Published in Procedia - Social and Behavioral Sciences 23, 229-230, 2011
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Longitudinal Assessment of Language and Reading in a Case of Corticobasal Syndrome

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Introduction

Corticobasal syndrome (CBS) is a neurodegenerative disease characterized by an asymmetric extrapyramidal disorder with progressive asymmetric apraxia, rigidity and cognitive impairment in frontal and parietal domains, including executive dysfunction, social disorder, and higher-order visual processing deficits. Due to its relative rarity and since, in many cases, the diagnosis can only be made after many years of evolution, little is known about language impairments in CBS. Most studies in the domain are cross-sectional, except for the case report by Tree and Kay (2008) which presented data on the evolution of memory and language in one man with CBS over a two-year period. That study revealed substantial impairments in episodic memory, semantic memory, naming and some aspects of reading and spelling. The authors drew attention to the presence of impairments in non-word reading and spelling that needed further investigation to determine if they constitute key features of CBS.

Case Study

In the present study, CN (a 72-year-old woman, native French speaker, with 11 years of education) was assessed twice at a twelve-month interval using a comprehensive test battery of language and neuropsychological functions. CN's performance was impaired on tests exploring visual-perceptual functions, praxis, semantic memory, working memory, and executive functions. Language impairments were identified in spontaneous speech and narrative discourse, picture naming, verbal fluency, and sentence comprehension. Repetition, reading of words and oral spelling were well preserved, whilst CN showed clear peripheral agraphia. This clinical portrait remained relatively stable over twelve months, except for narrative discourse (much less fluent) and written spelling, which became extremely slow and labored with almost no legible productions. Consistent with the results of Tree and Kay (2008), a non-word reading impairment, canonical of phonological dyslexia, was found in CN. At the second evaluation, we extensively explored the functional origin of this reading deficit. CN's ability was unimpaired in tasks requiring the activation and explicit manipulation of phonological representations, a result that suggests that phonological impairment was not the functional origin of her reading deficit. She could also read every non-word with consistent grapheme-to-phoneme conversion rules, but was impaired for those with context-sensitive conversion rules (see Table 1). To be pronounced correctly, these non-words require taking into account a wider proportion of the graphemic environment, and therefore could require more executive resources.

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Table 1. Number and percentage of correct responses on reading tasks of words and nonwords according to the nature and complexity of GPC rules for CN and mean (S.D.) for controls, at twelve month follow-up

	CN		Controls	
	Score (percentage correct)		Mean (standard deviation)	
	Words	Nonwords	Words	Nonwords
Consistent GPC rules (30)	30 (100%)	27 (90%)	30 (-)	26.6 (.91)
Digraphs and trigraphs (60)	59 (98.3%)	45 (75%)	56.3 (.80)	55.3 (.87)
Context sensitive rules				
C+I/E (10)	10 (100%)	2 (20%)***	10 (-)	10 (-)
G+I/E (10)	10 (100%)	4 (40%)*	10 (-)	9.83 (.41)
AIL (10)	9 (90%)	5 (50%)	10 (-)	10 (-)
EIL (10)	10 (100%)	4 (40%)*	10 (-)	9.25 (.96)
EUIL (10)	10 (100%)	4 (40%)*	10 (-)	10 (-)
OUIL (10)	10 (100%)	5 (50%)*	10 (-)	10 (-)
Total (60)	59 (98.3%)	24 (40%)***	60 (-)	59.08 (.23)

GPC: grapheme-phoneme conversion rules
Difference between words and nonwords: * $p \leq .05$; *** $p \leq .001$

Discussion

These results support the inclusion of non-word reading impairment in the clinical profile of CBS. They also raise the question of the linguistic or executive origin of language deficits in this disease.

Reference

Tree, J.J. & Kay, J. (2008). Longitudinal assessment of language and memory impairments in pathologically confirmed cortico-basal ganglionic degeneration. *Cortex*, *44*, 1234-1247.