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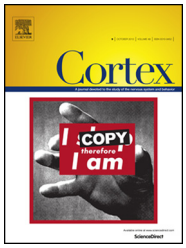

Research report

Intact reading in patients with profound early visual dysfunction☆

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ABSTRACT

Despite substantial neuroscientific evidence for a region of visual cortex dedicated to the processing of written words, many studies continue to reject explanations of letter-by-letter (LBL) reading in terms of impaired word form representations or parallel letter processing in favour of more general deficits of visual function. In the current paper, we demonstrate that whilst LBL reading is often associated with general visual deficits, these deficits are not necessarily sufficient to cause reading impairment and have led to accounts of LBL reading which are based largely on evidence of association rather than causation. We describe two patients with posterior cortical atrophy (PCA) who exhibit remarkably preserved whole word and letter reading despite profound visual dysfunction. Relative to controls, both patients demonstrated impaired performance on tests of early visual, visuoperceptual and visuospatial processing; visual acuity was the only skill preserved in both individuals. By contrast, both patients were able to read aloud words with perfect to near-perfect accuracy. Reading performance was also rapid with no overall significant difference in response latencies relative to age- and education-matched controls. Furthermore, the patients violated a key prediction of general visual accounts of LBL reading – that pre-lexical impairments should result in prominent word length effects; in the two reported patients, evidence for abnormal word length effects was equivocal or absent, and certainly an order of magnitude different to that reported for LBL readers. We argue that general visual accounts cannot explain the pattern of reading data reported, and attribute the preserved reading performance to preserved direct access to intact word form representations and/or parallel letter processing mechanisms. The current data emphasise the need for much clearer evidence of causality when attempting to draw connections between specific aspects of visual processing and different types of acquired peripheral dyslexia.

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1. Introduction

The concept of the visual word form is one that is well-established within the psychological literature. Cattel (1886) first documented 'whole word' reading by demonstrating how briefly presented words were easier to recall than briefly presented meaningless letter strings, and letters have subsequently been shown to be better identified when presented

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within a word than individually (Reicher, 1969; Wheeler, 1970) or within a non-word (Grainger et al., 2003). More recently, neuroimaging studies have identified an area within the left fusiform gyrus which is specialised for letter and word recognition and which may constitute the visual word form area (VWFA; Cohen et al., 2000). Given the recency of written relative to spoken language as a cultural invention, it is unlikely that a VWFA would have evolved specifically for reading. However, one suggestion is that accumulated reading experience promotes the specialisation of a pre-existing inferotemporal pathway for higher-order visual processing (McCandliss et al., 2003). The current paper emphasises the extent of this functional specialisation by demonstrating remarkably preserved reading in the context of profoundly impaired perception of non-word stimuli.

Neuropsychological evidence supporting the existence of highly-specialised processes for visual word recognition has been derived from patients exhibiting 'letter-by-letter reading' (LBL; also referred to as 'word form dyslexia' or 'pure alexia'; e.g., Shallice and Warrington, 1980; Farah and Wallace, 1991; Binder and Mohr, 1992; Warrington and Langdon, 1994; Hanley and Kay, 1996; Cohen et al., 2000). Such patients exhibit intact letter identification and relatively accurate, but slow, reading, whereby response latencies increase in a linear manner proportionate to word length. LBL reading has been suggested to reflect destruction or inaccessibility of a visual word form system, and is associated with damage to the VWFA (Warrington and Shallice, 1980; Cohen et al., 2000).

The attribution of LBL reading to a specific word form deficit has been challenged on two main grounds, namely that the condition and its characteristic word length effects can be accounted for by a general visual deficit and/or a letter identification deficit.

A general visual account of LBL reading suggests that reading, as a complex behaviour, can be disrupted by even the most subtle low-level visual deficits (Friedman and Alexander, 1984; Farah and Wallace, 1991; Price and Devlin, 2003), which propagate by a cascade process to the level of lexical and semantic representations within the visual system (Behrmann et al., 1998a, 1998b). A number of single case and case series studies of LBL readers have reported associated impairments on a range of perceptual tasks involving non-orthographic stimuli. For example, Friedman and Alexander (1984) identified an LBL patient who was impaired on tasks of letter identification, object recognition and had an elevated threshold relative to controls in detecting briefly presented pictures. Furthermore, Farah and Wallace's (1991) patient TU performed poorly on tasks involving the perception of non-orthographic stimuli under time constraints; these results were replicated by Sekuler and Behrmann (1996). More recently, Mycroft et al. (2009) found that seven LBL readers were similarly impaired for both linguistic and non-linguistic stimuli on tasks of visual search and matching, and the LBL group as a whole performed worse than the control group on a task of visual complexity. By contrast, there are documented cases of LBL readers with no discernible impairment in letter identification speed or the identification of rapidly displayed letters (Warrington and Langdon, 2002; Rosazza et al., 2007) or in a range of tasks assessing visual processing, such as complex picture analysis, visual short term memory and picture recognition from unusual views (Warrington and Shallice, 1980). However, proponents of pre-lexical theories of LBL reading tend to dismiss such cases as reflecting insufficiently sensitive assessment of visual processing skills or the use of non-reading tasks which are not making demands comparable to those involved in reading (Behrmann et al., 1998a, 1998b; Patterson, 2000).

Alternative accounts attribute LBL reading to an impairment of letter activation. Some accounts suggest that the critical letter processing deficits may be restricted to the identification of individual letters (e.g., Arguin and Bub, 1992, 1993; Reuter-Lorenz and Brunn, 1990; Behrmann and Shallice, 1995). Other accounts ascribe LBL reading to a deficit in the mechanisms responsible for rapid, parallel processing of letters, leading to the less efficient serial encoding of the component letters of a word (Patterson and Kay, 1982; Behrmann et al., 2001; Cohen et al., 2003). One such possible mechanism is the inability to use the optimal spatial frequency band for letter and word recognition, with letter confusability effects emerging at lower spatial frequencies (Fiset et al., 2006). It should also be noted that some authors have argued that deficits in letter processing are common to all LBL readers, while speculating that such deficits may be due to a more basic visual impairment (Behrmann et al., 1998a, 1998b).

One observation regarding both the general visual account of LBL reading is that the evidence base is largely associative in nature; that is, most studies claim that the co-occurrence of the characteristics of LBL reading (i.e., accurate but slow reading, with prominent word length effects) and a particular deficit (e.g., impaired perception of non-lexical stimuli) confers support for their chosen position. In addition, proponents of the general visual impairment account have claimed support for their position from control brain-damaged patients who show the complementary association of no perceptual deficit and no impairment of reading (e.g., patient OL; Mycroft et al., 2009). By contrast, in the current study it is argued that such evidence does not prove a causal link between general visual deficits and LBL reading behaviour. This is achieved by presenting evidence from two patients who exhibit profound visual dysfunction in the presence of accurate and rapid word reading. Rather than demonstrating a selective impairment to the visual word form system in the absence of general visual dysfunction, these patients' reading abilities are remarkably preserved despite grave and diffuse impairments to their visual system.

The two patients reported in this study have a diagnosis of posterior cortical atrophy (PCA), a neurodegenerative condition involving progressive visual impairment in contrast to relatively spared memory functions. The most frequent underlying pathology is Alzheimer's disease (AD), with PCA patients showing a greater distribution of senile plaques and neurofibrillary tangles in posterior regions of the parietal cortex, the occipital cortex and temporo-occipital junction relative to more anterior cortical areas (Rogelet et al., 1996; Ross et al., 1996; Tang-Wai et al., 2004). Characteristic symptoms of PCA include early visual processing deficits, and disorders of higher-order visuoperceptual and visuospatial processing (Benson et al., 1988; Mendez et al., 2002; Tang-Wai et al., 2004). Reading difficulties are often a prominent feature of PCA, occurring in about 80% of patients (Mendez et al., 2002)

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