Reframing pure alexia: Models of visual processing and implications for assessment and treatment strategies

Pure alexia (alexia-without-agraphia) is an acquired breakdown in reading ability with the preserved ability to write and to: (1) process linguistic stimuli via other modalities and (2) visually process stimuli other than written words (e.g., Rapcsak & Beeson, 2004). This disorder has been well-characterized with respect to both its neuroanatomical and cognitive psychological underpinnings, and this is the model with which most rehabilitation professionals are most familiar – and thus the approach from which behavioral assessment and treatment has been motivated (e.g., Warrington & Shallice, 1980).

In neuroanatomical terms, the domain-specificity of pure alexia is tied directly to what has been termed the "visual word form area" (VWFA; e.g., Cohen & Dehaene, 2004). According to the VWFA hypothesis, this region (left temporo-occipital cortex along the mid-fusiform gyrus) plays a critical role in both learning to read and in fluent literacy, which require rapid recognition of visual word forms. The role of this area in reading has received support from both functional neuroimaging and lesion-deficit studies (e.g., Brem et al., 2006; Schlaggar & McCandliss, 2007). Importantly, lesion-deficit studies across cohorts of patients with pure alexia have demonstrated a critical lesion area within the left middle fusiform gyrus (e.g., Damasio & Damasio, 1983); this area is spared in patients with left posterior cerebral artery infarcts and no reading deficits (Binder & Mohr, 1992).

It follows that if a critical cortical area subserves visual word form identification for reading, then pure alexia represents a domain-specific deficit; i.e., degradation of the representations themselves (substantiated neuroanatomically within the VWFA), or decreased ability to access the representations (i.e., white matter tracts leading to VWFA) (Dejerine, 1892; Geschwind, 1965). Accordingly, cognitive neuropsychological models of reading often identify a visual word form system or orthographic input lexicon (OIL) as a separate and specific functional module in the reading process (Beeson & Henry, 2008). Pure alexia, then, is conceptualized as a modality-specific inability to process orthographic input due to a dysfunction of or problem accessing the OIL.

An alternative view of pure alexia, however, is that it represents a modality-specific but domain-general visual processing problem (e.g., Behrmann et al., 1998). By this view, pure alexia is but one manifestation of a core visual processing deficit; that whole word reading requires fast, parallel letter identification results in reading being the most noticeably affected process (e.g., Price & Devlin, 2003). Thus, pure alexia may represent a problem present in any situation requiring visual processing of multiple items presented simultaneously (Starrfelt et al., 2009), and the seemingly material-specific deficit in pure alexia arises due to the unique demands reading places on the visual system, as opposed to processes that operate in a more serial fashion, such as number or object recognition.

The implications of the domain-general view of pure alexia for clinical assessment and treatment are clear: We see what we believe, and thus clinicians working with patients with pure alexia will be unlikely to assess for and thus treat the underlying visual deficits that may cause or contribute to the reading disorder. It is possible, however, that broadening our view of the problem will lead to more effective treatment measures. Therefore, we present a case study, MCL, to illustrate the spectrum of the visual processing impairment which may underlie pure alexia and the possible effectiveness of treatment strategies aimed at these broader visual and cognitive underpinnings of reading.

<u>Methods</u>

Participant. MCL is a 61yo right handed female psychology professor status-post a single, left-hemisphere CVA in the distribution of the posterior cerebral artery, resulting in a temporal-occipital lesion. Immediately following her stroke, she presented with global alexia without agraphia. She demonstrated recognition of single letters within days, and was able to read in a letter-by-letter strategy within a few weeks. There was no evidence of aphasia nor any other cognitive difficulties, although she did complain of "short term memory" problems.

MCL returned to teaching on a part-time basis at 6 months post-stroke, but continued to experience considerable difficulty reading her own slides and completing other literacy-based work activities (e.g., grading); therefore, she sought additional therapy for her alexia at 12 months post-onset.

Pre-Treatment Assessment. Following IRB approval and informed consent, MCL completed an initial test battery examining oral and written language, attention, memory, and executive function skills (Table 1). Test results were consistent with pure alexia without aphasia. There were no lexicality effects (Table 1), but clear word-length effects were evident; moreover, MCL used the tactile-kinesthetic strategy of "writing" words with her dominant hand (in the air or on the table), for longer and low-frequency words prior to reading aloud. Oral reading on the GORT-4 was relatively accurate albeit extremely slow. MCL demonstrated impaired performance on all visual, timed tasks; these domain-general impairments were supported by MCL's subjective description of her reading skills (Table 1). Written language was within normal limits (Figure 1).

Study Design. A single-subject, multiple-phase (A-B-C-A) treatment design was employed. Reading rate, decoding accuracy, and reading comprehension were regularly probed to examine the effects on reading proficiency of a text-level reading treatment alone versus direct attention training. A verbal fluency task served as a control probe. General treatment phases, established on a pre-determined timeline, were as follows:

Phase A (2 sessions): Baseline

Phase B (4 sessions): Direct attention training

Phase C (4 sessions): Reading treatment

Phase A (2 sessions): Return to baseline

Probe Tasks. A paragraph-level oral reading task was administered weekly to monitor decoding accuracy, rate, and comprehension. Materials were modified from two Graduate Record Examintion workbooks (Kaplan, 2003), and a different passage was used each week, with stimuli roughly equated for length (*m* = 237, *SD* = 43, range = 173-288 words) and complexity (calculated using the Flesch-Kincaid grade-level formula (Kincaid et al., 1983) (*m* = 12.6, SD = .31, range = 12.2-13.1).

Treatment Protocol. MCL received one, 90-min. treatment session per week for 12 weeks, with the first 4 sessions devoted exclusively to direct attention training (APT-II, Sohlberg & Mateer, 1998) and the next 4 sessions devoted to a text-level reading treatment (MOR; Beeson, 1998). An integral treatment component involved daily homework, designed to mimic treatment sessions, for one hour each day.

<u>Selected Results</u>

Probe performance. MCL's decoding accuracy remained stable throughout treatment, with variable comprehension and rate and without a discernable pattern or difference from the attention to text treatment protocols (Figure 2). Due to the high level of variability, we did not subject these data to statistical analysis. Verbal fluency performance was stable throughout and following treatment.

Standardized testing. Postitive gains in reading probe measures were supported by changes in decoding accuracy, rate and comprehension on the GORT-4 post-treatment (Figures 3 and 4). Noteably, the slope of change was linear, and in the expected direction (increased reading rate and decoding accuracy) following both the APT and MOR protocols.

<u>Discussion</u>

Our data partially support previous research demonstrating the utility of attention training (APT) for acquired alexia, and specifically for pure alexia. These data are consistent with the feasibility of a domain-general approach to pure alexia and demonstrate that careful clinical exploration may reveal domain-general visual problems beyond simply OIL access. We explore clinical and theoretical implications of these data and stress the need for clinicians to consider the complexity of visual functions supporting the reading process.

Table 1. Pre-treatment assessment

TEA	WNL: Untimed visual tasks (Visual elevator 1), Auditory tasks (Elevator count d reversal), Dual tasks (Telephone search with Counting) Below: Visual, timed tasks (Map Search 1 and 2, Visual elevator 2 (timed compo search)
RBMT	General memory index = 79 Percentile rank = 8 Below: delayed verbal memory (names, delayed story recall), delayed visual me delayed new learning (delayed novel task)
GORT-4 **Stories 7-14 (Form A)**	Visual errors: characterized → categorized, dismal→ dismantle, possibly→pote Rate score = 0 for stories 9-14
PALPA – nonwords and frequency/imageability-controlled word list	100% accuracy for words and non-words, obvious struggle/letter-by-letter stra words
MCL's subjective description of her reading	"I can't go back and look for a particular word in a passage. I can't scan left to right to see what's coming next. But… I also can't find items in a grocery store, or in my house, or on my desk. I have to track everything line by line and look at each item in detail. It won't just 'jump out'."

Figure 1. Written language example (BDAE written narrative), pre-treatment

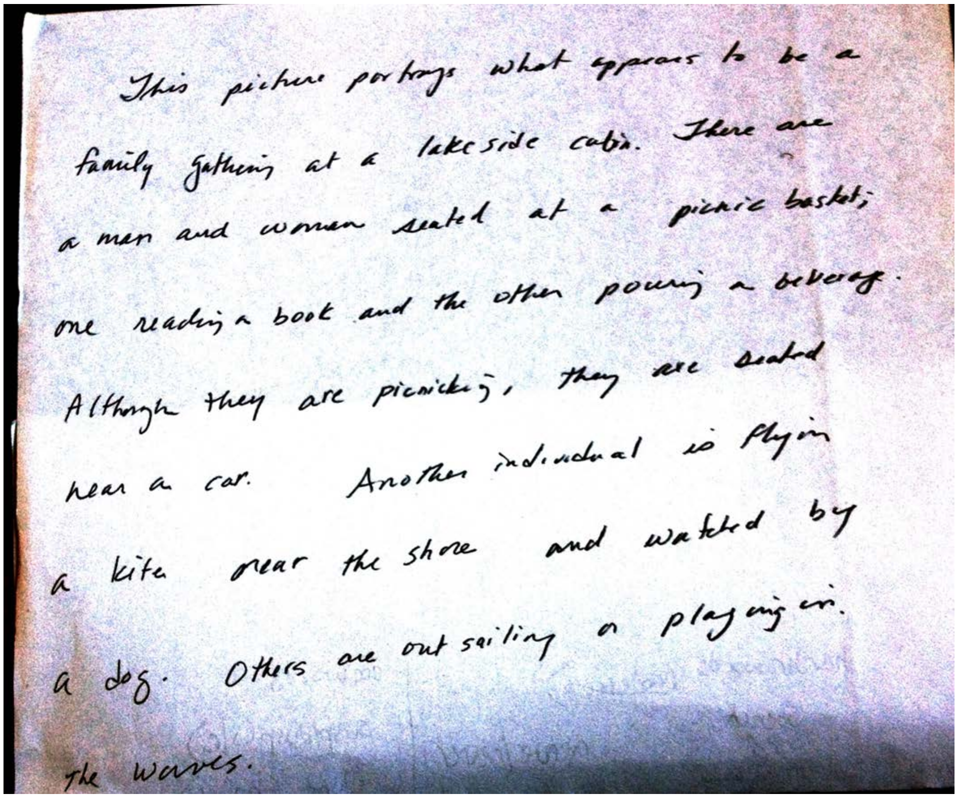
This picture portrays what appears to be a family gathering at a lakeside cabin. There are a man and woman seated at a picnic basket; one reading a book and the other pouring a beverage. Although they are picnicking, they are seated near a car. Another individual is flying a kite near the shore and watched by a dog. Others are out sailing or playing in the waves.

Figure 2. MCL Probe performance

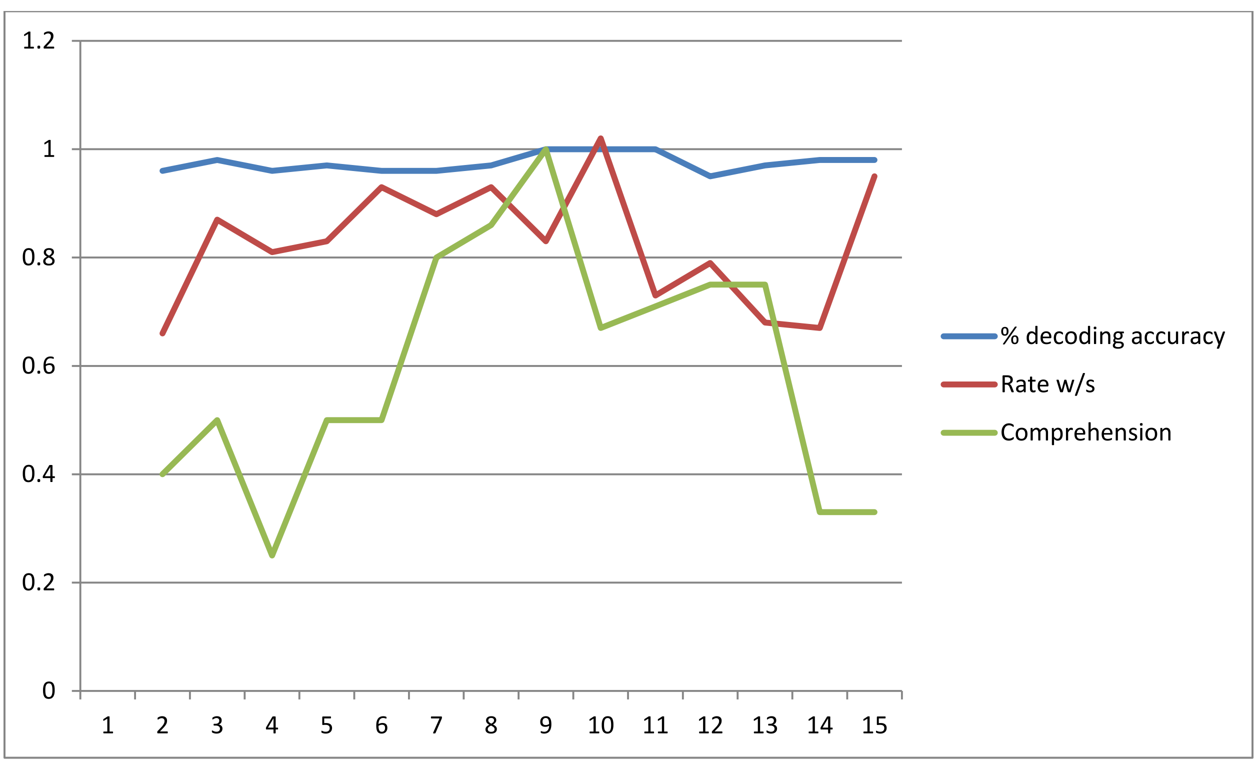

Figure 3.

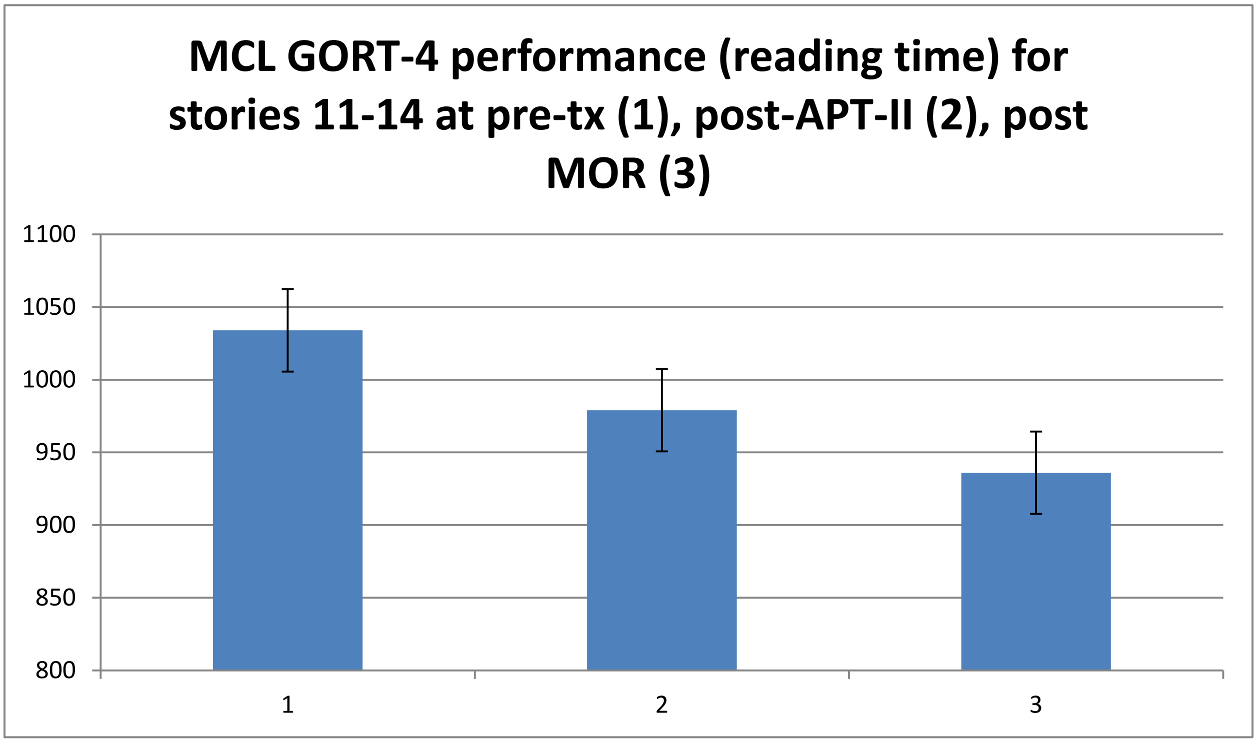

Figure 4.