Contents

Series preface ix
Preface xi
Preface to the second edition xiii

1 Introduction 1

Rehabilitation of cerebral visual disorders in the framework of visual neuroscience 1
Recovery of function: Restitution vs. substitution 5
The role of brain plasticity in the rehabilitation of cerebral visual disorders 8
The role of learning in visual rehabilitation 11
The significance of cerebral visual disorders for neuropsychological assessment and rehabilitation activities 15
Rehabilitation of cerebral visual disorders: Some methodological considerations 16
In search of evidence for the efficacy of treatment 18
About this monograph 20

2 The visual brain 23

3 Visual field disorders 29

Forms and frequency of occurrence 30
Spontaneous recovery of visual field from scotoma 35
Spontaneous adaptation and adaptability to the visual field defect 38
Awareness of visual field loss and subjective reports 61
Recovery from scotoma by systematic training 67
Substitution by oculomotor compensation 71
| Box 3.1 | Does site of parafoveal field loss matter? | 119 |
| Box 3.2 | Does type of parafoveal field loss matter for practice with reading? | 121 |
| Box 3.3 | Does time since brain injury matter? | 122 |
| Box 3.4 | Does age matter? | 124 |
| Box 3.5 | Does size of brain injury matter? | 126 |
| Box 3.6 | Practice effects in cases of bilateral field loss | 128 |

4 Visual acuity, spatial contrast sensitivity and visual adaptation

Visual acuity | 131 |
Spatial contrast sensitivity | 133 |
Light and dark adaptation | 138 |

5 Colour vision

Disorders | 141 |
Spontaneous recovery | 144 |
Practice with colour discrimination | 145 |

6 Visual space perception

Disorders | 152 |
Spontaneous recovery | 161 |
Practice with spatial localisation | 161 |
Balint’s syndrome | 173 |

7 Visual agnosia

Difficulties with definition of visual agnosia | 186 |
Forms of visual agnosia | 187 |
Spontaneous recovery | 190 |
Practice | 190 |
Long-term effects of treatment and control for efficacy | 203 |

8 Central scotoma

Disorders | 207 |
Spontaneous recovery | 208 |
Practice | 208 |
Long-term effects | 217 |
Comment | 222 |
## Contents

### Appendix

- Homonymous visual field disorders 223
- Spatial contrast sensitivity 225
- Colour vision 225
- Visual space perception 226
- Balint’s syndrome 226
- Visual recognition 227
- Central scotoma 228

### References 229

### Author index 255

### Subject index 267
1 Introduction

Rehabilitation of cerebral visual disorders in the framework of visual neuroscience

From the very beginning of neuroscience, vision research has mainly been concerned with the elucidation of the nature of various visual deficits and the identification of the location of brain injury responsible for these deficits (Zeki, 1993). Early clinical reports on patients showing selective loss of visual functions and abilities following acquired posterior brain injury have suggested a functional segregation of the visual cortex, a concept that many years later has been verified on the basis of combined anatomical, electrophysiological and behavioural evidence (Desimone & Ungerleider, 1989; Grill-Spector & Malach, 2004; Zeki, 1993). Enormous progress has been made in understanding the neurobiological basis of visual perception and the neuropsychology of vision is still a major topic in neuroscience. However, this progress is not reflected in the study of recovery of visual function in patients with acquired brain injury. The related findings are not included in neuropsychological rehabilitation, possibly because visual-perceptual disorders are not considered as cognitive deficits (see, for example, Cicerone et al., 2005; Halligan & Wade, 2005; Ponsford, 2004). At first sight, this is difficult to understand given the fact that about 30% of patients with acquired brain injury suffer from visual disorders (Clarke, 2005; Hier, Mondlock & Caplan, 1983a; Rowe et al., 2009; Sarno & Sarno, 1979; Suchoff et al., 2008). Furthermore, visual disorders often either directly affect cognitive performance or exacerbate cognitive deficits (Uzzell, Dolinskas, & Langfitt, 1988), which may interfere with the rehabilitation of other cognitive impairments and impede vocational rehabilitation efforts (Grosower, Cohen & Blankstein, 1990; Patel, Duncan, Lai, & Studenski, 2000; Reding & Potes, 1988; Rowe et al., 2009). However, it does not seem that a lack of interest in the recovery of vision or visual rehabilitation can account for the fact that the advances in understanding the neurobiological basis of visual perception have not led to greater progress in neuropsychological rehabilitation. As early as 1867, Zagorski reported the case of a 35-year-old lady who complained of loss of vision on the left side. Perimetric testing revealed a complete left-sided
hemianopia, probably caused by a right-sided occipital haemorrhage. Eight
days later the patient noticed return of light vision in her left hemifield;
6 weeks later she reported having full vision again. Weekly visual field mea-
surements were in agreement with the patient’s reports: the region of blindness
shrank successively and vision eventually returned to the left hemifield (see
Figure 1.1). Zagorski’s single case report is probably the first report on
recovery of vision after acquired brain injury. In their “Handbook for
Neurologists and Ophthalmologists” [author’s translation] Wilbrand and
Saenger (1917) dedicated a comprehensive chapter to the natural course of
complete cerebral blindness. According to their observations, vision recovered
first in one hemifield; a few cases later showed complete return of vision in
both hemifields. In most cases recovery of vision took place within hours
or days; in some patients, however, the process of recovery was much slower
and was not completed for several weeks. A similar course was observed
in subjects with homonymous hemianopia. In the same year (1917/1990),
Poppeleuter published his monograph on visual disturbances after occipital
gunshot wounds, in which he reported not only the results of his detailed
diagnostic visual disorders assessment, but also his observations on spon-
taneous recovery and the effect of systematic treatment. Poppeleuter pre-
ferred an experimental approach for assessing and treating his patients. In
his view, conventional assessment and treatment methods were too “crude”
(i.e., inaccurate), and systematic rehabilitation measures did not exist. His
approach was also very pragmatic, i.e., ecologically valid, which is exempli-
fied by his statement that “any intervention should, at the very least, have as
its aim that the man should again be able to converse comprehensibly, to
write his own letters, to read a newspaper, and to calculate his expenses by
himself” (p. 5). Poppeleuter pointed out that functional impairment in vision
in the acute stage may often be exaggerated since unspecific cognitive and
affective alterations can affect the use of spared visual capacities. Since com-
plete spontaneous recovery was the exception rather than the rule in his
patients, rehabilitation measures were required to reduce their visual handi-
cap and improve their independency in daily life activities and, thus, their
“usefulness for society”. Poppeleuter was aware of the difficulty of attribut-
ing an improvement unequivocally to the treatment: “Only exact control of
the effect [of treatment] offers a substantial argument for the systematic train-
ing effect over a short period of time, namely using a work task which
remains constant” (p. 240). He developed training methods that specifically
aimed at improving the reading impairments in patients with visual field loss
(i.e., hemianopic dyslexia), which have already been described by Mauthner
in 1881 and Wilbrand in 1907 (see Schuett, Heywood, Kentridge & Zihl,
2008a, for a comprehensive review). Poppelreuter correctly noticed that para-
foveal field loss is not only associated with impaired global text processing
but also with an impairment of the “co-ordination of the reading gaze-
shifts”, which becomes manifest as a distortion of the typical, staircase-like
oculomotor reading pattern. He therefore taught patients to compensate for
Figure 1.1 Spontaneous recovery of vision in Zagorski’s case with left homonymous hemianopia (1867). Left panel: Visual field of the left eye; right panel: visual field of the right eye. (A) Outcome of perimetric testing on 11 June 1867; (B) on 26 June 1867; (C) on 5 July 1867. On 27 July, Zagorski found complete recovery of vision in the left hemifield.
their field loss by systematically shifting their fixation from the beginning to the end of a line. The resulting improvement in reading performance is the first known example for the substitution of visual field loss by oculomotor activities. According to Poppelreuter, the substitution of an impaired function (the parafoveal visual field) by another, intact function (eye movements) critically depends on whether the replacement function also contributes to the visual capacity or performance in question under normal circumstance.

Yet, Poppelreuter’s observations on the recovery of vision after brain injury and his experiences with the systematic treatment of visually disabled, brain-injured patients as well as on the recovery of visual function in patients with occipital injury have largely been neglected in the literature. Even in the classic monograph by Teuber, Battersby, and Bender (1960) on visual disturbances, only few qualitative data are reported on the recovery of visual functions in a similar group of cases (Second World War soldiers) with missile wounds to the brain. Teuber (1975) reported follow-up observations in 520 soldiers with occipital gunshot injuries and confirmed Poppelreuter’s observation that vision can reappear in affected visual field regions.

In contrast to the diminishing interest in the study of recovery of vision after brain injury in humans after the end of the First World War, there was a growing interest among brain researchers in studying the effects of experimentally induced lesions to brain structures subserving vision in animals. It was Klüver (1942), especially, who studied the effect of experimentally induced brain lesions on vision in monkeys and found that bilateral occipital injury results in a profound but not total loss of visual information processing capacity. Yet, it required a “sufficiently long period of training” before the monkey was able to, for example, locate objects in space. A famous single case study on a rhesus monkey, Helen, from whom the striate cortex was almost totally removed bilaterally, and who was studied intensively over a period of 8 years (Humphrey, 1974), revealed that the monkey could regain an effective, though limited, degree of visually guided behaviour by practice in natural environments. Cowey (1967) and Weiskrantz and Cowey (1970) convincingly showed that practice can reduce the size of a cortical, but not a retinal, scotoma in monkeys, as defined by the animal’s ability to detect light targets. An even more complete and specific recovery of vision was demonstrated by Mohler and Wurtz (1977). Deficits in the detection of light stimuli in the affected field region as well as in saccadic localisation accuracy disappeared 6 weeks after the lesions had been induced. Again, systematic practice was necessary for recovery, which was mainly observed in the portion of the scotoma that had been subjected to practice. Thus, injury to the striate cortex does not always result in an irreversible, absolute loss of vision, but systematic treatment is required for its return. However, the results obtained in animal studies cannot directly be transferred to patients. It has, for example, been shown that patients can also accurately respond to light stimuli presented in their cortical scotoma. This phenomenon has been coined “blindsight”, since patients are never aware of the presence of the target. Demonstrating
this phenomenon requires special testing conditions and typically patients do not show it without systematic practice of the task (see Danckert & Rosetti, 2005; Weiskrantz, 2004, for comprehensive reviews). It is still an open issue whether this “residual visual capacity” is due to recovery, residual functioning of spared visual cortex, or represents a visual function that is based on extrageniculo-striate mechanisms that were not affected by injury. However, “blindsight” does not seem to reduce patients’ visual disability because they cannot make use of this sub- or unconscious visual function (Zihl, 1980).

Thus, although there is no doubt that in human subjects showing this capacity visual processing takes place in the “cortically blind” field region (like in monkeys with cortical scotoma), nobody would go so far as to consider a patient showing blindsight “visually rehabilitated”. Nevertheless, visual brain lesion research in primates has contributed substantially to our understanding of the recovery of visual function in humans. On the one hand, monkeys show considerable return of both elementary and complex visual function after intensive and systematic training to overcome the deficit. On the other hand, these experiments demonstrate that total and irreversible loss of a particular visual capacity is only to be expected if more than one structure of the neural network subserving this capacity is injured (see, for example, Frommer, 1978; Rothi & Horner, 1983; Stein, 1994, for reviews). As already shown by Mohler and Wurtz (1977), monkeys no longer recover from blindness after striate cortex injury when their ipsilateral colliculus superior has also been destroyed. Thus, both the site and the size of the lesion appear critical in the study of recovery of vision and adaptation to visual field loss after brain injury.

Recovery of function: Restitution vs. substitution

The first and foremost question in rehabilitation after brain injury is whether there is any recovery potential at all. If a particular visual function depends entirely on one single cortical structure, and if this structure is completely and irreversibly injured, then recovery of the affected visual function cannot be expected. Unfortunately, and despite enormous improvements in brain imaging techniques (Johansen-Berg, 2007), the definition of reversibility and irreversibility of brain injury is still an open issue. In cases of spontaneous recovery it is, of course, reasonable to assume that brain injury merely had reversible consequences (see Bosley et al., 1987). But does the opposite also always hold true, namely that the brain structure in question has really undergone irreversible injury when no spontaneous recovery occurs? Another important but similarly difficult question concerns the cortical representation of visual functions. “Functional specialisation” does not imply strict localisation of function. If it did, then injury to a particular cortical area would always destroy the function in question completely and irreversibly. However, the situation is yet more complicated, as the following case studies will demonstrate.
LM had lost most of her capacity to see motion following bilateral posterior brain injury due to sinus venous thrombosis (Rizzo, Nawrot, & Zihl, 1995; Zihl, von Cramon, & Mai, 1983; Zihl, von Cramon, Mai, & Schmid, 1991). She reported, however, somehow “seeing” objects in motion, provided that (1) only one stimulus was moving, (2) the speed of the moving stimulus did not exceed 6 degrees per second, and (3) objects were moving either horizontally or vertically. This “residual” movement vision could either constitute incomplete injury to V5 (the “visual motion” area) or be accounted for by other visual areas. The measurement of brain activity during the processing of moving visual stimuli in LM showed no evidence of activation of V5 in either hemisphere. Somewhat surprisingly, activation was observed in another visual area (V3) and in the superior parietal cortex (Brodmann’s area 7). Both areas, however, are not known to be “functionally specialised” to process visual motion signals and are not activated in normal subjects in the same experimental conditions, but are the likely candidates for LM’s residual movement vision (Shipp, de Jong, Zihl, Frackowiak, & Zeki, 1994; Zeki, 1991). Thus, movement vision is possible without V5, although under extremely restricted conditions. This “residual” movement capacity did not improve over the years and could not be used by LM to substitute the role of her injured V5, which would have reduced her severe daily visual handicaps. Nevertheless, LM learned efficient strategies to cope with her extraordinary visual disorder, mainly by avoiding conditions in which moving stimuli dominated or interfered with her activities.

DF, a patient reported by Milner et al. (1991), suffered a severe visual deficit of form recognition following bilateral posterior brain injury due to asphyxia while taking a shower as a result of a faulty gas water heater. She had great difficulties in discriminating, for example, simple shapes and line orientations. Despite poor performance in these tasks, she had little difficulty in everyday visually guided activities such as opening doors, shaking hands, eating meals, reaching out accurately for and grasping objects differing in form, size and orientation. The authors hypothesised that the preserved visuo-motor ability may depend on routes still functioning from the occipital lobe, where the analysis of visual forms is performed, to neural mechanisms in the parietal lobe that control visually guided movements of the hand and fingers. Explicit visual form perception and recognition is therefore not a crucial prerequisite for an appropriate visual guidance of hand and finger movements. In contrast to LM, who did not benefit from her residual visual motion perception, DF certainly could make use of her “residual” visual form processing capacity in her daily life activities.

Certainly, nobody would interpret the use of spared or substituted visual functions in these two cases as recovery (although it led to functional improvement). Yet, these and similar observations underlie the need for an accurate and detailed analysis of lost, impaired, spared, and substituted visual functions. Otherwise sparing or substitution of function could easily be confounded with recovery of function, especially if systematic practice is required.
to reveal a spared or substituted visual capacity. Systematic practice might be particularly important to reveal visual capacities in cases with denial of preserved vision (so-called negative Anton syndrome) as well as in cases with a reduction of initiative and self-generated activities due to concomitant depression or reduced motivation (e.g., Feibel & Springer, 1982; Richards & Ruff, 1989; Van de Weg, Kuik, & Lankhorst, 1999). The sites and sizes of brain lesions differ, of course, among patients, but this may not be reflected by the (initial) severity of a single visual deficit or a pattern of visual deficits. Recovery of visual function as well as functional improvement through compensation may, however, depend on the integrity of brain structures beyond the visual cortex and on fibre pathways interconnecting these structures. Thus, knowledge about the networks subserving complex visual capacities, which also include the involved cognitive components, is important in interpreting cerebral visual dysfunction as well as spontaneous recovery and improvement after practice.

Another difficulty arises from the fact that patients cannot (and must not) be kept in a “controlled” constant environment as animals can. Consequently, any kind of improvement of function can in principle also be attributed to confounding “environmental” variables. Finally, how much time should a brain function be given to recover “spontaneously”, and how long should treatment be continued before one can reliably state that no (further) recovery or spontaneous adaptation can be expected? If systematic intervention is started early after brain injury and the function in question returns (partially or completely) the primary and most obvious explanation is that the improvement might also have occurred without systematic intervention. This is a serious argument, but not easy to dismiss, as Poppelreuter (1917/1990) has already noted. Sceptics may even argue that even if treatment starts several weeks or even months after the onset of a functional deficit, an improvement after systematic practice may still represent spontaneous recovery. One possible way of avoiding this problem is to monitor the effect in the experimental group by using a control group. Although this seems to be an ideal methodological approach, one still faces the problem of heterogeneity of brain injury and the resulting differences in functional consequences as well as of controlling for environmental influences. For the patient it is, of course, irrelevant why improvement has occurred as long as it does occur and the outcome is beneficial. For the researcher, however, it is not irrelevant. There is agreement that the adoption of a method of treatment should be based on an underlying theoretical rationale and on the control of non-specific factors (e.g., motivation, emotional state, social support; Robertson, 1994). These factors may impair or enhance the improvement, although they do not represent and cannot substitute specific treatment procedures.

Thus, it seems that rehabilitation research in neuropsychology is a very laborious and difficult task, for which no satisfactory design exists. At the same time it is extremely risky because success cannot be guaranteed, even after a high expenditure of time, resources and energy. Of course, reports on
negative findings are just as important as reports on positive outcomes (Barlow & Hersen, 1985), not only for methodological reasons, but also because it allows for the extraction of criteria for making a valid decision about the efficacy of a particular treatment procedure in scientific and pragmatic (i.e., ecologic) terms. When developing and proving new methods, one can hardly predict their potential significance for rehabilitation at the same time. What is possible, however, is to plan, a priori, the development and evaluation of treatment methods, as well as to define patient eligibility criteria and select appropriate outcome measures in the context of behavioural benefit (Baddeley, Meade, & Newcombe, 1980). Using such measures allows the evaluation of treatment-related functional improvements in terms of whether they increase independence and life quality. This is not only important from the viewpoint of rehabilitation research but is also essential for the motivation of patients. The earlier the patient is aware of an improvement in everyday-life activities, the higher will be their motivation to co-operate and the earlier the patient will become an expert on the specific individual difficulties, and how to cope with them.

Studying the recovery of brain function as well as the mechanisms underlying substitution and compensation is not only of importance for neuropsychological rehabilitation but also contributes substantially to the understanding of the functional organisation and reorganisation of the brain and, thus, of brain plasticity. The better we understand dynamic brain–behaviour relationships as well as the potentials of brain plasticity and its underlying processes, particularly in pathological conditions, the more success we have in developing efficient rehabilitation methods and the more the patient will eventually benefit. Considering the prospects and limits of brain plasticity after acquired brain injury and appropriate models of intervention (including the important issue of treatment design) may help to improve and widen the scientific as well as the pragmatic (i.e., ecological) approach to neuropsychological rehabilitation. Scientific pragmatism does not substitute scientific research. However, scientific research without pragmatism is not helpful in functional rehabilitation since it lacks applicability. It is the interplay between basic and applied neuroscience that may allow us to successfully (or, at least, sufficiently) deal with the challenges in neuropsychological rehabilitation.

The role of brain plasticity in the rehabilitation of cerebral visual disorders

The term “brain plasticity” refers to the capacity of the brain to adapt successfully to changes in either its morphological and functional systems or in the environment. Morphological and functional alterations occur in normal and pathological conditions. “Normal” conditions refer to the developmental changes of the central nervous system (CNS) that occur particularly in the early and later phases in life. “Pathological” conditions refer to abnormal
brain development, abnormal brain ageing, and brain injury. It is well known that, even for the normally developing brain, two factors play a crucial role: adequate environmental stimulation and systematic practice; the terms environmental- and practice-dependent plasticity have been coined correspondingly (Nithianantharajah & Hannan, 2006). Brain plasticity consists of morphological, physiological, and functional principles, which all serve the successful adaptation of the CNS to challenges encountered during normal development or after acquired brain injury (de Magalhaes & Sandberg, 2005; Kolb & Whishaw, 1998). Recent evidence strongly suggests that the principles underlying adaptation and, thus, plasticity are similar for both conditions (Kelly, Foxe, & Garavan, 2006), except that in pathological conditions the “degrees of freedom” are reduced in the functional system affected. Regarding brain plasticity, the crucial question is which factors are advantageous and which are disadvantageous in dealing with the challenges arising from acquired brain injury.

Cognitive performance depends on an individual’s brain functions and capacities (“cognitive architecture”), successful acquisition of skills by learning (“cognitive reserve”), and on the intention to use these capabilities and skills in a systematic and regular manner in everyday life activities (e.g., Baltes, Staudinger, & Lindenberger, 1999; Hendrie et al., 2006; Reuter-Lorenz, & Lustig, 2005). Cognitive architecture consists of the neurobiological architecture of the CNS and in particular of the brain structures subserving the selection, coding and storing of information (information processing and attention; memory) as well as the formation of routines (strategies) and habits (procedural learning, procedural memory). Cognitive architecture requires plasticity for developing and optimising strategies for a flexible adaptation to task- and environmental-related changes and for strategic adaptation to challenges arising from functional losses that occur during ageing or after brain injury. The regular use of cognitive capabilities and skills is relevant for their maintenance and builds the crucial basis for establishing the cognitive reserve. Successful human behaviour is both highly flexible and highly stable; these two prerequisites rely on, at least, two parallel networks, i.e., a fronto-parietal network for optimal rapid adaptive control, and a cingulo-opercular network for stable set maintenance. This dual network control architecture also supports the overall resilience of top-down control in brain injury (Dosenbach, Fair, Cohen, Schlaggar, & Petersen, 2008). Thus, cognitive reserve may be understood as practice-dependent plasticity that can be used for successfully selecting and optimising functions and skills as well as for compensating if they are lost (Kramer, Bherer, Colombe, Dong, & Greenough, 2004). Within such a framework, the concept of the cognitive reserve may also serve as a viable model for explaining the compensatory capacity of the brain as a response to injury-related functional losses (Stern, 2009). Cognitive reserve in vision may be reflected by additional activation in prefrontal areas for visual cognitive activities such as visual search or scanning and reading, although there might exist a posterior–anterior activity.
gradient (Anderson et al., 2007; Hamker, 2005; Jung et al., 2008). Interestingly, healthy older, in contrast to younger, individuals activate additional or different brain structures (compensation vs. dedifferentiation, respectively) in these tasks to ensure “normal” performance (Cabeza, Anderson, Locantore, & McIntosh, 2002; Reuter-Lorenz, 2002; Stern et al., 2005). Thus, older subjects make use of a different, compensatory, neural network to maintain function despite age-related changes in the brain (Dickstein et al., 2007), which may be understood as an adaptation of functional brain networks (Stern, 2003). Learning by repetitive practice (training) is highly efficient when performed in an adequate environment. The combination of such “enriched experience” with neuropharmacological treatment may further improve therapeutic effectiveness (Will, Galani, Kelche, & Rosenzweig, 2004).

Summarising the neurobiological and neuropsychological evidence on brain plasticity shows that the most relevant role of brain plasticity is the adaptation of the functional systems to changing environments and to the various consequences of injury to these systems. In both cases, learning plays a crucial role (Hummel & Cohen, 2005; Will et al., 2004), and the acquisition and use of adaptation strategies depend on individual plasticity (Baltes et al., 1999). Interestingly, collective neuroimaging findings support the role of practice in facilitating recovery and reorganisation of function and, thus, use-dependent neuroplasticity (Levin, 2006). Thus, the individual cognitive architecture and cognitive reserve already present before brain injury, the spared functional plasticity after brain injury, as well as the environment and practice conditions for learning adaptation strategies determine which coping strategy is optimal for dealing successfully with the functional consequences of brain injury. The last two factors seem to be of greater importance because recovery of the affected function or capacity is the exception rather than the rule. Consequently, elucidating, understanding and considering the neurobiological and neuropsychological framework of brain plasticity in terms of functional adaptation/compensation as well as advantageous and disadvantageous environment and practice conditions play a central role in neuropsychological rehabilitation. The major concern in this field is undoubtedly establishing an adequate environment that “stimulates” the brain at the actual performance level of the injured functional system as well as selecting the appropriate type of practice. Only when both components are adequately chosen, i.e., when a task comprises both adequate stimulation and adequate behavioural demands, can plasticity fully materialise and ensure an efficient adaptation and, thus, a beneficial rehabilitation outcome (Kramer et al., 2004; Nithianantharajah & Hannan, 2006). Of course, the “environment” in a neuropsychological rehabilitation setting is and has to be restricted since it is impossible to realise the various, complex environments of everyday life and because patients are often unable to cope with a complex environment. It is, however, crucial that we “simulate” a prototypical environment in the neuropsychological rehabilitation setting, which allows the learning of adaptation strategies that can later successfully

be transferred to real-life situations with a high degree of generalisation. Concerning practice, one has to consider the best type of learning. Regaining efficient visual information processing and oculomotor strategies requires perceptual and skill learning; implicit/procedural learning appears therefore to be most appropriate (see below).

The role of learning in visual rehabilitation

Alleviating cerebral visual disorders is mediated by visual perceptual learning. Perceptual learning in the visual modality can be understood as practice-induced improvement in the ability to perform specific visuo-perceptual tasks. The underlying learning processes are top-down regulated (Ahissar & Hochstein, 2004), and enhance visual perceptual and visuo-motor flexibility even at low-level or “early” information-processing stages (Fahle, 2009). Because visual perception and action (including oculomotor activities) are guided by specific unifying principles for neural coding and computation across visual and motor domains, both visual perceptual learning and motor learning are assumed to be guided by similar brain mechanisms (Paz, Wise, & Vaadia, 2004). Therefore, practice-induced changes of impaired visual perceptual and visuo-motor activities are also suggested to depend on similar learning processes and, thus, brain networks.

In the rehabilitation of cerebral visual disorders, lower level visual functions and capacities (e.g., the visual field, visual acuity, contrast sensitivity, colour discrimination and visual localisation) may be understood as perceptual architecture, while higher level, visuo-cognitive capacities (e.g., text processing and recognition) involve the cognitive reserve. However, the flexible use of lower level visual functions and capacities also involves learning processes and executive functions and, thus, the cognitive reserve. Visual information-processing skills are based on learning and rehabilitation therefore has to focus on the acquisition of procedures to regain, for example, rapid global and selective local processing modes in scene and text processing, including the detection, localisation, discrimination and identification of relevant stimuli. At a more explicit level, the patient may supervise his activities and eventually decide whether the outcome of his activities was successful or not. This supervision comes into play as soon as the underlying procedures have become routine after intensive and systematic practice. Thus, practice largely takes place at the procedural level. The patient does not have easy access to the procedural level and possibly should not make use of it. Otherwise learning processes and supervisory activities may compete for attentional resources and interfere with each other, which may impede the implicit acquisition of the new skill required for successful adaptation to the visual perceptual task demands. Consequently, the patient should focus on the procedure in question and not be engaged in other cognitive activities. Immediate feedback on the appropriateness of the subject’s response may be preferred over inappropriate trial-and-error learning since it prevents errors that may
occur in systematic practice procedures (Clare & Jones, 2008; Mount et al., 2007; Sidman & Stoddard, 1967). Cognitive processes, particularly executive functions, are partly involved in procedural learning, at least in the initial learning phase (Beaunieux et al., 2006). In addition, repetition rate and passage of time of practice are crucial factors for the acquisition of perceptual skills (e.g., Ofen-Noy, Dudai, & Karni, 2003), and massive practice may be superior to distributed training (Beaunieux et al., 2006; Bilodeau & Bilodeau, 1961).

If skill acquisition is the major basis of learning new strategies to overcome functional deficits, it is important to consider whether a brain-injured patient is still capable of skill learning, i.e., whether the brain networks and functions that underlie this type of learning are sufficiently spared. Neurophysiological and brain-imaging studies in primates and humans have revealed that the “association” and “sensorimotor” regions of the basal ganglia contribute to the early and late stages of learning visuo-motor sequences (Miyachi, Hikosaka, & Lu, 2002). The dorsolateral prefrontal (DLPF) cortex and the presupplementary motor area (pre-SMA) are also involved; the latter structure is assumed to co-ordinate and adjust the contribution of the visual and motor loops for a final motor output (Nakahara, Doya, & Hikosaka, 2001; Pascual-Leone, Wassermann, Grafman, & Hallett, 1996). For the acquisition and adaptive use of oculomotor sequences, the frontal and supplementary eye fields as well as the DLPF cortex and the posterior parietal cortex play a significant role (e.g., Iba & Sawaguchi, 2003; Nobre, 2001). Supporting evidence stems from observations showing deficits in procedural learning in patients with basal ganglia disease (Koenig, Thomas-Anterion, & Laurent, 1999) or in those with injury to supplementary motor (Ackermann, Daum, Schugens, & Grodd, 1996) and prefrontal structures (Belderrain, Grafman, Pascal-Leone, & Garcia-Monco, 1999). In the context of skill acquisition it is important to note that a prefrontal dysfunction may particularly affect errorless learning (Sidman & Stoddard, 1967) and learning of visuo-motor sequences since it is associated with cognitive impairments in working memory and executive function (Pitel et al., 2006). Slow information processing (“cognitive slowing”), in contrast, does not seem to significantly affect skill learning (Timmerman & Brouwer, 1999). Ng, Stein, Salles, and Black-Schaffer (2005) reported cognitive deficits as major impairments after occipital stroke, and Nys et al. (2005) found long-term cognitive impairment in about one third of patients with stroke, but early neuropsychological assessment can provide valid information on affected and preserved cognitive capacities (van Zandvoort, Kessels, Nys, De Haan, & Kappelle, 2005).

Non-cognitive factors can also modulate the individual functional resources and their use and, thus, plasticity as well as establishing a cognitive reserve. Education and occupation are the most important factors. The enrolments in secondary education and literacy have been identified as crucial variables for functional adaptation (Charness, 2006). Higher education has a significant positive effect on most measures of cognitive performance, especially in
high-attention-demanding tests (Le Carret et al., 2003). Another important factor is motivation for cognition, i.e., the active search for task-relevant information, dealing with the challenge of finding an optimal or, at least, feasible solution for problems associated with unfamiliar or complex tasks as well as with the need to make flexible responses to changing environmental demands, and the compensation for losses by developing alternative strategies or changing goals (Ebner, Freund, & Baltes, 2006; Hultsch, Herzog, Small, & Dixon, 1999). Such adaptation is an active process, which relies on implicit and explicit learning, and is motivated and guided by individual internal and external factors, including motivation and openness to experiences. In the context of lifelong cognitive development, motivation for cognition seems to be essential, not only for improving but also for maintaining cognitive capacities and particularly their regulation with respect to functional compensation. Thus, the regular use of cognitive functions and engagement in cognitive activities may be a protective mechanism against losses in cognitive capacity that are associated with ageing or arise from brain injury. Cognitively stimulating experience also contributes to the cognitive reserve. Frequent cognitive activity is, for example, associated with higher perceptual speed and better semantic memory (Wilson, Barnes, & Bennett, 2003). Regular social activities can also be considered as cognitive activities since communication requires the use of a wide range of cognitive functions and capacities (Kramer et al., 2004). Gender, or rather the associated differences in self-estimations of cognitive performance, is another factor that can influence cognitive activities: men tend to overestimate, women to underestimate, their performance in cognitive tasks, a difference that appears to remain constant across the life-span (Pallier, 2003). Socioeconomic status and cultural background may also affect cognitive activities since they influence lifestyle and, thus, cognitive habits (e.g., Baltes et al., 1999; Kramer et al., 2004; Lehmann, Chiu, & Schaller, 2004; Seeman, Lusignolo, Albert, & Berkman, 2001). Cross-cultural differences in performance of cognitive tasks may therefore be substantial (Poortinga, van de Vijver, & van de Fons, 2004).

Personality traits play a significant role in cognitive reserve, because they “guide” and therefore influence cognition and motivation for cognition. It has been found that individuals with a higher degree of conscientiousness tend to underestimate their cognitive performance (Comijs, Deeg, Dik, Twisk, & Jonker, 2002; Kumar, Jorm, Parslow, & Sachdev, 2006). In association with a high level of openness, however, they are more efficient in information selection, problem solving, and dealing with challenges (e.g., David & Suls, 1999; Vollrath & Torgerson, 2000). Furthermore, openness and agreeableness are significantly correlated with cognitive abilities (Hultsch et al., 1999; Schaie, Wills, & Caskie, 2004). There is evidence that personality factors also influence the rehabilitation outcome in patients with brain injury. The Big Five personality model consists of the primary dimensions of individual personality differences, i.e., extraversion, agreeableness, conscientiousness, openness for experience and neuroticism (Matthews, Saklofske, Costa, Deary,
Openness, extraversion, and conscientiousness predict motivation to learn and developmental activity (Major, Turner, & Fletcher, 2006) and the use of coping strategies in daily life (Connor-Smith & Flachsbart, 2007). Consequently, considering individual personality traits and related learning style preferences is supportive for achieving learning targets (Jessee, O’Neill, & Dosch, 2006). It can therefore be assumed that openness may be helpful in accepting therapeutic interventions; conscientiousness may support a patient’s engagement in the rehabilitation measures. There is evidence that the Big Five personality traits remain relatively stable in adulthood (Rantanen, Metsapelto, Feldt, Pulkkinen, & Kokko, 2007), except for extraversion and openness, which seem to be negatively associated with age (Donnellan & Lucas, 2008). Thus, older patients may require more support with accepting treatment procedures and learning effective coping strategies, particularly when they are unfamiliar or less familiar with them.

Another factor that might influence rehabilitation outcome is mood. Depressive symptoms are the most frequent psychopathological symptoms after acquired brain injury. Prevalence rates range between 20 and 65% after stroke (Pohjasvaara et al., 1998; Pohjasvaara, Vajta, Leppävuori, Kaste, & Erkinjuntti, 2001) and between 30 and 40% after traumatic injury (Jorge et al., 2004). Although these rates may drop after 3 to 6 months, they are still about 30% (Jorge et al., 2004; Kotila, Numminen, Waltimo, & Kaste, 1999). Depressive symptoms are not only associated with reduced motivation and self-initiative behaviour but may also exacerbate cognitive dysfunction (e.g., Chamelian & Feinstein, 2006; van de Weg et al., 1999), which adversely affects the rehabilitation outcome (Ostir, Berges, Ottenbacher, Clow & Ottenbacher, 2008; Parikh et al., 1990; Shimoda & Robinson, 1998). The reduction of self-generated and self-initiative activities associated with depression or reduced motivation associated with brain injury may in addition have a negative impact on the individual rehabilitation potential (e.g., Richards & Ruff, 1989), and are therefore to be considered and often require additional therapy.

In summary, apart from the selection of training paradigms with appropriate stimulus (environment) and response (practice) conditions aiming at the acquisition of procedures that allow patients to compensate for cerebral visual disorders, sufficiently spared cognitive capacities (sustained, focused and divided attention, working and long-term memory, executive functions) are essential prerequisites for successful learning and thus for any kind of functionally oriented rehabilitation measures. Although basal ganglia as well as frontal and prefrontal structures are usually spared in posterior brain injury, posterior–anterior fibre connections may be affected, i.e., interactions between prefrontal and posterior structures may be interrupted, which are crucial for procedural learning and skill acquisition. Thus, the functional significance of white-matter lesions should also be considered when planning and conducting treatment in patients with cerebral visual disorders. Furthermore, personality traits and depressive states, which are known to reduce...
motivation and self-esteem, seem to determine the rehabilitation outcome (Ostir et al., 2008; Parikh et al., 1990; Shimoda & Robinson, 1998). Summarising the state of the art in neuropsychological rehabilitation demonstrates the necessity of a theoretical framework that considers not only the processes underlying the plasticity in the normal and the injured brain and the possibilities of influencing these processes by adequate environment and practice conditions, but also the factors modulating the potential of plasticity and, thus, of rehabilitation (Wilson, 2008). Since successful skill acquisition critically depends on these factors, as the study of cognitive plasticity in normal subjects indicates, this seems even more important.

The significance of cerebral visual disorders for neuropsychological assessment and rehabilitation activities

Adequate visual perceptual capacities are essential for many activities, including reading, writing, figure drawing, grasping and pointing, walking, cycling, and driving a car. Thus, cerebral visual disorders can affect visually guided activities in different ways and indirectly cause other associated handicaps, including basic daily activities (Warren, 2009). Secondary impairments might influence the outcome of tests that assess cognitive performance; thus, apparent impairments in the visual modality might be detected as a result of impaired cognitive performance or exacerbated cognitive deficits (Uzzell et al., 1988). Visual-field loss can affect performance in tasks that require global perception (e.g., visual problem solving in the Raven Standard Progressive Matrices (Raven SPM), colour sorting in the Farnsworth–Munsell (FM) 100-hue test; Zihl, Roth, Kerkhoff, & Heywood, 1988); moreover, reduced visual acuity and contrast sensitivity may affect facial recognition performance, judgement of line orientation, and visual form discrimination (Kempen, Kritchevsky, & Feldman, 1994; Skeel, Schutte, Van Voorst, & Nagra, 2006). In particular, visual spatial dysfunction may impair figure drawing and block construction. Of course, the outcome in tests measuring attention (Bruce, Bruce, & Arnett, 2007), visual working and long-term memory, cognitive flexibility, naming of colours and objects, etc., can be affected if the visual capacities required to perform such tests are inadequate.

Similar “secondary” effects of visual disorders may also influence a broad range of rehabilitation activities, in particular in the treatment of motor disorders in occupational and motor therapy and of language disorders in speech therapy. Many motor activities are visually guided, and visual guidance is known to be helpful for regaining performance (e.g., manipulation of objects, tool use, body posture control and walking).
Rehabilitation of cerebral visual disorders:
Some methodological considerations

Neurorehabilitation is the process of restoring the fullest possible degree level of functioning, and thereby of independence, of an individual who has suffered brain injury. It involves providing treatment or training and support, to enable such an individual to regain skills and abilities to compensate for the functional loss and to cope with everyday-life challenges.

It is a common popular belief that neuropsychological rehabilitation implies the return of the function that is impaired after acquired brain injury. Patients with cerebral visual disorders want to see “normally” again, i.e., to see as they did before brain injury. Thus, in common sense, recovery of function means the restitution of this function. It is sometimes very difficult to explain to patients and their relatives that functional improvement can be achieved by different means. To use Poppelreuter’s words, it is the outcome that counts, i.e., that the patient is again able to successfully cope with everyday life activities such as navigating without difficulties in less familiar or new surroundings, reading a newspaper, comprehending numbers with many digits, grasping objects and recognising faces (Poppelreuter, 1917/1991).

Interestingly, in a recent review on evidence-based cognitive rehabilitation (Cicerone et al., 2005), the only study that was considered in the discussion of visual rehabilitation investigated a restorative treatment method. Studies evaluating compensatory treatment methods were not mentioned, which is indicative of a certain absence of awareness of such approaches in the field of neuropsychological rehabilitation.

Rehabilitation measures aim at alleviating disability in the personal context as well as in the physical and social environment rather than simply aiming at reducing impairment (Wade, 2005). Improving the functional status in everyday life conditions by reducing the degree of handicap and increasing the degree of independency is crucial for regaining self-regulating autonomy (Ryan, Kuhl, & Deci, 1997), which significantly improves life quality. There are different means of achieving this goal. The affected function may recover, partially or totally, or may be substituted by another function, or the environment may be adapted to the patient’s disability and technical aids may be introduced to facilitate everyday life activities. Thus, evaluating rehabilitation measures involves the assessment of their ecological value and validity. The term “recovery of function” is neutral. Its meaning includes every form of return of a function or capacity, irrespective of the underlying processes or applied means. For example, the function “global perception” (or “visual overview”) may recover because vision returns in a previously blind.
field region, or because oculomotor scanning routines are learnt which successfully substitute the irreversible visual field loss. The patient benefits equally in both cases, although the former condition may be seen as more natural and the patient may therefore prefer and insist on measures that allow him to regain his full visual field. At the behavioural level, however, the patient may not be aware of the difference since he is able to quickly gain a complete overview of a scene as in the “old days”, as one patient with a complete left-sided hemianopia put it. He was convinced that his left visual hemifield reappeared after he had acquired a highly efficient compensation strategy after systematic practice with oculomotor scanning. Even objective performance measures in scanning or cancellation tests may not reveal any substantial differences between the two rehabilitation conditions. What is important, however, is that the brain is capable of “developing” and optimising a particular function in different ways. This is of particular significance in patients with acquired brain injury where the normal status is no longer available. This capacity depends on the interactions and synergisms in and between functional systems, e.g., the visual system, the oculomotor system, the attentional system and the executive system, and has been coined functional “equifinality” (Gottlieb, 2001). Restitution or restoration of a function should therefore be defined unequivocally and not be used for any kind of “functional amelioration”. At the behavioural level it appears rather difficult to use these terms unequivocally since even elementary functions such as the visual field or contrast vision are modulated by cognitive factors, in particular attention and learning (e.g., Russell, Malhotra, & Husain, 2004; Seitz & Watanabe, 2005). Any functional improvement can therefore at least partly be explained by the involvement of additional cognitive factors. The terms recovery of function and remediation appear more neutral but they do not indicate the underlying processes or the particular treatment rationale. To avoid semantic ambiguities, reports on practice-related functional improvement should always include a clear indication of the main means of treatment, i.e., restitution/restoration, functional substitution/compensation, or substitution by technical or other aids.

There is still an ongoing (and possibly never-ending) debate about the optimal as well as minimal methodological design for neuropsychological intervention that fulfils the scientific criteria for evidence-based recommendations. Undoubtedly, acquiring strict scientific evidence for the efficacy of a given training method is an important first step, which requires the exclusion or at least a fair consideration of non-treatment factors that may also significantly contribute to the treatment outcome. However, as Cicerone et al. (2005) correctly pointed out, we should move beyond the “simple” question whether and to what extent treatment measures are effective, and include the impact of treatment effects on functional activities, i.e., reduction of disabilities, as well as the examination of patient and therapeutic setting characteristics that may optimise (or diminish) the clinical outcomes of neuropsychological rehabilitation. We also agree with Coltheart, Brunsdon,
and Nickels (2005) that (cognitive-) neuropsychological rehabilitation should consider normal models of functioning for the development of assessment and specific treatment methods. Yet, this view needs to be complemented with the additional consideration of our understanding of pathological information processing and cognitive functions. We call for a reciprocal consideration of normal and pathological evidence since they represent two sides of the same coin. Understanding human brain–behaviour relationships should also include pathological conditions and it is also essential to use scientifically proven evidence on visual perception, visual perceptual learning, visual cognitive functions, and visually guided behaviour in the context of the personal, physical and social environment.

In search of evidence for the efficacy of treatment

It is widely agreed that the rehabilitation of cerebral visual disorders is of great importance since vision represents probably the most important sensory system in humans and is required for the guidance and control of a vast variety of motor activities. Thus, understanding and accurately identifying visual deficits as well as implementing specific treatment strategies are essential to maximise functional independence and self-regulation of patients (Anderson & Rizzo, 1995; Raymond, Bennett, Malia, & Bewick, 1996; Ryan et al., 1997). Yet, accepting training methods as rehabilitation measures requires us to determine their efficacy and functional impact. Figure 1.2 shows examples of possible treatment designs for neuropsychological rehabilitation. Design A is a more traditional design that involves comparing pre-treatment performance between the experimental group (EG) and the control group (CG). After treatment of EG (CG remains untreated), the performance of both groups is assessed and compared again. The performance difference between EG and CG indicates the treatment effect. Thus, CG serves as a “placebo” group. Design B represents a more pragmatic approach, which can be more easily applied in clinical rehabilitation settings. After patients’ initial assessment, a waiting period is introduced that lasts for several weeks and therefore allows controlling for spontaneous recovery or adaptation. At the end of the waiting period, a follow-up assessment is performed, which is also used as a pre-treatment performance measure. The treatment effect is assessed using a second follow-up measurement at the end of the training period. Comparing the changes in the respective visual function/capacity during the pre- and post-treatment waiting periods with those occurring during the training period allows valid determination of whether the treatment was effective, i.e., whether the changes in the respective visual function/capacity are significantly larger after the training period than after the waiting period. Design C represents a cross-over design and is appropriate when two (or more) visual functions/capacities are treated. For example, patients with homonymous hemianopia usually require both practice with oculomotor scanning and reading (see Chapter 3, p. 29). After a pre-treatment waiting
period, patients are randomly allocated to the scanning training group (1) or to the reading training group (2). After this training period, patients of both groups receive treatment for the other, hitherto untreated, visual function/capacity. If the treatment effects are specific, i.e., if the main significant improvement can only be found for the specifically addressed function/capacity (i.e., scanning or reading) after the respective training periods, we can reasonably conclude that specific practice is required for improvement and that practising visual activities in an unspecific (although systematic) manner is not sufficient. This design presupposes that the addressed capacities/functions are independent entities; otherwise a transfer effect would be expected. Conducting another follow-up assessment after treatment (e.g., after 6–8 weeks) is helpful to assess the stability and long-term effects of the treatment. Consistent with the recommendations of Cicerone et al. (2005), functional capacities in terms of activities of daily living (ADLs) or instrumental activities of daily living (IADLs) should be assessed before and after treatment as
well as at follow-up to examine the transfer and generalisation of treatment effects to daily living and/or occupational conditions. To minimise the effect of non-treatment factors and to increase transfer and generalisability, patient groups should be as homogenous as possible, also in terms of non-cognitive factors. Yet, the exact effect of such non-treatment factors is still unclear and remains to be investigated.

The design of the studies presented in this monograph is of types B and C (Figure 1.2). The minimum time interval between the initial assessment and beginning of treatment was at least 4 weeks (range: 4–192 weeks) to control for spontaneous recovery, or because of patients’ (or our) time constraints. In the majority of cases, waiting periods were longer than the training period. This design allows for the differentiation of treatment from non-treatment effects. However, it does not fulfil the criteria for a placebo-controlled clinical evaluation, because no controlled placebo treatment was administered in the pre-treatment waiting period and patients were asked to continue their “normal” everyday life activities. Pre- and post-treatment measures were conducted blind to intervention. There is no doubt that from a strict methodological point of view design type A would offer the highest level of evidence-based rehabilitation. However, it was not possible to use this type of design because neither national nor private insurance companies would have agreed to pay for a “placebo” treatment, and of course the “placebo” treatment can definitively not be declared as a rehabilitation measure. Because the “placebo” treatment also requires resources and thus produces costs, somebody would have to pay for it, either the patient or a funding institution.

The major challenge in the future of neurorehabilitation is a closer collaboration and co-operation between basic and applied neuroscience, and the improvement in the interaction between patients, clinicians, and researchers, so that all parties can differentiate their special priorities, skills, and concerns. Interactions of this sort hopefully create a research-friendly and patient-oriented, rather than only target-driven frame in rehabilitation research and services (Cumberland Consensus Working Group, 2009).

**About this monograph**

This monograph deals with the rehabilitation of visual deficits after acquired brain injury. Chapter 2 introduces the neurobiological foundations of visual perception. Each of the following six chapters (3 to 8) is dedicated to one of the major cerebral visual disorders and discusses their main features and frequencies of occurrence, their aetiologies and natural course (spontaneous recovery or adaptation) as well as assessment and treatment methods. Each chapter is introduced by a definition of the visual disorder as well as by a brief discussion of the significance of the respective visual capacity for human behaviour. This introductory section is followed by a description of the respective deficit(s) or disorder(s), their functional consequences for the patient in terms of disability and handicap, and observations on spontaneous
recovery. Subsequently, a detailed analysis of the rationale and outcome of treatment evaluations is provided. At the end of each chapter, a summary of assessment and practice measures and methods and suggestions for diagnostic and therapeutic procedures are presented. Before testing a patient with suspected cerebral visual disorders, a detailed neuro-ophthalmological examination is highly recommended to assess dysfunction of the peripheral visual system causing, for example, field defects and impaired colour vision, and of eye-movement abnormalities (e.g., oculomotor nerve palsies, defective accommodation). For the assessment of practice effects, all affected visual functions and abilities (see Appendix, Diagnostics, p. 225) should be re-tested. Pre–post comparisons allow the differentiation between specific (only the function or ability which was subjected to treatment shows an improvement) and unspecific treatment effects. The difficulty of tasks selected for practice should be tailor-made to the patient, i.e., adapted to the initial/actual level of the individual resources (e.g., attention, motivation). It may be very helpful for both the patient and the therapist to know the plan of treatment before the beginning of practice, and the (individual) steps as well as their significance with regard to the final goal of intervention. Appropriate feedback to the patient about the actual level of performance is also recommended.

For some visual disorders, results of larger patient samples were available; for others, evidence for the efficacy of systematic practice is still based on single cases. The majority of studies published since the first edition in 2000 have dealt with the rehabilitation of homonymous visual field disorders. Since visual field disorders are the most frequent cerebral visual disorders, the corresponding chapter is the longest of the monograph. Central visual field defects (central scotoma) are dealt with in a separate chapter. Although this visual field disorder is a rare condition, patients with a central scotoma typically show a combination of severe visual deficits and therefore need special rehabilitation measures. For other cerebral disorders such as impaired contrast sensitivity, achromatopsia (impaired colour vision), the Balint syndrome or visual agnosia, clinical treatment evidence is based on detailed single case studies. The observations reported in this monograph are nevertheless of heuristic value. They are helpful in developing and improving rehabilitation measures and will hopefully stimulate further research.

The recording of eye movements was a very useful tool in assessing a patient’s adaptation and compensation behaviour in global perception and scanning and searching tasks as well as in reading. In addition, it allowed the precise measurement of saccadic localisation performance and fixation accuracy in, for example, patients with visual-spatial deficits, Balint syndrome or central scotoma. Furthermore, analysing eye movements before and after treatment increased our understanding of the adaptive processes underlying the observed improvements. Eye movements can therefore be used to objectively assess practice effects. Interestingly, patients had a great interest in their eye-movement patterns in reading and scanning. Patients were surprised to

learn how laborious and time consuming their eye-movement pattern was before treatment, and how regular it became after training. The demonstration of the eye-movement recordings was helpful in explaining their individual disorder to patients as well as the need for and rationale of treatment. Moreover, it may also be useful to increase patients’ insight into their visual deficits, their treatment motivation and their compliance in following the recommended use of learned strategies in daily life activities. It was a great concern (as well as a challenge) for us to make all patients experts in their visual difficulties.

All patients reported in this monograph gave their informed written consent in accordance with the Declaration of Helsinki. They all received a detailed ophthalmologic and neurological examination before assessment and treatment. Patients with additional disturbances in the anterior visual pathways or of the oculomotor system, including vergence and accommodation, were excluded. Patients with severe deficits in memory, executive functioning, language, alexia or visual neglect were also excluded. For the assessment of cognitive functioning, the following tests were used: verbal short-term and working-memory spans, Auditory Verbal Learning Test (AVLT), logical memory, Token Test, verbal fluency (see Lezak, Howieson, & Loring, 2004, for details). For the diagnosis of visual neglect we used the criteria proposed by Halligan, Cockburn, and Wilson (1991). If, particularly in single case studies, descriptive statistics revealed sufficiently clear results, further statistical analyses are not reported since they did not add further information. Thus, in these cases with less complex data sets, we followed the old-fashioned saying that, “What you cannot see with the eyes, you cannot understand with statistics”.

Since this monograph is not intended to provide a meta-analysis of evidence-based treatment methods in the rehabilitation of patients with cerebral visual disorders, the studies cited have not been systematically evaluated in terms of their weaknesses and strengths. Rather, I have tried to present the positive evidence available but, at the same time, give my personal view about their utility as guidelines in practice and research; that also applies to my own studies. This approach reflects the fact that most studies on rehabilitation of patients suffering from cerebral visual disorders include only a small number of subjects. Accordingly, a strategy with general validity for treating patients with similar visual disorders cannot be recommended. Although I would personally like to suggest treatments based on evidence of greatest efficacy, the empirical evidence available does not allow that unless further research with larger numbers of subjects is undertaken.
The primate visual brain is organised in modules and consists of many distinct visual areas, and thus processing systems, which act in parallel. The primary visual cortical area (striate cortex, Brodmann’s area 17, visual area 1 or V1) is central to the visual brain. It receives its input from the retina via the lateral geniculate body (LGN) and possesses a highly accurate, topographically organised representation of the retina and thus of the visual field. The central visual field occupies a large proportion of the striate cortex; about half of the cortical surface is devoted to the central 10° of the visual field, which is only 1% of the visual field (Tootell, Hadjikhani, Mendola, Marrett, & Dale, 1998). V1 also distributes specific visual signals to the other visual areas, which are located in the surrounding cortex (see Bullier, 2003, for a review). This anatomical and functional organisation enables the visual brain to deal with the processing of global and local features of a visual scene. The result of processing at distinct levels of complexity at each stage can be flexibly and dynamically integrated into time- and space-coherent perception (Bartels & Zeki, 1998; Rainer & Logothetis, 2003; Tootell et al., 1998; Zeki, 1993; Zeki & Bartels, 1998).

Ungerleider and Mishkin (1982) have characterised the functional specialisation of the visual brain as consisting of two processing streams: The “where” or dorsal route, comprising occipito-parietal visual areas and connections, is specialised in space processing; the “what” or ventral route, comprising occipito-temporal visual areas and connections, is specialised in object processing (see Figure 2.1). A different view has been offered by Milner and Goodale (2006, 2008), who argued that information processed in the dorsal pathway is used for the implicit or unconscious visual guidance of actions while processing in the ventral stream is associated with conscious perception. Spatial and object information processing, as for instance in scene perception, requires co-operation and interaction between the various components of a distributed network consisting of the dorsal and ventral visual processing streams, the posterior parietal cortex, and prefrontal cortical areas (Corbetta et al., 1998; Saron, Schroeder, Foxe, & Vaughan, 2001). Both routes interact either directly or indirectly via attention involving the inferior parietal cortex (Singh-Curry & Husain, 2009) and working memory.
Figure 2.1 (A) Schematic drawing of the visual pathway from the retina to the striate cortex. BA 17: Brodmann area 17; V1: visual area 1. Note decussating of optic fibres at the optic chiasm. Injury to the left optic tract, optic radiation, or striate cortex causes right-sided homonymous visual field defects; injury to the right optic tract, optic radiation, or striate cortex causes left-sided homonymous visual field defects. (B) Schematic drawing of the dorsal (occipito-parietal) and ventral (occipito-temporal) cortical visual processing routes.
involving the prefrontal cortex (Goodale & Westwood, 2004; Oliveri et al., 2001). Spatial updating or remapping allows the brain to compensate for shifts in the retinal image resulting from saccades, which ensures coherent global perception (Merriam, Genovese, & Colby, 2007). Posterior parietal (Constantinidis, 2006), posterior cingulate (Dean, Crowley, & Platt, 2004), and prefrontal cortical areas (Barcelo, Suwazono, & Knight, 2000; Olson, Gettner, Ventura, Carta, & Kass, 2000) build an interactive network for the control of attention in visual space. Motivation and attention also contribute independently to orienting in space (Bendiksby & Platt, 2006); prefrontal mechanisms underlie the motivational modulation of (oculo-)motor activities in space (Roesch & Olson, 2003, 2004). Similar structures are involved in the visual guidance and control of eye movements, including scanning (Kennard, 2002; Lynch & Tian, 2006; Pierrot-Deseilligny, Muri, Ploner, Gaymard, & Rivaud-Pechoux, 2003; Schall & Boucher, 2007; Schiller & Tehovnik, 2001). Experience-dependent visual scanning requires hippocampal-dependent memory (Smith & Squire, 2008).

Eye movements play a crucial role in visual processing and thus in visual perception (see Martinez-Conde, Macknik, & Hubel, 2004, for a comprehensive review). Pflugshaupt et al. (2009) demonstrated that the gaze pattern during visual search reflects the functional specialisation of the visual field, with a clear bias of fixations in the centre of the field of view (or scene). The posterior thalamus and its reciprocal connections with cortical regions in the occipital, parietal and frontal lobes and with the limbic neocortex are assumed to form a cortical–subcortical network subserving attention as well as the intentionally guided and externally triggered saccadic movements that are involved in visual information processing (Andersson, Joliot, Perchey, & Petit, 2007; Corbetta, Miezin, Shulman, & Petersen, 1993; Dean et al., 2004; Himmelbach, Erb, & Karnath, 2006; Kustov & Robinson, 1996; Nobre, 2001; Olson et al., 2000; Pierrot-Deseilligny et al., 2003; Robinson, 1993; Robinson & Petersen, 1992; Schiller & Tehovnik, 2001, 2005; Selemon & Goldman-Rakic, 1988). The representation of objects and faces also requires the function of the extrastriate regions in the ventral visual pathway (Grill-Spector, 2003) as well as the top-down processes involving the prefrontal cortex that facilitate visual object recognition (Bar, 2003). Furthermore, there exists empirical evidence for category-specific processing of visual stimuli in humans (Ishai, Ungerleider, Martin, Shouten, & Haxby, 1999; Sigala, 2004; Wierenga et al., 2009). Yet, it is still unclear how the brain eventually computes and codes visual objects for accurate identification and recognition. Objects are selectively processed in a viewpoint- and size-dependent manner at intermediate cortical stages whereas higher order areas in lateral occipital and posterior parietal cortex are involved in object processing independent of image transformation. Thus, visual object information seems to be simultaneously represented in two parallel and hierarchically organised processing systems in the ventral and dorsal visual pathways (Konen & Kastner, 2008).
Visual processing is active, i.e., it is selective and flexible, and depends on the particular requirements of a task. Global perception allows continuous updating of the main spatial characteristics of a scene, which can serve as spatially and temporally coherent background information for visual selection and guiding fixation shifts. Local or selective processing does not generate a complete representation of a visual scene; it generates partial representations of aspects of a scene, which are task-relevant at a given time (Mather, 2006; see Figure 2.2). Selective visual disorders after brain injury and the specificity of deficits support the model of functional specialisation of the visual brain, albeit focal lesions are the exception rather than the rule (see Table 2.1). Patients with injury to the visual brain usually present an association of visual symptoms. These are caused by impairments at different levels of processing resulting from injury to visual cortical areas and fibre connections. Despite its significance, the aspect of communication between brain structures remains to be fully incorporated into neuropsychological models of brain functioning. New brain imaging methods such as diffusion tensor imaging (Bosnell, Giorgio, & Johansen-Berg, 2008; Guye, Bartolomei, &

![Figure 2.2](A) Schematic drawing of the pathways involved in overview (“vision at a glance”) and global visual perception. The first main processing stage is the posterior parietal cortex (PPCx), which builds a mental representation of the coarse spatial structure of a scene. This coarse spatial representation can serve as basis for fine, detailed processing, and is used by visual cortical areas for local processing as well as by prefrontal structures to guide and supervise attention and oculomotor activities (broken lines). (B) Schematic drawing of the “first” (Retina ➔ LGB ➔ V1 ➔ PPCx and OTCx) and “second” visual system (Retina ➔ CS ➔ Pulvinar ➔ PPCx). LGB: lateral geniculate body (corpus geniculatum laterale; CGL); V1: visual area 1; OTCx: occipito-temporal cortex; CS: colliculus superior; Pulvinar: nucleus pulvinar (posterior thalamus); PPCx: posterior parietal cortex.
Ranjeva, 2008) allow an accurate analysis of the white matter, and will help to further elucidate the important role of the connections within the visual brain and other brain structures involved in vision as well as in brain plasticity after brain injury (Carey & Seitz, 2007).

<table>
<thead>
<tr>
<th>Deficit/disorder</th>
<th>Sites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Homonymous visual field disorders</td>
<td>Postchiasmatic visual pathway (optic tract, lateral geniculate body, optic radiation, striate cortex)</td>
</tr>
<tr>
<td>Visual acuity</td>
<td>Postchiasmatic visual pathway (foveal projection; bilateral)</td>
</tr>
<tr>
<td>Spatial contrast sensitivity</td>
<td>Postchiasmatic visual pathway; occipito-temporal cortical areas</td>
</tr>
<tr>
<td>Visual adaptation</td>
<td>Postchiasmatic visual pathway</td>
</tr>
<tr>
<td>Dyschromatopsia, achromatopsia</td>
<td>Posterior regions of medial and lateral occipital gyri (lingual and fusiform gyri)</td>
</tr>
<tr>
<td>Visual spatial functions</td>
<td>(Left- and) right-sided parieto-occipital areas</td>
</tr>
<tr>
<td>Balint syndrome</td>
<td>Bilateral posterior parietal regions, including parieto-frontal fibre connections (white matter)</td>
</tr>
<tr>
<td>Visual agnosia</td>
<td>(Uni- or bilateral) occipito-temporal injury</td>
</tr>
</tbody>
</table>
Box 3.2 Does type of parafoveal field loss matter for practice with reading?

Case 1  Mr T., 47 years old; right-handed
Left-sided posterior cerebral artery infarction; time since brain injury: 14 weeks
Right-sided hemianopia (visual field sparing: 1°)
Hemianopic dyslexia, but no (pure) alexia
Practice with reading

Case 2  Mr H., 51 years old; right-handed
Left-sided posterior cerebral artery infarction; time since brain injury: 15 weeks
Right-sided paracentral scotoma (visual field sparing: 1°)
Hemianopic dyslexia, but no (pure) alexia
Practice with reading

Practice with reading

<table>
<thead>
<tr>
<th>Session</th>
<th>1</th>
<th>3</th>
<th>5</th>
<th>7</th>
<th>9</th>
<th>11</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>37/43</td>
<td>49/66</td>
<td>67/77</td>
<td>73/87</td>
<td>83/87</td>
<td>87/97</td>
</tr>
<tr>
<td>Case 2</td>
<td>43/50</td>
<td>57/70</td>
<td>73/87</td>
<td>83/90</td>
<td>90/97</td>
<td>—</td>
</tr>
</tbody>
</table>

Notes: Presentation time: 400 ms; 11 sessions in case 1, 9 sessions in case 2. Numbers refer to percentage of correct responses at the beginning and the end of each session (n. n = 20 trials).

Reading performance (in words per minute) before and after practice, and at follow-up

<table>
<thead>
<tr>
<th></th>
<th>Before</th>
<th>After</th>
<th>Follow-up</th>
<th>Cut-off</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>106</td>
<td>157</td>
<td>176</td>
<td>180</td>
</tr>
<tr>
<td>Case 2</td>
<td>124</td>
<td>168</td>
<td>179</td>
<td>180</td>
</tr>
</tbody>
</table>

Notes: Follow-up 8 weeks post practice.

Comment

Type of parafoveal visual field loss, i.e., homonymous hemianopia or paracentral scotoma, seems not to affect reading performance essentially, although reading performance before practice was lower in the subject with homonymous hemianopia. Improvement after practice was, however, comparable in both cases after a similar number of training sessions, as was reading performance at follow-up. At follow-up, both patients had nearly reached the respective cut-off scores. Note that text processing improved mainly in and not between training sessions.