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Multimodal alexia: Neuropsychological mechanisms and implications for treatment

Esther S. Kim[a,*], Steven Z. Rapcsak[b,c], Sarah Andersen[a,b], Pélagie M. Beeson[a,b]

[a] Department of Speech, Language, and Hearing Sciences, University of Arizona, P.O. Box 210071, Tucson, AZ 85721, USA
[b] Department of Neurology, University of Arizona, P.O. Box 245023, Tucson, AZ 85724, USA
[c] Neurology Section, Southern Arizona Veteran's Affairs Health Care System, 3601 S. 6th Ave., Tucson, AZ 85723, USA

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ABSTRACT

Letter-by-letter (LBL) reading is the phenomenon whereby individuals with acquired alexia decode words by sequential identification of component letters. In cases where letter recognition or letter naming is impaired, however, a LBL reading approach is obviated, resulting in a nearly complete inability to read, or global alexia. In some such cases, a treatment strategy wherein letter tracing is used to provide tactile and/or kinesthetic input has resulted in improved letter identification. In this study, a kinesthetic treatment approach was implemented with an individual who presented with severe alexia in the context of relatively preserved recognition of orally spelled words, and mildly impaired oral/written spelling. Eight weeks of kinesthetic treatment resulted in improved letter identification accuracy and oral reading of trained words; however, the participant remained unable to successfully decode untrained words. Further testing revealed that, in addition to the visual-verbal disconnection that resulted in impaired word reading and letter naming, her limited ability to derive benefit from the kinesthetic strategy was attributable to a disconnection that prevented access to letter names from kinesthetic input. We propose that this kinesthetic-verbal disconnection resulted from damage to the left parietal lobe and underlying white matter, a neuroanatomical feature that is not typically observed in patients with global alexia or classic LBL reading. This unfortunate combination of visual-verbal and kinesthetic-verbal disconnections demonstrated in this individual resulted in a persistent multimodal alexia syndrome that was resistant to behavioral treatment. To our knowledge, this is the first case in which the nature of this form of multimodal alexia has been fully characterized, and our findings provide guidance regarding the requisite cognitive skills and lesion profiles that are likely to be associated with a positive response to tactile/kinesthetic treatment.

1. Introduction

Damage to left temporo-occipital brain regions can give rise to a variety of alexia syndromes that differ in terms of severity, the nature of the underlying cognitive impairment, and neural substrates (Binder & Mohr, 1992; Cohen et al., 2003; Damasio & Damasio, 1983; Dejerine, 1892). The best documented of these syndromes is pure alexia, characterized by impaired reading in the context of relatively intact spelling. The hallmark of pure alexia is letter-by-letter (LBL) reading, wherein words are decoded by sequential identification of component letters. As a result, LBL readers demonstrate a "word length effect" in that reading latency increases and accuracy decreases as a function of the number of letters in the word. In cognitive neuropsychological terms, pure alexia is often described as a peripheral processing impairment involving letter identification, where visual information from written words fails to activate representations in the orthographic lexicon in a rapid, parallel manner (Behrmann, Plaut, & Nelson, 1998). Individuals with pure alexia typically compensate for their visual processing deficit by resorting to serial identification of letters. Insofar as letter naming is intact, this strategy provides a slow, but accurate, means of decoding words.

Despite the peripheral nature of the impairment, there is evidence that individuals with pure alexia can at least partially access lexical-semantic information for written words (see Behrmann et al., 1998 for a review). For instance, frequency and imageability effects in reading (Arguin & Bub, 1993; Coslett & Saffran, 1989; Kay & Hanley, 1991), as well as greater than chance performance on lexical decision or semantic categorization tasks (Bub & Arguin, 1995; Coslett & Saffran, 1994; Shallice & Saffran, 1986) have been demonstrated in several patients with this syndrome. In fact, of

* Corresponding author. Present address: Department of Speech Pathology and Audiology, Faculty of Rehabilitation Medicine, University of Alberta, 2-70 Corbett Hall, Edmonton, Alberta, Canada T6G 2G4. Tel.: +1 780 248 1542; fax: +1 780 492 9333.
E-mail address: esther.kim@ualberta.ca (E.S. Kim).

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the 57 reported cases of pure alexia reviewed by Behrmann et al. (1998), only 13 did not show any lexical or semantic effects on any of the reading measures, suggesting that once central reading processes are engaged, they provide top-down support to facilitate letter identification.

In addition to peripheral impairments, however, central impairments in the form of degraded orthographic lexical representations have also been documented in a subset of LBL readers. Due to the loss of word-specific orthographic information, these patients present with additional features of surface alexia/agraphia characterized by impaired processing of irregular words and over-reliance on a sublexical phoneme–grapheme conversion strategy in reading and spelling (Bowers, Arguin & Bub, 1996; Friedman & Hadley, 1982; Hanley & Kay, 1992; Patterson & Kay, 1982; Rapcsak & Beeson, 2004). To be clear, such individuals are not "pure alexics." Their parallel impairments of reading and spelling suggest that the same central orthographic representations mediate both tasks, consistent with a shared components model of written language processing (Tainturier & Rapp, 2001). Indeed, evidence from neuroimaging studies with normal readers corroborates that reading and spelling tasks produce overlapping activations in left inferior temporo-occipital cortex corresponding to the visual word-form area (VWFA; Cohen et al., 2000), suggesting that this region is a possible neural substrate of the orthographic lexicon (Beeson & Rapcsak, 2003; Cho, Rapcsak & Beeson, 2010; Rapp & Lipka, 2010). Consistent with this view, damage to the VWFA is a frequent finding in LBL readers, although the syndrome can also be caused by lesions that degrade or disrupt visual input to this region (Binder & Mohr, 1992; Cohen et al., 2003; Henry et al., 2005; Epelbaum et al., 2008).

Most LBL readers are reasonably accurate in decoding words by relying on a serial letter naming strategy. However, other patients with left temporo-occipital lesions have an additional letter identification/naming impairment that precludes the use of the compensatory LBL strategy, resulting in nearly total inability to read, or global alexia. It has been proposed that global alexia is attributable to a visual-verbal disconnection induced by damage to white matter pathways disrupting the transfer of visual information to left-hemisphere perisylvian language areas (Binder & Mohr, 1992; Cohen et al., 2004; Dejerine, 1892). The responsible lesions generally involve the inferior temporo-occipital regions implicated in LBL reading, but there is also evidence of dorsal extension into occipital white matter pathways producing damage to callosal fibers traveling in the splenium and forceps major (Binder & Mohr, 1992). Patients with global alexia typically present with dense right hemianopia and thus can only process visual information from the left visual field projected to the right hemisphere. Therefore, the severe letter naming impairment of these patients suggests that successful use of a compensatory LBL reading strategy requires the structural integrity of interhemispheric connections between right hemisphere visual association cortex and left-hemisphere perisylvian language areas (Cohen et al., 2003; 2004).

Treatment for patients with severe alexia and impaired letter naming has involved the use of kinesthetic or tactile techniques to facilitate letter recognition. In such treatments, the patient either traces letters with his/her finger (kinesthetic approach; e.g., Maher, Clayton, Barrett, Schober-Peterson & Gonzalez Rothi, 1998), or has a helper trace letters into his/her palm to facilitate identification of component letters in a word (tactile approach; e.g., Sage, Hesketh & Lambon Ralph, 2005), or a combination of kinesthetic and tactile input is provided as the patient traces letters into the palm of his/her own hand (e.g., Lott, Friedman & Linebaugh, 1994). The rationale for these methods is that the combined stimulation from tactile/kinesthetic and visual input is expected to improve letter naming and thereby facilitate access to the orthographic lexicon. Once letter identification has reached an acceptable level, training is then geared toward improving speed and accuracy of reading. The benefit of tactile/kinesthetic input to assist in letter naming was acknowledged by Goldstein (1948) and documented in a number of subsequent case reports across a variety of language systems (Kashiwagi & Kashiwagi, 1989; Kreindler & Ionasescu, 1961; LaPointe & Kraemer, 1983; Luria, 1970; Stachowiak & Poeck, 1976). Of particular interest are studies that experimentally controlled for the specific effects of tactile or kinesthetic treatment in alexia with impaired letter naming. This includes the eight English-speaking cases summarized in Table 1. All of these individuals had acquired alexia with persistent letter naming impairments that limited their ability to effectively use a LBL compensatory strategy. As shown, letter naming accuracy ranged from 0% to 77% at the outset, and following various forms of tactile/kinesthetic treatment, letter naming skills were improved in all but one (Sage et al., 2005). Additionally, all participants showed improved single-word reading accuracy and/or speed. Thus, the therapeutic value of tactile/kinesthetic treatments for global alexia appears to be relatively strong.

Response to tactile/kinesthetic reading treatment is dependent upon the ability to gain letter identity information via the tactile or kinesthetic modality. Relevant to this issue is the fact that there are some individuals for whom this skill is selectively impaired. A cohort of Japanese individuals with "kinesthetic alexia" who were unable to name letters or read words via the kinesthetic modality have been described by several investigative teams (Fukatsu, Fujii & Yamadori, 1998; Ihori, Kawamura, Fukuzawa & Kamaki, 2000; Ihori, Kawamura, Araki & Kawachi, 2002). Despite having intact visual reading and somatosensory function, some of these individuals demonstrated a kinesthetic-verbal disconnection attributed to damage to left parietal cortex and underlying white matter. Considered relative to the visual alexia literature, these observations suggest an apparent double dissociation between visual and kinesthetic impairments of reading. Specifically, damage to left occipito-temporal cortex results in defective visual identification of letters and words with preserved kinesthetic reading (e.g., Lott et al., 1994; Maher et al., 1998), whereas the opposite pattern is observed in patients with left parietal lesions (Fukatsu et al., 1998; Ihori et al., 2000; 2002).

In the present study, we explored the nature and treatment of an unusually severe case of acquired alexia accompanied by mild surface agraphia. Although the participant attempted to use a compensatory LBL reading strategy, success was limited due to her inability to correctly identify letters from visual input. At the outset, she was considered to be a good candidate for kinesthetic treatment; however, her modest response to treatment prompted further evaluation regarding the nature of her deficit. Detailed kinesthetic assessment and examination of her performance relative to a control group of participants with acquired alexia due to left temporo-occipital lesions further clarified the locus of the breakdown in cognitive functioning, and allowed us to hypothesize the regions of neural damage that contributed to her impairment. Ultimately, it became evident that the combination of deficits in this woman reflected a multimodal alexia profile not previously described in the literature.

2. Case history

2.1. Patient description

ST was a 74-year old right-handed female with 12 years of formal education. Prior to retirement, she worked as an office manager and reported no developmental history of reading or spelling difficulties. At the time of study, ST was 15 months post onset of a hemorrhagic stroke affecting left temporo-parieto-occipital brain regions. She had previously suffered a right hemisphere stroke 4.5