

## PERSPECTIVE

## Can visual function be restored in patients with homonymous hemianopia?

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It can be argued that vision is the major sensory input into the human brain, by virtue of the fact that about half of all afferent fibres projecting to the brain are from the eye and by the sophistication of the neural systems controlling visual processing. Brain damage disrupts these complicated processes, resulting in severe visual impairments including hemianopia. Patients with hemianopia complain mainly of difficulties with reading and scanning scenes fast enough to make sense of things as a whole. Consequently, they fail to notice relevant objects or avoid obstacles on their affected side and may collide with approaching people or cars. This has far reaching repercussions on their vocational and private lives. A small group are unaware of their defects until they are picked up on routine ophthalmic examination.<sup>1</sup> Given the relative importance of vision as a sense, the treatment of hemianopia should assume a priority role for neurological rehabilitation programmes. In clinical practice, however, this has not been the case.

### Pathogenesis of homonymous hemianopia

Forty per cent of homonymous hemianopias involve lesions in the occipital lobe, 30% the parietal lobe, 25% the temporal lobe, and 5% the optic tract and lateral geniculate nucleus (LGN).<sup>2,3</sup> The presence or absence of macular sparing is of no use in defining the site of the lesion and there is recent evidence to suggest that it simply represents a perimetric artefact.<sup>4,5</sup> Patients with additional temporo-parietal dysfunction, especially neglect, are particularly difficult to rehabilitate from the point of view of their hemianopia.

Data concerning the pathogenesis of lesions causing homonymous hemianopia must be interpreted with caution because they are influenced by the way patients are selected.<sup>6</sup> Allowing for this, 70% of lesions are arterial infarctions, 15% tumours, and 5% haemorrhages.<sup>1,2</sup> Males aged 50-70 are most frequently affected, reflecting the fact that homonymous hemianopia is primarily a consequence of vascular disease.

### Residual vision

Traditionally, geniculostriate lesions were considered to result in complete and permanent visual loss in the topographically related area of the visual field.<sup>7-9</sup> However, it has become clear that this is not necessarily the case. Work with monkeys and, later, humans demonstrated that despite destruction of the striate cortex, or even hemispherectomy, some patients retain certain visual functions.<sup>10-16</sup> Twenty per cent of patients retain the ability to make an accurate saccades to visual targets presented tachistoscopically in their blind hemifield.<sup>17</sup> This phenomenon was termed 'blindsight' to emphasise that these residual functions are not consciously perceived and

depend on the use of a forced choice technique. Its functions are thought to be primitive and concerned with the registration and localisation of objects, whereas the geniculostriate system is primarily concerned with their identification. Since these patients no longer have a functional striate cortex on their affected side, blindsight is probably mediated via subcortical structures such as the superior colliculus, or a sparse projection from the LGN to V5 identified in monkeys.<sup>18,19</sup>

### Spontaneous recovery

Over the years it has become clear that not only do the blind hemifields of some hemianopes actually retain certain visual functions, but that they also improve spontaneously. The degree of resolution depends on the underlying pathology.<sup>20-22</sup> Regarding field defects of vascular origin, the prognosis for spontaneous recovery is poor. Any recovery of a complete hemianopia occurs in the first 10 days after which further recovery is unlikely. Recovery of a partial defect is usually maximal within the first 48 hours. Less than 10% of patients recover their full field, and a proportion of these will, nevertheless, continue to complain about their visual function because of coexisting parietal lobe involvement. Up to 50% show spontaneous regression of varying degrees. The pattern of recovery largely depends on the extent of cerebral damage that is reversible and this, in turn, relies on the state of the collateral circulation. Further recovery is negligible after 10-12 weeks.<sup>23,24</sup> The extent of visual recovery correlates negatively with age, a history of diabetes or hypertension, and the presence of cognitive, language, or memory impairment. Vision returns to the perimetrically blind field in definite temporal stages starting with the perception of light, motion, form, colour and, finally, stereognosis.<sup>24-26</sup> Traumatic hemianopia behaves somewhat differently. Large areas of the visual field frequently recover, although the functions regained are unstable and often limited to the perception of light.<sup>20,27</sup>

The body of evidence would therefore suggest hemianopia is not always an absolute and permanent visual loss. This raises the question as to whether the natural course of a hemianopia can be advantageously manipulated by therapeutic intervention.

### Treatment of patients with hemianopia

The general aim is to reduce the disability resulting from the hemianopia, boost patients' confidence, and expedite their reintegration into an independent social and professional life. The most interesting work in this field involves psychophysical training techniques aimed at strengthening patients' attention for the blind hemifield and improving their ability to make eye movements towards that side.

However, before discussing these techniques we will mention the role of surgery.

### Surgical revascularisation procedures

In 1979, Roski *et al*<sup>28</sup> reported a patient with a 7 year old ischaemic field defect who recovered full vision following a superficial temporal artery-middle cerebral artery bypass. Since then there have been case reports describing the reversal of a variety of fixed neurological deficits, especially hemianopia, with bypass procedures.<sup>29 30</sup> Benzel and Mirfakhraee<sup>29</sup> hypothesised that following a stroke an area of non-functioning but viable tissue, which they termed the zone of penumbra, was created which could recover function when the blood flow was restored. Their claims were backed by angiographic evidence demonstrating the reperfusion of ischaemic areas of the brain coinciding with an expanding visual field on serial field plots. At present, however, there are no legitimate guidelines to identify those patients with fixed neurological deficits in the presence of viable tissue that might reverse with cerebral revascularisation. In current practice, bypass procedures are not performed since they are associated with excess mortality in patients with transient ischaemic attacks and critical arterial stenoses.<sup>31</sup> Endarterectomies are basically performed in these patients as secondary prevention against further neurological deficits, rather than as treatment of established ones.

### Optical aids

Customised spectacles, fitted with either mirrors or Fresnel prisms are designed to compensate for the hemianopic field of vision. They are primarily aimed at individuals with good central visual acuity who are otherwise neurologically intact.

#### HEMIANOPIC SPECTACLES

These involve an experienced optician mounting a small mirror onto the frame of a pair of glasses, at an angle that permits the patient to learn to look into the mirror and see the reflection of objects in the hemianopic field. It is placed beside the left eye in a left hemianope and vice versa, and is suspended so that it can be adjusted by the patient.<sup>32</sup>

#### HEMIANOPIC PRISMS

These work on the principle that a prism displaces the images of objects towards its apex. Therefore, considering a left hemianopia, the prism would be placed on the temporal side of the left lens of a pair of spectacles with its base to the left. The prism then displaces the images of objects in the left (hemianopic) field towards its apex and into the seeing nasal field of that eye. Only one lens is fitted with a prism because prisms reduce the acuity of the eye involved, and a reduction in acuity of both eyes would probably be more troublesome than the original hemianopia. Fifteen to 30 dioptre prisms are usually used, increasing the useful field of vision by a maximum of 15 degrees. This is useful for spotting objects. They require careful fitting and a small central area needs to be trimmed to avoid diplopia at fixation.<sup>33</sup>

Reports of positive effects with these optical aids are anecdotal. The only controlled trial of Fresnel prisms showed that although the prism treated group performed significantly better on visuospatial tests, no functional improvement in their activities of daily living could be demonstrated.<sup>34</sup> In addition, they confused patients while walking, required specialised opticians, and patients that were highly motivated to practise. Opinion was divided as to their efficacy and they have never come into general use.

### Cognitive rehabilitation techniques

Over the past 40 years several research groups have set out to determine whether patients with hemianopia have the ability to compensate for their field defect using eye movements, head movements, and extrastriate vision. Some have integrated this information into training techniques aimed at systematically reinforcing compensatory oculomotor strategies, thereby fortifying and enlarging the field of search. Other groups, inspired by the success of animal physiologists in restoring the lost visual field of non-human primates, attempted to reproduce the results in humans using similar training techniques.

#### COMPENSATORY OCULOMOTOR STRATEGIES

These enhance the patients' ability to explore their blind hemifield. Scanning eye movements are normal in only a minority of hemianopic patients. Their search times are usually longer owing to the repetition of search saccades and fixations, resulting in longer, unsystematic scanpaths<sup>35 36</sup> (scanpaths are series of saccades and fixations undertaken when viewing a structured scene). In addition, they dwell on their intact hemifield and their saccades are less regular, less accurate, and too small to allow rapid, organised scanning or reading. Consequently, objects or relevant parts of a scene located in the affected side are omitted.

In 1987 Ishiai *et al*<sup>37</sup> analysed the eye fixations of patients looking at simple patterns and found that whereas normal controls looked mainly at the centre, their hemianopic patients paradoxically concentrated on the blind side. Like Chédru *et al*<sup>35</sup> and Gassel and Williams<sup>38</sup> they considered this to be compensatory, since deviating the fixation point towards the hemianopic side brings the whole of the pattern into the seeing hemifield. Patients with additional neglect lacked this compensation. Meienberg *et al*<sup>39</sup> recorded the eye movements of hemianopes to visual targets presented in an unpredictable fashion. They highlighted a number of compensatory strategies that patients use when faced with unpredictable targets which are complementary to Ishiai *et al*'s<sup>37</sup> observation. One strategy involves waiting for the target with the eyes in mid-position, making repeated search movements towards the blind side; the other strategy is to direct the eyes towards the blind side and wait for the target where it is expected to appear. In a further strategy, they found that when presented with a target, normal subjects will make a single saccade directly to it; yet patients with hemianopia who lack information about targets in their blind hemifield have to search for them. The majority employ a staircase strategy consisting of a series of safe but slow stepwise saccadic search movements to bring the target into the seeing visual field (stairstep strategy). Less frequently, patients adopt an alternative more efficient strategy to try and 'catch' the target with a large saccade which overshoots the target, but brings it into the seeing hemifield. They then make a corrective glissade to foveate it (overshoot strategy).

Certain groups then attempted to train patients to adopt compensatory oculomotor strategies, focusing mainly on visual exploration and reading.

#### Visual exploration

Since hemianopes use both small amplitude and unsystematic saccades to scan, most training techniques involve two consecutive steps. Firstly, patients practise making large, quick saccades (of amplitude 30° to 40°) into their blind field, to enhance the overshoot rather than the stairstep strategy described previously. They are then taught to scan for targets among distracters on projected slides (of eccentricity 30° to 40°) in a systematic way, using

a visual search paradigm to improve the spatial organisation of their eye movements. The success of this systematic training is judged by the extent of normalisation of the eye movements and the degree of restoration of the visual search field. The search field is defined as the perimetrically measured area that a patient can actively scan via eye movements, but without head movements, when searching for a suprathreshold stimulus. The acquisition of compensatory oculomotor strategies appears to depend on some sort of systematic stimulation and practice, since the general stimulation from daily activities and even occupational therapy does not achieve the same effect.

In 1988 Zihl<sup>40</sup> trained 30 hemianopes by instructing them to practise making large saccadic eye movements. Within four to eight sessions their affected visual search field had apparently increased from 10° to 30°. Kerkhoff *et al*<sup>41</sup> validated these results with 92 hemianopic patients and 30 with additional hemineglect. Their training started with the practice of large saccades to targets in their blind hemifield, which were presented for a variable duration. They were encouraged to adopt a systematic scanning strategy, involving either horizontal or vertical scanning. Then they practised searching for targets on projected slides. After about 30 sessions (6 weeks), the mean search field size increased from 15° to 35° in the hemianope group. Those with additional neglect required 25% more training over 2–3 months to achieve a similar result. These improvements only occurred during the treatment phase of the study, and at mean follow up 22 months later there were no further significant changes. Internal controls showed that the magnitude of gain was independent of variables such as aetiology, time since lesion, type of field defect, field sparing, and patient age. Patients with the severest defects benefited most from training. Interestingly, the mean number of required treatment sessions increased dramatically with the frequency and extent of head movements during training. This clearly contradicts the assumption that head movements are helpful to the compensatory mechanisms for hemianopes as is sometimes claimed<sup>42</sup> and supports the view that they are deleterious.<sup>43</sup> In a further study, Kerkhoff *et al*<sup>44</sup> quantified the functional benefit of restoring oculomotor functions. After about 25 treatment sessions, their 22 patients showed a 50% reduction in the time taken to find objects on a table (table test), complementing the subjective improvement in a questionnaire rating their own disability. Following treatment, 91% of this group resumed part-time work. Zihl<sup>46</sup> made further contributions in this field by recording the eye movements of eight patients before and after similar treatment. He trained them to make large saccades and practise searching for targets. After about 26 sessions the patients' performance improved to within the normal range. He demonstrated that after training, the shorter search times were due mainly to fewer fixations and less repetition of the scanpath and of fixations. He concluded that patients can successfully adapt to their hemianopia with training.

### Reading

Reading has been the focus of rehabilitation attempts since the beginning of this century when Poppelreuter<sup>20</sup> trained first world war veterans with brain damage to overcome reading difficulties.

Patients with hemianopia have reading difficulties proportional to the extent of their field loss. Whereas the fovea possesses the acuity required to discriminate letters and words with sufficient clarity to read, the parafoveal visual field processes forthcoming text in advance of the fovea, in order to guide eye movements smoothly while

reading. Loss of the parafoveal field ruins this 'perceptual scan' and results in a characteristic reading disorder termed 'hemianopic dyslexia'. Left sided field loss handicaps the return eye movements required to find the beginning of a new line. Right sided hemianopia, however, is generally more disturbing in our culture, since we read from left to right, and is characteristically associated with prolonged fixations, inappropriately small amplitude saccades to the right and many regressive saccades.<sup>38 45 46</sup> With training, patients can improve reading eye movements. Basically, they are taught to perceive each word as a whole before reading it: left sided hemianopes are forced to shift their gaze first to the beginning of the line and the first letter of every word in that line, whereas right sided hemianopes are discouraged to read a word before they have shifted their gaze to the end of it. An electronic computer based reading system would easily lend itself to this method of training. Such a system, designed by Zihl, was used to train a group of 96 patients (quoted in Zihl and Kennard<sup>47</sup>). When trained, they were able to read faster with fewer errors. Eye movement recordings showed that the improvement was primarily attributable to the emergence of superior oculomotor strategies using fewer fixations, larger saccadic jumps, and shorter fixation periods. As one would expect, right sided hemianopes were more disabled than left, requiring more training sessions (33 compared with 26), and never quite reaching the same standard of improvement.<sup>48</sup> Using an identical protocol, Kerkhoff *et al*<sup>49</sup> had the same success with a group of 56 hemianopic patients after about 3 weeks (mean 13 sessions) of training. Both groups confirmed that at follow up (6 months to 2 years) the improved reading performance of their patients remained stable. Other studies suggest that patients with additional neglect do not develop adaptive oculomotor reading strategies.<sup>50</sup>

### Blindsight

Advocates of the existence of blindsight reasoned that since it has been shown that monkeys improve their ability to detect and localise light targets in their hemianopic field with practice, humans may also benefit from specific retraining.<sup>51 52</sup> Using a forced choice technique patients practised discriminating, without visualising, the position of targets presented tachistoscopically in their blind field. At first, target localisation was poor but was accurately restored with practice.<sup>53 54</sup> Supplying feedback on their performance precipitated the improvement.<sup>55</sup> Patients who improved felt more confident and less disabled, adding weight to the view that blindsight training may be of therapeutic value. This reasoning is incorrect, since the specific practice technique actually trains patients to make gaze shifts towards the affected side, using mostly large saccadic eye movements. Therefore, as a byproduct, the field of search becomes larger, and there is little evidence that the training of blindsight is itself useful therapy.

### Recovery of the visual field

The potential for recovery of the lost visual field has sparked a significant conflict of opinion. Preobrazhenskaya (cited by Luria<sup>56</sup>) trained hemianopic patients to read and noticed an enlargement in their visual field. Evidence began to accumulate suggesting that it was actually possible to shrink the scotoma of monkeys by systematically training them to detect and localise light stimuli.<sup>51 57</sup> Consequently, attempts were made to reproduce these results in humans. Using a psychophysical method, light thresholds were repeatedly determined at the visual field border in 12 patients. This induced an increase in the size of the

visual field that was confined to the trained area and showed intraocular transfer, indicating its central nature.<sup>58</sup> The workers then applied the same forced choice saccadic technique for training blindsight to enlarging the visual field of 55 patients, and were successful in the majority of cases. In most the field enlargement did not exceed 5° but there were individual cases with remarkable recovery. The recovered field included form and colour perception and remained stable after the treatment sessions had ended. This result could not be attributed to spontaneous recovery because no change occurred during periods without training. Many patients were symptomatically improved.<sup>59-60</sup> A mechanism was postulated whereby training induces selective attention of the defective field, so increasing neuronal activity in areas of the striate cortex surrounding the damaged area. In analogous experiments these results were duplicated by some groups,<sup>41-44</sup> but not by others.<sup>61-63</sup> Having failed to find any significant visual field increases in their patients Balliet *et al*<sup>62</sup> concluded that Zihl and Von Cramon's<sup>60</sup> results must have been artefactual and made four main criticisms, which were subsequently refuted in an exchange of letters that left the topic in a state of uncertainty. The answer probably lies in patient selection. As pointed out by Zihl and Von Cramon,<sup>60</sup> visual field enlargement was only found in those cases with partly reversible damage to the striate cortex, as evidenced by the sharpness of the visual gradient at the border of the field defect and the extent of brain damage on the computed tomography scan. Since sharply demarcated field defects are the commonest type seen in clinical practice it seems unlikely that restoration of vision is a prospect for most patients. Nevertheless, there has been renewed interest in this challenge, with the publication of encouraging preliminary results.<sup>64</sup>

## Conclusion

In conclusion, we have outlined rehabilitation techniques used to treat patients with hemianopia, in particular to reduce their main visual disabilities of reading and visual exploration. These methods are principally based on learning oculomotor compensation strategies, thereby strengthening the patients' attention for the blind hemifield, improving their ability to direct gaze movements towards that side and explore it efficiently. Recovering the lost visual field appears to be a less tenable prospect. Research into the rehabilitation of brain damaged patients with functional impairments is an intrinsically difficult and laborious task. It is fraught with methodological problems since the patients comprise an inhomogeneous group with respect to factors such as the nature and extent of brain damage, pattern of deficits, patient's insight, and motivation. The question of whether the acquisition of compensatory oculomotor strategies by patients translates to an improvement in their overall function is of crucial significance and must be answered. In the UK, research into this area of rehabilitation is notable by its absence. In view of the fact that at least 50% of neurological admissions to a general hospital are due to strokes and that 30% of these cases have hemianopia, there are ample patients in whom treatment procedures can be evaluated. We will then be able to make a pragmatic, scientific, and cost effective decision about whether or not we can reduce the degree of disability in our patients.

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