurgery, Queen Square, London WC1N 3BG, UK; gordon@plant.globalnet.co.uk

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Optic nerve

Optic nerve grey crescent

J B Jonas

Another feature in the morphological assessment of the optic nerve head

The optic nerve head has been defined as all areas inside the peripapillary scleral ring. Outside this ring in the parapapillary region, various features and abnormalities can be differentiated. In almost all eyes, the retinal pigment epithelium shows some histological irregularities close to the tip of Bruch's membrane at the border of the optic disc. It is the histological equivalent of the alpha zone of parapapillary atrophy which is present in almost all normal eyes. It can usually better be detected at the temporal disc margin than in other parts of the parapapillary region. The beta zone of parapapillary atrophy, present in about 25% of normal eyes, and in a higher percentage of glaucomatous eyes, reflects a complete loss of retinal pigment epithelium cells and an almost complete loss of retinal photoreceptors. Other abnormalities or changes in the parapapillary region include a hypertrophy of the retinal pigment epithelium mainly at the temporal optic disc border in eyes with a so called conus pigmentosus of the optic nerve head; proliferations of the retinal pigment epithelium such as in retinochoroidal toxoplasmotic scars or after subretinal parapapillary haemorrhages; and melanocytic lesions of the choroid such as choroidal naevi or a malignant choroidal melanoma.¹

The grey crescent of the optic disc as originally described by Shields² is another, mostly unrecognised, feature at the border of the optic nerve head. According to Shields, it is a slate grey crescent within the peripheral tissue of the optic nerve head. In his study, 12 out of 100 consecutive black patients revealed the grey crescent. The grey crescents were usually bilateral and were most often located along the temporal or inferotemporal disc margin. The clinical importance of the grey

crescent is that one may erroneously assume that the underlying tissue is not neuroretinal rim but parapapillary tissue. It will lead to a falsely small optic disc and neuroretinal rim area and, consequently, to falsely high measurements of the cup/disc diameter ratios. Additionally, it will markedly influence the assessment of the shape of the neuroretinal rim which normally follows the so called ISNT rule. The latter says that the smallest part of the rim is located in the temporal horizontal disc region, and that usually the widest part of the rim is located close to the inferior optic disc pole.¹

In their large population based study on the occurrence of the optic disc grey crescent in Iceland, published in this issue of *BJO* (p 36), Jonsson and colleagues found that the grey crescent was present in about 22% of the eyes examined. It was more commonly found in women, in hyperopic eyes, and in eyes without a small parapapillary atrophy. It was associated with a large optic disc, and it was usually located in the temporal region of the optic disc. The occurrence of the grey crescent was statistically unrelated to the prevalence of glaucoma. The authors have to be congratulated for their study and for renewing the interest in the grey crescent and for highlighting its importance for the morphological diagnosis of optic nerve abnormalities and diseases. One may, however, take into account the definition of the grey crescent as used by the authors. They defined the grey crescent as the "occurrence of a pigmented crescent that appeared, utilising a stereo viewer, to be located on or within the neuroretinal rim tissue—that is, inside the scleral lip of the disc whereby the scleral lip had to be clearly visible peripheral to the crescent." Since the alpha zone of parapapillary atrophy

is also characterised by an irregular pigmentation, and because the boundary between the alpha zone and the surrounding tissue usually follows a semilunar line, partially parallel to the peripapillary scleral ring (scleral lip), one must be aware not to confound the grey crescent with the alpha zone. The difference between both structures is that the alpha zone is located outside of the optic disc and may not be counted as neuroretinal rim, whereas the grey crescent is located inside of the optic disc and may partially or completely be regarded as neuroretinal rim. Not considering the differences between the alpha zone of parapapillary atrophy and the grey crescent will, therefore, markedly influence the morphological analysis of the optic disc.

The question arises what the histological equivalent of the grey crescent may be. Since the grey crescent is relatively dark, it may be associated with retinal pigment epithelium cells. These cells sitting on and forming Bruch's membrane, may partially be located in the optic nerve head region if Bruch's membrane extends internally to the peripapillary scleral ring. Future histological studies of the normal optic nerve head may be directed towards finding the clinical-histological correlate of the grey crescent, differentiating it histologically from the physiological alpha zone of parapapillary atrophy, and giving hints for the rate of the histological occurrence of the grey crescent in normal eyes. Optical coherence tomography may be an additional clinical method to analyse intravitreally the grey crescent and its spatial associations with the surrounding tissues.³

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Correspondence to: Dr J B Jonas, Universitäts-Augenklinik, Theodor-Kutzer-Ufer 1-3, 68167 Mannheim, Germany; Jost.Jonas@ma.augen.uni-heidelberg.de

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