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How pure is pure alexia? A neuropsychological analysis of a case series of patients with alexia due to left hemispheric strokes

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SUMMARY

Background: The aim of this study was to analyze a case series with acquired alexia after stroke within the posterior areas of the left hemisphere, in the context of the current criteria for pure alexia and their relevance to the set of symptoms observable in clinical practice.

Material/ Methods: Seven patients with ischemic strokes and an initial diagnosis of pure alexia were enrolled for detailed analysis. The evaluation consisted of neuropsychological assessment in the form of standardized tests and non-standardized reading tasks, while oculomotor activity during reading was measured. Language functions, visual object and space perception, verbal and nonverbal memory, and visuospatial constructional ability were among the domains assessed.

Results: In five of the participants, pure alexia was recognized based on significant and specific discrepancies between test scores, indicating primary abnormalities in the visual processing of letter strings as a basic mechanism of the disorder. In most of the patients, coexisting cognitive deficits were revealed; however, these were disproportionately milder and less functionally significant than reading disturbances.

Conclusions: Pure alexia is a relatively rare disorder after a stroke, but it considerably affects the quality of everyday independent functioning. Its clinical characteristics in practice rarely meet all the criteria proposed in the subject literature. The differential diagnosis of this form of alexia and other reading disorders requires detailed clinical analysis.

Key words: stroke, reading, agnostic alexia, cognitive disorder

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INTRODUCTION

Alexia is an acquired reading disorder typically recognized after a focal brain injury, mainly within the dominant hemisphere, in a patient whose premorbid reading abilities were normal. It is usually associated with various language disorders as an element of aphasia syndromes (Rapcsak & Beeson, 2011). The term central alexia (Leff & Starrfelt, 2014) describes this kind of condition, and it is further divided into three different variants, namely deep alexia, phonologic alexia and surface alexia. Another group of reading disorders that are not associated with or which occurr directly with language disturbances includes the peripheral alexias (Cohen et al., 2003): pure alexia, hemianopic alexia, neglect alexia, and attentional alexia.

The term "pure alexia" is commonly used to represent the classical syndromes of "alexia without agraphia" and "word blindness" (Bub, Arguin, & Lecours, 1993), or more recent labels such as "agnostic alexia", "word form dyslexia", and "letter-by-letter dyslexia" (Starrfelt & Shallice, 2014). This disorder—first described by Dejerine (1892)—is recognized in patients from different cultures who use different sign systems (Sakurai, Yagishita, Goto, Ohtsu, & Mannen, 2006). The core symptom is difficulty in the visual processing of strings of letters or even single graphical signs. It consequently leads to a decrease in reading speed (in the case of mild impairment) or to a complete loss of the ability to recognize visually presented text. However, writing single letters, words, and even phrases may be relatively preserved, and this significant discrepancy is not observed in patients with aphasias. Some errors may, however, be present – usually caused by the inability to recall the visual representation of a sign or difficulty with following what has been written, even by the writer themselves. Diagnostic problems often concern differentiating pure alexia and aphasia, especially if (early after onset) naming difficulties emerge. Additional cognitive problems may include comorbid visual perception and memory impairment, concerning both long-term memory and the learning of new information (Johansson & Fahlgren, 1979; Damasio & Damasio, 1983; Cohen et al., 2004).

A characteristic feature of pure alexia—suggesting that this disorder is modally specific – is that reading is facilitated by outlining letters with a finger or using other (such as somatosensory) modalities (Burns, 2004). Usually, patients can also correctly spell words presented auditorially. Sometimes certain fonts are easier to recognize than others. These deficits of written language recognition may justify the label "agnostic alexia". However, it must be stressed that pure alexia does not result from visual agnosia itself, and thus, individuals with an agnosia that more generally affects visual object recognition should not be considered as pure alexic. A very characteristic feature of pure alexia seems to be the word length effect, which means that reading time is linearly, positively correlated with the number of letters in a word (Coslett & Saffran, 2001; Leff, Spitsyna, Plant, & Wise, 2006). In healthy individuals, this effect is absent or minimal, and usually does not exceed 30 milliseconds, even if a word includes more than 10

letters (Barton, Hanif, Bjornstrom, & Hillis, 2014). However, a clear word length effect can be reproduced even in healthy subjects under special circumstances such as reading low-contrast words (Fiset, Gosselin, Blais, & Arguin, 2006). Even though the word length effect can also be found in other patients with reading disorders such as surface alexia, visual agnosia or hemianopia (Starrfelt & Shallice, 2014), many authors use the term letter-by-letter reading as a synonym for pure alexia, which seems controversial.

Patients with pure alexia frequently use some top-down strategies during reading to improve word recognition, and these strategies may reveal sensitivity to lexical information. Speed of reaction increases when a word has a higher frequency in the particular language (e.g., car vs. tar), when it is put in a context (a word-length effect might be weaker in the case of sentences when compared to single words), and when it refers to something real and easy to imagine (Johnson & Rayner, 2007). Patients also read real words faster than pseudo-words and tend to better recognize single letters when they are a part of a real word than a pseudo-word (Coslett & Saffran, 2001). These kinds of top-down strategies have a facilitating effect, and they have been observed to be even stronger compared to healthy individuals (Johnson & Rayner, 2007).

Most cases with pure alexia from the subject literature were associated with a vascular pathology in the occipito-temporal areas of the left hemisphere. classical understanding of this phenomenon assumes a mechanism of interhemispheric disconnection, and thus many authors believe that damage involving the corpus callosum (splenium) is necessary. Damasio & Damasio (1983) has pointed out, however, that some authors unreflectively repeated the false information that Dejerine was a strong supporter of this view. In fact, some cases can be found in the subject literature which has either supported the hypothesis of disconnection (Damasio & Damasio, 1983; Stommel, Friedman, & Reeves, 1991) or has indicated that pure alexia is possible without lesions affecting any part of the corpus callosum (Johansson & Fahlgren, 1979; Damasio & Damasio, 1983; Cohen et al., 2004; Leśniak, Soluch, Stępień, Czepiel, & Seniów, 2014). It is believed that the lesions crucial for pure alexia include the visual word form area (VWFA), which is located in the posterior part of the fusiform gyrus of the left hemisphere (Cohen et al., 2004; Leff et al., 2006). A similar effect can be observed after the disruption of nerve fibers along which information is transferred between the VWFA and other cortical areas, especially other visual areas and language-sensitive structures of the same hemisphere (Cohen et al., 2004; Henry et al., 2005; Leff et al., 2006). The most commonly described pathology, based on CT and MRI scans, includes lesions to the fusiform, lingual, hippocampal, and parahippocampal gyri of the left hemisphere, as well as the left thalamus.

Because of the typical lesion location and its extension to the primary visual cortex, right-sided hemianopia is frequently observed (Damasio & Damasio, 1983). However, neither visual field disorders nor other sensory deficits can be responsible for the characteristic symptoms of pure alexia, because some patients do not have these deficits (Leff et al., 2006; Leśniak et al., 2014). Never-

theless, hemianopia itself may affect the reading process (Leff et al., 2000). According to Pambakian et al. (2004), hemianopia can be present in roughly 30% of patients who have suffered a stroke. Although it primarily affects the vision of contralateral stimuli, it can disturb bilateral visual processing as well (Rizzo & Robin, 1996). Thus, the real-life problems of patients in natural situations concern driving, moving in crowded places, perceiving fast-changing images, and reading. Apart from visual field deficits, abnormalities of visual search using either eye movements (Zihl, 1995a) or head movements (Zangemeister, Meienberg, Stark, & Hoyt, 1982) increase the severity of reading problems. Pambakian et al. (2000) have noticed that in patients with hemianopia, a target of saccadic eye movements is mostly unrelated to image features. Thus, their oculomotor pattern appeared more random than in healthy individuals. Shorter fixation times are likely to reflect a lack of information from the peripheral visual fields essential for planning successive eye movements.

However, there are also some differences between left- and right-sided hemianopia. While in left hemianopia a decreased amplitude and increased number of leftward saccades is observed, patients with right hemianopia experience difficulties with systematic, organized reading saccadic behavior. It is likely that both bottom-up and top-down processes of eye movement control are affected, yet higher-order (i.e., cognitive) controlling processes seem more important than the text features themselves (Rayner, Liversedge, White, & Vergilino-Perez, 2003). In this case, the cognitive control relies on peripheral information, which normally supports the reading process. However, the exact mechanism of so-called parafoveal preview is debatable. It either provides raw information that is stored as an iconic representation, or the visual information is partly processed and activates lexical units and/or semantic networks related to the word (Findlay & Gilchrist, 2003). Besides, this information may be used to direct the eyes to the most appropriate position of the following word (McConkie, Kerr, Reddix, Zola, & Jacobs, 1989). Hence, it seems that reading disorders in patients with hemianopia may result from the deficit of global spatial information necessary to plan eye movements, as observed in children with developmental dyslexia (Crawford & Higham, 2001), and different profiles of visual behaviors should be expected between individuals with pure alexia and those who present only hemianopia.

Starrfelt & Shallice (2014) have proposed several criteria for pure alexia, including the presence of an acquired reading disorder (deficit in word reading, the presence of a word length effect), correct writing and spelling, a lack of aphasia, dementia, or visual agnosia, and a lesion located in the posterior areas of the left hemisphere. From a practical point of view, the question remains as to how often a clinician may expect to see a patient who meets all the above criteria and to what extent other perceptual or cognitive symptoms contribute to the whole clinical picture. In an attempt at an answer, we analyzed seven post-stroke patients with reading difficulties as primary symptoms in the clinical picture.

MATERIALS AND METHODS

Participants

This case series included seven patients (six men and one woman, all right-handed, using Polish as their native language) hospitalized in the 2nd Department of Neurology due to an ischemic stroke. None had dementia or other significant cognitive or emotional disabilities just before infarction. They all reported serious reading disorders as a primary post-stroke deficit (decreased reading speed at least 2 SD below the average score for normal controls from the same age group), and a lesion in the posterior areas of the left hemisphere was confirmed by CT or MRI. The examined patients had neither consciousness, nor general intelligence, nor behavior control disturbances, something confirmed in the clinical assessment.

Procedures

All participants underwent a neuropsychological assessment including language, visuospatial and visual recognition functions, and memory. Their consciousness, thinking, criticism, and behavior control was assessed based on non- standardized clinical tasks.

Following the psychometric cognitive evaluation, reading speed and the magnitude of the word length effect was measured.

Methods

Measurement cognitive tools:

– Boston Diagnostic Aphasia Examination (BDAE; Goodglass, Kaplan, & Barresi, 2000) to assess five language-related areas: free narration and dialogue, auditory comprehension, expression (including repetition and naming), reading, and writing.
– Visual Object and Space Perception battery (VOSP; Warrington & James, 1991) to assess visual object perception and space perception.
– Rey's Complex Fig. Test (RCFT; Copy and Immediate Recall; Meyers & Meyers, 1995) to assess visuospatial constructional ability and visuospatial nonverbal memory.
– Auditory Verbal Learning Test (AVLT; Strauss, Sherman, & Spreen, 2006) to assess the person's ability to encode, store, and recall verbal information.

Reading assessment

The reading assessment was based on a non-standardized reading speed test comprised of a text containing 100 words. The reading time was recorded and then used to calculate a simple speed expressed by the number of words per minute. The reference group comprised 10 healthy individuals aged 52–82 years. They were asked to read the text aloud at a natural pace. The mean reading speed in this control group was 122 words per minute (SD = 20).

In order to estimate the magnitude of the word length effect, eye-tracking data were recorded using either the Iota XY-1000 eye tracker (Iota SE, Sweden) or the Hi-Speed SMI eye tracker (SensoMotoric Instruments, Germany). The recording was preceded by a nine-point calibration. Participants were allowed to move their eyes freely while the position of their heads was fixed using a headrest. Stimuli were presented on a 19″ computer screen situated 60 cm from the eyes. The stimuli included 50 words, printed in black Arial font of 1° size, presented in five blocks (three- to seven-letter words). The total dwell time (fixations and saccades inside the area of interest) was recorded and used as the reading time for each word.

RESULTS

The results for each patient obtained in the psychometric tests and the reading assessment are presented in Table 1 and Table 2.

Table 1. Scores obtained by the study patients in Boston Diagnostic Aphasia Examination

	Patient						
BDAE subtests	**P1/AE**	**P2/WC**	**P3/AM**	**P4/JS**	**P5/KT**	**P6/MN**	**P7/JZ**
Auditory comprehension							
Word Discrimination (max 72)	65	63	70	71	48.5*	62.5	59.5
Body Part Identification (max 20)	19	20	20	20	16.5	20	20
Commands (max 15)	15	15	14	15	15	15	15
Complex Material (max 12)	11	9	8	11	10	10	11
Expression							
Oral Expression (max 12)	12	12	12	12	9	12	11
Verbal Ability (max 14)	14	13	14	12	11	14	13
Automatic Sequences (max 8)	8	7	7	8	7	8	7
Repetition							
Repetition of Words (max 10)	10	10	10	10	10	10	10
Repetition of Sentences (max 16)	16	16	16	16	14	16	16
Naming							
Responsive Naming (max 30)	30	28	30	30	30	30	30
Confrontation Naming (max 105)	96	77	105	105	85	101	89
Animal Naming	9	8	12	21	5*	24	15
Reading							
Symbol Discrimination (max 10)	9	6 **	10	10	10	9	9
Word Recognition (max 8)	8	8	8	8	6*	7	8
Word-Picture Matching (max 10)	9	1 ***	10	10	4**	10	6*
Word Reading (max. 30)	18	1 **	27	30	7*	21	11*
Sentence Reading (max. 10)	7	1 **	9	10	0**	9	8
Reading Sentences and Paragraphs (max. 10)	3 *	0 **	7	10	0**	9	7
Writing							
Writing Mechanics (max. 21)	20	17	18	21	16	21	20
Serial Writing (max. 47)	47	37	39	47	35	47	45
Primer-Level Dictation (max. 15)	15	15	15	15	14	15	14
Spelling to Dictation (max. 10)	10	6	9	10	8	10	7
Written Confrontation Naming (max. 10)	9	8	10	10	6	10	8
Sentences to Dictation (max. 12)	12	7	11	12	2*	12	3

Note. Numbers indicate raw scores obtained in each subtest;
* = z score in a range -1 to 0;
** = z score in a range -2 to -1;
*** = z < -2

Table 2. Results of reading and cognitive tests obtained by the study participants

	Patient						
Cognitive measure	**P1/AE**	**P2/WC**	**P3/AM**	**P4/JS**	**P5/KT**	**P6/MN**	**P7/JZ**
Reading speed (words/min.)	4.7****	0****	54***	79.2**	2.9****	10.5****	13.3****
Word length effect (sec/letter)	2.01****	3.04****	0.03	0.21****	1.5****	1.09****	1.52****
RCFT							
Copy	36	29 **	32	29	32	33	30*
Immediate recall	16	15	6,5 *	7,5	13	21	14
AVLT							
Sum 1-5	14 **	27 **	36 *	26	18**	32*	26*
Delayed recall	0 **	0 **	3 **	0 **	0**	0**	0**
Delayed recognition	0 **	11	5 **	4 **	6*	11	6*
VOSP - object perception							
Incomplete letters	17	17	18	17	18	20	15 **
Silhouettes	8 **	20	14 **	15	18	16	15
Object decision	16	18	14 **	11 **	13**	17	17
VOSP - space perception							
Dot counting	9	9	10	8	7 **	6 **	10
Position discrimination	19	14 **	19	19	20	18	19
Number location	9	7	2 **	9	9	8	7
Cube analysis	9	10	4 **	8	5**	9	10

Note. RCFT = Rey's Complex Fig. Test; AVLT = Auditory Verbal Learning Test; VOSP = Visual Object and Space Perception battery;
* - score in a range 1 – 2 SD below the mean;
** - score in a range 2 – 3 SD below the mean;
*** - < 3SD;
**** - < 4SD;

Patient 1. (AE)

A 73-year-old male with a secondary education experienced an ischemic stroke resulting in right hemianopia. CT revealed a lesion in the left hemisphere (60 x 50 x 45 mm) involving the lingual gyrus, cuneus, fusiform gyrus, parahippocampal gyrus, and hippocampus (Fig. 1. A). A second, minor lesion (20 mm in diameter) was localized in the left middle frontal gyrus.

The patient complained of difficulties with reading and memory. He identified single letters with great effort, but was unable to read even simple written information, e.g., newspaper titles.

The neuropsychological examination did not find any evidence of aphasia, but in the BDAE – in addition to reading impairment – difficulties with color naming were revealed. His reading was slow and laborious with many errors (substitutions of visually or spatially similar letters such as "p", "d", and "b"), which resulted in problems with understanding the whole text. However, because his verbal working memory was severely impaired, it was probably not solely an effect of alexia. On the other hand, his visuospatial (nonverbal) memory appeared normal. Apart from that, the patient experienced slight difficulties with recognizing the shapes of animals and objects in the VOSP, yet his ability to evaluate spatial relationships and his visuo-constructive abilities were intact.

The diagnosis of pure alexia was made based on an extremely low reading speed (roughly five words per minute) and a considerable word length effect (over two seconds per letter). The result of the BDAE suggested relatively intact spelling and writing.

Patient 2. (WC)

A 58-year-old male, professionally active (archivist), was referred to the stroke unit after complication of a left carotid endarterectomy. Neurological examination revealed a slight right hemiparesis and right hemianopia. CT showed a large (50 x 70 x 60 mm) lesion involving the fusiform gyrus, parahippocampal gyrus, hippocampus, precuneus, splenium, and lateral thalamus (Fig. 1. B). A second, minor (10 mm) lesion was situated in the subcortical white matter of the left middle frontal gyrus.

During his stay in the acute treatment unit, the patient noticed that he was unable to read and reported that the letters seemed to be “meaningless zigzags”. A language assessment revealed occasional difficulties in naming (concerning mainly colors) and mildly decreased verbal fluency (effectiveness of searching semantic memory resources). However, the weakest and most striking were the results of the reading subtests of the BDAE. The patient had severe difficulties with reading not only words and sentences but also single letters. Fairly good writing skills contrasted with the latter symptoms. Sporadically occurring errors included substitutions and letter omissions, although he could correctly spell even the incorrectly written word.

Memory impairment involved learning of the verbal material. Mild visuospatial difficulties were revealed in one subtest of the VOSP and in the copy of the RCFT.

Pure alexia was recognized based on a considerable discrepancy between reading ability and writing skills. The patient was unable to correctly read any word from a text during a 10 minute period, although he put great effort into this task. He also displayed a large word-length effect of over three seconds per letter.

Patient 3. (AM)

A 69-year-old right-handed female with a secondary education was admitted to the stroke unit after her first ischemic stroke. Neurological examination revealed right hemianopia, central facial nerve palsy, mild right hemiparesis, and somatosensory disorders. A large (70 x 30 x 45 mm) ischemic lesion in the left occipital area and ventromedial temporal lobe was detected on CT scans. The lesion involved the lingual gyrus, cuneus, and parahippocampal gyrus (Fig. 1. C). There was also a second smaller (20 mm) lesion in the lateral part of the rostral thalamus. The patient complained mainly of visual abnormalities (including reading impairment) and difficulties with memorizing new information. No symptoms of aphasia were revealed. However, a decreased reading speed (54 words per minute) was noticeable, although without any difficulties in the reading of single words or letters. In the BDAE she obtained nearly maximum scores in the reading subtests. Similarly, the word length effect was negligible (30 milliseconds). Consequently, the diagnosis of pure alexia was ruled out, and the reading disorder was interpreted rather as a hemianopic alexia. The patient showed significant memory deficits regarding both verbal and nonverbal material. She also produced abnormal scores in both parts of the VOSP battery.

Patient 4. (JS)

An 81-year-old man (retired academic professor) was admitted to the neurological unit due to transient speech disorders and vision disturbances. The neurological examination revealed right hemianopia and right facial nerve paresis. CT revealed an ischemic lesion (30 x 60 x 45 mm) involving the left lingual and parahippocampal gyri as well as the lateral thalamus (Fig. 1. D). Small (3–4 mm) lesions were also visible in the white matter of the upper part of the right cerebellum. The patient reported memory difficulties (psychometrically confirmed by AVLT) and vision disturbances interfering with everyday activities such as reading books and watching TV. Some shape recognition difficulties were seen in the VOSP, but spatial perception was normal. He produced nearly maximum scores in the BDAE, including in the reading and writing subtests. His reading speed was reduced (79 words per minute), but no problems with single word reading or letter recognition were revealed. The word length effect was slightly elevated (210 milliseconds). Based on these results, hemianopic alexia, but not pure alexia, seemed to be the proper diagnosis.

Patient 5. (KT)

A 59-year-old male was hospitalized due to sudden right-sided vision loss and difficulties in reading with comprehension. Five years earlier, he had suffered a left hemispheric hemorrhage with hemiparesis and aphasia. These symptoms had resolved themselves almost completely. The current neurological assessment revealed right hemianopia (further confirmed by perimetry examination) and central facial nerve palsy. An ischemic lesion in the left occipital and temporal lobes was revealed by CT (Fig. 1. E). The patient's complaints included mainly reading difficulties. A neuropsychological assessment using the BDAE revealed a mild language disorder (mainly naming difficulties) and overt reading problems of a severity disproportionate to other language-related deficits. The patient also had a significantly impaired verbal memory, while his visuospatial memory was in the normal range. Based on that clinical picture, a diagnosis of pure alexia was proposed, even considering residual mild symptoms of aphasia.

Patient 6. (MN)

A 52-year-old male, an art historian, currently working as a theatre art director, with hypertension but no neurological history, suddenly experienced a stroke. His initial complaints included severe headache, visual disturbances, and difficulties in recognizing letters and numbers to the point of completely losing the ability to read. The neurological examination revealed right-sided hemianopia (confirmed in the perimetry examination) and sensory disturbances in the right upper limb. An MRI scan showed an acute ischemic lesion including the medial and lateral occipitotemporal (fusiform) gyrus, primary visual cortex, parahippocampal gyrus, hippocampus, and splenium (Fig. 1. F, G).

He was admitted to the outpatient rehabilitation clinic two months after the stroke's onset, where the initial neuropsychological examination had been carried

out. The BDAE revealed no typical aphasia syndrome, but the subtests *Sentence Reading* and *Reading Sentences and Paragraphs* were possible only when time was unlimited (reading one sentence/paragraph lasted approximately 4–7 minutes).

The patient read a 100-word text in 9 minutes 40 seconds, which was significantly below normal. Psychometric assessment revealed a considerable deficit of auditory-verbal memory (short-term and learning) and defective performance in dot counting (but not in the other VOSP subtests). Visuospatial memory was preserved, though copying time was abnormally long (above 6 minutes).

The diagnosis of pure alexia was made based on the discrepancy between the patient's undisturbed ability to write and the severe difficulties in the visual recognition of letters and words.

Patient 7. (JZ)

A 63-year-old male, an electrical technician, was admitted to the Neurorehabilitation Department three weeks after a left hemispheric ischemic stroke. His first stroke had taken place six years earlier, causing aphasia and right-sided hemiparesis, but a complete remission of those symptoms was noted. As a result of the severe second stroke, a relatively isolated alexia occurred. He was unable to recognize single letters and to read multi-word text. Moreover, he complained of occasional trouble with visually identifying commonly known objects and his personal belongings (e.g., he would misrecognize a piece of clothing until he touched it).

Neurological examination showed a slight right-sided hemiparesis (mostly involving the upper limb), a slight sensory disturbance in the right upper limb, and a right hemianopia. An MRI scan revealed an acute ischemic lesion (63 x 33 mm wide) in the left hemisphere, including the medial and lateral parts of the occipitotemporal (fusiform) gyrus, optic radiation, striate cortex, hippocampus, parahippocampal gyrus, splenium, and part of the thalamus (Fig. 1. H). Several smaller heterochronous lesions were also identified in different regions of the brain.

In the neuropsychological examination conducted a month after the second stroke, no typical aphasia syndrome was recognized, but only an increased latency in word finding. In the BDAE, major difficulties involved letter/word identification and reading aloud. Writing was relatively well preserved, with sporadic paragraphias and elisions (mostly in writing from dictation). Tasks assessing naming yielded mixed results: word finding in response to the examiner's questions was intact, while the naming of visually presented objects/colors/activities was disturbed. This discrepancy was interpreted as resulting from the patient's visual agnostic problems (complaints regarding object recognition in natural situations). Interestingly, the symptoms of visual object agnosia were not overt in the VOSP, although the patient scored in the bottom threshold of normal in the majority of the VOSP object perception tasks. The only clearly abnormal performance was found in the *Incomplete Letters* subtest. The psychometric examination revealed a considerable deficit of auditory verbal memory (short-term and learning). Visuospatial memory was only slightly limited, but the patient's copying speed was decreased.

Pure alexia was diagnosed as the main deficit based on the clear discrepancy between a relatively preserved ability to write and severe difficulties in visual recognition and the reading of letters and words, in the absence of an aphasia syndrome (Table 3).

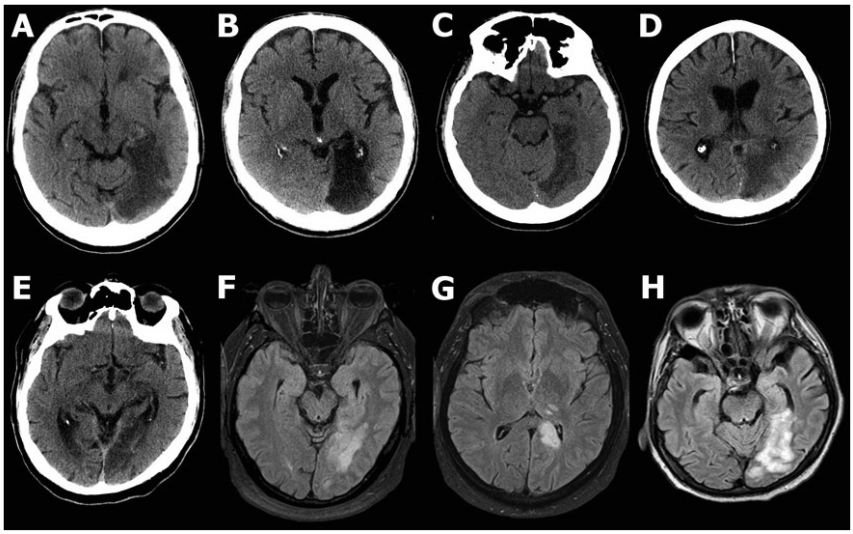

Fig. 1. Patients' brain imaging data (computed tomography or magnetic resonance imaging) showing the location of stroke.
A = patient AE; B = patient WC; C = patient AM; D = patient JS; E = patient KT; F and G = patient MN; H = patient JZ

Table 3. Patients' results in the context of criteria of pure alexia as proposed by Starrfelt and Shallice (2014)

	Patient						
Criterion of pure alexia	**P1/AE**	**P2/WC**	**P3/AM**	**P4/JS**	**P5/KT**	**P6/MN**	**P7/JZ**
1. Acquired reading disorder	yes	yes	yes	yes	yes	yes	yes
2. Correct writing and normal oral spelling	yes	mild problems	mild problems	yes	mild problems	yes	mild problems
3. No aphasia, dementia or visual agnosia	mild anomia	mild anomia	mild agnosia	yes	mild anomia	yes	mild anomia, mild agnosia
4. Deficit in word reading (prolonged RT, word length effect) or unable to read at all	yes	yes	no	marginal word length effect	yes	yes	yes
5. Lesion in the posterior left hemisphere	yes	yes	yes	yes	yes	yes	yes
Final diagnosis of reading disorder	pure alexia	pure alexia	hemianopic alexia	hemianopic alexia	pure alexia	pure alexia	pure alexia

DISCUSSION

The diagnoses of pure alexia in five of the seven cases described above were made based on the neuropsychological data analysis with particular attention to the considerable discrepancy between reading and writing skills, which is a core feature of the syndrome. The word length effect, considered a very characteristic feature of the disorder, was present in all patients except one (AM), who was diagnosed with hemianopic alexia. A slight word length effect was also observed in another patient with hemianopic alexia (JS), which means that this phenomenon is not specific for pure alexia. However, when compared to the result observed in patients with pure alexia, the difference seems striking. It is worth noting that only one of our patients (KT) had some mild language difficulties that could have been classified as aphasic, nevertheless the severity of reading impairment was disproportionate to other language-related deficits. The absence of aphasia syndrome in most of our patients with pure alexia confirmed the known clinical fact that these two deficits are distinct.

Pure alexia seems to also be independent of visual object agnosia (both apperceptive and associative). A patient can have significant difficulties with object recognition while not presenting symptoms of agnostic alexia (patient AM), and pure alexic difficulties may occur without visual object agnosia (the absence of agnostic symptoms in the case of pure alexia is one of the diagnostic criteria). Patients MN and WC showed considerable pure alexia, and at the same time, relatively preserved visual object recognition and space perception. However, this relationship is rarely clear-cut. Most cases in clinical practice seem instead to rather resemble patients AE and JZ, who, while being alexic, experienced some symptoms of object agnosia (visible during formal testing as well as in everyday life). Comorbidity of deficits probably also depends on the stage of stroke recovery.

All our patients with pure alexia had some additional cognitive deficits. The most frequent involved verbal memory. Co-occurrence of these dysfunctions is consistent with data from the subject literature (Johansson & Fahlgren, 1979; Damasio & Damasio, 1983; Cohen et al., 2004; Pąchalska 2019) and the common understanding of functional neuroanatomy – all patients had stroke lesions including the left hippocampus or adjacent structures. Visuospatial and nonverbal memory were usually better preserved. However, some visuospatial and constructional difficulties were observed in WC's and JZ's copies on the RCFT. Visuospatial abnormalities in several cases were revealed in the *Space Perception* VOSP subtests (some might have resulted from hemianopia). It is also worth noting that many of our patients made mild mistakes in writing – a fact that contradicts the classic definition of pure alexia (Starrfelt & Shallice, 2014), according to which writing and spelling should be correct.

The reading process might also be affected by the sole presence of hemianopia (Leff et al., 2000). In our case series, we identified two patients with such difficulties. Both complained of reading problems in natural situations, and their reading speeds when measured during the neuropsychological evaluation were decreased. However, the word length effect, even if slightly elevated in relation

to the normal (as in the case of JS), was negligible. Both were also able to read single letters and words. Their reading speeds were reduced, but were several times faster than those observed in other patients. Thus, the diagnosis of pure alexia was ruled out, and the observed reading difficulties were attributed to hemianopic alexia. A faulty performance in the object and space perception tasks could not be considered an additional differentiating factor, as both hemianopic patients scored below normal in some subtests of the VOSP.

CONCLUSIONS

Our conclusions are as follows: 1) agnostic alexia is a rather uncommon phenomenon; 2) pure alexia in practice rarely meets all the criteria proposed by Starrfelt & Shallice (2014); 3) a slight other type of language disturbance (e.g., anomia) can often co-occur with reading difficulty, but the latter is incomparably more severe; 4) pure alexia may co-occur with object agnosia (sometimes only concerning selected types of stimuli, e.g., atypical angle and perspective, fragmented shape); 5) other cognitive impairments (verbal memory deficits, constructional and visuospatial disturbances) are not uncommon in patients with alexia; 6) reading impairment may be secondary to a visual field deficit, which additionally hinders differential diagnosis; 7) the word length effect, considered to be symptomatic of pure alexia, can also be present in other cases, e.g., in hemianopic alexia; 8) the differential diagnosis of reading disorders requires a careful analysis of the whole clinical picture, which is especially important when establishing the basis for neuropsychological rehabilitation.

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