

Rehabilitation of Executive Functioning After Focal Damage to the Cerebellum

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Executive dysfunction accounts for significant disability in patients with many types of brain injury in many locations. Clinical reports have described impaired executive functioning after damage to the cerebellum, and anatomical and neuroimaging studies have identified the likely basis for this effect: a cortico-ponto-cerebellar network through which the cerebellum is densely connected to areas of frontal cortex. The patterns of executive impairment attributable to cerebellar damage have been extensively described in the past 15 years, but there has been no assessment of the efficacy of rehabilitation in this patient population. Here, the use of a cognitive rehabilitation technique, Goal Management Training, in a patient with persisting executive dysfunction after a right cerebellar hemorrhage is described. The patient made and maintained modest gains on measures of sustained attention, planning, and organization that translated into significant improvement in real-life functioning. This is the first report on the rehabilitation of impaired executive functioning following focal damage to the cerebellum and in the presence of intact frontal cortex.

Key Words: *Neuropsychology—Cognition—Attention—Strategy application—Goal neglect.*

The cerebellum is conventionally associated with motor functions such as coordination and balance, but in the last 2 decades nonmotor effects

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of cerebellar lesions have been recognized, with difficulties in the broad cognitive domains of executive functions and attention. Impaired executive functioning and attention have been demonstrated with a number of tasks such as verbal fluency,¹ planning,² response shifting,³ divided attention,⁴ and spatial attention.⁵

Functional neuroimaging studies have reported cerebellar activations during performance of executive function and attention-based tasks. Many of these studies were initially aimed at identifying prefrontal regions of activation and almost inadvertently demonstrated that when a cognitive task increases activation in prefrontal cortex, parallel activation of contralateral neocerebellar cortex is often observed.⁶⁻⁸ Although these results have been called into question because of the motor or speech output typically required of these tasks, activation of cerebellar cortex independent of motor movements during an attention-shifting task has been demonstrated.⁹ Furthermore, research has reported distinct motor and attention roles in different regions of the cerebellum.⁷

These findings suggest that cognitive impairments broadly described as executive or attentional can result from cerebellar damage and occur in the absence of motor demands; these may be mediated by disruptions to critical frontocerebellar networks.¹⁰⁻¹²

To our knowledge there is no research on the rehabilitation of executive dysfunction following focal damage to the cerebellum. Goal Management Training (GMT)^{13,14} is a rehabilitation approach that relies on verbally mediated, metacognitive strategies to systematically target disorganized behavior resulting from executive and attentional impairments. The effectiveness of a brief GMT intervention has been demonstrated in a randomized experimental probe of participants with traumatic brain injury¹⁴ and healthy elderly adults.^{15,16} The objective of the present study was to assess the efficacy of a validated executive-rehabilitation technique in a patient with executive impairments after focal damage to the cerebellum.

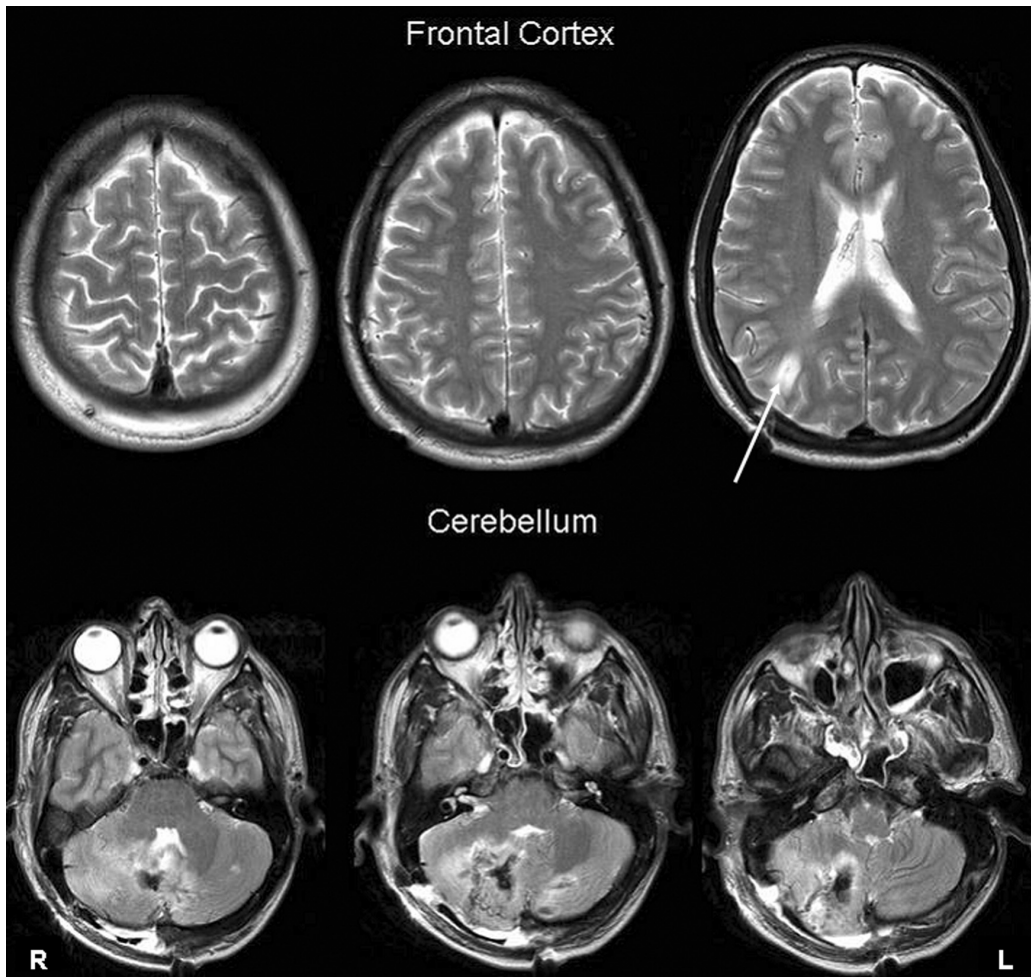


Figure 1. Magnetic resonance imaging (1.5 T) scan taken 27 days after right, arterial venous malformation hemorrhage. Top of figure reveals the intact frontal cortex, and the bottom reveals focal damage to the right hemisphere of the cerebellum (arrow represents entry for surgical evacuation of hemorrhage).

CASE REPORT

The patient is a 41-year-old right-handed male with 21 years of education employed as a high-level bank executive. Previous medical history was unremarkable. In May 2005, he suffered a right-sided, cerebellar arteriovenous malformation (AVM) rupture. Magnetic resonance imaging (1.5 T) data were reviewed by a senior neurologist (M.P.A.) and revealed isolated damage to the cerebellum with no damage to supratentorial areas of cortex (see Figure 1). The ventricles were not enlarged, and no blood was ever in the fourth ventricle.

During his hospitalization at an acute care hospital, the patient's documented difficulties included right-sided dysmetria, dysarthria, poor attention to task, poor self-monitoring, and reduced insight. He was also described as "very perseverative." He was discharged to

a rehabilitation hospital 12 days following surgery, where he underwent 5 weeks of in-patient rehabilitation that focused on improving his walking and speech. At 27 days postrupture, the patient was referred to behavioral neurology for an assessment. The neurological assessment made note of deficits in motor timing, "frontal slowing," and deficits in executive functions. The patient was referred for a detailed neuropsychological assessment 61 days postrupture. His complaints at that time included dizziness triggered by sudden movements, slightly slurred speech, an inability to return to work because of slowed information processing, and difficulties with organization. He denied any problems with his memory or his mood.

Table 1 (top half) summarizes the patient's performance on various neuropsychological measures. The clinical assessor was blinded to the study. The patient showed impaired performance on verbal fluency, which stood in

Table 1. Summary of Performance

Clinical Neuropsychological Testing	Baseline (61 Days Postinjury), Scaled Score ^a	Clinical Follow-up ^b (266 Days Postinjury), Scaled Score	
Fluency			
Verbal	6	—	—
Semantic	13	—	—
Design	9	—	—
Trail Making Test (D-KEFS)			
Motor speed	11	9	9
Letter sequencing	5	10	10
Number sequencing	5	11	11
Letter–number sequencing (switching)	9	10	10
Ruff 2 and 7			
Easy (automatic) search speed	7	—	—
Accuracy	12	—	—
Effortful (controlled) search speed	5	—	—
Accuracy	8	—	—
Rey-Osterrieth Complex Figure			
Immediate recall	17	—	—
Delayed recall (45 min)	17	—	—
Brief Visual Memory Test (BVM-T-R)			
Acquisition	9	—	—
Delayed recall	10	—	—
California Verbal Learning Test (CVLT-II) ^c			
Acquisition	12	17	17
Delayed recall	12	13	13
Semantic clustering	13	18	18
Intrusions ^d	14	13	13
Perseverations ^d	13	10	10
Experimental outcome measures	Baseline (127 days postinjury)	Postrehabilitation (218 days postinjury)	Follow-up (322 days postinjury)
Sustained Attention to Response Task (SART)			
Commission errors	9	3	2
Omission errors	9	4	1
D-KEFS Tower Test			
Total achievement score	19/30	21/30	21/30
Rule violations	2	0	0
Cognitive Failures Questionnaire			
Overall	0	7	8
DEX (Self Assessment)			
Overall	1	2	0
DEX (Significant Other)			
Overall	11	—	0

D-KEFS = Delis-Kaplan Executive Function System; DEX = Dysexecutive Questionnaire.

a. Expected age matched scaled score is 10 (SD = 3). Measures do not correct for education levels.

b. Only CVLT and D-KEFS Trails were repeated during routine clinical neuropsychological follow-up.

c. Alternate forms of the CVLT were used at baseline and follow-up testing.

d. Higher scaled scores with respect to intrusions and perseverations on the CVLT denote greater impairment.

sharp contrast to his spared design fluency. Moreover, a fine-grained analysis of his letter fluency performance indicated reduced switching, defined as the ability to flexibly shift from 1 subcategory to another, an ability that is impaired in patients with frontal lobe lesions.¹⁷ His acquisition and retention of a set of 6 designs and their spatial locations were normal. His acquisition of the word list (California Verbal Learning Test), however, was characterized by a high number of double recalls and intrusions indicative of poor source recall and response monitoring. The patient's pattern of performance on the word list indicated a reduction in the executive control of memory. Overall, test results were in keeping with a degree of compromise in functions subserved by the left anterior hemisphere, including compromise in aspects of executive functioning.

Rehabilitation: GMT

GMT has been identified as a viable method for cognitive rehabilitation through the use of self-instructional strategies, simulated real-world tasks, self-monitoring exercises, and homework assignments.¹⁴ Patients are assisted in developing awareness of attentional lapses in everyday life and strategies to overcome these lapses. The primary focus of GMT is the acquisition of a simple self-command ("stop") to interrupt ongoing automatic behaviors that occur during attentional lapses in order to resume executive control. Once the opportunity for more controlled processing has been established, patients are encouraged to refocus on the goal of the task at hand, to divide the task into manageable steps, and to continuously check their progress toward the goal (see Levine et al¹⁴ and Robertson¹³ for a more detailed description).

Research Design and Outcome Measures

This single-case study was part of a larger randomized control trial investigating the efficacy of 2 viable rehabilitation therapies (1 of which was GMT) following traumatic brain injury, stroke, or benign tumor excision. The current rehabilitation protocol consisted of 7 weekly 2-hour sessions. The efficacy of GMT was assessed via a battery of standardized and experimental tests of executive functioning and attention, including the Sustained Attention to Response Task,¹⁸ Delis-Kaplan Executive Function System Tower Test,¹⁹ the Revised-Strategy Application Test (R-SAT),²⁰ and the Hotel Task.²¹ Questionnaires were also used, namely the Dysexecutive Questionnaire (DEX) Self Assessment and Significant Other versions²² and the Cognitive Failures Questionnaire (CFQ).²³ These tests were administered

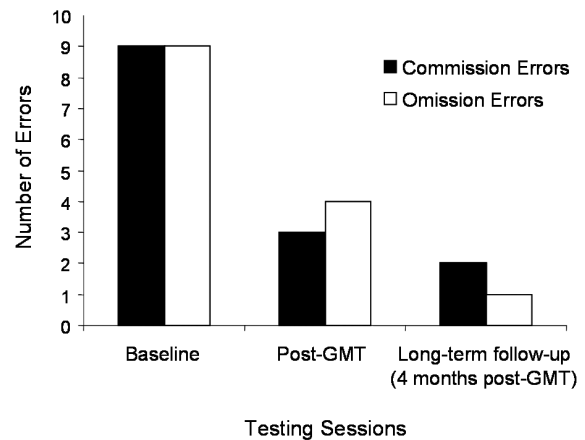


Figure 2. Error performance on the Sustained Attention to Response Task (SART) tested immediately before and following Goal Management Training (GMT). Long-term efficacy of GMT was also assessed 4 months following the last GMT session.

to the patient prior to rehabilitation (127 days postinjury) and after the 7-week rehabilitation program (218 days postinjury). Long-term efficacy of treatment was assessed approximately 4 months (322 days postinjury) after the final rehabilitation session.

RESULTS

Experimental Outcome Measures

See Table 1 (bottom half) for performance summary. See Figure 2 for error performance on the Sustained Attention to Response Task. The patient's performance on 2 behavioral measures, the R-SAT and the Hotel Task, was at ceiling for all items; there was therefore no possibility for improvement as assessed by these measures, and they are not reported in the table.

On self-report measures, the DEX and CFQ, the patient endorsed very few items prerehabilitation. This may be an indicator of poor awareness of his difficulties, particularly in light of the fact that the patient's spouse endorsed several more items on the DEX than the patient at initial assessment (DEX-Self overall = 1, DEX-Significant Other overall = 11). Interestingly, immediately postintervention and at long term follow-up, the patient was more aware of his difficulties because he endorsed more items on the CFQ. On the DEX, the patient's self-ratings remained low, comparable with his initial reports. At long-term follow up, the patient's spouse reported no difficulties on the DEX Significant Other assessment.

Clinical Neuropsychological Follow-up

At 48 days postrehabilitation (266 days postinjury), the patient was seen for a routine clinical reassessment using tests not part of the experimental outcome battery. This information allowed us to determine if any concomitant changes on the clinical measures corresponded with the improvements documented on the experimental outcome measures (see Table 1). At follow-up, improvements were documented in letter and number sequencing and the switching between letters and numbers. The acquisition of new verbal information with respect to the executive aspects of memory also improved postrehabilitation. Specifically, the patient used a highly efficient organizational strategy and was better able to monitor his response as indicated by a reduction in double recalls.

Functional Outcome

Soon after completing the rehabilitation program, the patient felt confident enough to return to work, initially on a part-time basis. Within a month he had resumed full-time duties in an intellectually challenging occupation. At latest follow-up, he is performing well. As noted above, his wife has noticed complete resolution of the dysexecutive signs that concerned her before the program.

DISCUSSION

To our knowledge, there is no research on the rehabilitation of cognitive impairments following focal damage to the cerebellum. The present study reports the successful use of a focused executive-rehabilitation program in a patient with executive dysfunction following a right cerebellar hemisphere AVM hemorrhage. The implications of the present results are two-fold. First, these results provide further evidence that lesions to the cerebellum can cause executive impairments that can significantly affect daily functioning, in the presence of intact supratentorial cortex. Second, our results provide evidence that GMT is a practical rehabilitation technique for executive impairments in patients with cerebellar damage and other patient populations. Our patient made gains in areas specifically targeted by GMT. These benefits were maintained at long-term follow-up and coincided with his return to a demanding profession. In the patient's words, GMT helped him become more aware of situations in which he was likely to make errors, realize the need to reflect on things and double-check his work, and stay focused on what he wanted to achieve in a day's work.

Although therapeutic gains were modest as measured by test performance, the patient's reports of real-life gains in relation to return to work were significant. In high-functioning individuals such as this patient, it is not unusual for subtle changes as measured by test performance to be experienced as a significant departure from premorbid functioning. The patient was at or near ceiling for several tests, yet he reported marked real-life deficits pretraining that improved posttraining. Test performance may therefore underestimate the degree of therapeutic benefit derived from GMT. The patient's gains are unlikely to be accounted for by spontaneous recovery, because he was already in the chronic phase of recovery at the time of the intervention.

Attention and executive functions have generally been associated with prefrontal areas of cortex. Anatomical studies have revealed important contralateral connections between the cerebellum and prefrontal cortex. Prefrontal lesions have been shown to cause hypometabolism to the contralateral cerebellum,²⁴ and cerebellar damage can cause frontal hypometabolism.²⁵ The dissociation between impaired verbal fluency and preserved design fluency is consistent with the expected effects of contralateral dorsolateral frontal lesions. The patient's unilateral damage to the right posterior lobule of the cerebellum preferentially affected a critical neural pathway directly linking the cerebellum with language areas of cortex. Taken together, the neuropsychological profile and the patients' own spontaneous complaints of organization and planning difficulties implicate impaired executive functioning. Interestingly, these deficits are in the presence of intact frontal cortex.

Although this was an uncontrolled single case, several factors support the claim that the treatment was effective: (1) the profile of cognitive deficits is now known to be specific to the lesion site, (2) the relevant deficits remained quite severe even in the chronic phase of recovery, (3) improvements in cognition were seen in the domains targeted by the treatment, (4) the targeted behaviors vanished after treatment according to his spouse, and (5) the patient was able to return to a demanding job within weeks of completion. GMT is a viable treatment option that can be used in a conventional rehabilitation setting. We are currently investigating this therapeutic approach in a randomized control trial in groups of brain-damaged patients.

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