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CASE REPORT

COMMUNICATING USING THE EYES WITHOUT REMEMBERING IT: COGNITIVE REHABILITATION IN A SEVERELY BRAIN-INJURED PATIENT WITH AMNESIA, TETRAPLEGIA AND ANARTHRIA

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We describe here a case of cognitive rehabilitation in a young patient with closed head injury, who had dense anterograde amnesia and such disabling neurological defects (tetraplegia and anarthria) that the condition evoked some features of an incomplete locked-in syndrome. After a prolonged period of no communicative possibility, the patient underwent a specific training, based on principles of errorless learning, with the aim of using a computerized eye-tracker system. Although, due to memory disturbances, the patient always denied ever having used the eye-tracker system, learned to use the computerized device and improved interaction with the environment. This favourable outcome may serve as a stimulus for devising new training approaches in patients with complex patterns of cognitive impairments, even when associated with severe motor impairments.

Key words: traumatic brain injury, amnesia, locked-in syndrome, eye-tracker, cognitive rehabilitation.

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INTRODUCTION

Severe traumatic brain injury (TBI) usually determines diffuse axonal damage, with volume loss of both grey and white matter, mainly in the prefrontal regions (1), possibly associated with discrete brain lesions. As a consequence, when patients with TBI regain consciousness they generally show attention, memory, and executive impairments (2), as well as severe motor impairments if descending motor pathways are disconnected. In particular, traumatic (or secondary vascular) brainstem lesions may cause tetraplegia and anarthria, thus generating a clinical picture resembling locked-in syndrome (LIS) (3). Patients with LIS, affected by a selective lesion in the ventral brainstem (typically in the pons), show preserved consciousness, and unimpaired linguistic as well as intellectual and emotional abilities, but can only perform vertical eye movements (4–5). Some patients with LIS may present other residual voluntary movements besides vertical eye movements, and can be thus classified as affected by "incomplete" LIS (4), but they nonetheless experience extreme difficulties in communicating. For this reason patients with LIS can actively interact with the environment only by means of complex communicative systems, based for instance on brain-computer interface devices (6).

Patients with TBI with extremely severe motor disturbances (tetraplegia and anarthria) face the same difficulties as patients with complete or incomplete LIS, but, in contrast to patients with LIS, they show the typical cognitive defects observed after a TBI. Therefore, any rehabilitative treatment in such patients will be difficult and discouraging, even when it is aimed only at restoring interaction with the environment.

We describe here a young male patient affected by a severe pathological combination (TBI with extremely severe motor disturbances and cognitive impairment) in whom, after a prolonged period of no communicative possibility, a specific training to use a computerized eye-tracker system improved his interaction with the environment. Although the patient learned to use the eye-tracker system, he always denied ever having used it, due to his memory disturbances.

CASE REPORT

A 27-year-old right-handed employed male graduate sustained a severe closed head injury in a road accident. The patient was in severe coma (Glasgow Coma Scale < 8) for approximately one month, presenting repeated episodes of paroxysmal autonomic and respiratory insufficiency, but eventually he opened his eyes and started to present roving ocular movements. Ten months after the accident, he was judged to be in a vegetative state (Glasgow Outcome Scale-Extended (7), GOS-E = 2; Disability Rating Scale (8), DRS = 27).

Immediately after the trauma computerized tomography (CT) scanning revealed multiple micro-haemorrhagic lesions in cortical and subcortical areas of both hemispheres and in the brainstem. Subsequently, serial CT scans documented diffuse bilateral cortical and subcortical atrophy. Two years after onset, a magnetic resonance imaging (MRI) scan confirmed the presence of diffuse bilateral atrophy involving the cortex (particularly the enthorinal cortex), the corpus callosum and all sections of the brainstem; diffuse white-matter hyperintensities, mainly in the left fronto-parietal region, were also observed.

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When the patient was admitted to our rehabilitation department he was still in a vegetative state. One year after onset inconsistent but reproducible ocular movements towards acoustic or visual stimuli began to be observed, and the patient was diagnosed to be in minimally conscious state (9). Sixteen months after onset, the patient was able to direct his head and gaze, although for brief periods of time, towards people calling him by name or towards people entering his room, but still presented spastic tetraplegia and anarthria (GOS-E = 3; DRS = 25). From that time on, the patient managed to use an eye-code communication system, with eyes wide open to mean affirmative responses and eyes shut to express negative responses. The patient's ocular responses were not sufficiently consistent to be able to adopt a double-checked agreed system of interpretation (10), but the patient proved able to understand simple auditory and written verbal commands (e.g. to look at specific objects or towards specific directions, to open or close his eyes). However, he was densely amnesic: he could recognize his close relatives (parents and brothers) but not friends, could not recall relatives' ages or occupations and could not answer questions about everyday events or his past history. As far as episodic memory could be tested by means of the eye-code system, retrograde amnesia for at least the past 5 years and profound anterograde amnesia were detected.

Eighteen months after onset, the patient performed conjugated eye movements in all directions and could make small lateral movements of his head, but he breathed through a tracheostomy tube, was dysphagic, amimic, tetraplegic, anarthric, and densely amnesic; he also showed bursts of pathological laughing and crying (GOS-E = 3; DRS = 24).

Since the eye-code system did not allow the patient to communicate his feelings or needs, we decided to start a rehabilitative training for enhancing his communicative skills by means of an eye-tracking system. Because of the concomitant memory defects, we adopted an errorless learning procedure (11).

Rehabilitative training

As a first step, we verified whether the patient could interact with a computerized infra-red eye-tracking system (MyTobii, Tobii Technology, Danderyd, Sweden). To calibrate the system onto patient's eye movements we asked him to pursue a slowly-moving coloured circle on the monitor with his eyes and to fix his eyes on it whenever it stopped and flashed (5 locations tested per trial). On the first day, patient's attempts to follow and fix on the target ranged from 2–3 correct performances (mean correct fixations: 2.3/5, over 20 trials); meaning that he could not use the computerized eye-tracking system successfully in a 40-min session. The procedure was repeated on the following days, and each time before starting the patient was asked whether he had ever used an eye-tracker and whether he had previously met the psychologist administrating the eye fixation procedure. Using the usual eye-response code the patient replied negatively to both questions. Nonetheless, he proved able to fix on 3–5 points correctly during the calibration procedure (mean correct fixations: 3.8/5, over 20 trials). On this basis it was possible to start using simple communication software based on fixation of 2 regions of the monitor to mean positive or negative responses. The patient was then required to use the yes/no eye-tracking response procedure to answer basic 2-choice questions assessing object and colour recognition (e.g. the examiner presented a pen and the patient was asked "is this a pen?" or "is this a key?"; 20 questions were given for each task with a correct/wrong ratio of 50%). The patient was 85% and 90% correct with objects and colours, respectively; a performance very similar to that obtained using the eye-response code. From the fifth session on, the patient performed the calibration procedure with only occasional errors (mean correct fixations: 4.8/5, over 20 trials), and practised the infra-red eye-tracking system for yes/no questions. However, after 6 sessions he still denied ever having used or even seen the monitor with the infra-red devices, or ever having met the examiner.

The encouraging observations about the patient's steady improvement in using the eye-tracking system (despite his dense anterograde amnesia) led us to plan a rehabilitation programme with 2 main aims: (*i*) to teach the patient to use a simple eye-commanded writing software (based on an enlarged keyboard shown on the monitor); and (*ii*) to make him autonomous in using the eye-tracking system, i.e. in launching and shutting down the desired software (by navigating through simplified directories), and in shifting among available computer programs (writing software; augmentative communication software – a collection of simple screenshots showing symbols for basic needs and desires; 2 simple games based on visual perceptual matching and on visual short-term memory).

To achieve these goals, a 2-month training programme was planned, with 3 40-min sessions per week. Each session was divided into 2 main parts: in the first we administered copying and writing upon dictation tasks (with letters, two-letter syllables, and words) and word completion tasks (incomplete words were shown and the patient had to identify the missing letter); in all these tasks the patient had to fixate his eyes on the target letter on the screen. These exercises were intended to stimulate the patient's ability to manipulate phonological and graphemic representations, to scan the monitor systematically, and to attend to prolonged tasks.

The second part of each session was aimed specifically at teaching basic instructions for using a personal computer (PC) via the eye-tracker system. The patient had already used a PC in his premorbid life for simple leisure activities, but, prompted by specific questions, he could not recall any relevant information. The rehabilitative training aimed to teach sequential commands for efficient use of the PC: after repeated demonstration of commands, the patient was required to repeat the same steps by fixating his eyes on the appropriate boxes on the monitor. In the recall phase, we strictly followed an errorless procedure, by which errors in patients' responses are minimized (11).

RESULTS

During the training period the patient gradually improved his performances, but with very different levels of efficiency in writing and in computer-related tasks.

In most writing tasks the patient clearly improved (Table I), but he never succeeded in writing a single word correctly upon dictation, whereas he found copying relatively easier. However, a relevant change in error type was observed: at the baseline most errors were perseverations (50% of errors), unrelated responses (30%) and graphemic errors (20%), whereas at the end of the training perseverations (15%) and unrelated responses (5%) had drastically reduced, and most errors were graphemic in nature (omissions, insertions, transpositions or substitutions of letters).

A favourable outcome was achieved in computer usage. The patient became autonomous in using the eye-tracker to launch the calibration software spontaneously and on request, to navigate through directories, to find the desired software, and to shift between different programs. The formalized assessment (Table I) revealed that the patient became fully efficient after a few sessions and progressively required less time and effort to use the system. However, during an informal debriefing at the end of the training, the patient still denied being able to use, or even having ever seen, an eye-tracking system, and also did not recognize the psychologist who had assisted him during the rehabilitative programme.

DISCUSSION

The young patient described here was affected by a combination of severe pathological conditions, such that dense anterograde amnesia and selective cognitive impairments were associated with extremely severe motor impairments. At the end of the rehabilitation programme the patient made frequent graphemic errors in copying and in writing to dictation, with a clear length-effect, a pattern consistent with diagnosis of a specific writing impairment (defect of the graphemic output buffer) (12), while reading and auditory verbal comprehension were spared, at least as far as could be assessed by informal testing procedures. Other cognitive tasks, such as object recognition or short-term retention of visuospatial information, were performed without relevant difficulties, but the systematic neuropsychological assessment was beyond the scope of the present study.

Table I. *Patient's performance in the training tasks before, during and after the rehabilitative programme*

	Baseline Correct/ total	First month Correct/ total	End treatment Correct/ total
Writing			
Writing letters to dictation	2/20	14/20	18/20
Writing two-letter syllables to dictation	1/20	7/20	14/20
Writing words to dictation	0/20	0/20	0/20
Copying letters	6/20	18/20	20/20
Copying two-letter syllables	4/20	8/20	13/20
Copying words	0/20	1/20	2/20
Word completion	4/20	13/20	15/20
Computer usage			
Launching programs	0/5	2/5	4/5
Program shifting	1/5	2/5	4/5
Keyboard functions usage	1/5	2/5	5/5

Our main aim was to verify whether a specific training programme could improve the patient's communicative skills by means of a computerized eye-tracker, despite the dramatic defect of anterograde episodic memory. For this purpose we adopted an errorless training procedure that is thought to rely on implicit memory (13) and to be the most effective rehabilitative method in amnesic patients (14). At the end of the training the patient had learned to use the eye-tracker system, and had achieved sufficient skills to launch and use the computer device; these results support the usefulness of errorless learning methods. Amnesia in patients with TBI is often related to difficulties in applying efficient strategies in learning or retrieval processes (15); here, we could only demonstrate the relative sparing of implicit memory, thus confirming the possible fractionation of memory subsystems in patients with amnesia.

During the cognitive rehabilitation period, the patient's neurological conditions showed a further trend towards improvement: the patient had recovered some volitional movements of his head and right arm (proximally). However, he still presented anarthria and very severe motor impairments, which prevented efficient communication. This disabling condition was relieved (at least partially) by the eye-tracking system that allowed selection of communicative symbols by eye movements over the screen. The improvements in computer skills (despite persistent anterograde amnesia) were probably made possible by progressive reduction in attention defects (as demonstrated, for instance, by the reduction in perseverative errors in writing tasks), and this could be ascribed partially to the rehabilitative training itself. However, the parallel improvement in neurological conditions did not allow us to make strong inferences about specific effects of the training. On the other hand, this rehabilitative study was not performed in controlled conditions: the patient was tested before and after the treatment, but no multiple baseline observations were collected and no control treatment was attempted. Therefore, we can only claim here that the additional cognitive defects in our patient did not preclude the use of the rehabilitative programme aimed at exploiting his communicative skills.

The present favourable outcome thus serves as a starting point and a stimulus for devising new training approaches in patients with TBI, even when the lesion generates severe motor impairments (a sort of incomplete "locked-in state") hampering interaction with the environment. While patients with "pure" LIS can learn to use novel electronic devices for augmentative communication (6) without extensive practice, thanks to the lack of associated cognitive impairments, we demonstrated here that specific rehabilitative strategies can allow patients with TBI with extremely severe motor defects to achieve a more active role in their life.

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