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CLINICAL PSYCHOLOGY & NEUROPSYCHOLOGY | CASE REPORT

Posterior cortical atrophy: Impact on daily living activities and exploration of a cognitive rehabilitation approach

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Abstract: Posterior cortical atrophy (PCA) is a neurodegenerative disease affecting the posterior region of the brain. Little is known about both the impact of PCA on functioning and how to support patients on a daily basis. The purpose of this study was to describe the functional profile of DD, a woman diagnosed with PCA, as well as to explore a pilot cognitive rehabilitation program designed to optimize functioning in daily living. The *ADL Profile* was used to assess the daily tasks that DD chose to undertake. Four operations, i.e. formulate a goal, plan, carry out and verify goal attainment, were scored for each task. Difficulties were observed during the execution of all tasks, as she struggled to find items or showed unsafe behaviors. Impairments were also seen in formulating a goal and planning, especially for less routine tasks. DD identified two tasks to be addressed in rehabilitation: setting the table and dealing cards. Learning was optimized using errorless learning and compensatory aids when setting the table, while dealing cards received no intervention. Only setting the table improved significantly with time. Further studies should be conducted to portray a wider functional profile of people living with PCA and develop effective rehabilitation programs.

Subjects: Behavioral Sciences; Allied Health; Health Conditions

N. Bier

ABOUT THE AUTHOR

N. Bier is an associate professor of occupational therapy at the Université de Montréal. She is also a researcher at the Research center of the Institut universitaire de gériatrie de Montréal. The main goal of Nathalie Bier's research program is to better understand the impact of cognitive deficits in aging and dementia on everyday living and functioning, as well as to develop interventions to promote aging in place – such as the use of cognitive rehabilitation and new technology. This project is thus in line with both exploring the impact of dementia, in this case PCA, on everyday living and the development of a new rehabilitation approach to support functioning.

PUBLIC INTEREST STATEMENT

Posterior cortical atrophy (PCA) is a neurodegenerative disease affecting the posterior region of the brain, therefore impacting the ability to orient oneself in space and to perceive the objects of the environment. Little is known about the impact of PCA on functioning in everyday living and how to support persons suffering from it on a daily basis. The purpose of this study was to describe the functional profile of DD, a woman diagnosed with PCA, as well as to explore a rehabilitation program designed to optimize functioning in daily living. Difficulties were observed during the execution of all tasks, as DD struggled to find items and showed unsafe behaviors. With the help of a rehabilitation program, DD was able to relearn how to do a significant activity (setting the table). Further studies should be conducted to portray a wider functional profile of people living with PCA and develop rehabilitation programs.

Keywords: posterior cortical atrophy; activities of daily living; performance-based assessment; visuo-spatial abilities; executive functions; cognitive rehabilitation

1. Introduction

Posterior cortical atrophy (PCA) is a neurodegenerative condition characterized by a progressive and relatively selective decline of visuo-spatial abilities (Caixeta et al., 2013; Chan et al., 2015). It is estimated that Alzheimer's disease (AD) pathology is the cause of this rare and atypical dementia in more than 80% of cases (Alladi et al., 2007). Visuo-spatial deficits are generally first reported as early as age 50 or 60; thus PCA is considered to be an early-onset dementia (Mendez, Lee, Joshi, & Shapira, 2012). Recently, a number of studies, including meta-analyses, have been published to characterize the neuropsychological profile and pattern of cortical atrophy in PCA, and to compare this condition with other dementias (e.g. Alves, Soares, Sampaio, & Gonçalves, 2013; Borruat, 2013). However, a better understanding of the impact of PCA on everyday life is also crucial and may significantly contribute not only to differential diagnosis, but also to intervention planning and patient management.

Visuo-spatial abilities have been identified as important for everyday functioning, as they play a key role in the execution of daily tasks including self-care (Nijboer, de Port, Schepers, Post, & Visser-Meily, 2013) and mobility (Maeshima, Itakura, Nakagawa, Nakai, & Komai, 1997; Nijboer et al., 2013). To our knowledge, only one study by Shakespeare, Yong, Foxe, Hodges, and Crutch (2015), has investigated the impact of visuo-spatial difficulties on activities of daily living (ADL) in the specific context of PCA. In that study, functional independence was documented using a questionnaire filled out by the caregiver. Overall, patients with PCA were shown to have more difficulties than patients with AD who have a typical amnestic presentation in certain ADL, most specifically in self-care (basic activity of daily living: BADL) and in instrumental activities of daily living (IADL) that require more elaborate visuo-spatial abilities or calculations; IADL that were most impacted included activities such as writing, using the phone, making a hot drink, handling money, and using electrical appliances. In contrast, patients with AD were shown to be more affected in tasks requiring memory (day-to-day memory, repetitive questioning). Patients with PCA also had more sleep problems and presented some behavioral disinhibition (Shakespeare et al., 2015).

Cognitive interventions to improve functional independence in PCA are also rare, but promising. A recent review (Weill-Chounlamountry, Alves, & Pradat-Diehl, 2016) identified only three case studies on the topic. Roca, Gleichgerrcht, Torralva, and Manes (2010) presented a 5-week intervention program aimed at improving ADL selected with the patient and his family, namely locating objects in space, reading written messages left by his family and pouring drinks. Their intervention program combined education about the disease, training in object recognition with salient features, visual scanning, tactile direction of movements and letter recognition. The patient showed improvement on neuropsychological testing after the program, which is in line with the type of tasks practiced during training. The patient and his wife also noted subjective improvement. Weill-Chounlamountry et al. (2012) reported results of a very similar cognitive training program, that also included teaching the use of an external compensatory strategy and physical therapy. After 6 months of global therapy, they observed improvement in some visual abilities and stable neuropsychological testing. They also noted subjective improvements in ADL. Finally, in a third study, Alves et al. (2015) used global cognitive training comprising reality orientation, cognitive stimulation, and cognitive training exercises, and observed only modest effects on neuropsychological testing post-intervention, with no measure of the impact on ADL.

Therefore, to date, no studies have provided evidence of improved independence in ADL after cognitive rehabilitation interventions in PCA. A *cognitive rehabilitation approach* (Clare & Woods, 2004) may offer promising results in PCA, though no studies have yet explored this. It consists of selecting specific daily tasks and optimizing performances with the help of methods such as errorless learning (promoting learning by limiting the amount of errors produced). Such techniques

support cognitive abilities that have been preserved or relatively spared from impairment and have a rapid impact on independence in ADL.

This single-case study had two main objectives. First, it aimed to further describe the impact of PCA on independence in ADL, using a performance-based functional assessment tool. Such a tool has the advantage of allowing the identification of essential components related to the person's performance, such as patterns of errors and difficulties that cannot be captured by proxy questionnaires and that can support planning of cognitive interventions. Second, this study aimed to explore the impact of a *cognitive rehabilitation approach* on relearning of significant ADL, based on results obtained through the performance-based evaluation.

2. Methodology

2.1. General design

In order to document the impact of PCA on independence in ADL, a descriptive case-study was undertaken. To explore the impact of a cognitive rehabilitation approach, we used a multiple baseline design with training in one activity offered and another serving as a control condition. We first present the case of DD along with her neuropsychological, imaging and oculomotor testing results, followed by a description of the impact of PCA on her independence in ADL (part 1). We then present the intervention study (part 2).

This research project was approved by the Research Ethics Committee of the *Institut universitaire de gériatrie de Montréal*, and DD gave informed written consent to participate in the two parts of the study.

2.2. Case description

DD, 69 years of age, was a French-speaking right-handed woman who has been retired for 8 years. She was living in a two-story house with her husband at the time of the study. She completed 18 years of schooling. Her professional activities included various positions and mandates related to improving health interventions.

DD described herself as a sociable and relaxed person, who could always find a solution to any situation she encountered. She mentioned that she was always considered to be an active person, mainly referring to her many travels around the world. She first experienced some visuo-spatial difficulties during an overseas trip. She noticed that these difficulties worsened over the following year and consulted a neurologist. She was referred for a neuropsychological exam and brain imaging. Six months later, she was referred for a functional examination and ocular screening. Because of her symptoms, DD mentioned that she had ceased many of her activities and that her main occupation now consisted of watching television. She reported difficulties in dressing, preparing hot meals and watering her plants. She also mentioned that grocery shopping were now mainly realized by her husband, although she was sometimes joining him.

The diagnosis of PCA was done by the patient's neurologist on the basis of the results of the neuropsychological testing and the presence of bilateral parietal atrophy on MRI. The diagnosis of PCA-pure (classification level 2) was consistent with the criteria of the Consensus Classification of posterior cortical atrophy described by Crutch et al. (2017) including clinical, cognitive and neuroimaging findings.

2.3. Neuropsychological profile

The neuropsychological profile of DD was documented through an extensive clinical evaluation (see Table 1). The MMSE score (Mini Mental State Examination; Folstein, Folstein, & McHugh, 1975) suggested that the disease had mildly affected her global cognitive abilities. This decline was associated with moderate agitation, anxiety and depression (NPI ten—Neuro-Psychiatric Inventory 10 items; Cummings, Mega, Gray, Rosemberg-Thompson, & Gornbein, 1994; and GDS 15 items version—Geriatric Depression Scale; Sheikh & Yesavage, 1986).

Table 1. Neuropsychological data for DD on standardized tests

Domain	Test name	Measures	Score	Impairment
Global cognition				
	MMSE	Total	21/30	***
Mood and neuropsychiatry				
	GDS-15	Total	8/15	mild depression
	NPI 10	Total	9/120	–
Attention & Working memory				
	Digits span	Forward	6	–
		Backward	2	***
	BTA	Total	2/20	***
Executive functions				
	TMT Oral	Time/errors—Part A	7 sec./0	-/-
		Time/errors—Part B	80 sec./2	***/-
	Fluency (90 sec)	Category Switching	7 (ss: 3)	***
Memory				
	Logical memory	Immediate recall	25/50	–
		Delayed recall	24/50	–
	RAVLT	Sum free recall (1–5)	49/75	–
		Delayed free recall	8/15	–
		Delayed recognition	14/15	–
Semantic memory				
	BNT	Without cue	26/30	–
		With cues	30/30	–
	Fluency	Animals	10 words	**
		Fruits and vegetables	8 words	***
	PPTT	Visual form	40/52	***
Visuo-spatial perception				
	Hooper	Total	19/30	***
	VOSP	Screening test	20/20	–
		Incomplete letters	14/20	***
		Silhouettes	13/30	*
		Object decision	13/20	*
		Progressive silhouettes	12	–
		Dot counting	6/10	***
		Position discrimination	10/20	***
		Number location	2/10	***
		Cube analysis	0/10	***
	Clock drawing	On command	4/10	***
		Copy	2/10	***

*Notes. Sec = seconds;— = within normal limits; * = z-scores between −1.5 and −2; ** = z-scores between −2 and −3; *** = z-scores below −3; ss = scale score.*

More specifically, DD presented a severe visuo-spatial deficit that was particularly evident in visual exploration and spatial orientation tasks (number location, dot counting, position discrimination and cube analysis from the VOSP—Visual Object and Space Perception Test; Warrington & James, 1991). She also performed poorly in reading and written calculations, as well as in visual imagery, which could partly explain her impaired performance on backward digit span (Wechsler,

2008) and her difficulties with mental calculations. Her performance in clock drawing (poorer under copy than under command) and writing suggested a mild left unilateral neglect. Yet, DD did not exhibit any simultagnosia on a picture description task (Boston Cookie Theft picture; Goodglass, Kaplan, & Barressi, 1983), and visual object perception was only mildly impaired (VOSP).

The examination also revealed a mild finger agnosia and an ideomotor apraxia for both symbolic gestures and pantomimes. She showed a marked dressing apraxia that was spontaneously reported by both her and her husband. Her left-right spatial disorientation (Culver right-left discrimination test; Culver, 1969; and designation) may have contributed to her dressing apraxia.

DD exhibited a mild dysexecutive syndrome that went beyond her visuospatial deficits (Verbal fluency: category switching; TMT Oral—Trail Making Test; Ricker & Axelrod, 1994). More specifically, it was very difficult for her to switch from one semantic category to another in the verbal switching fluency task. She also failed the go-no-go tapping task, despite the fact that instructions had been repeated twice.

Semantic memory was mostly within the normal range as long as it was assessed with non visuo-spatial material such as the Similarities subtest from the WAIS IV or knowledge of famous people and events (Benoit, Rouleau, Langlois, Dostie, & Joubert, 2018; Langlois, Joubert, Benoit, Dostie, & Rouleau, 2015). Verbal episodic memory results were within normal limits (logical memory; Wechsler, 1997; and RAVLT, Rey Auditory Verbal Learning Test; Schmidt, 1996).

In summary, DD exhibited a predominant deficit in visuo-spatial tasks but her visual object recognition and semantic processing were preserved. This dissociation suggests that the visual dorsal stream of the brain ("where") was impaired, whereas the visual ventral stream ("what") remained functional. Executive functions were also impaired, even in tasks which did not involve any visuo-spatial material. However, her verbal memory was globally preserved. These results and her score on the MMSE support the idea that DD was probably in the early to moderate stage of her disease.

2.4. Magnetic resonance imaging (MRI)

A structural MRI of the brain was performed at the time of the neuropsychological evaluation. The exam revealed an atrophy of the parietal lobes, predominantly on the right side of the brain. There were no focal lesions, nor any mass effect.

2.5. Ocular and oculomotor screening

When assessed with the Subjective Symptom Questionnaire: Acquired Brain Injury (Scheiman, 2011), DD explained that she experienced excessive eyestrain and could not avoid skipping lines while reading. Binocular confrontational visual fields (Bégin, Boudreault, & Sergerie, 2009) were roughly normal in the four quadrants, but the visual extinction test was positive in both left quadrants. Binocular visual acuity was normal at 0.5 m at 30 cm with glasses (Minnesota Low-Vision Reading Test 1-MnRead 1; Senécal, Gresset, & Overbury, 2006) at 100 words per minute, but DD had to be cued to complete the task (she was saying "is there anything else?"), which was consistent with the extinction results. During this test, DD also skipped some words (mostly in the biggest lines), suggesting disorganized saccades during reading. Ocular motility tests (NSUCO/Oculomotor test; Maples, 1994) showed impairments in ability, accuracy and head movement during saccades (7/15), mainly during pursuit (4/15). Convergence insufficiency was also observed in the "Near point of convergence test". Finally, she struggled with the Developmental Eye Movement—C test (Garzia, Richman, Nicholson, & Gaines, 1990), showing disorganized visual exploration and focusing mainly on the right-hand side. These measures revealed the apparent normality of basic ocular functions (visual acuity, visual field and contrast sensitivity were also found to be normal in a standardized examination), but oculomotor control was severely impaired.

3. Part 1: impact of PCA on independence in ADL

3.1. Design

A descriptive case study, using qualitative and quantitative data, was used to document the impact of PCA on independence in ADL. This design is suitable for studies that aim to conduct an in-depth exploration of a phenomenon within its real-life context, as well as to inform future research and clinical practice (Yin, 1981, 2011). More specifically, we used a non-structured, performance-based assessment of ADL.

3.2. Measurement of independence in ADL

The *Activities of Daily living Profile* (*ADL Profile*) (Dutil, Bottari, Auger, & Auger, 2017; Dutil, Bottari, Vanier, & Gaudreault, 2005; Dutil, Forget, Vanier, & Gaudreault, 1990) was used to evaluate DD's performance in her ADL. The *ADL Profile* is a non-structured, performance-based tool administered in the individual's home and community environments in order to evaluate independence in ADL. The *ADL Profile* is designed to detect deficits in BADL and IADL related to four prominent operations involving goal-directed behaviors (executive functions): 1) formulating a goal regarding a specific task, 2) planning, 3) initiating and carrying out the task, and finally 4) verifying attainment of the goal. Task performance is evaluated according to these four operations. Scoring and its interpretation take into account patient behaviors as well as assistance provided by the examiner.

DD was seen in her home by two trained occupational therapists (NB and CB; to evaluate by consensus) shortly before lunchtime and was asked, as per the administration guidelines of the *ADL Profile*, to engage in her routine activities, as she would normally do at this time of the day if no one was examining her. The purpose of these instructions was to encourage goal formulation and planning, as these operations are very important for everyday functioning (Bottari, Dassa, Rainville, & Dutil, 2009a, 2009b; Lezak, 1982). No further specifications were given to DD except for general indications to ensure that she would formulate more goals and engage in subsequent activities. Also, the amount of verbal interaction and assistance provided by the examiner remained minimal throughout the assessment as one of the tool's goals consists of observing the initiation of a chosen task, as well as the self-identification and correction of errors. In fact, DD had to decide by herself which ADLs she would perform during the evaluation (goal formulation) and how she would execute them (planning); she also needed to execute them (carry out the task) and decide when the task was over (verify goal attainment), in order for the examiners to fully assess the four operations in relation to each task. She was clearly informed throughout the evaluation that she was free to continue or stop at any time. From the perspective of maintaining a rigorous single case methodology, DD's performance was videotaped to help scoring and permit a more in-depth analysis of her performance via written transcriptions.

A substantial number of psychometric studies have been conducted with this test among individuals with severe cognitive deficits. Generally, good psychometric properties have been reported for internal consistency, test-retest reliability, inter-rater reliability, and content and factorial validity (Dutil et al., 2017, 1990; Rousseau, Dutil, & Lambert, 1994a, 1994b). A complete description of the psychometric properties can be found elsewhere (Bottari et al., 2009a).

3.3. Data analysis

Based on video recordings, DDs performance was transcribed into verbatim reports of all her behaviors as well as the assistance received from the examiners. The verbatim reports were transcribed by AES and NB and cross-validated by CB. Main difficulties were identified and organized according to the four operations, for each task. For each difficulty, assistance received was noted. Assistance not requested by DD was also highlighted. Together, this information aimed to support the scoring of each operation, for each task. Analysis was done by AES and NB and cross-validated by CB (triangulation).

DD's level of independence was scored following the tool's ordinal scale, ranging from independent with no difficulty (score of 3) to dependent (score of 0). Scoring was assigned for each of the four

operations in each activity (i.e. formulating a goal, planning, carrying out the task, and verifying attainment of the goal) and the lowest score on any one operation became the independence score on the task as a whole. A score of 3 indicated that she performed the operation/task independently. A score of 2 meant that she completed the operation/task independently, but with difficulty (e.g., she struggled or repeated the same action a few times before succeeding, the quality of execution was poor), while a score of 1 indicated that assistance was needed from the examiner (1v for verbal assistance, 1p for physical assistance and 1vp for both) in order for her to complete the operation/task. A score of verbal assistance was given when assistance was required to compensate for underlying cognitive difficulties. A score of physical assistance indicated an aid responding to a physical need (ex. balance), while a score of verbal and physical assistance meant that both physical and cognitive deficits were at issue. A score of 0 was given when DD could not complete the task by herself, showed unsafe behaviors or needed constant direction and assistance from the examiners to get through the operation/task adequately. The rating was done by AES and NB, and discussed with CB to reach a consensus.

3.4. Results of the evaluation of independence in ADL

DD chose to perform seven tasks: 1) preparing a light meal (meal preparation—familiar task); 2) preparing a hot meal (meal preparation—with elements of novelty as she no longer carries out this IADL); 3) watering plants (housecleaning—familiar task); 4) using the washroom (hygiene—familiar task); 5) putting on boots and a winter coat (dressing—familiar task); 6) walking outdoors to go grocery shopping (outdoor mobility—elements of novelty as her husband is now responsible for grocery shopping) and 7) shopping (use of public services—elements of novelty as her husband is now responsible for grocery shopping). We had to limit our evaluation to these tasks, because she mentioned being very tired after shopping. Insisting would have led to an increasing degree of frustration and fatigue.

Her level of independence in these tasks, as measured by the *ADL Profile*, is presented in Figure 1. A chart providing an illustration of DD's performance during the evaluation, including her actions, main errors and main types of assistance required for every task, is presented in the Supplementary Material.

3.5. Scores on the four operations

Scores indicate that DD presented deficits on all four operations. Goal formulation was only impaired in less familiar tasks; DD needed a lot of verbal assistance to think about preparing

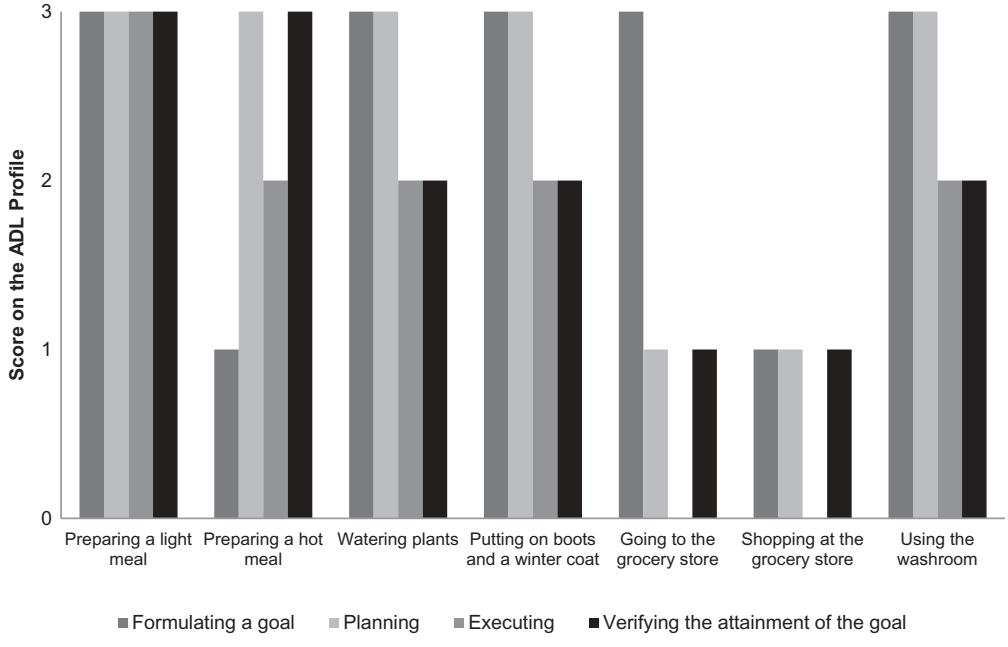

Figure 1. DD quantitative independence scores for 7 ADL tasks. A score of 3 stands for independent with no difficulty. Scores of 2 were given when DD completed the operation (e.g. formulating the goal, planning, carrying out or verifying the attainment of the goal) independently but with difficulty. Scores of 1 were used when assistance from the examiner was needed (1v for verbal assistance, 1p for physical assistance and 1vp for both). Score of 0 indicates that despite assistance, DD was not able to complete the task or operation (dependent).

a hot meal and to go grocery shopping. In fact, she told the examiner that she had stopped cooking elaborate meals. Planning was also impaired in these less familiar tasks.

Carrying out the task and verifying the attainment of the goal were the most impaired operations. Indeed, with the exception of preparing a light meal, DD scored between 2 (independent with difficulty) and 0 (dependent) on all tasks. For the operation "verifying the attainment of the goal" (with the exception of preparing light and hot meals), she could hardly understand and explain the difficulties she encountered, although she verbalized several times that she was having trouble and was not as effective as she used to be.

3.6. Scores for each task chosen by DD

3.6.1. Preparing a hot meal, watering plants, putting on boots and a winter coat
Although no assistance from the examiner was needed (score of 2), DD struggled to execute these three tasks. For example, she required three trials and considerable time to put on her winter coat. Also, she had difficulty finding cooking equipment in the kitchen drawers and adequately using the watering can to water the plants. She had a tendency to only water the plants located on the right side of her visual field, consistent with the mild left neglect noted in the neuropsychological evaluation. Additionally, DD mentioned being uncertain of her performance throughout completion of the tasks; she also frequently mentioned that they were difficult for her.

3.6.2. Walking outdoors, shopping
She obtained a score of 0 (dependent) for carrying out both tasks. Her behavior was particularly unsafe when walking outdoors. The examiner had to intervene on two occasions to ensure her safety, once when she failed to notice a rapidly oncoming car on her left, while crossing the street, and a second time when she walked in the street (rather than on the sidewalk) while cars were driving by rapidly, just inches away.

At the grocery store, she was unable to find the items she was looking for and requested the examiner's and employees' verbal assistance on several occasions. The score of 0 (dependent) given for carrying out this task is based on her inability to find certain items despite repeated explicit assistance from the examiner. For example, after having been told by an employee in which aisle she could find chicken noodle soup, she walked up and down the aisle three times, ineffectively searching for the soup. In fact, she was mainly looking to the right and top shelves of the aisle (while the soup was slightly to the left and lower down) and no visual scanning was observed. After several minutes of vainly walking up and down the aisle, and given DD's mounting frustration, the examiner first provided open-ended implicit cues that failed to help her find the soup. In the end, the examiner had to stop DD right in front of the exact spot where the soup was located and suggest that she look directly in front of her, but DD was still unable to spot the soup. Verbal assistance was then provided through such comments as "look down a bit" and "move to your left", but was not effective. DD constantly limited her visual search to the upper shelves, whereas the soup was directly in front of her, at eye level. The examiner finally decided to place her hand directly in front of the soup; this level of assistance allowed the participant to find the soup she was looking for. At the checkout counter, she requested assistance from the cashier as she was struggling with handling her money, holding her purse and gloves, and talking to the cashier to get through the purchase. The cashier had to go so far as to directly ask her to place the items from her shopping cart onto the counter for checkout purposes.

4. Part 1: discussion

The purpose of this first part of the study was to describe the impact of PCA on independence in ADL. DD, a 69-year-old woman presenting with this rare form of dementia and living at home with her husband, was observed while carrying out seven tasks with a performance-based tool, the *ADL Profile* (Dutil et al., 2005). Results showed that her cognitive deficits had a major influence on all aspects of daily living. She struggled with formulating goals, planning and verifying attainment of her goals for many basic and instrumental ADLs. Carrying out the tasks was, however, the most

difficult operation, and her difficulties were even more evident in less familiar settings and visually crowded environments (e.g. outside, in the grocery store).

Neuropsychological testing and brain imaging indicated that DD exhibited impairments that are characteristic of PCA. She showed predominant visuo-spatial disorders, in addition to some deficits in verbal working memory (backward digit span) and a mild left unilateral neglect. Furthermore, she presented a deficit in right-left discrimination, finger agnosia, ideomotor apraxia and dressing dyspraxia. These impairments are consistent with a bilateral parietal atrophy predominant on the right side, as supported by neuroimaging data. The testing also indicated relatively well-preserved verbal episodic memory and semantic processing, as she demonstrated good naming and verbal processing skills by being able to recognize famous people. These results are also consistent with the cognitive profile of patients with PCA, in whom semantic and episodic memory remain generally intact (Andrade et al., 2010; Benson, 1988; Biotti, Pisella, & Vighetto, 2012; Crutch et al., 2012, 2013; Tang-Wai et al., 2004). In contrast, DD showed difficulty on arithmetic and calculation tasks, as well as tasks involving visual episodic memory. Moreover, she presented difficulties on tasks assessing executive functions (TMT Oral, Stroop test, conflictual tapping).

Results of the present study suggest that these cognitive symptoms were a good reflection of her difficulties in ADL, in particular visuo-spatial and executive deficits. Some of these were accentuated during tasks executed in less routine situations, or in environments with abundant visual stimuli. These findings underline the usefulness of conducting a broad functional evaluation in addition to neuropsychological testing in order to thoroughly explore the difficulties experienced by persons with dementia in their ADL. Indeed, leaving the comfort and familiarity of the home exposes the person to a larger spectrum of challenges that cannot be fully evaluated with pen and paper tests (Bottari et al., 2009a, 2009b). In some instances, DD was unaware of her problems and engaged in unsafe behaviors (e.g., not looking left and right while crossing the street). Factors like familiarity of the environment and fatigue may have also affected her ability to perform the tasks. Therefore, a complete performance-based assessment can highlight important difficulties that may go unnoticed with ADL questionnaires or shorter ADL performance-based evaluations.

The analysis of DD's performance in the *ADL Profile* and the results obtained by Shakespeare et al. (2015) both suggest that visuo-spatial difficulties in patients with PCA have an important impact on independence in ADL. In fact, DD presented difficulties in finding and accurately handling everyday objects, even with frequent and direct verbal assistance. In addition to her marked visuo-spatial deficits, DD's examination also showed an oculomotor control deficit. These deficits, related to the dorsal ("where") stream, could explain her difficulties in efficiently finding and handling objects, as well as the mistakes she made when carrying out her ADLs. Results from DD's evaluations are also in line with findings in a study exploring patients' and their caregivers' perceptions about their conditions (PCA), with qualitative in-depth interviews (Harding et al., 2018). Patients reported being markedly restricted in dressing themselves, cooking as well as in engaging in a wide range of hobbies.

Results of this study also support the observations that deficits in executive functions appear as important symptoms in various forms of dementia and seem to be crucial for independence in this population. The link between executive dysfunctions and difficulties in ADL has been suggested in AD (Martyr & Clare, 2012) and fronto-temporal dementia (Bier et al., 2012; Mioshi, Hodges, & Hornberger, 2013; Mioshi et al., 2007). The present study also highlights this link in PCA. The contribution of executive functions to independent living should thus be considered in dementia (Bier et al., 2016). This means that evaluations should go beyond the execution or carrying out phases of tasks, by using an approach that allows goals to be formulated, tasks to be planned and errors to be detected and corrected by the patient and not by the examiner (Bier et al., 2016; Bottari et al., 2009a, 2009b). Furthermore, evaluations should include situations that are more complex or novel to the patient, in which executive functions are particularly solicited (Burgess, 2000). If they are to be conducted in the

persons' home and community environments, it should be noted that the evaluators have to be specifically trained to observed but also to manage events were the person's security may be compromised; such as occupational therapists. Observations collected with tools such as the *ADL Profile* are very rich but should be carefully conducted.

The combination of visuo-spatial and executive function deficits may explain why DD also tends to spend her day watching television rather than engaging in more complex, less familiar tasks. While DD described herself as having been an active person previously, her routine had become sedentary due to her cognitive decline, associated with exacerbation of her PCA symptoms.

A cognitive rehabilitation approach should be implemented with such patients in order to minimize the decline of ADL abilities and avoid deconditioning (Weill-Chounlamountry et al., 2016). For DD, observation in her home and community with the *ADL Profile* showed that she performed better in routine tasks than in less familiar tasks. For example, she was able to prepare the same meal for lunch every day without difficulty, even if it consisted of locating several ingredients in a stocked refrigerator and placing them on a plate.

Thus, the potential usefulness of learning routinized activities, perhaps supported by procedural memory, may be relevant in PCA. In fact, participants in a study by Harding et al. (2018) commonly reported relying on a routine response to the environment. They also described a tendency to simplify tasks and rely on familiarity. The second part of this study therefore aimed to explore the potential of developing a training program to teach DD specific simple tasks that would support her independence in ADL.

5. Part 2: cognitive rehabilitation

5.1. Design of the study

An ABC multiple-baseline design (Ottenbacher, 1986) was selected in order to assess DD's performance throughout the intervention. This design involves taking outcome measures at baseline (A) and during the intervention comprising two conditions, i.e. B and C. Post-intervention measures could not be taken because DD's condition deteriorated during the study, and she decided to stop the intervention process. The multiple baseline involves comparing a target activity with a control activity. The control activity is not expected to change because it is not trained. Therefore, it is used to assess the effectiveness of the intervention, as it helps determine whether improvements are solely due to the intervention or to other independent variables, such as time or simple repetition (Krasny-Pacini & Evans, 2018).

5.2. Activity selection

In order to select the tasks in which DD was to be trained, an interview was conducted with her and her husband. She revealed personal concerns regarding two activities that were skipped over during the functional evaluation. For instance, DD mentioned encountering difficulties when using her computer, specifically only being minimally able to navigate on the Internet or locate all the visual stimuli on the computer screen. Moreover, both she and her husband reported major difficulties setting the table appropriately, a task which had become part of her daily routine since she had ceased preparing elaborate meals. In fact, setting the table was something DD spontaneously said she wanted to improve. Also, when discussing leisure, the participant shared with the examiners her interest in playing cards with her two grandchildren when they visit her. She then reported having difficulty dealing cards, mentioning that she was hardly able to distribute the correct number of cards at the appropriate place in front of each player.

Thus, based on her interests and struggles, the cognitive intervention targeted "setting the table". In parallel, "dealing playing cards" represented the control task, for which no intervention was provided. Moreover, these two tasks are comparable in terms of complexity and cognitive demands, as they both involve visuospatial organisation and item distribution. Furthermore, they

were both meaningful and deemed priority tasks by DD, which allows us to suppose that she was similarly motivated to improve her ability to carry out both.

5.3. A—baseline measures

Four baseline measures were completed for the target activity and three for the control activity (sessions number 1 and 2). Performance was scored using a scale of 0 to 10 for each activity: 10 items to be set for a dinner for two on the dining table (5 items per person), and 10 playing cards, i.e. 5 for each of two players, to deal. Emphasis was placed on execution of the tasks, since we were looking for a routinization of the steps involved. Scoring was based on DD's ability to place the items appropriately.

5.4. B and C—cognitive intervention for setting the table

The intervention consisted of two conditions: B) an external visual aid and C) errorless learning.

B) A placemat showing the items to be placed (a large and a small plate, a knife, a fork and a wine glass) was used to help DD's visuospatial organisation and appropriate distribution of the tableware. Two identical placemats were provided to the participant, one for her husband and one for herself. Three sessions were dedicated to training her on the use of this compensatory aid (sessions number 2–4).

C) Errorless learning was added to the training in the use of the external visual aid for two other sessions (sessions number 5 and 6). Errorless learning is a method used to minimized or reduce the production of errors by offering maximal guiding during the leaning phase (Fish & Wilson, 2018). Although generally used with persons presenting with severe episodic memory deficits, it was recognized as a usefull learning method for learning of procedural tasks with motor components (Poolton, Masters, & Maxwell, 2005). To apply the method, the experimental task was divided into ten steps. These particular steps were selected according to the order in which DD habitually placed the tableware. The sequence was executed repetitively following the same order, with the help of errorless learning to ensure proceduralization of the task. More specifically, the task consisted of placing the right items on top of the outline/silhouette of the same shape on the placemat. Verbal guidance was provided while she was placing the objects in order to avoid errors, as well as positive reinforcement for correct actions. When DD made a mistake, which was rare, she was told right away with a demonstration of the right placement and verbal guidance. This second part of the cognitive rehabilitation was developed with the intent of eventually decreasing the need for the external visual aid. The objective was to allow DD to increasingly rely on a routine and then, progressively remove the external aid.

Every intervention session was separated into three steps: 1) first, DD practiced the target activity (setting the table), within conditions B or C; 2) she was evaluated while performing this activity (score out of 10) and then, 3) while executing the control activity (dealing playing cards, score out of 10). One measure was taken at each session numbers 3–5 for the target activity and three measures were taken for the control activity. No measure could be taken during session 6 since DD stopped her participation in the project, by specifying that she was not interested anymore.

5.5. Data analysis

DD's performance scores were first analyzed by means of visual analyses of graphed data (Franklin, Gorman, Beasley, & Allison, 1997). Tau-*U* statistics were then used to measure improvement within each task, by comparing baseline data with the intervention phases. Results of interventions B and C were combined since there was not enough data to compare them separately. Derived from Kendall's Rank Correlation and Mann-Whitney *U* tests, Tau-*U* is a non-parametrical statistic that measures the amount of overlap between two phases, while controlling for potential trends during baseline (Parker, Vannest, Davis, & Sauber, 2011). Calculations were performed with the Tau-*U* calculator website (Vannest, Parker, & Gonen, 2011: http://singlecaseresearch.org).

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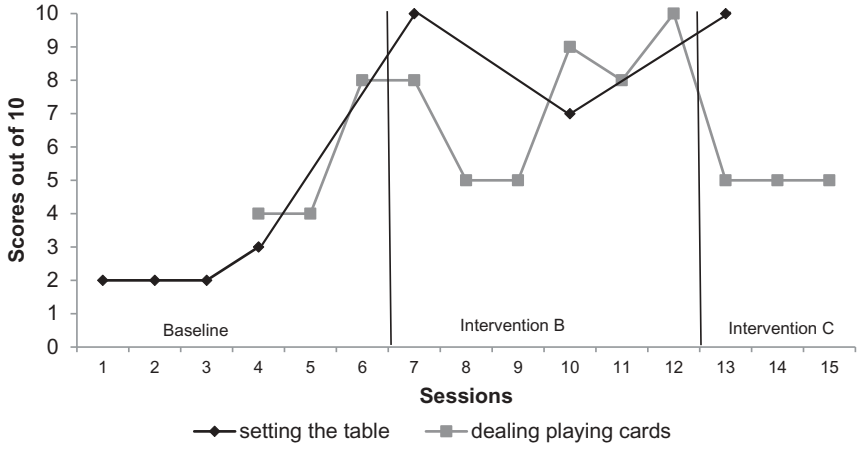

Figure 2. DDs score out of 10, for the experimental (setting the table) and control tasks (dealing playing cards).

5.6. Results

5.6.1. Setting the table

DD's performance for setting the table throughout the course of the study is presented in Figure 2. At baseline, DD scored an average of 2.5 out of 10, meaning that between 2 and 3 items were appropriately placed on the kitchen table. When the placemat was first introduced, a rapid increase in performance can be seen, as DD carried out the target task without any errors. However, the improved performance was not maintained at the following session. With the introduction of errorless learning, performance returned to 10. Overall, DD scored an average of 8.5 during the intervention phase, while an average score of 2.5 was reported at baseline (variation of 6 points). The difference was significant, the Tau-*U* statistic indicating no overlap between the two phases (Tau-*U* = 1, $p = .034$).

5.6.2. Dealing playing cards

Visual analysis shows an improvement for the control task across sessions, although the performance is inconsistent. Overall, DD scored an average of 5.3 during the baseline phase and maintained an average score of 6.7 throughout the intervention phases (variation of 1.4 points). The difference was not significant, the Tau-*U* statistic indicating important overlap between the two phases (Tau-*U* = .56, $p = .17$), therefore equivalent performance between baseline and intervention.

6. Part 2: discussion

The purpose of this second part of the study was to investigate the effectiveness of a cognitive rehabilitation approach aimed at helping DD, a woman with a diagnosis of PCA, engage in a meaningful activity. Results suggest that the use of a placemat combined with errorless learning improved DD's performance, as she was able to adequately place six items more following the intervention than previously.

As for the placemat, immediately after its introduction, DD reported being more successful at locating the edges of the table and organising the items conveniently in front of each seat. Thus, implementing this visual aid in DD's home environment represented a landmark for her and the execution of the target task was facilitated. External visual aids were also used by Weill-Chounlamountry et al. (2012) with a patient with PCA, and the authors suggest that it was an effective support for dressing. Based on the principle that errors must be avoided to enhance learning in persons with severe cognitive impairments, errorless learning has been described as a successful tool and is considered as a gold standard in cognitive rehabilitation (Clare et al., 2010; Haskins, 2012). To our knowledge, no other study has explored the use of this learning method in PCA. The present study did not extend over a sufficient number of sessions to be able to draw conclusions about the efficacy of errorless learning for routinizing the procedure to support independence in ADL on a long-term basis.

However, these preliminary results are consistent with findings in the field of cognitive rehabilitation in different forms of dementia. For example, errorless learning combined with practice helped patients diagnosed with different forms of dementia learn how to use cell phones and a tablet computer (Bier et al., 2015; Bier, Paquette, & Macoir, 2018; Imbeault et al., 2018; Imbeault, Langlois, Bocti, Gagnon, & Bier, 2018), prepare meals (Bier et al., 2011), and relearn people's names (Clare, Wilson, Carter, Roth, & Hodges, 2002; Dunn & Clare, 2007). Thus, providing a learning context that optimizes preserved cognitive functions and solicits impaired cognitive functions less can improve the performance of ADL tasks in many forms of dementia. Finally, results by Weill-Chounlamountry et al. (2012) suggest that a holistic rehabilitation approach, including neuropsychological rehabilitation, occupational therapy and physical therapy, may be promising in PCA. Other studies have also suggested the importance of using psycho-education to better inform patients and their families about the condition and its potential impact on ADL (Roca et al., 2010; Videaud, Torny, Cartz-Piver, Deschamps-Vergara, & Couratier, 2012). Future studies may thus address hollistic rehabilitation programs to support PCA.

This study represents an important first investigation of the impact of cognitive rehabilitation on ADL in PCA. However, the study presents some limitations. First, the relatively small number of intervention sessions and assessments of DD's performance limits interpretation of our findings, especially in regard to long-term retention. Second, the participant's interest in the target tasks diminished progressively during the intervention, as her priorities shifted to other tasks (e.g. using the computer) that she also reported finding difficult. Moreover, DD showed occasional mood swings throughout the therapy sessions. This is in fact consistent with findings on the neuropsychiatric profile of PCA patients, in whom aspects of irritability and anxiety have been documented (Isella et al., 2015; Suárez-González et al., 2016). Although no studies have yet established a correlation between psychiatric components in PCA and independence in ADL, the reduced motivation and irritability displayed by the participant may have influenced her participation and her performance during the rehabilitation process. Thus, non-cognitive functions should be assessed in PCA, in view of the influence they may have on involvement in a cognitive rehabilitation process (Sohlberg & Mateer, 2001). Also, carefully planning for possible drop-out during the course of an ecological cognitive rehabilitation program may help in documenting the reasons why a person may want to cease her or his participation; for example, progression of the disease, less impact on everyday skills than expected, shift of priorities, etc. Future studies could also document the participants' experiences of the rehabilitation approaches as they undertake the intervention, to better identify and anticipate potential negative experiences. To our knowledge, no studies have rigorously documented those aspects and it should be done in the future.

7. General conclusion

This study represents the first investigation of the impact of PCA on ADL in an ecological context, and of the effect of a cognitive intervention approach in this disease. It describes a detailed functional profile of a person presenting with PCA, affecting mainly the dorsal stream, using a performance-based assessment administered in the patient's home and community environments. It also describes an intervention based on the *cognitive rehabilitation approach*. Findings provide a better understanding of the impact of visuo-spatial deficits and executive dysfunctions on independence in ADL in PCA. This study also showed that the settings and environments used during evaluations may considerably influence participants' performance. Although these results must be confirmed by future studies involving more participants, given that PCA is recognized as a heterogeneous disease (Borruat, 2013; Crutch et al., 2012), an improved understanding of the functional profile of PCA is a necessary first step to guide interventions aimed at maintaining participation and quality of life in this population (Harding et al., 2018).

As for the intervention that was implemented with DD, this is the first study using a combination of compensatory aids and errorless learning in PCA in order to improve performance in a specific and meaningful ADL. Further studies should involve more participants in order to validate the effectiveness of this approach in PCA. However, despite the lack of collected data, the results are promising with regard to enhancing performance in specific ADL and, therefore, self-efficacy and wellbeing of people living with PCA.

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