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Selective alexia and agraphia sparing numbers—a case study

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Abstract

We report a patient (MT) with a highly specific alexia affecting the identification of letters and words but not numbers. He shows a corresponding deficit in writing: his letter writing is impaired while number writing and written calculation is spared. He has no aphasia, no visuo-perceptual or -constructional difficulties, or other cognitive deficits. A similar pattern of performance has to our knowledge only been reported once before [Anderson, S. W., Damasio, A. R., & Damasio, H. (1990). Troubled letters but not numbers. Domain specific cognitive impairments following focal damage in frontal cortex. *Brain*, 113, 749–766]. This study shows that letter and number reading are dependent on dissociable processes. More interestingly, it points to a common mechanism subserving the perception and production of letters. We suggest that a deficit in a visuo-motor network containing knowledge of the physical shape of letters might explain the pattern of performance displayed by MT.

Keywords: Alexia; Agraphia; Reading and writing; Letter reading; Number reading; Exner's area; Visuo-motor interaction; Number writing; Third alexia

1. Introduction

Since Dejerine's (1892) description of the selective loss of reading ability (alexia without agraphia), it has become widely accepted that reading and writing are dissociable cognitive processes. Yet in most cases, alexia is associated with agraphia as well as aphasia. In addition to describing patients with alexia with and without agraphia caused by posterior lesions, Dejerine also reported a third variety of alexia, occurring in patients with anterior lesions and Broca's aphasia (Dejerine & Mirallié, 1885). More recently Benson (1977) has shown that the third alexia can be dissociated from pure alexia and the central alexias, and that it therefore deserves to be seen as a clinical entity. In 1990 Anderson, Damasio & Damasio reported a case of isolated alexia and agraphia for letters caused by a lesion in Exner's area in the left premotor cortex. To our knowledge, this is the first patient on record suffering from the third alexia in pure form, without associated aphasic deficits. This patient was completely unable to identify single letters, while being perfectly able to read Arabic numerals. Her writing of letters was slow and laborious, while on the other hand, she was perfectly able to write numbers and perform written arithmetic. We report a patient (MT) showing a similar pattern of performance. MT is severely impaired in reading and writing of letters and words, while his number reading and written arithmetic is intact. He has no aphasia or other cognitive deficits. The pattern of performance displayed by these patients represents some interesting challenges to our understanding of the cerebral organization of visual identification and written output.

2. Case report

MT suffered a head trauma in a car accident in April 1999, when he was 18 years old. He was discharged from hospital after 24 h with a diagnosis of concussion. No post-trauma symptoms were noted. When MT got home he realized that he could no longer read. He was examined by an ophthalmologist, who found that MT had severe problems with letter-identification, but that he could still read numbers. His writing was observed to be clumsily formed. On June 8th, 1999, MT went totally blind for about 10 min,

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followed by dizziness, reduced balance and headache. He was admitted to hospital, where bilateral peripheral visual field defects were found. No other neurological symptoms were noted. This was interpreted as an ischemic attack. Six months later his visual fields were found intact. MT is fully right handed (+100 at the Edinburgh handedness inventory (Oldfield, 1971)). MT gave informed written consent according to the Helsinki Declaration to participate in this study.

Before the accident, MT was a student at a commercial college and his grades were on an average level. Of particular interest is his grade in Danish, where he got a B on his latest exam before the accident, indicating that his reading and writing abilities were not below normal premorbidly. There was no report of dyslexia or other learning disabilities, and MT had no history of neurological or psychiatric disease. MT reported that he did not read much outside school before the accident. He was able to use a computer and keyboard before the injury, but mainly used this for games, not writing.

CT- and MRI-scans, performed in June 1999, showed no abnormalities. A hyperintensity was noted on MR-angiography, in the mid portion of the basilar artery, but this might not be abnormal. Seeing that our patient's pattern of performance resembled that described by Anderson, Damasio, and Damasio (1990), we performed a new MRI-scan in November 1999, looking explicitly for a lesion in Exner's area. No abnormalities were found at that time. We also performed a SPECT scan which indicated a small flow defect in the parieto-occipital area of the right hemisphere, which may include posterior temporal regions. It should be noted that this finding is somewhat uncertain, as it is at the limit of the resolution.

2.1. Neuropsychological evaluation

The neuropsychological examination and experimental investigation was mainly undertaken in June and July 1999. MT was also examined on two later occasions, at which point he was in a language training program which may have affected the test-results. Results obtained after July 1999 will therefore be marked in the following. All test-results are shown in Table 1.

2.1.1. General abilities

The neuropsychological assessment of MT revealed a verbal IQ below the normal average and a performance IQ in the upper normal range. MT's monozygotic twin brother (BT) was also assessed with a set of tests of general abilities, and a comparison between the two reveals an almost identical pattern (see Table 1). This suggests that MT's low scores on verbal subtests reflect his premorbid abilities, and are not a result of his injury. MT is fast and efficient in tests demanding high psychomotor speed, like Block Design and Trails A. His digit span, both forwards and backwards, is above normal, and also superior to BT's.

Table 1
(A) Test results in standardised tests: MT and BT (twin brother)

	MT	BT
WAIS-subtests—raw scores (standard-scores)		
Information	14 (8)	14 (8)
Vocabulary	30 (4)	30 (4)
Similarities	11 (5)	11 (5)
Digit span	15 (17)	11 (12)
Picture arrangement	30 (12)	28 (10)
Digit symbol	36 (4)	—
Picture completion	16 (11)	20 (15)
Raven advanced progr. matrices, set I		
Scoring first response	4/12	5/12
Self corrected responses	3/12	2/12
Sum correct	7/12	7/12
Other[a]		
Sentence repetition	16/22[b]	16/22[b]
Mental arithmetic	18/20	12/20
Block design, correct	12/12	12/12
Block design, time	13 s	12 s
Trail making test A	26 s	—

(B) Results of neuropsychological tests, MT	
Language	
Boston naming	48/60[b]
BORB picture naming (low freq. items)	14/15
Famous faces naming	14/15
Colour naming	10/10
Visual perception	
VOSP, shape detection	10/10
VOSP, dot counting	10/10
BORB, unusual views	13/15
Street completion test	19/20
Poppelreuter overlapping figures	15/15
Colour recognition	10/10
BIT—star cancellation	54/54
Visuoconstructive tests	
Rey's figure, copy	35/36
BORB, drawing from memory	9/9
Copy of house	3/3
MMSE, copy of figure	1/1
Memory	
Rey's figure, recall	32/36
RBMT, face recognition	10/10
Other	
Stroop—simple version	47/50[b]

VOSP, The Visual Object and Space Perception Battery (Warrington & James, 1991); BORB, Birmingham Object Recognition Battery (Riddoch & Humphreys, 1993); RBMT, Rivermead Behavioral Memory Test (Wilson, Cockburn & Baddeley, 1985); BIT, Behavioral Intattention Battery, (Wilson, Cockburn & Halligan, 1987).

[a] These tests were selected from the standard neuropsychological test-battery from Copenhagen University Hospital.

[b] This result is >2 standard deviations below the mean compared to Danish norms.

2.1.2. Visuoperceptual and -constructional abilities

The results on the visuoperceptual tests show that MT does not have difficulties in recognizing objects, faces or colors. He shows no problems with integrating fragmented pictures (Street), separating overlapping pictures (Poppelreuter) or mentally transforming images (BORB: Unusual